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# Vestibular hypersensitivity to sound in superior canal dehiscence: Large evoked responses in the legs produce little postural sway

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**Abstract**

*Objective:* Patients with superior canal dehiscence (SCD) typically have enhanced sound-evoked vestibular reflexes, such as vestibulo-collic and vestibulo-ocular reflexes. We wished to investigate whether sound-evoked lower limb EMG responses and postural sway are also enhanced in this condition.
*Methods:* Eight patients with CT confirmed SCD (11 affected ears) and 8 age-matched normal controls participated. Three sound-evoked responses were measured; vestibulo-collic reflexes (i.e. vestibular-evoked myogenic potentials, VEMPs), lower limb vestibulo-spinal reflexes and body sway (centre of pressure in mm). Sound stimuli were 500 Hz air-conducted tone bursts of varying lengths (VEMPs: 2 ms; vestibulo-spinal: 20 ms; sway: 1 s and 200 ms) set at fixed levels above each subject's VEMP threshold.
*Results:* SCD patients had very large VEMP and vestibulo-spinal responses following high intensity stimulation, but at the matched intensity of 15 dB above threshold amplitudes were similar in both SCD patients and controls. The amplitude of both responses increased linearly with increasing stimulus intensity in both groups. Large (∼20 mm), stereotyped sway responses were present in only one (atypical) patient with high intensity stimulation. Small (∼2 mm) sway responses were present in the remaining patients, and began immediately following the vestibulo-spinal responses.
*Conclusions:* Despite the presence of large vestibular reflexes, there is usually very little body sway in response to loud sounds in SCD patients.
*Significance:* Large short-latency vestibulo-spinal reflexes in SCD do not necessarily evoke large sway responses.


*Keywords:* Tullio phenomenon; Superior canal dehiscence; Vestibular; VEMP; Sway

## 1. Introduction

Superior canal dehiscence (SCD) is a recently described condition in which a dehiscence, or hole, in the temporal bone overlying the superior semicircular canal produces vestibular hypersensitivity to sound and pressure (Minor et al., 1998). SCD patients typically experience sound-evoked vestibular symptoms (the Tullio phenomenon) and have enhanced sound-evoked vestibular reflexes (Minor, 2005). In particular, vestibulo-collic reflexes (vestibular-evoked myogenic potentials, or VEMPs, recorded from the neck) in SCD have pathologically low thresholds (Brantberg et al., 1999; Watson et al., 2000), and sound-evoked ocular responses are large (Aw et al., 2006; Halmagyi et al., 2003; Rosengren et al., 2008; Welgampola et al., 2008).

However it is unclear to what extent other sound-evoked vestibular reflexes are enhanced in SCD. Dieterich et al. (1989) and Colebatch et al. (1998) both described patients with the Tullio phenomenon who had sound-evoked lower limb reflexes (vestibulo-spinal reflexes). Colebatch et al. (1998) showed that the reflexes inverted with left vs. right head rotation and were present with subjects standing

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but not sitting, features shared with galvanic-evoked vestibulo-spinal reflexes (Britton et al., 1993). The patient described by Dieterich et al. (1989) also had a marked, stereotyped postural disturbance when exposed to loud sounds, thought to be caused by mechanical stimulation of the otoliths by a hypermobile stapes footplate. Given that SCD is now considered the most common cause of the Tullio phenomenon, it is likely therefore to also be a cause of sound-evoked postural disturbance. Our interest was stimulated by a patient with confirmed SCD who complained of prominent sound-evoked postural movements. In this and seven other SCD patients we compared three sound-evoked responses: vestibulo-collic reflexes, vestibulo-spinal reflexes and body sway.

## 2. Methods

### 2.1. Subjects

Eight patients with symptomatic SCD participated (4 males, 4 females; aged 41–82 years, Table 1). Only patient 1 was noted during clinical assessment to have postural responses to sound. The diagnosis of SCD was made on the basis of clinical features (such as vestibular hypersensitivity to sound or pressure and conductive hyperacusis), pathologically low VEMP thresholds and evidence of dehiscence shown by high-resolution CT imaging reformatted off-line into the superior canal plane as described by Belden et al. (2003) and Minor (2005). Three had bilateral dehiscence, four right side only, and one left side only (11 affected ears: 7 right, 4 left). Eight normal controls also participated (3 males, 5 females; aged 43–73 years). The control subjects were matched where possible to within 5 years of the patient's age. However, the control for patient 7 was 6 years younger and for patient 2 was 9 years younger. Two additional SCD patients (patients 9 and 10), were included in a supplementary set of observations comparing body sway to vestibulo-spinal reflexes directly. Written, informed consent was obtained from all participants and the study was approved by the local ethics committee.

### 2.2. Vestibulo-collic reflexes (VEMPs)

VEMPs were recorded following monaural stimulation of the affected or matched side/s in all participants. Subjects lay supine on a chair, with the backrest tilted to approximately 30 degrees from the horizontal, and lifted their heads to activate the sternocleidomastoid (SCM) muscles. Rectified EMG was monitored and recorded to ensure that SCM contractions were similar between trials. VEMP thresholds were obtained by reducing the intensity in 3 or 6 dB steps over successive trials and were accurate to 3 dB. VEMP thresholds were used as a reference and stimulus intensity was subsequently set at fixed levels above each subject's individual VEMP threshold, e.g. 15 dB above threshold ("+15 dB").

The sound stimuli were 500 Hz, 2 ms air-conducted (AC) tone bursts, delivered with alternating polarity via calibrated headphones (TDH 49, Telephonics Corp., Farmingdale, NY, USA): maximum peak intensity 10 V pp, 142 dB peak SPL, equivalent to approximately 124 dB nHL for 2 ms tone bursts. A total of 100–256 stimuli were delivered at a rate of 5 Hz. Surface EMG was recorded from the SCM muscles bilaterally with the active electrodes on the middle of the SCM belly, reference electrodes on the medial clavicle and earth on the sternum. The sampling period was from 20 ms before to 100 ms following stimulus onset. VEMP peak-to-peak p13 and n23 amplitudes were corrected by dividing by the mean level of pre-stimulus rectified EMG activity.

Table 1
Demographic characteristics and VEMP thresholds of SCD patients and normal controls

| SCD patient | Age | Affected side | VEMP T SPL | VEMP T nHL | Normal control | Age | VEMP T SPL | VEMP T nHL |
|---|---|---|---|---|---|---|---|---|
| *Main group*[a] | | | | | | | | |
| 1 | 62 | R | 91 | 77 | C1 | 58 | 112 | 98 |
| | | L | 86 | 72 | | | 118 | 104 |
| 2 | 82 | L | 97 | 83 | C2 | 73 | 121 | 107 |
| 3 | 61 | R | 88 | 74 | C3 | 59 | 115 | 101 |
| 4 | 49 | R | 91 | 77 | C4 | 50 | 112 | 98 |
| 5 | 41 | R | 85 | 71 | C5 | 43 | 115 | 101 |
| | | L | 91 | 77 | | | 109 | 95 |
| 6 | 61 | R | 100 | 86 | C6 | 62 | 112 | 98 |
| 7 | 68 | R | 100 | 86 | C7 | 62 | 109 | 95 |
| | | L | 106 | 92 | | | 106 | 92 |
| 8 | 60 | R | 91 | 77 | C8 | 54 | 112 | 98 |
| Mean | 60.5 | | 93.4 | 79.3 | | 57.6 | 112.8 | 98.8 |
| *Additional patients* | | | | | | | | |
| 9 | 41 | R | 97 | 83 | | | | |
| 10 | 54 | L | 100 | 86 | | | | |

[a] Eight patients and controls participated in the main experiment, and two additional patients were recruited for further sway testing. *Abbreviations:* VEMP T, VEMP threshold; SPL, peak sound pressure level; nHL, hearing level.