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Case Report

Electrocochleography summating potential seen on auditory brainstem response in a case of superior semicircular canal dehiscence

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Abstract

Background: Superior canal dehiscence syndrome (SCDS) is a condition in which an abnormal communication between the superior semicircular canal and the middle cranial fossa causes patients to hear internal noises transmitted loudly to their affected ear as well as to experience vertigo with pressure changes or loud sounds. Patients with SCDS can have an elevated ratio of summating potential (SP) to action potential (AP) as measured by electrocochleography (ECochG). Changes in this ratio have been observed during surgical intervention to correct this abnormal communication.

Case Description: We present a case of SCDS along with history, physical examination, vestibular function testing, and computed tomography imaging. Due to the disabling symptoms, the patient elected to undergo surgery for plugging of the superior semicircular canal by middle cranial fossa approach. Simultaneous intraoperative ECochG and auditory brainstem response (ABR) were performed. Changes in SP/AP ratio, SP amplitude, and ABR wave I latency were observed during surgery, with a large ECochG SP amplitude generating a new wave, identifiable on the ABR and preceding the traditional wave I. The patient's symptoms resolved after surgery, and no long-term detriment to hearing was observed.

Conclusions: This case demonstrates the intraoperative changes in ECochG during surgery for repair of a SCDS. The substantial intraoperative changes in the summating potential can create a novel wave on intraoperative ABR.

Key Words: Electrocochleography, auditory brainstem response, superior canal dehiscence syndrome

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INTRODUCTION

Superior canal dehiscence syndrome (SCDS) is a clinical syndrome described by Minor *et al.* in 1998, in which a hole in the bone overlying the superior semicircular canal causes a collection of symptoms including hearing the body's internal noises transmitted loudly (i.e., autophony), hearing one's own pulse (i.e., pulse-synchronous tinnitus), and sound or pressure-induced vertigo and nystagmus.[11,12] Physiologic findings include the presence of bone

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conduction hyperacusis on pure tone audiometry, decreased thresholds in response cervical vestibular-evoked myogenic potential (VEMP), and elevated ocular VEMP amplitudes. The collection of symptoms and physiological findings is attributed to abnormal inner ear physiology induced by the presence of a "third mobile window" on the inner ear—in addition to the oval and round windows.[16] SCDS is diagnosed by the presence of a dehiscence on computed tomography (CT) imaging, clinical symptoms, and at least one physiologic test consistent with the condition. Many patients with SCDS are content simply to have an explanation for their unusual symptoms; however, some have significant functional impairment. To address the underlying pathophysiology, a surgery was developed to plug and resurface the affected superior semicircular canal via the middle cranial fossa approach. Patients undergoing the surgery report postoperative improvement in both autophony and dizziness symptoms,[6,7] however, patients also experience expected deficits in the function of the plugged canal,[5] and may experience a mild sensorineural hearing loss.[19]

Electrocochleography (ECochG) is an electrophysiologic test used for the diagnosis of endolymphatic hydrops, especially as seen in Ménière's disease.[8,9,15,17] Patients with SCDS have recently also been found to have an elevated summating potential to action potential (SP/AP) ratio, as measured by ECochG,[2] and this abnormal ratio appears to often correct after surgery for plugging the dehiscent semicircular canal.[1,14,22] Intraoperative monitoring procedures such as ECochG or auditory brainstem responses (ABR) are commonly used during vestibular schwannoma surgery. We have been recording ECochG and ABR to assess for intraoperative changes in inner ear physiology during plugging of the superior semicircular canal.[22] We present a case of SCDS in which the intraoperative summating potential becomes so large that it forms a new wave detectable on the ABR.

CASE REPORT

A 49-year-old woman presented with the chief complaint of "I can hear my eyes move." Her symptoms included chronic dizziness, sensitivity to loud sounds, and autophony. Approximately 10 years prior to presentation, she began noticing that she would feel vertigo when near sirens, during applause in stadiums, or when the radio volume was high while driving in the car. She would experience momentary dizziness with coughing or straining, and could hear her heartbeat constantly in her left ear. In addition to hearing her eyes move, she heard her back cracking in her left ear and each swallow transmitted loudly to that ear. On physical examination, her ears appeared normal by otoscopy. She had normal vestibulo-ocular reflex function to rapid head impulses in the planes of each semicircular canal. Dix–Hallpike positioning maneuvers did not elicit any positional nystagmus or vertigo. Using infrared goggles, however, vertical nystagmus was seen in the plane of the left superior semicircular canal when 500 Hz and 750 Hz tones were applied to the left ear at 110 dB hearing level (HL). Simultaneous head movements in the plane of the left superior semicircular canal were also observed with application of these tones.

An audiogram showed normal hearing bilaterally, with a 20 dB gap between air conduction and bone conduction thresholds at 250 Hz in the left ear and a bone-conduction threshold of −10 dB HL indicating bone-conduction hyperacusis [Figure 1a]. Ocular VEMPs were performed and showed abnormally large amplitudes in response to 500 Hz tone bursts applied to the left ear (70.9 μV, Figure 1b), suggesting a diagnosis of SCDS.[23] Cervical VEMPs in response to click stimuli revealed a reduced threshold of 70 dB nHL in the left ear and a normal threshold in the right ear (90 dB nHL). CT scan of the temporal bone was performed and demonstrated a dehiscence of the left superior semicircular canal [Figure 2]. She elected to proceed with surgical plugging and resurfacing of the affected canal by the middle cranial fossa approach.

During surgery, ECochG and ABR were simultaneously recorded, and are shown in Figures 3 and 4. A large elevation of SP resulted in an SP/AP ratio >1 [Figure 3a], which persisted throughout the case as shown by the stacked traces and the post-plugging result [Figures 3b, c and e]. This is in contrast to the conventional SP/AP ratio on the contralateral side [Figure 3d]. Note the typical wave I of the ABR for the contralateral side [Figure 4a] but the appearance of a separate wave preceding wave I on the affected side, which appears to represent the large SP. The elevated baseline SP and wave I of the ABR increased further in amplitude during the case, and remained elevated at the conclusion of surgery.

She recovered well from surgery, with complete resolution of her symptoms of autophony and sound or pressure-induced vertigo. As expected, she transiently had oscillopsia with head movements in the plane of the superior semicircular canal that symptomatically improved after vestibular physical therapy. Follow-up audiogram 3 months after surgery demonstrated resolution of the air-bone gap and bone conduction hyperacusis, with no sensorineural hearing loss [Figure 1c]. Similarly, the elevated ocular VEMP amplitude was normal after surgery [Figure 1d].

METHODS

Preoperative evaluation included high resolution CT imaging, ocular VEMP responses to 500 Hz tone burst stimuli and cervical VEMP in response to clicks.[13] During

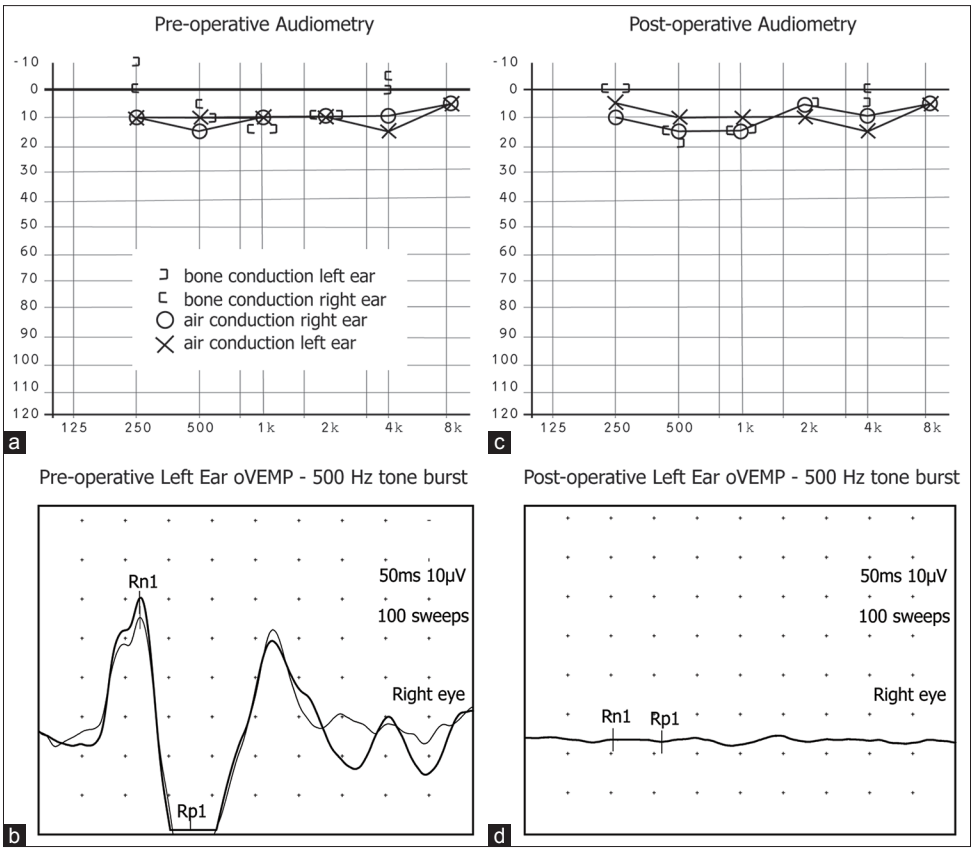

Figure 1: Preoperative audiometry and ocular VEMP data (a and b) demonstrated a low-frequency air-bone gap for the symptomatic left ear, as well as elevated ocular VEMP amplitudes in the contralateral eye in response to 500 Hz tone bursts in the symptomatic ear. Three months after surgery (c and d) the low-frequency air-bone gap decreases, without any sensorineural hearing loss, and the ocular VEMP amplitudes normalize, both consistent with the patient's resolution of symptoms

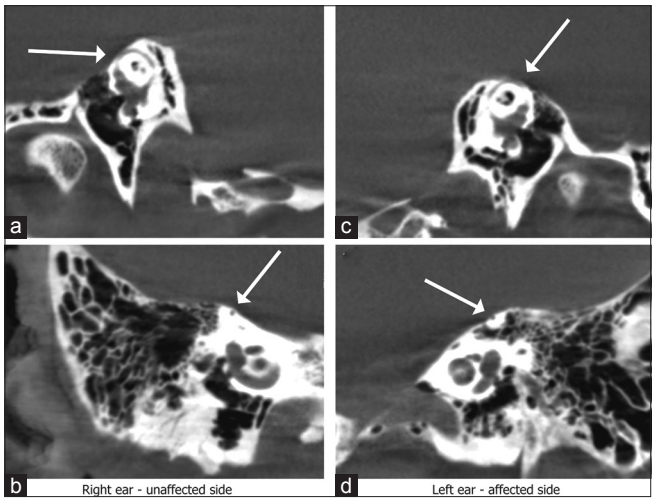

Figure 2: High-resolution computed tomography (CT) demonstrated a thin but intact covering of bone over the right superior semicircular canal on reformation in the plane of the affected canal (a) and orthogonal to that plane (b). A dehiscence was seen affecting the superior semicircular canal on the left side (c and d)

surgery, ECochG and ABR (EndeavorTm CR IOM system Viasys Healthcare, Dublin, OH) were simultaneously recorded. Following induction of general anesthesia, earphones with gold foil coating (Nicolet -Viasys Electrode - Gold Tiptrode 10 and 13mm Nicolet-Viasys Healthcare, Dublin, OH) were inserted and coupled to the audiometric transducers. Conductive gel was applied directly to the posterior aspect of the tympanic membrane under the operating microscope to provide a continuous line of this conductive gel along the canal to the gold foil coating. Before incision, baseline recordings for both monitoring procedures were obtained. For ECochG unfiltered clicks of 100 µs duration were presented at an intensity of 85 dB nHL. Two replications of averaged responses elicited by 1500 clicks presented at a rate of 11.7 per second were obtained. For ABR clicks of 100 µs duration with an intensity of 100 dB nHL at a rate of 11.9 Hz during two cycles of 1000 stimuli were delivered. Responses for both techniques were band pass filtered (ECochG 20–1500 Hz/ABR 1–3000 Hz) and averaged. The contralateral ear was stimulated with white noise at an intensity of 60 dB nHL for both procedures. For ECochG the summating potential to action potential (SP/AP) ratio was calculated. SP/AP ratio greater than 0.4 was defined as abnormal for purposes of this study, based on commonly used standards for clinical testing.[10]

Intraoperative image guidance was used to localize the dehiscent canal. To obliterate the lumen of the canal, the

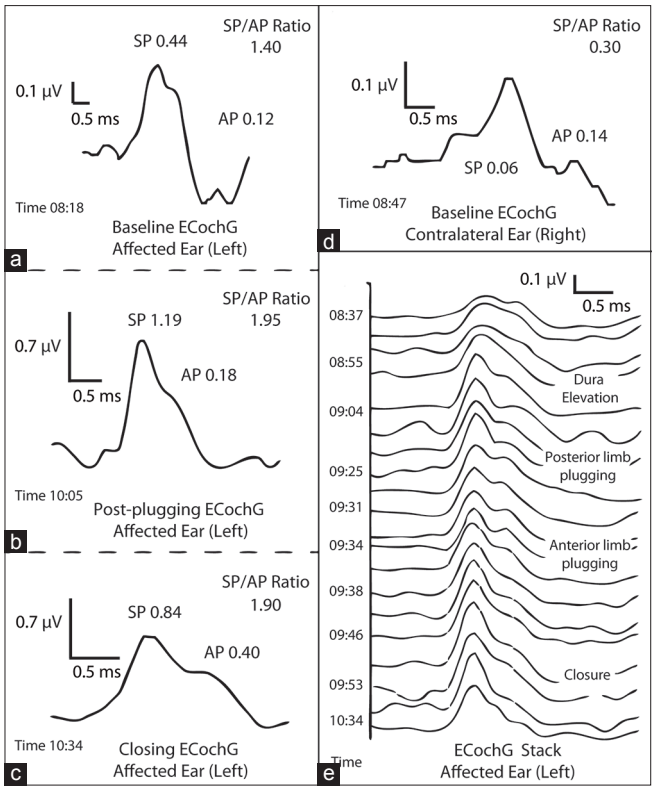

Figure 3: Intraoperative ECochG revealed significantly elevated SP/AP ratio that did not decrease after plugging of the affected superior semicircular canal. In fact, the elevated SP/AP ratio further increased during surgery. (a) Baseline ECochG prior to plugging, followed by (b) ECochG immediately after plugging the canal and (c) at the completion of surgery (each window is labeled with time of day). (d) Contralateral ear demonstrates a normal (<0.4) SP/AP ratio. (e) ECochG tracings stacked over an approximately 2-hour intraoperative time interval. Time of day and notable intraoperative events are labeled

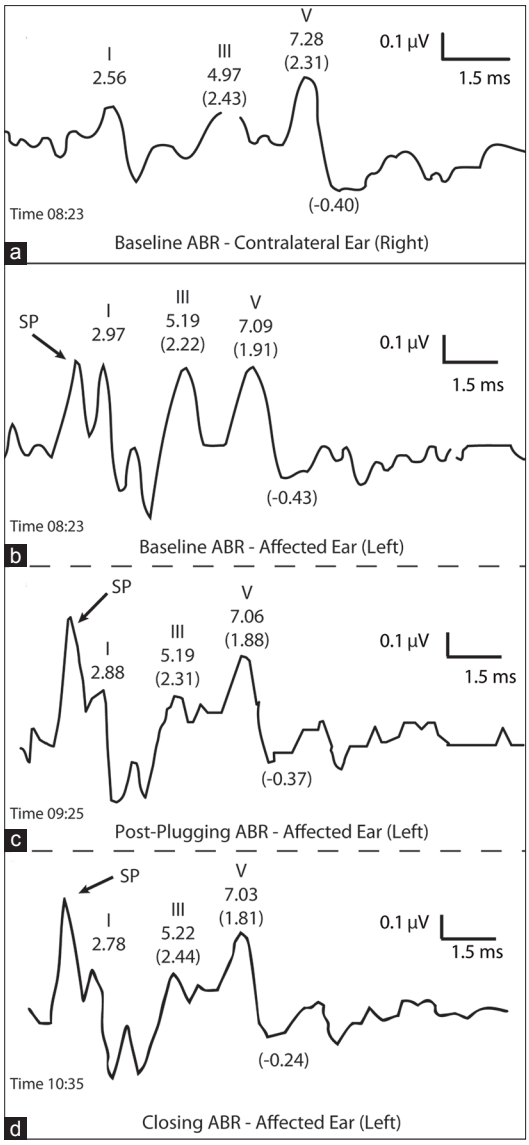

Figure 4: Intraoperative ABR is normal for the contralateral ear (a) and shows the appearance of a separate wave prior to wave I, and corresponding to the ECochG summating potential in the affected ear (b). Similar to the SP on ECochG, the new wave becomes elevated during the case and remains elevated at the conclusion of surgery (c and d, each window is labeled with time of day)

lumen of the dehiscent canal was plugged for 2–3 mm beyond either end of the dehiscent opening using fascia strips, bone dust, and bone chips. Care was taken not to induce unnecessary force or suction near the membranous labyrinth. The area was covered with hydroxyapatite cement, followed by a layer of fascia and fibrin glue.

This study was a review of existing clinical data with patient identifiers removed. It qualified for exemption from an institutional review board protocol on the basis of the United States Department of Health and Human Services criteria 45 CFR 46.101(b4). This exemption was approved by the Johns Hopkins Institutional Review Board.

DISCUSSION

ECochG as a diagnostic and intraoperative adjunct has been reported in cases of SCDS.[2,20] Intraoperative correction of an elevated SP/AP ratio has also been described.[1,22] The clinical implications of these intraoperative changes in SP/AP ratio are unclear, however, they may reflect real-time changes in inner ear physiology as we discuss in a separate manuscript describing the group outcomes from these patients.[22] Here, we focus on one curious observation that we occasionally noted in the ABR data, namely, that large summating potentials seen in ECochG could in some cases be appreciated in ABR recordings as well. In other cases, the ABR did not demonstrate this dramatic separation between SP and wave I. Rather, a fusion of them appeared to move the latency of wave I of the ABR to smaller values. The case presented here is one example in which a large intraoperative summating potential on ECochG caused a distinct, new wave on the ABR recording, preceding the usual first wave on the ABR. Interestingly, despite this unusual intraoperative finding,

this patient had an uneventful postoperative course, with complete resolution of her symptoms and no unexpected sequelae. Had this separate wave representing the SP of ECochG – or a fusion product of it and wave I – been mistaken for wave I itself, a reduction of this wave's amplitude might have been reported to the surgeons as evidence of compromise of inner ear function. On the contrary, the reduction of SP encountered in several other cases is not necessarily an indicator of inner ear damage but likely represents the salient effects of closure of the labyrinthine third window.[22]

ECochG has been historically a test used for the assessment of endolymphatic hydrops, associated with Ménière's disease.[8-10,17] Only recently has ECochG been studied in cases of SCDS. The SP/AP ratio is commonly elevated, and often decreases after surgical plugging of the superior semicircular canal.[22] Other physiologic abnormalities associated with SCDS are corrected after surgical plugging, including the elevated low-frequency air-bone gaps identified on pure tone audiometry,[19] decreased cervical VEMP thresholds,[21] and elevated ocular VEMP amplitudes. Each of these findings is attributed to abnormal inner ear physiology reflecting the presence of a third mobile window on the inner ear, in addition to the oval and round windows. The plugging of the extra or "third" mobile window, restores normal inner ear physiology, and is shown in the normalization of these tests. Elevations in the SP/AP ratio seen here and in other cases of SCDS likely also reflect abnormal physiology caused by the third mobile window. How the presence of a dehiscence leads to elevations in summating potential is unclear. One potential explanation is that the opening into the middle cranial fossa creates a pressure differential between endolymphatic and perilymphatic compartments of the inner ear, such that a hydrops *ex vacuo* due to lower pressure perilymphatic compartment leads to biasing of the basilar membrane and increased summating potential.[2] An alternative theory is that sound-sensitive hair cells in the vestibular system contribute to the summating potential, similar to the increased vestibular sensitivity to sound observed in the abnormal cervical and ocular VEMP responses in patients with SCDS.[20] Finally, a recent theory has proposed that patients with SCDS may have endolymphatic hydrops,[18] although the pathophysiology of this is unexplained. It is curious that the SP/AP ratio was noted to rise during surgery, prior to manipulating the labyrinth, as shown in the ECochG stack traces here. During exposure of the superior semicircular canal by middle cranial fossa approach, the dura is retracted away from the labyrinth, with more retraction required as the superior semicircular canal is approached. This finding could reflect elevations of intracranial pressure transmitted to the labyrinth, as has been proposed by Büki *et al.* in measuring otoacoustic emissions, another measure that like ECochG reflects hair cell function.[3,4] The use of ECochG in cases of SCDS may provide insight into the origin of the summating potential, leading to new applications of this electrophysiologic measure.

CONCLUSION

We present a case of superior semicircular canal dehiscence syndrome during which intraoperative ECochG recordings showed an SP/AP ratio >1, and a summating potential large enough to be identified as a new wave on the ABR. While the clinical consequences of a large spike in the summating potential is unknown, this finding likely reflects a change in inner ear physiology and may lead to new understandings of origin of the ECochG summating potential.

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Conflicts of interest
There are no conflicts of interest.

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