



## Bilateral superior canal dehiscence syndrome

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Superior canal dehiscence (SCD) syndrome is a newly recognised condition<sup>1-4</sup> where dehiscence of bone over the superior semicircular canal can lead to unusual auditory and vestibular symptoms and signs.

In the 1880s, Ewald demonstrated that pressure applied to surgically fenestrated canals in pigeons could cause a nystagmus in the plane of the stimulated canal. This has long been recognised as a possible complication of cholesteotoma eroding bone over the horizontal canal accounting for horizontal nystagmus induced by applying alternating pressure to the external ear canal, called Hennebert's sign.

In 1929, Tullio<sup>5</sup> showed that in dogs with surgically fenestrated superior canals loud sounds could effect a nystagmus in the plane of the canal. Also, in rabbits and pigeons with intact labyrinths loud sounds could induce vestibular responses with disconjugate rotation of the eyeballs, tilting of the head and leg flexion. These vestibular responses are now called the Tullio phenomenon.

More recent electrophysiological studies in humans showed that a myogenic response to a loud click stimulus occurs in the sternomastoid muscle.<sup>6</sup> The response—named the vestibular-evoked myogenic potential (VEMP)—is generated by the saccule and is non-hearing dependent. Ears affected with the Tullio phenomenon have an abnormally low VEMP threshold.<sup>7</sup>

### Case report

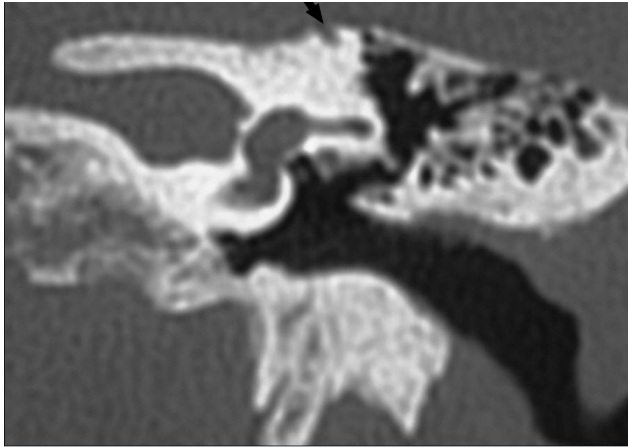
A 58-year-old female presented complaining of intermittent dizziness, oscillopsia and aural symptoms. For 2 years there had been pulsatile tinnitus in both ears, particularly the right ear. More recently stooping, nose-blowing or a sneeze would elicit brief vertigo and oscillopsia. Also she had noticed that when walking on firm ground she could hear her footsteps: "my footsteps just echo inside my body".

On examination ear drums, hearing and acoustic reflexes were normal. There was no spontaneous nystagmus. Vestibulo-ocular reflexes were normal. When the patient performed a valsalva manoeuvre a brief down beat and slightly right-torsional nystagmus occurred. This was reproduced and recorded in infrared light by a right eye camera (Figure 1 – mpeg video-clip accessible at <http://www.nzma.org.nz/journal/123-1321/4307/video.mpg>).

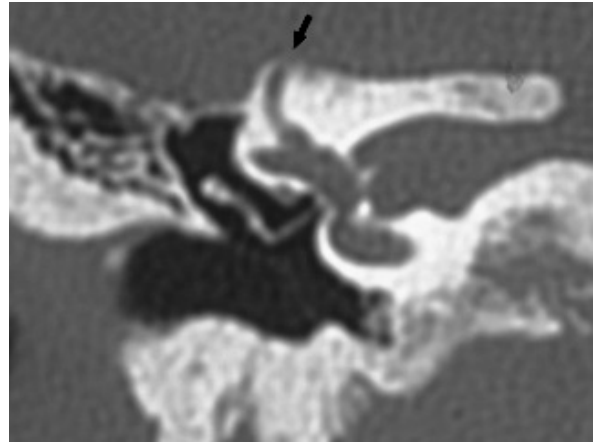
Further provocative tests for Tullio phenomena were done. However, straining against a closed glottis, alternating pressure in the external ear canal, or a 0.5kHz tone at 90dB and a 1kHz tone at 120dB did not induce symptoms or nystagmus.

CT scans with 0.625m slices in the coronal plane showed a bony dehiscence of both superior semicircular canals (Figure 2). MRI scans imply an abnormal absence of bone between the membranous portion of the superior canals and the overlying dura (dark space), in comparison with a subject with a normal bony covering (Figure 3).

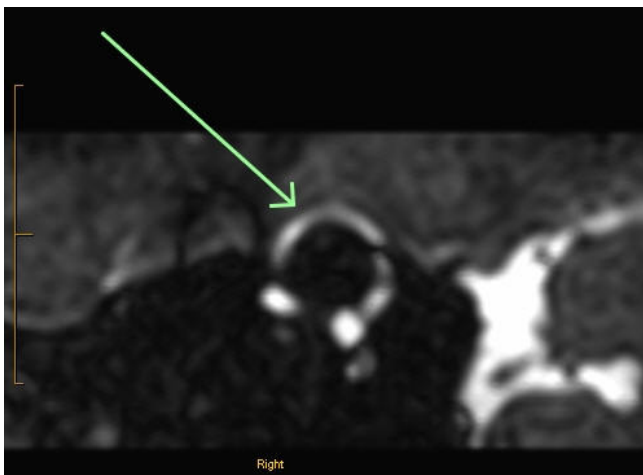
**Figure 2. Coronal CT scan right ear. Arrow indicates absence of bone over the superior canal.**



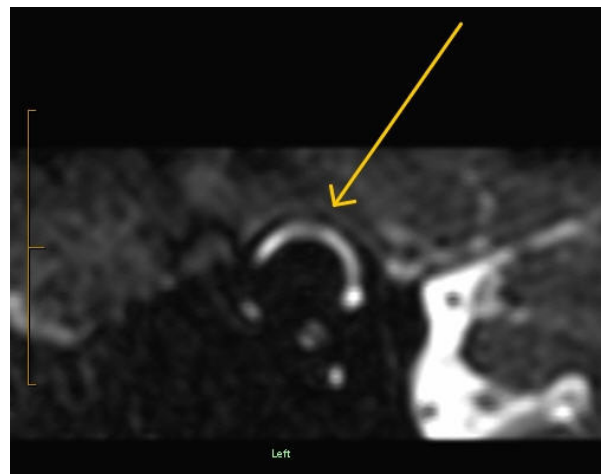
**Coronal CT scan left ear. Arrow indicates absence of bone over the superior canal.**



**Figure 3. Right coronal MRI scan. Arrow indicates the closeness of dura to superior canal perilymph.**



**Left coronal MRI scan. Arrow indicates the closeness of dura to superior canal perilymph.**

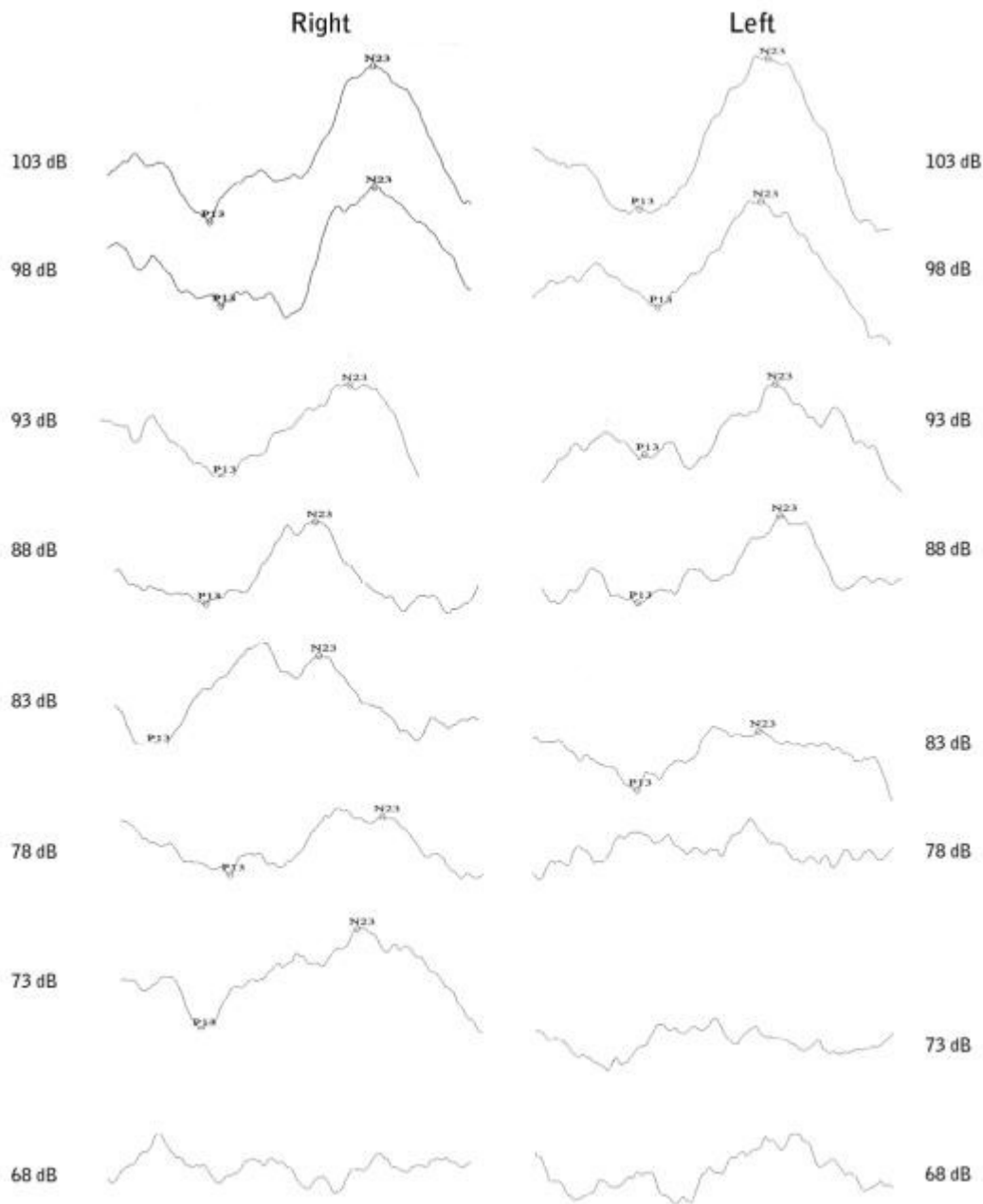


**Right coronal MRI scan of a normal ear. Arrow indicates bone separating dura from the superior canal.**



Cervical VEMPS were measured with surface electrodes over the sternum and each flexed sternomastoid muscle, using 0.1 msec clicks from a headphone in 5 dB descending steps. On the right the threshold for a clear N13/P23 wave-form was 73dB, and 83 dB on the left, both significantly below the normal ( $96\pm 5$  dB)<sup>8</sup> (Figure 4).

**Figure 4. Cervical VEMPS**



In this patient there is both CT scanning and VEMPS evidence of bilateral SCD syndrome.

## Discussion

Bony dehiscence over the superior canal can result in vestibular or auditory symptoms and signs, or both. The reasons for the differences are not known, but the dehiscence creates a so-called “mobile third window” effect. The vestibular symptoms are vertigo and oscillopsia.

A valsalva manoeuvre with closed nostrils, positive pressure in the ear canal cause an ampullofugal deflection (excitatory) of the superior canal cupula. In contrast—a valsalva manoeuvre against a closed glottis—bilateral jugular venous compression and a negative pressure in the ear canal can cause an ampullopetal (inhibitory) cupula deflection. The auditory symptoms can be autophony (hearing one’s own voice loudly in the ear), hypersensitivity for bone-conducted sounds, a blocked ear or pulsatile tinnitus.

In a microscopic study of 1000 temporal bones sectioned vertically in the plane of the superior canal, 0.5% had a complete dehiscence and 1.4% had bone so thin it could appear dehiscent even on high resolution CT scanning.<sup>9</sup> Conventional CT scans are displayed in the axial and coronal planes. False positive dehisces can be reduced with 0.5-mm-collimated helical scans with reformation of the images in the plane of the superior canal.<sup>10,11</sup>

The onset of SCD symptoms is typically in adulthood when, presumably, abnormally thin bone over the canal is disrupted by trauma or eroded by overlying structures. In most patients (as in this report) the symptoms are mild, and are prevented by avoiding the stimuli that cause them. In rare cases with disabling symptoms (sound-induced vertigo, pulsatile oscillopsia) surgical treatment is justified, either by closure of the defect with fascia and bone cement<sup>12</sup> or by superior canal plugging.<sup>13</sup>

In summary, SCD syndrome can cause unusual auditory and vestibular symptoms. Elicitation of nystagmus by a valsalva manoeuvre, sound stimulation or external canal pressure and a CT scan implying dehiscence are strongly suggestive, but a reduced VEMP threshold is required for certain diagnosis.

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