■ CASES STUDIES

Characterization of Bilateral Superior Canal Dehiscence

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In the superior semicircular canal (SSC) dehiscence syndrome, patients can have sound- or pressure-induced vertigo and oscillopsia. They may also present conductive hearing loss or higher than normal bone conduction thresholds.
Clinical manifestations are due to the effect of a third mobile window in the inner ear created by the dehiscence. Diagnosis is based on clinical manifestations, vertical and rotatory nystagmus induced by sound and pressure reflecting SSC stimulation, reduced threshold and increased amplitude of vestibular evoked myogenic potentials (VEMP) and temporal bone CT scan images showing the SSC dehiscence. Characteristic eye movements can be recorded with the scleral search coil technique.

Key words: Bilateral dehiscence. Superior semicircular canal. Vertigo. Oscillopsia

Caracterización de la dehiscencia bilateral del conducto semicircular superior
En la dehiscencia del conducto semicircular superior aparecen vértigo y oscilopsia inducidos por sonidos intensos o cambios de presión intracraneal o del oído medio. Puede aparecer hipoacusia de transmisión y aumento del umbral auditivo por vía ósea.
Las manifestaciones se explican porque la dehiscencia crea una tercera ventana móvil en el oído interno. El diagnóstico se establece por la clínica, el nistagmo vertical-rotatorio desencadenado por sonido o presión, que es propio de la estimulación del conducto semicircular superior (CSS) correspondiente, la disminución del umbral y el aumento de la amplitud de los potenciales evocados miogénicos (VEMP) y las imágenes de dehiscencia del CSS en la tomografía computarizada de peñascos. Los movimientos oculares característicos desencadenados se pueden estudiar mediante la técnica de la bobina escleral (BE) en campo magnético (scleral search coil).

Palabras clave: Dehiscencia bilateral. Conducto semicircular superior. Vértigo. Oscilopsia.

A 57-year old male presented at the Department of Otolaryngology and Head and Neck Surgery of the Johns Hopkins Hospital (Baltimore, Maryland, United States) reporting, imbalance and a feeling of vertical shift of objects whenever he coughed, cleared his throat, sneezed, strained, or lay down. He felt pressure in his ear and reported bilateral tinnitus, more intense on the right side.

During the Valsalva manoeuvre with a closed nose (open glottis) included in the otoneurological examination, mixed downward vertical nystagmus was observed along with a torsional effect with the superior ocular pole facing right (clockwise). Eye movements are not triggered with bilateral presentation of 250 to 4000 Hz sounds or when applying positive or negative pressure in the external auditory canal.

The tonal audiometry revealed the existence of slight bilateral sensorineural hearing loss for sharp frequencies.

The results of the electronystagmography using the conventional binaural, bithermal stimulation technique were normal. A high resolution computerized tomography (HRCT) of the temporal bone was performed with 0.5-mm slices and reconstruction of the SSC plane and it revealed a bilateral dehiscence of the SSC (Figure 1).

The methodology described by other authors[1] was used for evoked myogenic potentials (VEMP). Characteristic p13 and n23 waves were seen on the VEMP. With stimulation in the right ear, response appeared at the 85 dB threshold and with stimulation of the left ear, response was obtained at 103 dB.

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Figure 1. High resolution computerized tomography of the temporal bone with 0.5-mm slices and reconstruction of the SSC plane in which bilateral dehiscence of the SSC can be seen.

With the aim of contributing to identifying the side on which the patient's symptoms originate, eye and head position were recorded using double scleral coils (SC) (Skalar, Delft, the Netherlands) located inside a magnetic field. This system consists of 3 pairs of metal coils placed in a cubic frame that generate 3 orthogonal magnetic fields. Binocular movements are recorded in 3 dimensions by means of double SC comprising 2 orthogonal coils inside a single silicone ring. The ring is placed on the surface of the eye. Thus, 2 vectors defined by the voltages induced inside the coils are generated by the 3 magnetic fields. The test was conducted in the dark, except for the diode the patient must look at while the test is being carried out. The system used has been described in depth by Migliaccio et al.[2] The patient performed the Valsalva manoeuvre with closed glottis several times while looking at the illuminated diode in front of him. The SSC-recorded eye movements revealed the presence of mixed vertical-torsional nystagmus. During the Valsalva manoeuvre with closed glottis, the nystagmus was downward vertical-torsional with clockwise slow phases (with respect to the patient). This reflects inhibition of the SSC. Nevertheless, during the passive phase, the nystagmus observed was vertical with upward counter-clockwise slow phase, which means that the SSC was excited. With this technique the eye's axis of rotation can be calculated during stimulation of each canal.[3] The vector of eye movement was seen to be aligned mainly in the RALP plane (right anterior left posterior) (Figure 2).

DISCUSSION

The symptoms reported by this patient are consistent with SSC dehiscence syndrome previously commented on. Moreover, these patients may present Tullio's phenomenon or Hennebert's sign. The Valsalva manoeuvre may trigger vestibular symptoms and signs, as well as characteristic eye movements in the direction of the semicircular canal affected and these can be studied using the magnetic-field SC technique.[4] Increased amplitude and a decreased threshold for the appearance of VEMP have been observed in this syndrome.[4,5] The positive predictive value of the HRCT in the diagnosis of dehiscence increases by setting a slice thickness of 0.5 mm and reconstruction in the SSC plane.[6] The nystagmus seen in SSC dehiscence as a result of SSC stimulation is caused by the contraction of the ipsilateral superior rectus and the contralateral inferior oblique muscle. This gives rise to a nystagmus whose slow phase faces vertically upward and counter-clockwise on stimulation of the right SSC and vertically upward and clockwise on stimulation of the left SSC.[7]

In the presence of SSC dehiscence, exposure to loud noises, positive pressure in the external auditory canal, and/or the Valsalva manoeuvre with closed nose (open glottis) all cause ampullofugal (excitatory) deflection of the cupula in the SSC. This provokes conjugated eye movements with an upward vertical slow-phase component and a torsional component in which the superior ocular pole moves toward the unaffected side. However, application of negative pressure in the external auditory canal, the Valsalva manoeuvre with closed glottis and jugular compression give rise to ampullopetal (inhibitory) deflection in the cupula in the SSC; the eye movement evoked is downward vertical with a torsional component towards the affected ear.[4] These findings are in line with both the otoneurological examination as well as the SSC eye movement recording in this patient. In the otoneurological examination, during the Valsalva manoeuvre with closed nose (open glottis), mixed clockwise

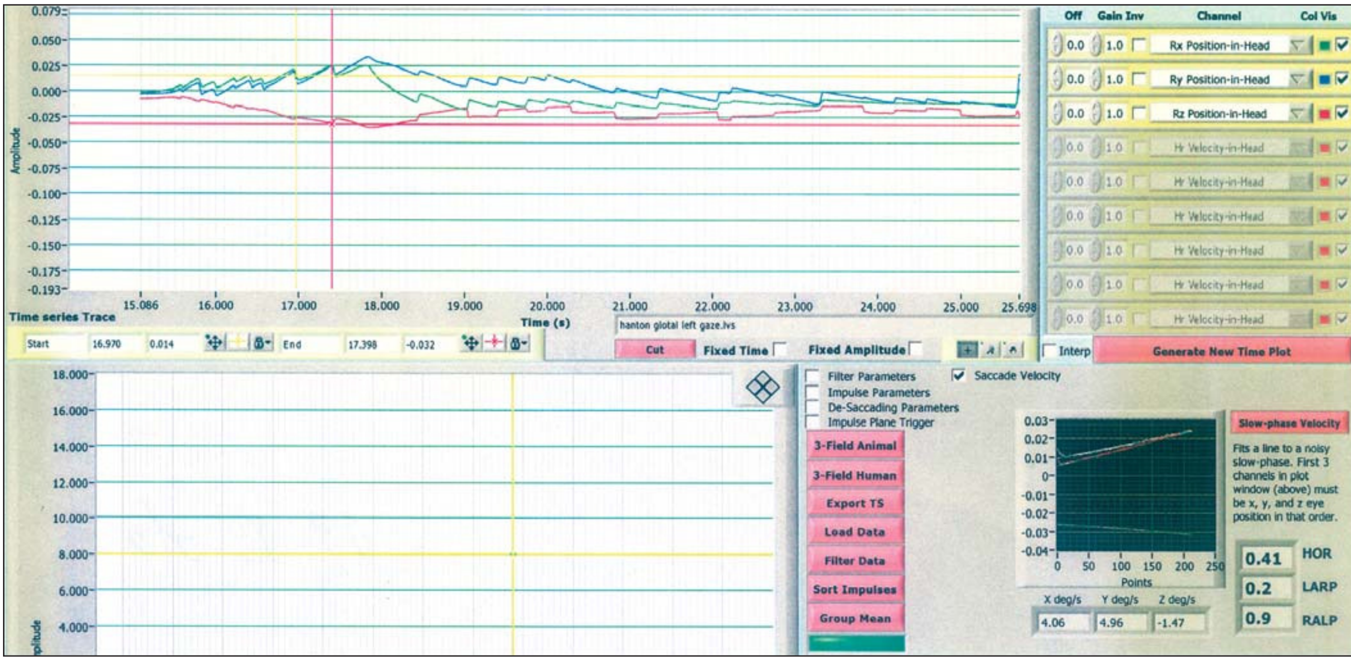

Figure 2. Valsalva manoeuvre with closed glottis. Active phase (as far as the red cursor): slow phases of downward, clockwise vertical nystagmus. Passive phase: slow phases of upward, counter-clockwise vertical nystagmus. Vector of eye movement mainly aligned in the RALP plane (higher value in RALP than in HOR and LARP). As per convention, 3-D recording of eye movements in axes x, y, and z, a positive wave deflection recording represents eye movements that are, respectively, clockwise, downward and to the left.

downward vertical nystagmus was observed (superior ocular pole toward the right), reflecting stimulation of the right SSC. With respect to SSC-recorded eye movements during the active phase of the Valsalva manoeuvre with closed glottis (inhibitory stimulus for the SSC), nystagmus was seen with a clockwise downward vertical slow phase, which in effect, means that there is inhibition of the right SSC. During the passive phase of the Valsalva with closed glottis (excitatory stimulus for the SSC), nystagmus was observed with a counter-clockwise upward vertical slow phase, which in turn corresponds to excitation of the SSC, also on the right. It has been demonstrated that in patients with dehiscence of the semicircular canal, eye movements are aligned with the plane of the affected canal.[3] In this case eye movements are aligned in the RALP plane. In order to distinguish between stimulation of the right SSC (RA) and posterior left CSS (LP), one must consider that in the first case, counter-clockwise upward vertical eye movements will appear, whereas in the second case, eye movements are clockwise and downward vertical. In the light of this and the results mentioned, we are facing stimulation of the right SSC. The images of the HRCT reveal bilateral SSC dehiscence; hence, this test corroborates the diagnosis but does not reveal any information about the ear that is causing the symptoms. The results of the VEMP are characteristic of dehiscence of the right SSC, since on the right the response occurs at a lower threshold than usual. It appears that the mechanism causing this decrease in the threshold of appearance of the VEMP in SSC dehiscence has to do with decreased impedance for sound and pressure transmission as a result of a third mobile window in the inner ear.[4] Therefore, the clinical manifestations and complementary testing commented above help to identify the side where the symptoms arise in cases of bilateral dehiscence of the SSC.

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