RESPONSE TO “MICHAEL YONG, ERICA ZAIA, BRIAN WESTERBERG, AND JANE LEA. DIAGNOSIS OF SUPERIOR SEMICIRCULAR CANAL DEHISCENCE IN THE PRESENCE OF CONCOMITANT OTOSCLEROSIS”. OTOL NEUROTOL 2017;38:1071–1075

To the Editor: In the article “Diagnosis of Superior Semicircular Canal Dehiscence in the Presence of Concomitant Otosclerosis” reported by Yong et al., the authors review a series of three patients with concomitant otosclerosis and superior semicircular canal dehiscence (SCDS) (1).

All three patients presented with conductive hearing loss and normal tympanic membranes, but also with symptoms and signs potentially attributable to a third mobile window lesion. The latter prompted further evaluation, including vestibular-evoked myogenic potentials and temporal bone computed tomography (CT) of the temporal bone. This eventually resulted in the diagnosis of concomitant fenestral otosclerosis and SCDS.

The authors provide an important argument for a well-executed history, audiovestibular testing, and imaging to properly diagnose patients with conductive or mixed hearing loss and an intact tympanic membrane. We fully agree with the authors’ analysis on proper patient evaluation and counselling in case of concomitant otosclerosis and SCDS, which should reflect the current state-of-the-art. We would also like to thank them for sharing case number 2, where the patient elected to go ahead with stapedotomy without satisfactory result. This is very useful additional information (2) whether or not to combine or stage procedures to treat otosclerosis or third mobile window lesions (such as large vestibular aqueduct syndrome, SCDS, etc.).

We have had a similar experience with bilateral otosclerosis and bilateral large vestibular aqueducts, as a reflection of concomitant third mobile window lesions, where we opted not to proceed with stapedotomy (3). However, we also encountered a patient with bilateral SCDS and bilateral cochlear cleft of the otic capsule, a finding not that well known by clinicians and radiologists. This patient particularly did not report the symptoms quite typical to SCDS, e.g., autophony, pulsatile tinnitus, etc. She only complained of a hearing loss, which turned out to be a bilateral conductive hearing loss. An important aspect in this specific case is the presence of ipsilateral and contralateral stapedial tendon reflexes.

The presence of a cochlear cleft—mimicking an otosclerotic focus on CT scan—should be considered in the differential diagnosis when audiometric findings do not agree or other radiological manifestations potentially indicate a conductive hearing loss of inner ear origin.

A cochlear cleft is a small, nonosseous space occupying a short segment of a spiral plane in the middle of the otic capsule, parallel to the basal turn of the cochlea. The cochlear cleft is a radiological finding and is frequently observed (in up to 41%) in magnified temporal bone CT images in children. It is found less often with advancing age (4,5).

What needs to be emphasized though is that in many of the otosclerosis cases no lucency of the fissula ante fenestram or any other findings consistent with otosclerosis are found (6). This is particularly the case in early stages of otosclerosis, although stapedial tendon reflexes are absent and middle ear inspection would confirm stapes fixation. It should also be highlighted that (in contrast to what Table 2 in the article by Yong et al. might suggest) there is a considerable variation in SCDS symptomatology, ranging from asymptomatic patients over patients only presenting with vestibular or auditory symptoms, and the full-blown clinical picture where both are reported (7,8). In fact, the absence of “pathognomonic” conductive hyperacusis, sound-induced nystagmus or vertigo or pressure-induced nystagmus or vertigo does not preclude the diagnosis of SCDS (9).

The take-home message for the clinician is to be aware of the variation in symptomatology of SCDS, the importance of stapedial tendon reflexes and vestibular-evoked myogenic potentials in discriminating both disorders more easily, needs to be able to interpret the CT and/or magnetic resonance imaging and be aware of its limitations with regards to both SCDS and otosclerosis, and be able to communicate these considerations to the patient to provide proper counselling.
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RESPONSE TO WEAVER TS, SHAYMAN CS, HULLER TE. THE EFFECT OF HEARING AIDS AND COCHLEAR IMPLANTS ON BALANCE DURING GAIT. OTOLOGY & NEUROTOL
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To the Editor: In the article “The effect of hearing aids and cochlear implants on balance during gait” reported in 2017 by Weaver et al. (1) the authors did not find an overall effect of the use of hearing aids or cochlear implants on spatio-temporal parameters of gait nor on gait variability expressed as coefficients of variation. Nevertheless, the authors themselves stress the considerable variation among participants and highlight that some individuals might indeed benefit from sound as an additional source of sensory input to supplement balance. However, their study did not give insight regarding which individuals might present with this benefit.

Recently, we performed a study (2) with a research hypothesis very similar to that from Weaver et al. (1); adults wearing a cochlear implant (CI) and presenting with bilateral caloric areflexia walked overground at self-selected speed in three different conditions: with CI turned on and no additional sound source, with CI turned on and an additional sound source, and with CI turned off. Gait parameters significantly improved in the condition with sound compared with walking with the CI turned on but with no additional sound source. By increasing the motion of the pelvis, the knee, and the ankle, stride length significantly increased (mean difference 7.8 ± 1.2 cm) while stride time decreased (mean difference 0.059 ± 0.016 s).

At first sight, these results seem contradictory to the findings from Weaver et al. (1). However, some individuals in their study also improved their gait pattern in the condition where auditory feedback was available. Surprisingly, four out of five of the individuals that did improve their gait pattern belong to the cochlear implant group and thus represent the individuals in our sample. An explanatory hypothesis related to cochlear implantation is the increased possibility for vestibular compensation through spread of current from the CI to the vestibular system. Parkes et al. (3) showed that electrical stimulation from a CI could elicit vestibular evoked myogenic potentials responses.

Furthermore, all participants in our study had confirmed vestibular loss by bilateral areflexia on caloric testing. Weaver et al. (1) used 30 seconds standing Romberg test on solid surface with eyes closed to assess normality of vestibular function. However, several studies indicate this test is not sensitive enough to detect loss of balance or vestibular function in the age range (age 19–95) under investigation (4,5). It is, therefore, possible that some of his participants did show some degree of vestibulopathy. It cannot be ruled out that these were the top performers that showed most benefit from the auditory feedback.

Finally, attention should be paid to the sound source under consideration. Weaver used broadband white noise (0–4 kHz) combined with the sound of natural rain whereas in our study music was played at a comfortable sound level. Music inherently possesses a rhythm component that might serve as a cue to adjust both rhythm (stride time) and pace (stride length) of gait. These beneficial effects of cues are lacking when using broadband white noise.

In conclusion, while one study on its own cannot give insight regarding which individuals can benefit from sound as an additional source of balance related feedback, a combined look at the studies from Weaver et al. and our study should give a better understanding of how to approach auditory stimulation in the future.