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*Opinion Article*

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# Role of Vestibular Rehabilitation in Patients with Superior Semicircular Canal Dehiscence Syndrome

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**ABSTRACT**

Superior canal dehiscence syndrome (SCDS) described in 1998 is a disorder characterized by vertigo in response to loud sounds, pulsatile tinnitus, conductive hyperacusis, autophony and chronic disequilibrium. While resurfacing and/or plugging of the superior canal may improve autophony and reduce dizziness handicap, it can result in immediate and long-term deficits in VOR function, necessitating vestibular rehabilitation (VR) during the early post-operative recovery period. Case description: a 34- year-old man with a history of autophony, pulsatile tinnitus and audible eye movements in his right ear. He denied any vertigo or disequilibrium. Immediately after surgical plugging he presented severe vertigo and disequilibrium as well as deterioration of right hearing loss. Due to persistent instability, VR was performed for five months after surgery with improved outcomes. Our purpose is to emphasize and increase awareness in surgeons, especially in Latin America, the importance of VR immediately after surgery. Additionally, we make some comments about the surgical decisions in two patients with otosclerosis and SCDS and the importance of VR.

**Keywords:** Superior Semicircular Canal Dehiscence, Chronic Disequilibrium, Vestibular Rehabilitation

## Introduction

Superior semicircular canal dehiscence syndrome (SCDS) was first reported by Lloyd Minor and colleagues in 1998 [1]. They described a series of patients with symptoms of chronic disequilibrium with sound or pressure, and also, induced vertigo and nystagmus in the plane of the superior semicircular canal. This marked a period of heightened interest in both the diagnosis and treatment of SCDS.

Over time, the pathophysiology of the third mobile window syndrome was also described. This theory explained that sound pressure entering the oval window is dissipated through the labyrinth. For air-conducted sound, a loss of acoustic energy could explain the increase in hearing thresholds. For bone conducted sound, low impedance levels could allow bone-conducted sound to access the perilymph of the inner ear via the bone labyrinth, resulting in hearing bone-conducted sounds better than normal [2].

Patients with SCDS can have symptoms of autophony, pulsatile tinnitus, audible eye movements, footsteps, chewing and bowel movements. These patients could also experience aural fullness and chronic disequilibrium. Moreover, vertigo likely arises from the flow of endolymph in the superior canal ampulla, a consequence of distinct pressure gradients between the oval window and the dehiscence.

Importantly, clinical manifestations may vary from patient to patient. Some SCDS patients may present with a myriad of these symptoms, meanwhile other patients refer primary audiological symptoms without sound or pressure induced vertigo. Moreover, few patients refer only chronic disequilibrium [3].

Definitive surgery, such as resurfacing and/or plugging of the superior semicircular canal, have the goal to eliminate the third window pathophysiology [4]. Patients with debilitating symptoms may be candidates to surgery. Furthermore, surgery may improve autophony and dizziness handicap [4].

After surgical plugging, patients are expected to have a reduction in the function of the superior semicircular canal [5,6]. Unfortunately, one third of patients have a temporary pan-labyrinthine hypofunction and approximately 15% have plugging material that also impacts function of the posterior canal [7,8]. Additionally, benign paroxysmal positional vertigo

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(BPPV) has been reported in up to 25% of patients post-surgery and approximately 25% of patients develop a high-frequency sensorineural hearing loss [8,9].

Although surgery for SCDS improves patient´s symptoms, it could also affect vestibular function both acutely and chronically. Patients should be warned of the risk of vestibular hypofunction after surgical repair of the dehiscence [9]. Furthermore, following surgical repair of the dehiscence, patients often experience dizziness and imbalance [10].

Both post-surgical BPPV and temporary labyrinthine hypofunction require vestibular rehabilitation (VR). Vestibular rehabilitation, especially in Latin America, is rarely indicated after surgery of SCDS. We must be able to communicate these considerations to the patient to provide proper counselling. Specific interventions included vestibular habituation and adaptation exercises, balance and gait training, and patient education is needed [11].

Perioperative steroids are given variably across reports, often for longer durations if signs of sensorineural hearing loss, labyrinthine hypofunction, or facial paresis are detected postoperatively.

Early postoperative persistence of slight decline in auditory and vestibular function may be due to fluid, blood, or air at the surgical site, and must be follow up clinically [12].

Preoperative and postoperative audiometry and vestibular testing allow objective tracking of symptoms and outcomes.

SCDS is well known as a great mimicker [13]. Most of the radiographically diagnosed SCDS are asymptomatic and does not necessarily imply an active third inner window. SCDS is a great audiologic mimicker and the differential diagnosis must be made with Meniere´s disease, otosclerosis vestibular migraine.

The clinical and audiometric presentations of patients with concurrent otosclerosis and SCDS are often indistinguishable from those of patients with only otosclerosis [14]. The presence of both, otosclerosis and SCDS can mimic the conductive hearing loss of otosclerosis, this clinical situation has been a well described cause of stapedectomy failure reported in the literature. Stapedectomy in these patients typically results in a persistent conductive hearing loss.

Surgery (stapedectomy) may turn a non-active 3rd window into an active one. Sound energy could be attenuated by the fixed stapes, and hence, the drive for auditory symptoms is smaller [15]. It may also change the mechanism of energy dissipation in the inner ear preventing vestibular symptoms. In the face of a 3rd window, the inner ear may have lower resistance to sound. Fenestration or removal of the stapedial footplate could transfer some abnormal energy to the inner ear and could increase the higher risk of inner ear damage during operation. Vestibular rehabilitation following surgical repair focusing on central compensation and habituation is needed [16,17]. Patients may be advised about risks and may avoid surgery, preferring instead the use of hearing aids. Patients need to be aware of VR after superior canal surgery.

Many patients with SCDS also have migraine. SCDS is an effective migraine trigger [18]. Prior to pursuing surgery, control of migraine is critical to avoid exacerbation of migraine after surgery and to distinguish treatable symptoms that are unliked to be helped by repairing a dehiscence.

**Objective**

Our purpose is in a brief and concrete communication to justify and emphasize the need for early intervention of vestibular rehabilitation in cases of SCDS. To this end, we describe two clinical cases that exemplify different prognoses when vestibular rehabilitation is performed in time. We describe the effects of physical therapy involving vestibular rehabilitation exercises such as gaze stabilization, sensory reweighting, adaptation, and proprioceptive training in the treatment of patients after surgical repair of SCDS.

**Clinical Case 1**

34-years-old man who referred a history of eighteen months of autophony, pulsatile tinnitus associated with effort and physical activities as well as audible eye movements in his right ear. He had no vertigo, dizziness or disequilibrium. His audiological test was normal but we noticed conductive hyperacusis with thresholds of -10 dB in bone conductive response in low frequencies that are compatible with tuning forks. Air conducted cVEMPs and bone conducted oVEMPs with tone burst at 4000 Hz were positive for SCDS. Temporal bone CT demonstrated dehiscence of the right SCC. He underwent surgery in another institution. Immediately after surgical plugging he presented severe vertigo and disequilibrium as well as sensorineural medium right hearing loss. Two months later he was treated with intratympanic dexamethasone with partial recovery of his right sensorineural hearing loss. We saw him five months later due to persistent instability and right SN hearing loss. (Figure 1). Instead of low VOR gain just in the plane of superior SCC he had a low gain (0.78) and asymmetric VOR of 21% in the horizontal plane in HIMP protocol and 33% of asymmetry and 0.68 of horizontal VOR gain in SHIMP protocol (Figure 2) Dizziness handicap inventory (DHI) was 48 (moderate handicap), dynamic gait index (DGI) was 17/24 and dynamic visual acuity (DVA) showed 3 line difference [19]. We started VR focusing on the principles of central compensation and habituation. Improved outcomes after three months of VR: DHI 3/100, dynamic gait index 24/24, DVA no line difference. vHIT showed normalization of VOR gain with symmetric response and no compensatory saccades.

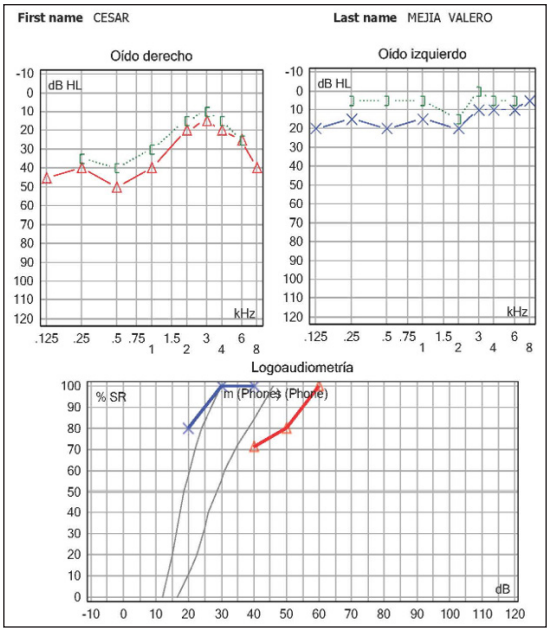


**Figure 1:** Sensorineural right hearing loss after surgery

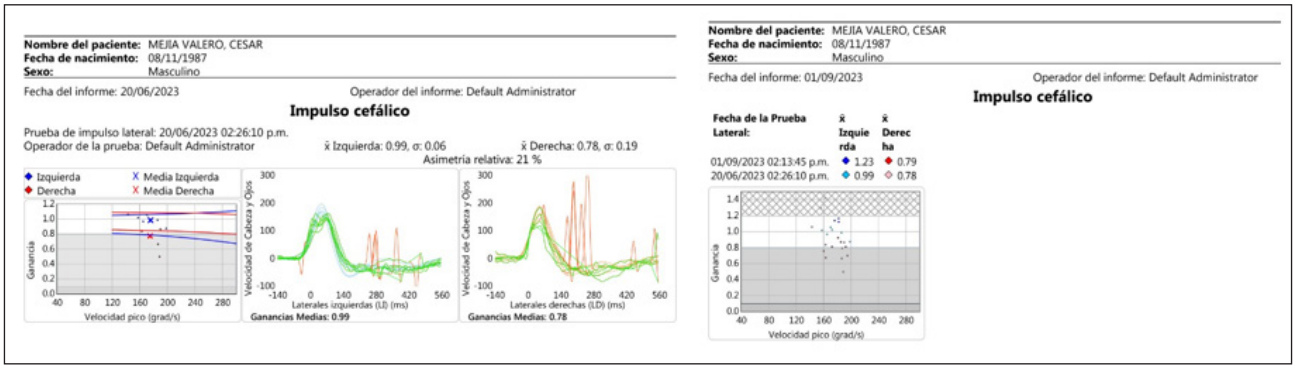


**Figure 2:** Low and asymmetric right horizontal VOR with uncovered saccades in vHIT, both in HIMP and SHIMP protocols

## Clinical Case 2

47-year-old male with a long history of bilateral conductive hearing loss. Right stapedectomy in a different institution. Persistent air-bone gap postoperatively. CT scan confirmed good placement of the prosthesis and bilateral SCCD. Left stapedotomy. Progressive right hearing loss. Loud autophony and aural fullness in the right ear become permanent. He experienced dizziness and vertigo trigged by physical activity, rapid head movements and band. He was submitted to right SCC plugging via MFA. We started VR as soon as he left the hospital. At 3 months post operative dizziness had completely resolved, his DHI was 1/10, DGI 0/24, normal DVA. His hearing was unchanged. (Figure 4)

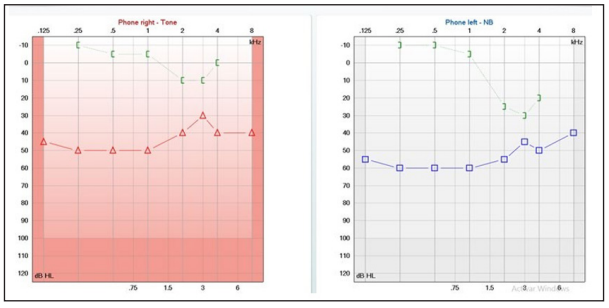

**Figure 3:** Pre op audiometry

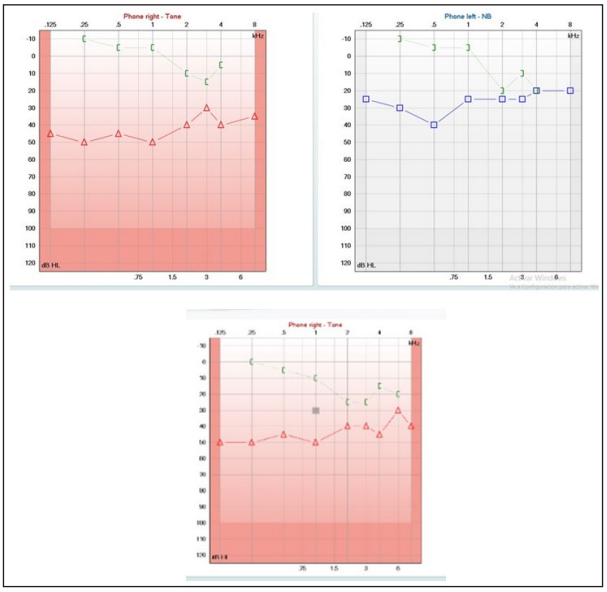

**Figure 4:** Post right SCC repair

## Discussion

We present two different cases of simple SCDS and concomitant SCDS and otosclerosis emphasizing the need of early intervention and vestibular rehabilitation. Although surgery is the gold treatment for SCDS, it is not harmless. Surgery can

cause temporary or chronic labyrinthine hypofunction that may benefit from early VR.

A systematic literature review aimed to assess available evidence on the comparative effectiveness of different surgical treatment for SCDS showed noise-induced vertigo improved in the majority of cases following plugging plus resurfacing. Surgery also improved pressure-induced vertigo in 85% of patients and decreased mean DHI, One study reported a transient increase in vertigo in the majority of patients for several weeks after surgery and other refer temporary disequilibrium in all cases [20]. This supports the need for vestibular rehabilitation in the immediate postoperative period.

In special cases of concomitant otosclerosis and SCDS, surgery may turn a non active 3rd window to an active one, sound energy is attenuated by fixed stapes and hence, the drive for auditory symptoms is smaller. It may also change the mechanism of energy dissipation in the inner ear preventing vestibular symptoms. In the face of a third window, the inner ear may have lower resistance to sound penetrance- fenestration or removal of the stapedial foot place transfer some abnormal energy to the inner ear and could increase the higher risk of inner ear damage during operation.

Yong et al. has previously described the main differences between otosclerosis patients vs SCDS patients vs otosclerosis and SCDS patients [15]. Mainly, the last group is characterized by an absent stapedial reflex, absent air conduction VEMPs but present bone conduction VEMPs, and supra-normal bone conduction levels are observed.

Dizziness after SDSC surgery is not commonly reported on the literature, nevertheless objective measures such as Dynamic gait index, dynamic visual acuity and subjective measures such as Dizziness Handicap Inventory are crucial in order to accurately evaluate our patients pre and post surgery [11]. Follow up using these same measures is capital.

Additionally, vestibular rehabilitation focusing on central compensation and habituation is recommended [16,17].

**Conclusion**

We present with two clinical cases the need to start VR immediately after surgery of SCDS. With early VR, patients can compensate for vestibular hypofunction both as a component of the disease itself and as a consequence of surgery. In special cases of concomitant otosclerosis and SCDS, patients may be advised about risks of surgery and be aware that a reduction in the vestibular function after SCDS surgery is possible as well as the need of prompt vestibular rehabilitation.

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