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Clinical Records

Superior semicircular canal dehiscence simulating otosclerosis

G. Michael Halmagyi, M.D., F.R.A.C.P., Swee T. Aw., M.B., B.S., Ph.D., Leigh A. McGarvie, B.E., M. Biomed.E., Michael J. Todd, B.E., M.Biomed.E., Andrew Bradshaw, B.E., B.Sc., Robyn A. Yavor, R.N., Paul A Fagan, M.D., F.R.C.S., F.R.A.C.S.

Abstract
This is a report of a patient with an air-bone gap, thought 10 years ago to be a conductive hearing loss due to otosclerosis and treated with a stapedectomy. It now transpires that the patient actually had a conductive hearing gain due to superior semicircular canal dehiscence. In retrospect for as long as he could remember the patient had experienced cochlear hypersensitivity to bone-conducted sounds so that he could hear his own heart beat and joints move, as well as a tuning fork placed at his ankle. He also had vestibular hypersensitivity to air-conducted sounds with sound-induced eye movements (Tullio phenomenon), pressure-induced nystagmus and low-threshold, high-amplitude vestibular-evoked myogenic potentials. Furthermore some of his acoustic reflexes were preserved even after stapedectomy and two revisions. This case shows that if acoustic reflexes are preserved in a patient with an air-bone gap then the patient needs to be checked for sound- and pressure-induced nystagmus and needs to have vestibular-evoked myogenic potential testing. If there is sound- or pressure-induced nystagmus and if the vestibular-evoked myogenic potentials are also preserved, the problem is most likely in the floor of the middle fossa and not in the middle ear, and the patient needs a high-resolution spiral computed tomography (CT) of the temporal bones to show this.

Key words: Otosclerosis; Bone Conduction; Semicircular Canals; Vestibular Function Tests

Introduction

Absence of bone between the apex of the superior semicircular canal and the middle cranial fossa – superior semicircular canal dehiscence (SCD) – has been recently described as a cause, perhaps the most common cause, of sound- and pressure-induced vestibular symptoms such as imbalance, vertigo and oscillopsia.[1–5] Patients with SCD show nystagmus[1,5] or other eye movements[6] in response to pressure or sound – the Tullio phenomenon – and abnormally high-amplitude, low-threshold vestibular-evoked myogenic potentials (VEMPs) in response to clicks.[7–9] As well as showing vestibular hypersensitivity to air-conducted sounds, SCD patients show cochlear hypersensitivity to bone-conducted sounds.[2–4] The characteristic audiological pattern in SCD is an air-bone gap with intact acoustic reflexes;[3,4] the air-bone gap is at least in part due to a conductive hearing gain rather than a conductive hearing loss. If the symptoms of SCD become intolerable, surgical resurfacing of the superior semicircular canal via the middle cranial fossa approach can be effective treatment.[1] Here we report a patient with SCD who was thought by others, largely on the basis of the audiologic findings, to have otosclerosis, and underwent a stapedectomy elsewhere 10 years ago. This case is reported to draw attention to the way in which SCD can simulate otosclerosis and to simple tests that can distinguish between the two.

Case report

A male now aged 65, was first seen, about loss of balance, in June 2002. He had first noticed a hearing problem in his 30s whilst serving in the Royal Australian Navy. He first sought help for it in 1985, aged 48. He remembers being told at the time that he had a bilateral high-frequency hearing loss due to noise (gunnery) and 'mild otosclerosis' – the old records are lost. He was advised against surgery, which he duly put off until 1992. At that time he remembers being told that he had 'a 90 per cent chance of having better hearing after operation and a 10 per cent chance of having worse'. After a stapedectomy on the left ear there was no improvement in hearing and he remembers being told by the surgeon that 'when he broke the stapes there was no clear break and more bone came out than should have'. Five years later, in 1997, the left hearing deteriorated over four weeks and the ear felt blocked; there was also some vertigo. Another surgeon found what he thought at the time was 're-obliteration of the stapes footplate and subluxation of the stapes

From the Neurology Department, Royal Prince Alfred Hospital, Sydney, and the Otolaryngology Department, St Vincent's Hospital, Sydney, Australia.
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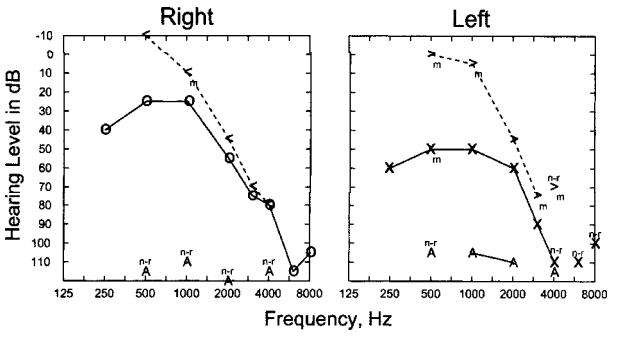

FIG. 1

Audiogram. Following the left stapedectomy and two revisions there is still a large air-bone gap at 500 and 1000 Hz on the left as well as on the unoperated right side. The contralateral acoustic reflex, the sound stimulus in the operated left ear and the volume probe in the unoperated right ear is present at 1 kHz and 2 kHz (A–A) and absent at 0.5 and 4 kHz. It is absent at all frequencies with the stimulus in the unoperated right ear and the volume probe in the thrice-operated left ear. There is also a severe high frequency loss due to noise damage. Speech comprehension at 65 dB was 95 per cent on the right and 85 per cent on the left. (m = masked; n-r = no response).

footplate'. The vertigo improved after surgery and so did the hearing – back to the previous level. In August 2001, after a violent sneeze, he felt his left ear pop and his hearing drop. A computed tomography (CT) scan was reported as showing fluid in the left middle ear. At operation (under local anaesthetic) the same surgeon who carried out the first revision found that the long process of the incus had necrosed and the prosthesis was lying free; attempting to remove it produced intense vertigo. A Goldenberg stapes-incus device was then inserted and tucked under the malleus handle and left on the mobile oval window membrane. The hearing improved after operation but not quite to the level it was before the drop after sneezing; the vertigo slowly disappeared.

On specific questioning about his hearing in June 2002, he volunteered that his own chewing has always sounded too loud, his hearing was always unusually good underwater and that when sitting on a park bench he could hear through his buttocks! Although he had no pulsatile tinnitus sometimes he could hear his own joints move. He started using hearing aids after the first revision surgery and while he did think that he derived some benefit from them his family had noted that with his hearing aids out his voice did not become any louder.

On examination there was no spontaneous, gaze-evoked head-shaking or positional nystagmus, no deficiency of vestibulo-occular reflexes on impulsive testing, and no abnormalities of stance or gait. Sound- and pressure-induced nystagmus was sought with infra-red videonystagmoscopy. In this way it was possible to observe that a nasal Valsalva manoeuvre would produce a slight counterclockwise ocular torsion and glottal Valsalva would produce slight clockwise torsion. A Hennebert test on the unoperated right ear produced slight counterclockwise torsion with raised pressure and slight clockwise torsion with lowered pressure. The Hennebert test was negative on the left (operated) ear. A 0.5 kHz and a 1.5 kHz tone at 110 dB NHL produced no nystagmus from either ear. (For kinematic consistency the direction of a torsional eye movement has to be defined, just like the direction of a horizontal eye movement – left and right, from the patient's and not from the observer's point of view. This means that a torsional eye movement that rotates the 12 o'clock point of the eye, as viewed by an observer, towards the patient's right is called clockwise whereas one that

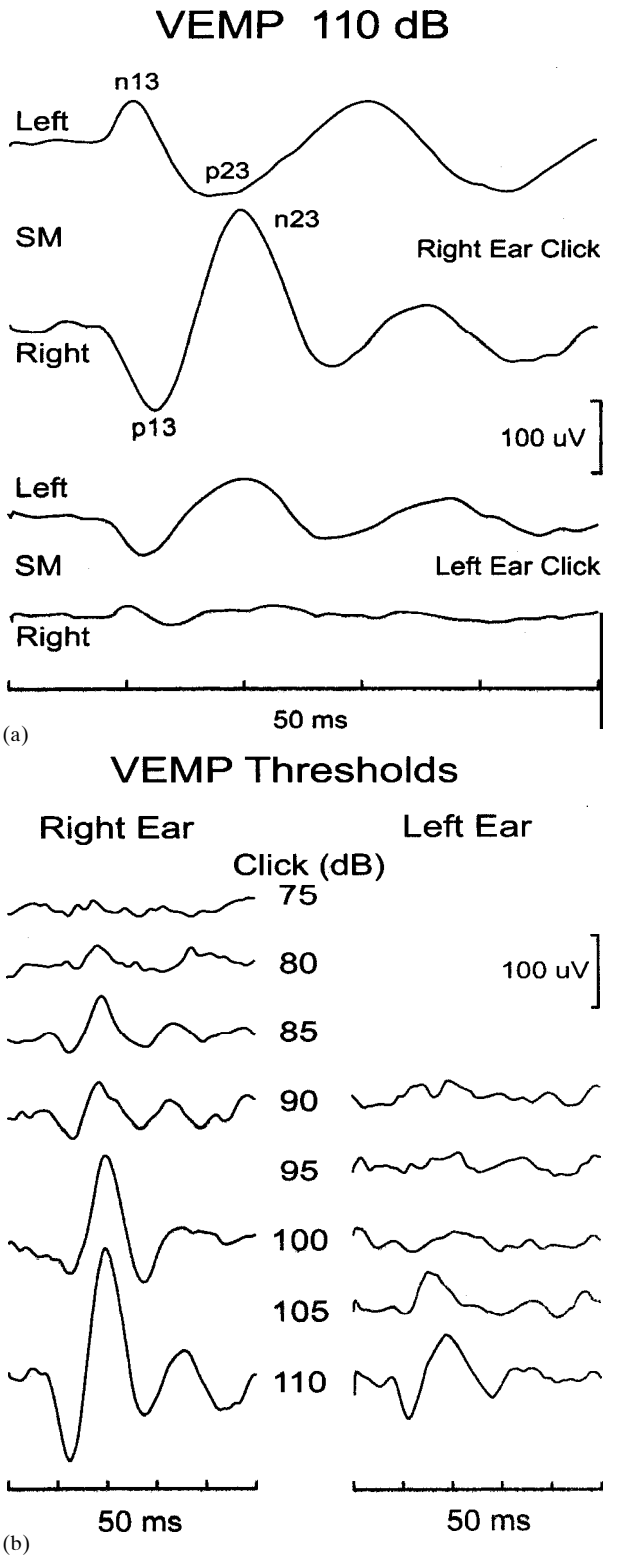

FIG. 2

Vestibular evoked myogenic potentials. (a) Shows that despite the significant air-bone gap, p13-n23 responses to 110 dB clicks are still present. The VEMP from the left ear is normal in amplitude but smaller (115 μV) than from the right (307μV) indicating that there is some conductive loss on the left (as a result of the three operations) as well as a conductive gain. The response from stimulating the right ear is abnormally large and is accompanied by a contralateral inverse response, n13-p23. (b) Shows that the VEMP threshold is normal from the left ear (with a conductive loss the VEMP should be absent) and abnormally low (80 dB; normal >95 dB) from the right ear.

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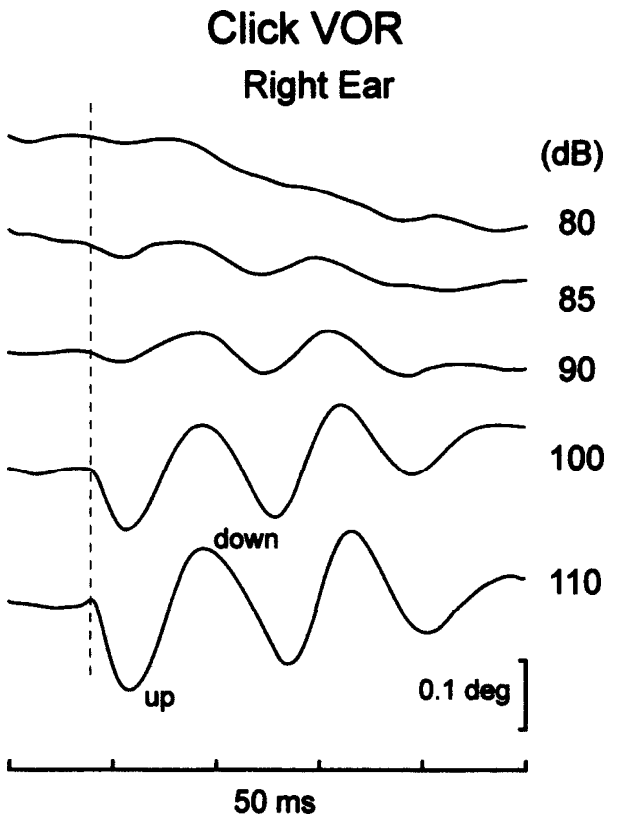

FIG. 3

Click-evoked vestibulo-ocular reflex. Averaged vertical EOG responses of the left eye to 256, 100 μ sec, 110 dB clicks to the right ear. There is a vertical vestibulo-ocular reflex with a latency to onset (dotted vertical line) of 7.5 msec, a peak-to-peak displacement of about 0.3 degrees and a threshold of 85 dB. Normal subjects have responses of only 0.03 degrees or less with a threshold of 100 dB or more.[6]

rotates the 12 o'clock point to the patient's left is called counter-clockwise. This convention means that what looks clockwise to the observer is counter-clockwise for the patient.)

The Weber test lateralized to the left; the Rinne test showed better bone than air conduction bilaterally. The patient could also hear a 256 Hz tuning fork when it was placed on his ankle.

The audiogram showed a severe, bilateral, symmetrical high frequency loss as well as a bilateral low-frequency air-bone gap more marked on the left than on the right (Figure 1). On the left the bone threshold at 0.5 kHz was 0 dB NHL and on the right it was -10 dB. The acoustic reflexes from stimulating the right ear were present ipsilaterally but not contralaterally (recording from the thrice operated left ear); from stimulating the operated left ear the ipsilateral acoustic reflex was absent but the contralateral reflex was present at 100 dB at 1.0 and 2.0 kHz. In other words acoustic reflexes were present in the probe right conditions (1 kHz ipsilateral; 1 kHz and 2 kHz contralateral) and absent in the probe left conditions consistent with the previous left stapes surgery.

VEMPS were present from both sides in response to the standard 110 dB NHL click stimulus (Figure 2 (a)). The ipsilateral p13-n23 amplitude was 307 μV from the right ear and right sternomastoid and 115 μV from the left ear. There were bilateral contralateral n13-p23 responses, larger from stimulating the right ear than the left. The VEMP threshold was 80 dB in the right ear (N>95 dB) and 100 dB in the left (Figure 2 (b)). The caloric test showed a mild (28 per cent) left canal paresis.

The click-evoked vestibulo-ocular reflex (VOR) was recorded by averaging the vertical electro-oculogram (EOG).[6] A 0.3 deg click-evoked VOR could be recorded from the unoperated right ear at a threshold of 85 dB, close to the VEMP threshold of 80 dB (Figure 3), and at a

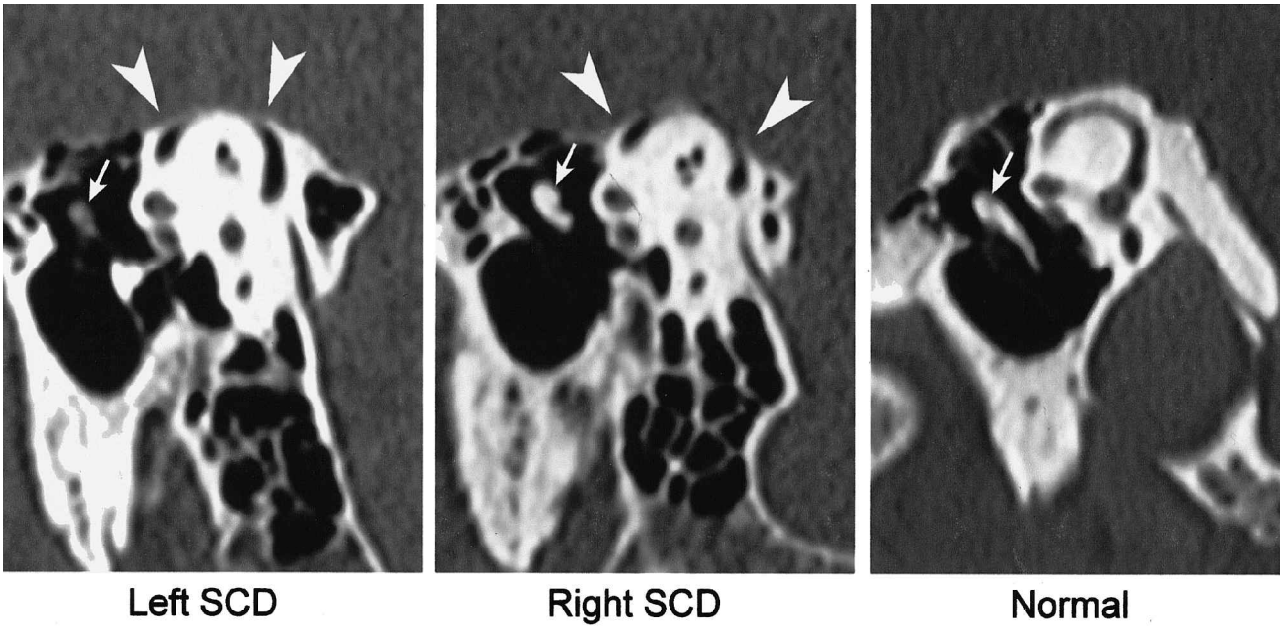

FIG. 4

High-resolution spiral CT of the temporal bones, reconstructed in the plane of each superior semicircular canal, showing a bilateral superior semicircular canal dehiscence (large white arrowheads). CT from a normal subject is shown for comparison. The TORP device is clearly seen abutting the oval window on the left. (Courtesy of Dr John Harding-Smith, Central Sydney Imaging)

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threshold 110 dB from the operated left ear. Normal subjects have a click-evoked VOR of only 0.03 deg or less at a threshold greater than 100 dB.[6]

- **The patient originally presented with an air bone gap thought 10 years ago to be a conductive hearing loss due to otosclerosis and was treated with a stapedectomy**
- **It now transpires that the patient actually had a hearing gain due to superior semicircular canal dehiscence**
- **The methods of investigation including vestibular evoked myogenic potentials and high-resolution spiral CT of the temporal bone are described**

A high-resolution, spiral CT of the temporal bones at 0.5 mm slice thickness confirmed bilateral SCD with no evidence of otosclerosis (Figure 4). A three-dimensional reconstruction showed that when viewed from the middle fossa the SCD was 5.2 mm in length (Figure 5).

The possibility of superior semicircular canal resurfacing[1] was discussed with the patient but at this stage he has declined further surgery and is happy to monitor progress.

Discussion

SCD allows sound, raised middle-ear pressure and raised middle cranial fossa pressure to activate the vestibular system. This can be shown by nystagmus or other eye movement responses to sound or pressure or both – the Tullio phenomenon,[1,5,6] and by low-threshold, high amplitude VEMPs in response to clicks.[2–4,7,8] In SCD, loud (>100 dB NHL), low-frequency (0.5–1.5 kHz) air-conducted sounds can induce a torsional-vertical nystagmus due to activation of the superior SCC.[1,5] The vertical component of the nystagmus slow phase rotates the eyes upwards, while the torsional component rotates the upper, i.e. 12 o'clock, poles of the eyes away from the stimulated ear. Raised middle-ear pressure from a nasal Valsalva manoeuvre or from raised external ear canal pressure (Hennebert's sign) can induce similar nystagmus. Conversely raised middle cranial fossa pressure from a glottal Valsalva manoeuvre or from jugular vein compression and lowered external ear canal pressure can produce nystagmus in the opposite direction to that produced by a nasal Valsalva. Although many patients with SCD show nystagmus responses to both pressure and sound, some will show responses only to pressure but not to sound, e.g. Minor *et al.*[1] Cases 3, 6 and Streubel *et al.*[4] *Cases 1–4* and the present case, whereas others will show responses only to sound but not pressure, e.g. Minor *et al.*[1] *Case 8.* Some patients even have pulse-synchronous spontaneous torsional pendular nystagmus.[4,10]

Many, perhaps most, patients with SCD show not only the excessive vestibular sensitivity to air-conducted sound described above but also excessive cochlear sensitivity to bone-conducted sounds.[2–4,8,9] For example they can hear their own joints move, their own eyes move (different to gaze-evoked tinnitus), their own heart beat (pulsatile tinnitus) and heel-strike during walking. They can also hear a vibrating tuning fork placed on a distant bony prominence such as the ankle.

Some, perhaps most, patients with SCD show an air-bone gap on the pure-tone audiogram that looks like a conductive hearing loss but is in fact a conductive hearing *gain*.[2–4] Moreover, the bone-conduction threshold is often negative, as it was in our patient's unoperated right ear.

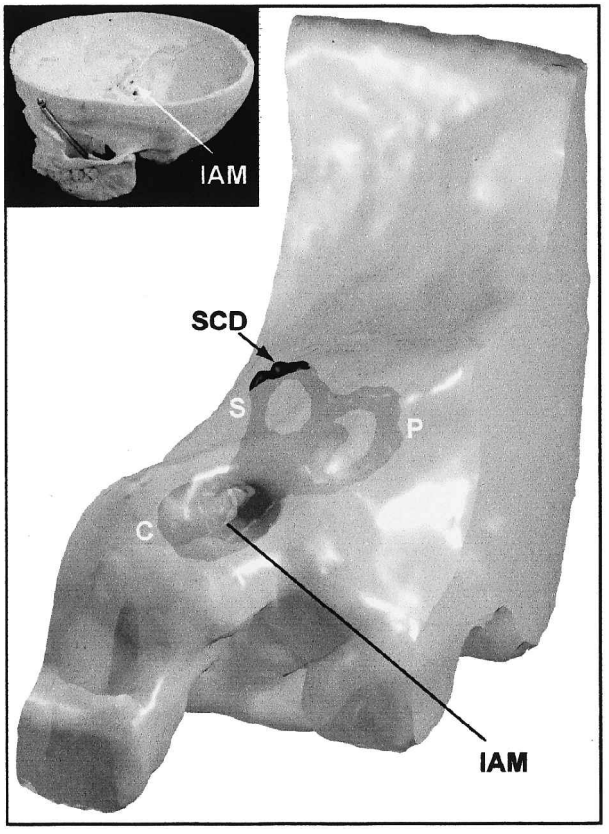

FIG. 5

Three-dimensional surface-rendered reconstruction of the temporal bone CT showing the location of the SCD in the floor of the middle cranial fossa. The rendering has been made translucent so that the entire inner ear is visible within the temporal bone. IAM, internal auditory meatus; C, cochlea; S, superior semicircular canal; P, posterior semicircular canal.

Two possible explanations for the air-bone gap are: a dissipation of sound energy through the third window, creating a sensorineural loss, or a pre-existing sensorineural loss and improved bone but not air thresholds through a third window.[4] An additional window might also be the reason for the appearance of an air-bone gap in some patients with large vestibular aqueduct syndrome who have reasonable air-conduction thresholds.[11]

It is important to note that in patients who have an air-bone gap due to SCD the acoustic reflexes are preserved, unlike in patients with otosclerosis.[3] An air-bone gap is the cardinal feature of ossicular chain fixation due to fenestral otosclerosis and in these cases the acoustic reflexes are expected to be absent.[12]

A large amplitude, low-threshold click-evoked VEMP with a prominent contralateral reversed potential (n13-p23) is characteristic of SCD.[2–4] In contrast, a conductive hearing loss reduces and, if severe enough, abolishes the VEMP due to a loudness effect.[13,14] Therefore both VEMPs as well as acoustic reflexes should be absent in a patient with an air-bone gap due to otosclerosis, whereas the acoustic reflex should be preserved and the VEMP should not only be preserved but be abnormally large in SCD.

Our patient presented with a bilateral hearing loss and was found to have, in addition to a severe symmetrical high-frequency loss, what was considered elsewhere to be a mild bilateral low-frequency conductive loss. At stapedectomy seven years later although the footplate would have been found to be mobile the rest of the ossicular chain

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seems not to have been checked for fixation. There was no change in the air-conduction threshold after surgery and the subsequent auditory and vestibular problems that developed, requiring revision surgery, were a consequence of the original surgery and not of the original disorder, which was always in the middle fossa floor and not in the middle ear. Although neither SCD nor VEMPs had been described at the time of the original surgery the preservation of acoustic reflexes in a patient with an air-bone gap worth closing should suggest that ossicular chain fixation might not be the problem.

VEMPs are normally abolished by conductive hearing loss; for example a 15 dB loss at 1 kHz will reduce the VEMP amplitude by 70 per cent.[13] The last of the three operations on our patient's left ear produced a slight conductive hearing loss so the fact that a VEMP could be recorded at all from that ear indicates that the response would have been bigger before any surgery. There is a similar previous report of two patients with SCD being diagnosed as having otosclerosis and being treated with a stapedectomy.[4]

The major lesson from this case is that any patient with a significant air-bone gap but intact acoustic reflexes could have SCD, a well-known anatomical abnormality[15], rather than otosclerosis and should have sound and pressure-induced nystagmus sought and VEMPs tested. If there is nystagmus in response to sound or pressure and if the VEMP is intact then the patient needs high resolution CT scanning of the temporal bones to assess the bony tegmentum over the superior SCC.[16] It should also be noted that SCD can not only mimic stapes fixation as in this case, but might also mimic stapes hypermobility. In the cases reported by Deecke *et al.*[17] and by Dieterich *et al.*,[18] Tullio phenomenon was attributed to a hypermobile stapes and the patients were said to have been helped by surgical stapes fixation. However, in neither case were the audiometric or CT data reported and in retrospect both probably had SCD.

Note added in proof

After this paper was accepted for publication Minor *et al.* reported the audiological findings in four patients with SCD and apparent conductive hearing loss, three of whom had stapedectomy for suspected otosclerosis.[19]

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Address for correspondence:
Dr G. M. Halmagyi,
Neurology Department,
RPA Hospital,
Camperdown, Sydney
NSW 2050, Australia.

Fax: +61 9515 8347
E-mail: Michael@icn.usyd.edu.au

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