CASE REPORT

Enhanced otolithic function in semicircular canal dehiscence

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Abstract

The enhanced sound- and vibration-induced vestibular evoked myogenic potentials (VEMPs) and their lower threshold in patients with a thinning of the bony wall of the superior semicircular canal (superior canal dehiscence, SCD) have been interpreted as being due to the dehiscence allowing sound and vibration to activate, unusually, the receptors of the dehiscent semicircular canal. We report a patient with bilateral SCD, as verified by high resolution CT scans, who had bilaterally decreased superior semicircular canal function, as shown by rotational tests of canal function. This patient also showed enhanced VEMPs and reduced thresholds. We conclude that in this patient the enhanced VEMP responses are thus probably due to enhanced otolithic stimulation by sound and vibration after dehiscence.

Keywords: Ocular vestibular evoked myogenic potentials, VEMPs, vestibular, otolith, utricular, saccular

Introduction

In 1998 Minor et al. [1] described a new clinical entity, superior canal dehiscence syndrome (SCD), characterized by the onset of nonspecific vestibular and cochlear symptoms due to hypersensitivity of labyrinthine receptors caused by a bony defect, usually located in the external wall of the superior semicircular canal (SSC). The symptoms were idiosyncratic and included dizziness, Tullio phenomenon, positional vertigo, pulsatile tinnitus, and conductive and/or neurosensory hearing loss. The possibility of SCD must be evaluated on the basis of particular bedside signs (such as nystagmus induced by sound or pressure, Valsalva maneuver or Hennebert sign) and reinforced by the results of certain tests, essentially cervical vestibular evoked myogenic potentials (cVEMPs) and ocular VEMPs (oVEMPs) to air-conducted sound (ACS) or bone-conducted vibration (BCV) with threshold and amplitude analysis [2–4]. In SCD patients pressure stimulation of the middle ear or an increase in intracranial pressure [5,6] can evoke a typical nystagmus, detected as a specific oculomotor pattern recorded by three-dimensional scleral search coils [7,8], usually aligned in the plane of the affected canal, while analysis of ACS- and BCV-evoked cVEMPs and oVEMPs [9] demonstrates increased VEMP amplitude and depressed detection threshold for cVEMPs to ACS [2]. Final confirmation is only obtained by high-resolution computed tomography (HRCT) with sagittal reconstructions following and mapping the circumference of the SSC [10].

Recently these enhanced myogenic potentials in SCD patients have been attributed to the activation of SSC receptors by ACS [11]: ‘The oVEMP and VOR in superior semicircular canal dehiscence predominantly represent activation of the dehiscent canal’ (p.165). The results for the SCD patient we report here are inconsistent with this suggestion because she had absent superior canal response, but still had an enhanced oVEMP n10.

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Here we report the case of a patient who experienced postural instability and feelings of vertigo when sound, vibration or pressure were applied to the ear, mastoid or external canal, respectively, but for whom objective measures of the vestibulo-ocular response of the SSCs showed a bilaterally decreased canal response, although the HRCT showed clear bilateral SCD.

Case report

A 55-year-old female primary school teacher was referred to our tertiary outpatient otoneurology center with dizziness, oscillopsia, and hearing loss, particularly on the left, associated with disabling bilateral tinnitus, interspersed with aural fullness during the day, more in the left ear than in the right. She also complained of particularly irritating and disabling pulsatile tinnitus in both ears on changing head position. Case history identified oscillopsia related to intense exertion and Tullio phenomenon evoked by sounds, often when children were playing noisily at school. The patient did not recall any history of head injury. Otoscopy findings were normal. Audiological study (pure-tone audiometry) revealed mixed hearing loss sparing stapedial reflexes, and normal auditory brainstem responses. She was tested with written consent, and the procedures described below have been approved by the local ethics committee. After a series of diagnostic procedures, she was diagnosed with bilateral SCD. That diagnosis was confirmed by HRCT imaging.

Vestibular symptoms were evaluated with a standardized set of tests, including bedside examination with a video oculography (VOG) recorder. The pattern of horizontal, torsional, and vertical eye movements was evaluated and recorded with three-dimensional infrared VOG (50 Hz sampling; Torsio VNG Ulmer, Synapsys, Marseille, France). The patterns investigated were: spontaneous nystagmus (absent), head-shaking nystagmus (no nystagmus at cessation of head shaking), positional nystagmus (Dix-Hallpike maneuver and head roll maneuver, which failed to reveal any abnormal eye movement). The Valsalva maneuver (by VOG), Tullio’s phenomenon after exposure to sound of 500–3000 kHz at intensities of 100–110 dB, and mastoid vibration at 100 Hz also failed to evoke the expected oculomotor responses in the plane of the affected canals. This absence of vestibular symptoms in the clinic was in contrast to the patient’s reports of oscillopsia and Tullio phenomenon in everyday life.

The function of each canal was evaluated with the video head impulse test system (vHIT), recently validated [11] as a clinical tool for identifying peripheral semicircular canal deficit. Horizontal and vertical head-impulse tests were recorded by vHIT (250 Hz). The vertical head impulses were passive, unpredictable head rotations in the planes of the SSC (also known as anterior semicircular canals): left anterior – right posterior (LARP) and right anterior – left posterior (RALP). vHIT recordings correspond very closely to search-coil recordings for measuring VOR deficits and detecting overt or covert catch-up saccades, which indicate inadequate canal function [12].

We also evaluated cVEMPs and oVEMPs in response to BCV applied to the midline of the forehead at the hairline (a location called Fz), carried out using a hand-held mini-shaker (Brul & Kjaer, model 4810; Naerum, Denmark), fitted with a 2 cm bolt (M5) terminated in a bakelite cap 1.5 cm in diameter. The mini-shaker was driven by computer-generated signals, usually consisting of 50 repetitions of a 500 Hz tone burst lasting a total of 7 ms (including a 1 ms rise and 1 ms fall with zero-crossing start). Application of BCV at Fz is special because it ensures approximately equal and simultaneous stimulation of both labyrinths [4]. For ACS and BCV, the electromyogram (EMG) signals were amplified by two independent differential amplifiers (filter cut-offs: 20 Hz to 500 Hz), and the unrectified signals were averaged (n = 50 presentations) and simultaneously acquired from the two eyes or the two sternocleidomastoid muscles using a Medelec AMPLAID MK12 averager (Milan, Italy), sampling rate 20 kHz.

Results

vHIT testing showed catch-up saccades in the plane of both anterior semicircular canals, a clear indication of bilaterally deficient VOR function of the SSCs (Figure 1a, b). Caloric tests showed a normal pattern (Fitzgerald-Hallpike stimulation). ACS cVEMPs were carried out with 50 mini short tone bursts of 500 Hz lasting 2 ms and a repetition frequency of 4 Hz. With these parameters, a clearly detectable response would be expected at 100 dB sound pressure level (SPL), while no response would be detected at <85 dB SPL. These values, which have proven reliable in our laboratory, are generally accepted as standard values under the conditions described. In this case, we found bilateral lowering of the detection threshold, the p13-n23 complex being evoked by the unusually low intensity of 70 dB SPL stimuli bilaterally (Figure 2).

cVEMPs and oVEMPs to Fz BCV stimulation showed a large oVEMP response with a very prominent n10 component, which has been attributed to utricular functioning. In addition the threshold for
evoking the n10 component was much lower than found in healthy subjects (Figure 3).

The patient was then referred to a tertiary radiology center for magnetic resonance imaging (MRI) of the posterior cranial fossa with paramagnetic contrast enhancement, which revealed bilateral dehiscence of the SSC. These findings were fundamental for explaining the vestibular symptoms and confirming the suspicion of defective labyrinthine capsule (Figure 4a, b).

**Discussion**

More than a decade since the first description of SCD, much has been discovered about the evolution of the disease, onset of symptoms, and diagnosis. Much more research will be necessary to understand why SCD may be radiologically and clinically manifest, with reproducible objective test results, but subjectively silent. The question that clinicians ask is why patients do not complain of a whole series of symptoms, but only some of the symptoms characterizing the syndrome? These patients must be differentiated from other patients with evident clinical signs and symptoms of dehiscence in order to be diagnosed with SCD, after radiological confirmation.

To solve this clinical dilemma, two types of anatomical situation must be considered. The first is that the SSC is not really dehiscent. The temporal bone that envelops the semicircular ducts may be so thin or absent as to provide radiological evidence of dehiscence, while being insufficient to manifest as a clinical syndrome with symptoms affecting daily quality of life. This situation is presumably linked to the elasticity of the dura mater, which protects the membranous labyrinth and prevents its exposure, as well as to the position of bone loss.

Another possibility is that the dura mater herniates into the dehiscent SSC, compressing the membranous labyrinth and reducing canal function, while still allowing the symptoms caused by sound and vibration. This new clinical condition is due to autoplugging, as if these patients had a partial syndrome of SCD. Patients with autoplugging can still have sound-, vibration- (acceleration-) or pressure-induced vertigo, but have reduced rather than enhanced semicircular canal function. This was the situation in the present case.

Suspicion of SCD in a patient with some or all signs and symptoms of third mobile window, or with vertigo and oscillopsia evoked by loud noises (Tullio phenomenon) and/or by maneuvers that change middle ear or intracranial pressure (Valsalva maneuver), or hypersensitivity to bone-conducted sounds (autophony), or sometimes with exclusively auditory symptoms and signs, is currently reinforced by medical history and certain fundamental tests, especially VEMPs. Until 2005, the main VEMPs performed and used clinically were eVEMP evoked largely by ACS. This clinical test of vestibular function has mainly
been performed by ACS, seldom by BCV using a tendon hammer or bone vibrator (B-71) or a mini-shaker (Bruel & Kjaer model 4810). It is now universally accepted that when patients with SCD are studied by ACS cVEMPS they show increased amplitude and low threshold [2].

Recently a new generation of oVEMPs was devised [13], recording myogenic potentials from eye muscles evoked by sound or vibration (by tapping or BCV). oVEMPs are a simple new measure of otolithic function. They indicate mainly utricular function [14]. They are very easy to perform in a clinical setting. The electrodes are placed on the inferior margin of the orbit and, unlike for cVEMP, the patient only has to look upwards to cause contraction-activation of the extraocular muscles of the inferior orbit (for a review see Curthoys et al. [15]). oVEMPs testify to the activity of the muscles of the inferior rectus (IR) and inferior oblique (IO) contralateral [14] to the stimulated labyrinth. This new method has enriched the diagnostic tools for detecting SCD by specialist otoneurologists, because also in this case patients with SCD, stimulated by ACS or BCV, show a negative and therefore excitatory component that appears after about 10 ms (hence defined as n10) [15]. However, the oVEMP is a crossed otolith-ocular response – the n10 is greatly increased in amplitude beneath the eye on the side contralateral to the SCD with respect to the n10 evoked ipsilaterally on the affected side.

In the literature, the n10 is mainly attributed to activity of the contralateral utricular macula [3,14,16], although some researchers studying SCD have attributed generation of the negative component to abnormal stimulation of a ‘roofless’ semicircular canal [11] or to increased susceptibility of the saccular macula to the hypersensitivity of the ‘open’ system [17]. Actually, interpretation of results obtained with a large cohort of patients (133 subjects) with superior vestibular neuritis [16] shows that n10 is largely generated by activation and therefore by the response of the utricular otolithic receptor that projects its information onto the superior vestibular nerve [18]. Indeed, in subjects with superior vestibular neuritis, the cVEMP is intact, whether evoked by ACS or BCV, confirming the neuroanatomical and neurophysiological evidence [18–20].

The present case study is therefore particularly important for a number of reasons, as follows. (i) It is a case of bilateral SCD. (ii) It clearly confirms that in SCD patients cVEMPs evoked by ACS have high amplitude and low threshold. (iii) The outcome of oVEMPs in relation to the function of the two SSCs is highly significant because the n10 component due to 500 Hz BCV oVEMP is present and increased in
amplitude, as predicted. For the first time, a reduction in the BCV stimulus threshold necessary to activate n10 has been demonstrated in the presence of the dehiscence. (iv) The n10 component of the oVEMP to BCV is present and enhanced (Figure 3), despite evidence that the SSC has bilateral deficient VOR gain and clear, reproducible, catch-up saccades, which indicate a deficient canal response [12].

Since the caloric test response was normal, indicating normal horizontal semicircular canal function on both sides, a reasonable hypothesis for bilateral SSC dysfunction is spontaneous autoplugging by the meninges (dura mater) on the membranous labyrinth of the canal (Figure 4).

A fundamental question now arises. If activation of the dehiscent SSC, hypersensitive to vibratory

![Figure 3](image_url)  
Figure 3. Ocular VEMPs in response to 500 Hz bone-conducted vibration (BCV) stimulation at Fz at threshold. The n10 response is present and increased in amplitude under both eyes. Reducing the stimulus intensity shows the very low threshold necessary to activate n10 caused as a result of the presence of the dehiscence. The low threshold to BCV is similar to the low threshold with air-conducted sound (ACS) cVEMPs.

![Figure 4](image_url)  
Figure 4. High resolution CT scan: (a) right and (b) left temporal bone. Off-axis oblique reformations view through the superior portion of the temporal bone, which demonstrates bilateral superior semicircular canal dehiscence (the two large gray triangles in a and b show the area of absent bony covering); the small white arrows show where the lumen of the canal has been plugged.
stimulation, is necessary to generate the n10 component of the oVEMP, we should not detect the n10 component under the patient’s eyes. However, in the present case the component was indeed evoked by BCV at Fz (Figure 3) and had high amplitude and a low threshold. The most likely possibility is that the n10 of the oVEMP was produced by activation of the utricular macula, hypersensitive to vibration applied to Fz on the patient’s skull [3,14]. After opening the labyrinth Tulio himself observed a very clear pattern of movement of the utricular macula [21], and the results of this patient seem to correspond to Tulio’s observation. This rare case is of great interest because it provides new evidence of the role of the utricular macula in the generation of the n10 component of oVEMPs.

**Conclusion**

Enhanced VEMP responses in a patient with CT-verified bilateral SCD, but with reduced superior canal function, strongly imply that otolith function after dehiscence is enhanced.

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**References**


