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SHORT COMMUNICATION

Hearing eyeball and/or eyelid movements on the side of a unilateral superior semicircular canal dehiscence

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Abstract Hearing of eyeball movements has been reported in superior semicircular canal dehiscence (SSCD), but not hearing of eyelid movements. Our main objective was to report the hearing of eyeball and/or eyelid movements in unilateral SSCD. Our secondary objective was to access its specificity to SSCD and discuss the underlying mechanism. Six patients with SSCD who could hear their eyeball and/or eyelid movements were retrospectively reviewed. With the aim of comparisons, eight patients with an enlarged vestibular aqueduct (EVA), who share the same mechanism of an abnormal third window, were questioned on their ability to hear their eyeball and/or eyelid movements. Three patients with SSCD could hear both their eyeball and eyelid movements as a soft low-pitch friction sound. Two patients with SSCD could hear only their eyelid movements, one of whom after the surgery of a traumatic chronic subdural hematoma. The latter remarked that every gently tapping on the skin covering the burr-hole was heard in his dehiscent ear as the sound produced when banging on a drum, in keeping with a direct transmission of the sound to the inner ear via the cerebrospinal fluid. One patient with SSCD, who could hear only his eyeball movements, had other disabling symptoms deserving operation through a middle fossa approach with an immediate relief of his symptoms. None of the eight patients with EVA could hear his/her eyeball or eyelid movements. Hearing of eyeball and/or eyelid movements is highly suggestive of a SSCD and do not seem to occur in EVA. In case of radiological SSCD, clinicians should search for hearing of eyeball and/or eyelid movements providing arguments for a symptomatic dehiscence. The underlying mechanism is discussed particularly the role of a cerebrospinal fluid transmission.

Keywords Superior semicircular canal dehiscence · Enlarged vestibular aqueduct · Eyeball movements · Eyelid movements · Blink · Surgical repair · Bone conduction · Cerebrospinal fluid · Third window

Introduction

In 1998, Minor et al. described a new syndrome caused by a dehiscence of the bone overlying the superior semicircular canal (SSC) at the level of the middle cranial fossa [1]. This syndrome is characterized by vestibular symptoms typically induced by sound and pressure stimuli and/or auditory dysfunction with conductive hearing loss [1, 2]. In addition, patients can perceive their own body sounds, such as their voice, footsteps when walking, cracking of the cervical spine, rarely their eyeball movements [2–4]. To the best of our knowledge, the hearing of eyelid movements, i.e., blinking has never been reported. We present the features of six patients who could hear their eyeball movements, their eyelid movements or both on the side of a unilateral superior semicircular canal dehiscence (SSCD).

Materials and methods

Six patients seen between 2011 and 2016 with a unilateral SSCD were included as they could hear their eyeball and/or eyelid movements on the affected ear. They had a detailed otoneurological examination, pure tone audiometry, and stapedial reflexes. Cervical vestibular-evoked myogenic

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potentials (cVEMPs) were performed with air conduction tone bursts stimuli and considered in keeping with a SSCD if present at a threshold of 70 dB or less. High-resolution computed tomography (CT) scans of the temporal bones were reformatted to provide Poschl views, and SSCD was defined as a bone defect of the SSC of 2.5 mm or more (see Fig. 1).

These six patients were compared with eight patients with an enlarged vestibular aqueduct (EVA), defined as more than

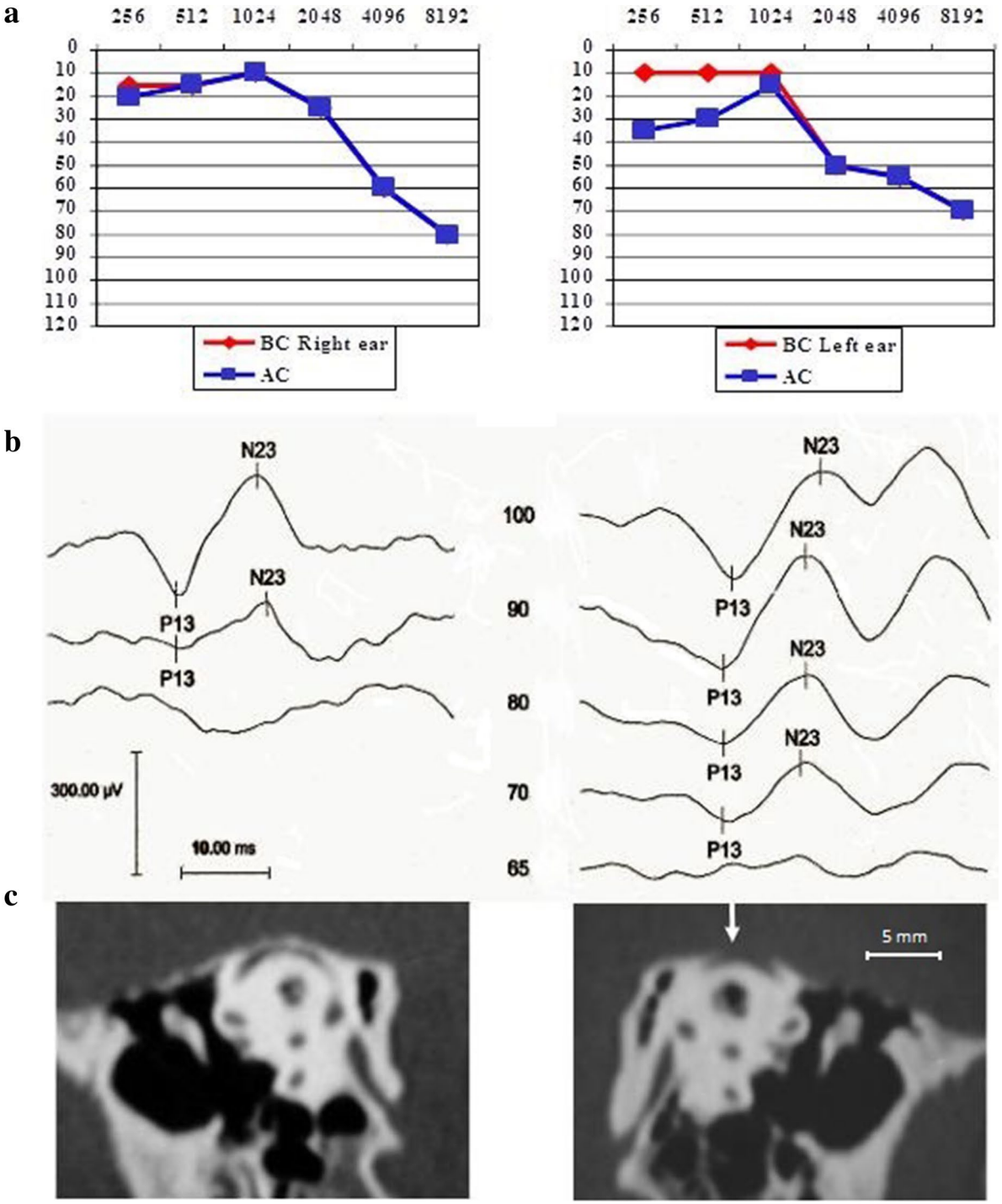

Fig. 1 Audiometry, cVEMPS and CT scan in patient 1 before surgery. **a** Pure tone audiometry showing high-frequency sensorineural hearing loss on the right and mixed hearing loss on the left. *BC* bone conduction, *AC* air conduction. **b** cVEMPs showing a normal threshold on the right (90 dB) and a lowered threshold on the left (70 dB) in keeping with a left SSCD. **c** CT scan in the Poschl's plane showing a 4 mm bone dehiscence at the level of the left superior semicircular canal (white arrow)

2 mm diameter at its midpoint on CT scan with axial views. These eight patients were questioned on their ability to hear their eyeball and/or eyelid movements, as EVA and SCCD share the same mechanism of an abnormal third window [5].

Results

The features of the six patients (mean age 51, range 26–67 years old) with unilateral SSCD are presented in Table 1. One patient could hear only his eyeball movements (patient 1), two patients could hear only their eyelid movements (patient 2 and 3), and three patients were able to hear both their eyeball and eyelid movements (patient 4, 5 and 6). Symptoms were revealed by surgery of a traumatic subdural hematoma (patient 2) or immediately after a head trauma (patient 3). Five out of six patients complained of various vestibular symptoms, including one patient who had dizziness when trying a continuous positive airway pressure machine for obstructive sleep apnea (patient 3). Four patients had hearing loss corresponding to a mixed hearing loss in 3 (see Fig. 1) and a conductive hearing loss in another patient. cVEMPs were suggestive of the diagnosis with a low threshold on the side of the SSCD in 4 out of 6

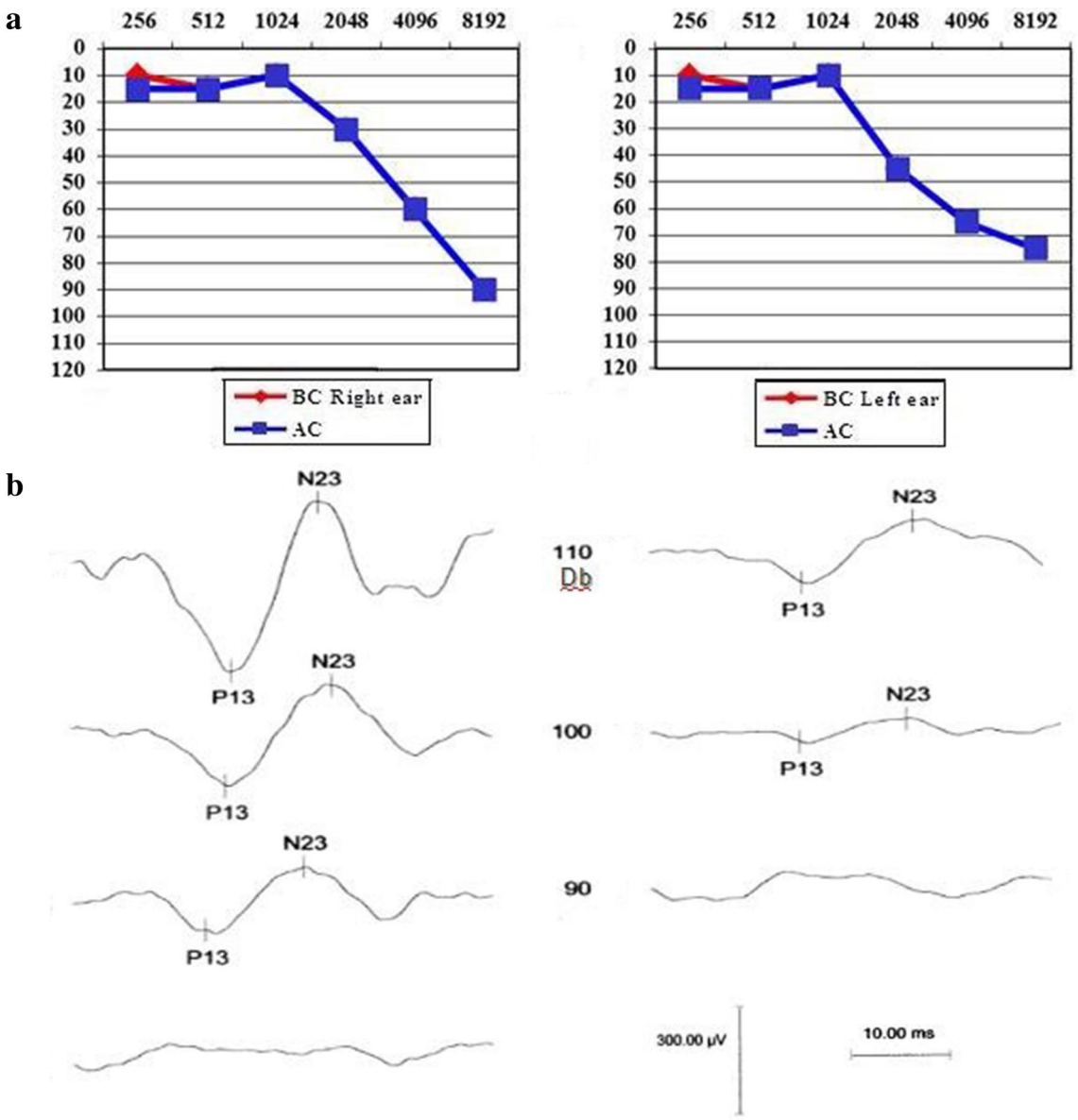

Fig. 2 Audiometry and cVEMPS in patient 1 after surgery. **a** Pure tone audiometry showing bilateral high-frequency sensorineural hearing loss with disappearance of the conductive component on the left side. *BC* bone conduction, *AC* air conduction. **b** cVEMPs showing normal thresholds on both sides (post-operatively, the threshold is higher and the amplitude is lower on the left side)

patients (see Fig. 1). The diagnosis was confirmed in all patients by a CT scan with reconstructions parallel to the SSC (see Fig. 1).

The following two case histories illustrate representative patients.

Patient 1

A 61-year-old man insidiously developed tinnitus and hearing loss on the left side about 7 years prior to his presentation. Three years before presentation, he noted that objects in the visual field seemed to move whenever he spoke and that his voice resounded in his left ear. He complained of vertical visual instability when walking and of hearing his footsteps in his left ear. These symptoms improved when using shoes with a soft sole. He experienced vertigo for a few seconds when sneezing or coughing. He heard in his left ear his heartbeats as well as the cracking of his cervical spine during head motion. Curiously, he also complained of hearing the movement of his eyeballs in his left ear and compared this noise to a soft low-pitch friction sound. He could not hear his eyelid movements. Pure tone audiometry revealed a bilateral symmetrical sensorineural hearing loss in the high frequencies with a superimposed conductive hearing loss on the left side (Fig. 1). cVEMPs revealed abnormally low thresholds on the left side (Fig. 1). CT scan showed a 4 mm bone dehiscence of the left SSC (Fig. 1).

The symptoms were so disabling with a Dizziness Handicap Inventory (DHI) score at 68 that the patient had to stop working and got depressed. He was operated on through a middle fossa approach with covering the defect by bone wax, gently tucked into the open roof of the canal, followed by a layer of temporalis fascia and a bone graft harvested from the middle fossa bone flap. When awaking in the operating room, he immediately noticed that he could not hear anymore the movement of his eyeballs or cervical spine. When re-assessed 2 years after surgery, all his symptoms had disappeared, apart from his initial left tinnitus, and his DHI score was 6. His left hearing loss improved by disappearance of the conductive component and cVEMPs regained normal thresholds (Fig. 2).

Patient 2

A 55-year-old man was operated on in June 2014 for the drainage of a right hemispheric chronic subdural hematoma, 10 weeks after a head trauma during a brawl. He had to be re-operated on 1 month later for a residual hematoma. Surgery was performed under general anesthesia through a 14 mm parietal burr-hole with a drain left in place within the subdural space during 3 days post-operatively. The bone flap was not replaced at the end of surgery. After this second surgery, the patient complained of fullness in his left ear, and of hearing his footsteps when walking, his heartbeats when walking rapidly or lifting heavy loads, as well as the resounding of his voice whenever he spoke. He noticed that he could hear every eyelid movements in his left ear as a soft low-pitch friction sound with no simultaneous movement of the tympanic membrane on otoscopic examination. He could not hear his eyeball movements. Interestingly, he remarked that every gently tapping on the skin covering the burr-hole was immediately transmitted to his left ear and described the resulting noise as the one produced when banging on a drum. Tapping the skull around the burr-hole was not transmitted to his left ear. Routine neurological examination was normal. Pure tone audiometry revealed a bilateral sensorineural hearing loss in high frequencies, with a superimposed conductive hearing loss on the left side. A CT scan showed a 2.5 mm bone dehiscence of the left SSC and was normal on the right side (see Table 1).

None of the eight patients (mean age 42, range 17–73 years old) suffering from an EVA was able to hear either their eyeball or eyelid movements.

Discussion

The unique feature in these six patients with a SSCD was the hearing of eyeball and/or eyelid movements on the side of the affected ear. Four patients were able to hear their eyeball movements as a soft low-pitch friction sound. This phenomenon should be differentiated from the occurrence of a tinnitus in lateral gaze occurring after surgery for a brainstem lesion, typically a vestibular schwannoma [4, 6, 7]. In this latter circumstance, the tinnitus is usually heard when moving the eyes towards the diseased ear [4, 6, 7]. However, none of our patients had a previous history of a brainstem lesion and all of them were able to hear their eyeball movements when moved in any direction. Hearing of eyeball movements has already been detailed in two patients with a bilateral SSCD that were not operated on [3, 4]. It is noteworthy that our first patient had an immediate relief of hearing his eyeball movements after surgery which confirms the relationship between this phenomenon and the SSCD.

In our series, five patients could hear their eyelid movements on the side of the affected ear which to the best of our knowledge has never been reported before. These patients reported the hearing of a soft low-pitch friction when blinking and the sound was similar to the one produced by the movements of their eyeballs for the three patients that can hear both. Although subjective, the sound seems different from the hearing of a click in the ear (muscular tinnitus) that patients complain after eye closure in oculostapedial synkinesis. The latter rarely occurs spontaneously but typically occurs after facial palsy or hemifacial spasm [8–11]. Concomitantly to facial movements,

Table 1 Summary of features in the six patients (mean age 51, range 26–67 years old)

	Patient 1 (61 years)	Patient 2 (55 years)	Patient 3 (67 years)	Patient 4 (57 years)	Patient 5 (26 years)	Patient 6 (42 years)
Medical history	None	None	Cardiac arythmia	Kidney tumor	Personal = none Familial = his father had a unilateral SSCD	Migraine Fibromyalgia
Age at onset	52 years	54 years	66 years	56 years	24 years	37 years
Triggering factor		Since surgery of a subdural hematoma	Since minor head trauma			
Audiological symptoms						
Hearing						
Eye movement	Yes	No	No	Yes	Yes	Yes
Eyelid movement	No	Yes	Yes	Yes	Yes	Yes
Personal voice	Yes	Yes	Yes	Yes	No	Yes
Footstep	Yes	Yes	Yes	Yes	Yes	Yes
Heartbeat	Yes	Yes	Yes	Yes	Yes	Yes
Cervical spine	Yes	Yes	No	Yes	No	No
Others	No	Yes (tapping on the cranial flap is transmitted to his left ear)	No	No	No	No
Fullness	Yes	Yes	No	No	Yes	Yes
Tinnitus	Yes	No	Yes	No	Yes	Yes
Vestibular symptoms		No				
Pressure (sneezing, coughing)	Yes		No	No	No	Yes
Tullio	No		No	No	No	Yes
Others	Movement of the visual field when speaking or walking		Dizziness when using sleep apnea machine precluding the use of this apparatus	Instability aggravated in darkness Oscillopsia when running	Dizziness during physical exertion (tennis, jogging) One drop attack	Positional and spontaneous dizziness
Audiovestibular explorations						
Pure tone audiometry	Left mixed HL	Left mixed HL	Right mixed HL	N	N	Right conductive HL
Stapedial reflexes	Yes	No	Yes	No	Yes	Yes
VEMP	90 dB (right ear) versus 70 dB (left ear)	90 dB on both sides	55 dB (right ear) versus 100 dB (left ear)	95 dB (right ear) versus 80 dB (left ear)	55 dB (right ear) versus 95 dB (left ear)	55 dB (right ear) versus 90 dB (left ear)
Caloric stimulation	N	NP	NP	N	Left hypovalence (29%)	NP
CT scan	Left SSCD (4 mm)	Left SSCD (2.5 mm)	Right SSCD (6.5 mm)	Left SSCD (3 mm)	Right SSCD (5 mm)	Right SSCD (6 mm)
MRI scan	N	Subdural hematoma	NP	N	NP	N
Treatment	Surgical repair via middle cranial fossa approach	None	None	None	None	None

HL hearing loss, *NP* not performed, *N* normal, *SSCD* superior semicircular canal dehiscence

otoscopic examination can reveal a retraction of the tympanic membrane which is mainly detected by changes in middle ear compliance during impedance audiometry [8, 10, 11]. None of our patients had a previous history of facial palsy or hemifacial spasm or, on clinical examination, a simultaneous movement of the tympanic membrane during blinking.

From a pathophysiological point of view, it is challenging to explain why patients with a SSCD can hear their eyeball and/or eyelid movements. In animal experimentation, it has been demonstrated that the selective application of a vibrator to the eye was able to induce brainstem auditory evoked potentials after complete immobilization of the ossicular chain (air conduction route suppression), stapes footplate and round window [12]. Thus, activation of the cochlea was not based on relative motion between the stapes footplate and oval window with fluid volume displacements between the oval and round windows but by other mechanisms which need to be elucidated [12]. As the muscles of the eyeballs and eyelids are attached to the skull bones (eye socket), it can be assumed that, when these muscles move, there is a friction that can be heard by the patient as a noise in the ear, via a bone conduction route, as demonstrated in animals [12]. There are at least five factors contributing to bone conduction hearing, one of which, the cerebrospinal fluid (CSF) transmission is considered to be a poor contributor in physiological conditions [13]. In the case of SSCD, transmission of sounds to the inner ear can obviously be facilitated through an abnormal third window in direct contact with the CSF space. This putative mechanism is well illustrated by patient 2 in whom tapping gently on the skin over the burr-hole (and not around) was heard in his dehiscent ear as the sound produced when banging on a drum. Similarly, a direct transmission through intracranial pressure (known to be pulse regulated) explains exceptional cases of pulse-synchronous nystagmus occurring in SSCD and resolving after surgery [14, 15]. Conversely, transmission of sounds from the eyeball and/or eyelid to the CSF space cannot be simply explained except if one considers a conduction through adipose tissue of the orbits, optical nerve sheaths or their surroundings with meningeal extensions [16]. Indeed, an anatomical study shows that the optic nerve sheath is continuous with the dura mater of the brain and surrounded by the arachnoid space containing CSF [17]. The reason why patients with EVA do not hear their eyeball or eyelid movement is unclear. It can be suggested that for SSCD, the third window is in the middle fossa close to the anatomical structures of the eyes, while for EVA it is in the posterior cranial fossa. In addition, the intrinsic mechanism of the third mobile window is probably not comparable in these two entities regarding a different anatomy and function between the SSC and the vestibular aqueduct.

Conclusion

Hearing of eyeball and/or eyelid movements as a low-pitch sound in the ear should draw clinician's attention to SSCD. This phenomenon does not occur in EVA although additional reports on malformations or other pathologies of the inner ear will be necessary to define its specificity to SSCD. Conversely, in case of radiological SSCD, clinicians should search for hearing of eyeball and/or eyelid movements providing arguments for a symptomatic dehiscence.

Compliance with ethical standards

Funding The authors declare no funding.

Conflict of interest The authors declare no conflict of interest.

Ethical approval This is an anonymous retrospective study. For this type of study formal consent is not required.

References

1. Minor LB, Solomon D, Zinreich JS et al (1998) Sound-and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal. Arch Otolaryngol Head Neck Surg 124:249–258
2. Minor LB (2000) Superior canal dehiscence syndrome. Am J Otol 21:9–19
3. Schmuziger N, Allum J, Buitrago-Téllez C et al (2006) Incapacitating hypersensitivity to one's own body sounds due to a dehiscence of bone overlying the superior semicircular canal. A case report. Eur Arch Otorhinolaryngol 263:69–74
4. Albuquerque W, Bronstein AM (2004) "Doctor, I can hear my eyes": report of two cases with different mechanisms. J Neurol Neurosurg Psychiatry 75:1362–1366
5. Merchant SN, Rosowski JJ (2004) Conductive hearing loss caused by third-window lesions of the inner ear. Otol Neurotol 29:282–289
6. Coad M, Lockwood A, Salvi R, Burkard R (2001) Characteristics of patients with gaze-evoked tinnitus. Otol Neurotol 22:650–654
7. Biggs ND, Ramsden RT (2002) Gaze-evoked tinnitus following acoustic neuroma resection : a deafferentation plasticity phenomenon. Clin Otolaryngol Allied Sci 27:338–343
8. Donne AJ, Homer JJ, Woodhead CJ (2000) Oculostapedial synkinesis following Bell's palsy. J Laryngol Otol 114:135–136
9. Kim P, Fukushima T (1984) Observations on synkinesis in patients with hemifacial spasm. Effect of microvascular decompression and etiological considerations. J Neurosurg 60:821–827
10. Ohki M, Kato H (2012) Idiopathic tinnitus concomitant with eye closure. Otol Neurotol 33:267–269
11. Fujiwara K, Furata Y, Fukuda S (2014) Idiopathic oculostapedial synkinesis without history of facial nerve palsy. Otol Neurotol 36:510–512
12. Perez R, Adelman C, Sohmer H (2011) Bone conduction activation through soft tissues following complete immobilization of

the ossicular chain, stapes footplate and roud window. Hear Res 280:82–85
13. Stenfelt S, Goode RL (2005) Clinically oriented basic science review. Bone-conducted sound: physiological and clinical aspects. Otol Neurotol 26:1245–1261
14. Tilikete C, Krolak-Salmon P, Truy E, Vighetto A (2004) Pulse-synchronous eye oscillations revealing bone superior canal dehiscence. Ann Neurol 56:556–560
15. Younge BR, Khabie N, Brey RH, Driscoll CLW (2003) Rotatory nystagmus synchronous with heartbeat: a treatable form of nystagmus. Trans Am Ophthalmol Soc 101:113–118
16. Bhutta MF (2015) Eye movement autophony in superior semicircular canal dehiscence syndrome may be caused by trans-dural transmission of extraocular muscle contraction. Int J Audiol 54:61–62
17. Killer HE, Laeng HR, Flammer J, Groscurth P (2003) Architecture of arachnoid trabeculae, pillars, and septa in the subarachnoid space of the human optic nerve: anatomy and clinical considerations. Br J Ophthalmol 87:777–781