Published in final edited form as:

Otol Neurotol. 2015 January ; 36(1): 139–145. doi:10.1097/MAO.0000000000000642.

Intraoperative Neuromonitoring for Superior Semicircular Canal Dehiscence and Hearing Outcomes

Angela Wenzel^{*}, Bryan K. Ward^{*}, Eva K. Ritzl[†], Sergio Gutierrez-Hernandez[†], Charles C. Della Santina^{*}, Lloyd B. Minor[‡], and John P. Carey^{*}

^{*}Departments of Otolaryngology–Head and Neck Surgery [†]Neurology, Johns Hopkins School of Medicine, Baltimore, Maryland [‡]Stanford University School of Medicine, Stanford, California, U.S.A

Abstract

Background—Recent findings in patients with superior semicircular canal dehiscence (SCD) have shown an elevated ratio of summing potential (SP) to action potential (AP), as measured by electrocochleography (ECoChG). Changes in this ratio can be seen during surgical intervention. The objective of this study was to evaluate the utility of intraoperative ECoChG and auditory brainstem response (ABR) as predictive tools for postoperative hearing outcomes after surgical plugging via middle cranial fossa approach for SCD syndrome (SCDS).

Methods—This was a review of 34 cases (33 patients) in which reproducible intraoperative ECoChG recordings were obtained during surgery. Diagnosis of SCDS was based on history, physical examination, vestibular function testing, and computed tomography imaging. Simultaneous intraoperative ECoChG and ABR were performed. Pure-tone audiometry was performed preoperatively and at least 1 month postoperatively, and air-bone gap (ABG) was calculated. Changes in SP/AP ratio, SP amplitude, and ABR wave I latency were compared with changes in pure-tone average and ABG before and after surgery.

Results—Median SP/AP ratio of affected ears was 0.62 (interquartile range [IQR], 0.45–0.74) and decreased immediately after surgical plugging of the affected canal to 0.42 (IQR, 0.29–0.52; $p < 0.01$). Contralateral SP/AP ratio before plugging was 0.33 (IQR, 0.25–0.42) and remained unchanged at the conclusion of surgery (0.30; IQR, 0.25–0.35; $p = 0.32$). Intraoperative changes in ABR wave I latency and SP amplitude did not predict changes in pure-tone average or ABG after surgery ($p > 0.05$).

Conclusion—This study confirmed the presence of an elevated SP/AP ratio in ears with SCDS. The SP/AP ratio commonly decreases during plugging. However, an intraoperative decrease in SP/AP does not appear to be sensitive to either the beneficial decrease in ABGs or the mild high-frequency sensory loss that can occur in patients undergoing surgical plugging of the superior

Address correspondence and reprint requests to John P. Carey, M.D., Johns Hopkins Outpatient Center, 6th Floor, Department of Otolaryngology–Head and Neck Surgery, 601 North Caroline St, Baltimore, MD 21287, U.S.A.; jcarey@jhmi.edu.

The authors disclose no conflicts of interest.

semicircular canal. Future work will determine the value of intraoperative ECochG in predicting changes in vestibular function.

Keywords

ABR; Electrocochleography; Superior canal dehiscence syndrome

Superior semicircular canal dehiscence syndrome (SCDS) is a clinical syndrome described in 1998 in which a hole in the bone overlying the superior semicircular canal causes manifold symptoms, including autophony, pulsatile tinnitus, bone-conduction hyperacusis, and sound- or pressure-evoked vertigo and nystagmus (1,2).

One of the most studied surgical methods used for repair of the dehiscence canal is plugging via the middle cranial fossa approach. Individuals who have undergone this surgery report improvements in both autophony and dizziness handicap inventory scores (3,4). However, a mild high-frequency sensorineural hearing loss persists in up to 25% of patients, with a few patients having more severe losses (5–7). Variations on this surgical technique include resurfacing the canal or approaching it via the mastoid cavity; however, postoperative outcomes from these approaches are less studied (8–10).

Currently, it is unknown if there are any intraoperative predictors of postoperative hearing in patients undergoing this surgery. Historically, electrocochleography (ECochG) has been well studied as an electrophysiologic test for presumed endolymphatic hydrops (11–14). Patients with SCDS have also been shown to have an elevated summing potential to action potential (SP/AP) ratio, as measured by ECochG (15), and this finding appears to correct after surgical plugging of the dehiscence semicircular canal (16). ECochG and auditory brainstem reflexes (ABRs) have been monitored during some neurotologic procedures like cerebellopontine angle tumor surgery (17–19) as prognostic tools for postsurgical hearing outcomes (20–22). Their prognostic role in surgical repair of SCDS is unclear.

The purpose of this study was to investigate whether intraoperative changes in ECochG and ABR are associated with postsurgical hearing outcomes in patients undergoing repair of SCD.

Materials and Methods

Study Population

Clinical data from patients undergoing surgical repair of SCD via the middle fossa approach from July 2009 until October 2013 were reviewed retrospectively. Patients were diagnosed as having SCDS based on history and physical examination and at least one abnormal physiologic test consistent with SCDS, including vestibular evoked myogenic potentials (VEMPs) or sound- and/or pressure-induced nystagmus. Patients must also have had evidence from high-resolution computed tomography of SCD on reformation in the plane of the affected canal and orthogonal to that plane (23,24). To warrant surgical repair, patients must have reported that symptoms of SCDS (autophony, chronic disequilibrium, pulsatile tinnitus, or sound- and pressure-induced vertigo) were disabling. 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 U.S. Department of Health and Human Services criteria 45 CFR 46.101(b4). This exemption was approved by the Johns Hopkins Institutional Review Board.

Surgical Technique

The middle cranial fossa surgical approach was performed in all cases with image guidance (LandMarX surgical navigation system; Medtronic Xomed, Jacksonville, FL, USA) to localize the dehiscence canal, which was plugged using fascia strips, bone dust, and bone chips, as previously described (25,26), then resurfaced with hydroxyapatite cement (Hydroset; Stryker Leibinger GmbH & Co. KG, Freiburg, Germany). Intra-operative neural monitoring was performed for the facial nerve, ABR, ECoG, and somatosensory evoked potentials.

ECoG and ABR

Intraoperative ECoG and ABR (Endeavor CR IOM system Viasys Healthcare, Dublin, OH, USA) were simultaneously recorded binaurally. After general anesthesia induction, insert earphones with gold foil coating (Nicolet-Viasys Electrode-Gold Tiptrode 10 and 13 mm; Nicolet-Viasys Healthcare, Dublin, OH, USA) were inserted and coupled to audiometric transducers. Conduction was further enhanced in the latest recordings by applying conductive gel directly to the posterior tympanic membrane via operating microscope and providing a continuous line of conductive gel along the canal to the gold foil coating. Before incision, baseline recordings for both monitoring procedures were obtained. For ECoG, unfiltered clicks of 100-microsecond duration were presented at 85 dB nHL. Two replications of averaged responses elicited by 1,500 clicks presented at a rate of 11.7 per second were obtained. For ABR, clicks of 100-microsecond duration with an intensity of 100 dB nHL and a rate of 11.9 Hz during two cycles of 1,000 stimuli were delivered. Responses for both techniques were band-pass filtered (ECoG, 20–1,500 Hz; ABR, 1–3,000 Hz) and averaged. While recording, the contralateral ear was stimulated with masking white noise at 60 dB nHL for both procedures. For ECoG, the SP/AP ratio was calculated. An SP/AP ratio greater than 0.4 was defined as abnormal for this study based on commonly used standards for clinical testing (27). For ABR, waves I, III, V, latencies and amplitudes and interwave latencies from I to III and I to V were recorded and calculated.

Audiometry

Audiometric tests were performed at our institution on all patients before surgery and at least 1 month afterward. Pure-tone hearing thresholds were obtained using both air conduction (AC) and bone conduction (BC). A four-frequency pure-tone average (PTA) was calculated across 500, 1,000, 2,000, and 4,000 Hz for AC and BC as well as air-bone gap (ABG) for 250, 500, 1,000, 2,000, and 4,000 Hz preoperatively and post-operatively. Speech discrimination was also tested using the Northwestern University Auditory Test No. 6 word list. Because of the high prevalence of hemotympanum and middle ear effusion during the immediate postoperative period, only changes in BC thresholds were used for analysis.

Data Analysis

Summary statistics were performed for demographics, presenting symptoms, ECoG SP and AP amplitudes, and SP/AP ratio. ABR summary statistics were performed for waves I and III, amplitudes, and latencies, as well as for wave V latency and waves I to III and III to V interwave latencies. For ECoG between-group comparisons, unaffected contralateral ears were used as controls. For ABR, a control group consisted of 22 cases of vestibular schwannoma surgery, using data from the normal ear. Equipment, settings, and recording protocols were unchanged between groups. Mean age of controls (45.9 yr; SD, 9.9 yr) was not different from SCD patients (44.5 yr; SD, 8.8 yr; $p = 0.6$).

Normality of each variable was assessed with the Shapiro-Wilks test, with a value of $p < 0.10$, suggesting a non-normal distribution. ECoG and ABR variables were not normally distributed; so nonparametric tests were performed for between-group differences. Kruskal-Wallis one-way analysis of variance was used to assess differences in ECoG and ABR variables between affected and control ears before and after plugging. Wilcoxon ranked sum tests were then performed for pairwise comparisons if significant. Based on previous observations of intraoperative changes in SP/AP ratio and wave I latency, a priori planned comparisons were performed for between-group differences in SP, AP, SP/AP ratio, and wave I latency for affected ears compared with control ears and for affected ear at end-of-surgery compared with affected ear at start-of-surgery values. Planned comparisons were limited to $k-1$ total comparisons for each Kruskal-Wallis test statistic. Age, PTA, ABG, and changes in PTA met criteria for normal distribution. Paired t tests were therefore performed for PTA and ABG before and after surgery, and between-group t tests were used for comparing age and hearing outcomes across groups. Univariate and multiple linear regression analyses were performed between postoperative PTA and ABG and changes in intraoperative ECoG and ABR values. For all statistical analyses, associations were considered statistically significant for two-sided statistics with a value of $p < 0.05$. SPSS Statistics 17.0 (SPSS Inc., Chicago, IL, USA) was used for all analyses.

Results

Illustrative Case

The following case illustrates the observation that correction of the SP/AP ratio is not necessary to achieve a successful outcome in SCDS surgery. This 35-year-old man had a 5-year history of symptoms in the right ear, including autophony, ability to hear his eye movements, and disequilibrium caused by loud sounds as well as by coughing, straining, or sneezing. When performing a nasal Valsalva maneuver, he had nystagmus characteristic of excitation of the right superior canal, with the eyes moving slowly upward and rolling to his left. In addition, he had a movement of the head in the right SC plane when a loud 500-Hz tone was presented to his right ear.

Audiometry before surgery showed a 40-dB ABG at 250 Hz with -10 dB bone conduction threshold. In addition, smaller ABGs were seen through 1,000 Hz (Fig. 1A). His ocular VEMP amplitude was high (35.6 μ V) for air-conducted sound stimulation of the right ear but was normal at 1.4 μ V for the left ear. Because he found his symptoms debilitating, the

patient chose to have surgical plugging of the dehiscent canal. At the start of surgery, the SP/AP ratio was elevated (0.68) for the affected ear (Fig. 1B), increased abruptly with dural elevation to 2.39 (Fig. 1C), and remained an elevated abnormal ratio of 1.18 after completion of the plugging and resurfacing (Fig. 1D). Nevertheless, when the patient returned 4 months after surgery, he noted resolution of all SCDS symptoms, and postoperative hearing test showed normal thresholds without ABGs (Fig. 1A).

Patient Characteristics

Thirty-four cases (33 patients) of SCDS plugging via the middle cranial fossa approach between July 2009 and October 2013 had reliable intraoperative ECoChG recordings for the affected ear and were included. One subject with bilateral SCDS developed contralateral symptoms after the initial surgery and subsequently underwent contralateral surgical plugging. Both cases were included in the analysis. This represents 53.9% of all performed SCDS surgeries ($n = 63$) at this center during the study period. The remaining 29 cases performed during the study period were excluded because of technical difficulties of the monitoring techniques, including loss of ECoChG signals during surgery attributed to fluid entering the middle ear or displacement of the electrodes, leading to smaller amplitude, less reproducible responses, or to absent responses altogether. Demographic data and presenting symptoms for all included cases are shown in Table 1.

ECoChG Findings

Median intraoperative ECoChG results are summarized in Table 2. Median preplugging SP/AP ratio in affected ears was 0.62 (range, 0.14–1.90), with 26 (76.5%) of affected ears having an abnormally elevated ratio of greater than 0.4 (27). Between-group differences were identified for SP/AP ratio (Kruskal-Wallis test, $p < 0.001$) and AP amplitude (Kruskal-Wallis test, $p = 0.02$) but not SP amplitude (Kruskal-Wallis test, $p = 0.08$). The SP/AP ratio for affected ears significantly decreased to 0.42 (range, 0.12–1.90) during surgery immediately after plugging (Wilcoxon rank-sum test, $p < 0.001$), with 24 (70.6%) of 34 affected ears demonstrating intraoperative decreases in SP/AP ratio, and 15 (44.1%) declining to a ratio less than 0.4. After plugging, SP amplitude decreased significantly (Wilcoxon rank-sum test, $p = 0.03$) and there was a trend toward increasing AP amplitude at the end of surgery (Kruskal-Wallis test, $p = 0.10$). In seven cases (20.6%), elevated preplugging SP/AP ratio increased further during surgery and remained above baseline at conclusion of the case (see Fig. 1 for a sample case).

For the subset of cases in which reliable bilateral ECoChG recordings were obtained ($n = 19$), median SP/AP ratio of contralateral ears before plugging was 0.33 (range, 0.20–0.55) and remained unchanged at the conclusion of surgery (Wilcoxon rank-sum test, $p = 0.32$). Preoperative SP/AP ratio was elevated in affected ears compared with contralateral ears ($p < 0.001$; Table 2). In two patients, the contralateral ear demonstrated an SP/AP ratio greater than 0.4 at the time of surgery. One of these patients had been diagnosed as having bilateral SCD.

ABR Findings

Reproducible ABR recordings were achieved in 19 of the affected ears that also had reproducible ECochG, and these cases were included for study. Median descriptive intraoperative ABR findings for affected ears and for controls (contralateral ears of patients with vestibular schwannoma) are shown in Table 3. Although median values for waves I and III amplitudes and interwave latency of waves I to III in affected ears appear to show a trend toward decreased amplitude and shorter latency at the end of surgery, these changes were not significant. No significant between-group differences were identified for ABR wave latency or amplitude; therefore, no pairwise comparisons were performed.

Audiometric Correlations

Mean preoperative and postoperative hearing outcomes are presented in Table 4. There was a significant increase in four-frequency PTA of affected ears between preoperative and postoperative audiograms for bone conduction (t test, $p < 0.01$) and for air conduction (t test, $p < 0.01$), a finding consistent with our previous report of early hearing changes in SCD surgery (5). Speech discrimination scores of affected ears were unchanged after surgery (t test, $p = 0.09$). Average low-frequency ABG decreased significantly at least 1 month postoperatively (paired t test, $p < 0.001$).

Some subjects demonstrated intraoperative changes in ABR wave I latency or SP amplitude during the canal exposure and plugging steps of the procedure (Fig. 1). As these changes may have reflected real-time changes in physiology, we hypothesized that they might predict hearing outcomes for the operated ear. By univariate analysis, however, intraoperative changes in SP amplitude, AP amplitude, SP/AP ratio, and ABR wave 1 latency were not associated with changes in PTA or ABG at least 1 month after surgery ($p > 0.05$).

Discussion

The role of ECochG as diagnostic and intraoperative adjunct has been reported in cases of SCDS, and intra-operative correction of an elevated SP/AP ratio has also been described (15,16). To our knowledge, this is the first study to examine intraoperative ABR findings in these patients and whether intraoperative changes in ECochG and ABR have functional consequences for patients undergoing repair of SCD. The aim of this study was to evaluate the utility of intraoperative monitoring techniques during superior semicircular canal plugging via the middle cranial fossa approach and to investigate if intraoperative findings might provide useful predictive information about postsurgical hearing outcome.

One of the study's lessons is that intraoperative auditory evoked responses can successfully be performed in many individuals undergoing the middle fossa approach for repair of SCD. However, this is not universally true, and one limiting technical factor is the propensity of fluids to enter the middle ear and dampen sound delivery to the cochlea. SCD is frequently accompanied by dehiscences in the bone of the tegmen mastoideum and tegmen tympani (28), and these serve as conduits for blood and irrigation fluids to enter the middle ear. Further technical difficulties may include electrode displacement from the ear canal, leading to smaller amplitudes, less reproducible responses, or to absent responses altogether. This

resulted in exclusion of nearly half of the performed cases. Indeed, we found that there was a learning curve requiring close cooperation between surgeons and intra-operative monitoring team members. Critical to success of the ECoChG recordings was application of the conductive gel directly to the tympanic membrane by the surgeons before placement of the foil-coated ear canal probes. Electrical coupling of the foil electrodes to the conductive gel on the tympanic membrane was essential for optimizing signal-to-noise ratio, but the gel volume had to be kept minimal so as not to dampen tympanic membrane vibration or occlude the sound delivery tube inside the foam tip of the probe.

Our data corroborate the presence of an elevated SP/AP ratio in a majority (76.5%) of patients with SCD. The median SP/AP ratio of affected ears in this study (0.62) is equivalent to the mean (0.62) reported in the series by Adams et al. (16). Prior reports by this group identified a decline in SP/AP ratio in the majority of patients after plugging. Arts et al. (15) speculated that a dehiscence might cause bias in the basilar membrane, leading to an elevation in the SP. They also identified an increase in compound action potential amplitude after plugging the dehiscent canal in some patients.

These findings may reflect alterations in pressure transmission away from the round window in cases of a dehiscent superior semicircular canal. The third mobile window in the perilymphatic compartment might reduce perilymph pressure (hydrops ex vacuo) instead of raising endolymph pressure, which is what has been proposed as the cause of the elevated SP/AP ratio in patients with Ménière's disease (29,30). The intraoperative changes in SP and AP amplitude may reflect increasing pressure imposed throughout the labyrinth that can occur with canal-plugging procedures. Changes in SP/AP ratio and in SP amplitude might therefore be interpreted as a quantitative indicator of pressure imposed on the labyrinth at the time of surgery. This pressure might also derogate cochlear hair cell function, endocochlear potential, or basilar membrane mechanics.

ABR may also reflect the effects of pressure transmission in the labyrinth as shorter wave latencies at the end of surgery, and changes in wave amplitudes were noted during plugging. It should be kept in mind, however, that ABR is a far-field intraoperative technique that might not detect subtle changes occurring during plugging that near-field ECoChG can measure. In addition, ABR has known technical limitations, especially in vestibular schwannoma surgery (21,31). Electrical noise in the operating room can be considerable, and even small changes in this noise, in the patient's blood pressure, temperature, or anesthesia, or surgical maneuvers may affect ABR amplitudes and latencies. Especially during noisy steps of surgery like the use of electrocautery or drilling, ECoChG has proven to be more useful than ABR because it offers more reliable higher-amplitude potentials (21). Nevertheless, ABR remains a useful monitoring technique, providing additional information about the entire auditory pathway.

Although both inner and outer hair cells have been proposed to contribute to the SP, the precise cause of the SP remains unclear. Recent work on sound sensitivity of saccular and utricular afferents suggests that even these structures may contribute to the SP (32,33). Possible contributions of other labyrinthine hair cells to the SP may help explain the elevated SP/AP ratio in these patients. A third mobile window that leads to pressure

transmission across these structures could result in elevated SP that may decrease after dehiscence repair, analogous to the effects on VEMP responses (34). The influence of these organs on the SP warrants further investigation.

In seven cases, the SP increased abruptly on dura elevation from the dehiscence. This again may reflect a change in perilymphatic pressure because this event opens the perilymphatic compartment to air. We cannot conclude that a correction of an abnormal elevated SP/AP ratio during plugging of the dehiscent canal is important for postoperative hearing outcomes. ECoChG evoked by clicks may simply be insensitive to mild changes in AC thresholds at low frequencies. Fortunately, most patients enjoy relief of their conductive hyperacusis symptoms (e.g., autophony), with the mild changes in AC thresholds needed to reduce or resolve their low-frequency ABGs. Likewise, when sensorineural hearing loss does occur after SCD plugging, the degree is usually mild, and click-evoked ECoChG may be insensitive to this. ECoChG may be useful in predicting individual instances of more severe sensory hearing loss, but these are fortunately rare. Furthermore, there may be value in predicting vestibular end-organ function after surgery, and that will be the subject of future work.

Conclusion

The results of this study confirm that surgical plugging of SCD typically results in a reduction of SP and often results in normalization of the SP/AP ratio. Intraoperative changes in ECoChG and ABR, however, did not predict changes in postoperative hearing, with some patients having large intraoperative increases in SP/AP ratio, yet normal postoperative hearing. Thus, it does not appear that a goal of this surgery should be normalization of the SP/AP ratio per se. Visual confirmation of complete closure of the communication between the intracranial space and the canal lumen should remain the goal of SCD repair. Intraoperative auditory monitoring—particularly ECoChG—appears to provide useful information in real-time about pressure transmission in the labyrinth.

Acknowledgments

The authors thank “Chely” Nirma Carballido Martinez and Clinton Ogega for assistance with acquiring intraoperative electrocochleography and auditory brainstem response data for review.

References

1. Minor LB, Solomon D, Zinreich JS, Zee DS. Sound- and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal. *Arch Otolaryngol Head Neck Surg*. 1998; 124:249–58. [PubMed: 9525507]
2. Minor LB, Cremer PD, Carey JP, Della Santina CC, Streubel SO, Weg N. Symptoms and signs in superior canal dehiscence syndrome. *Ann N Y Acad Sci*. 2001; 942:259–73. [PubMed: 11710468]
3. Crane BT, Minor LB, Carey JP. Superior canal dehiscence plugging reduces dizziness handicap. *Laryngoscope*. 2008; 118:1809–13. [PubMed: 18622314]
4. Crane BT, Lin FR, Minor LB, Carey JP. Improvement in autophony symptoms after superior canal dehiscence repair. *Otol Neurotol*. 2010; 31:140–6. [PubMed: 20050268]
5. Ward BK, Agrawal Y, Nguyen E, et al. Hearing outcomes after surgical plugging of the superior semicircular canal by a middle cranial fossa approach. *Otol Neurotol*. 2012; 33:1386–91. [PubMed: 22935810]

6. Limb CJ, Carey JP, Srireddy S, Minor LB. Auditory function in patients with surgically treated superior semicircular canal dehiscence. *Otology Neurotol.* 2006; 27:969–80.
7. Phillips DJ, Souter MA, Vitkovic J, Briggs RJ. Diagnosis and outcomes of middle cranial fossa repair for patients with superior semicircular canal dehiscence syndrome. *J Clin Neurosci.* 2010; 17:339–41. [PubMed: 20074960]
8. Vlastarakos PV, Proikas K, Tavoulari E, Kikidis D, Maragoudakis P, Nikolopoulos TP. Efficacy assessment and complications of surgical management for superior semicircular canal dehiscence: a meta-analysis of published interventional studies. *Eur Arch Otorhinolaryngol.* 2009; 266:177–86. [PubMed: 18953551]
9. Fiorino F, Barbieri F, Pizzini FB, Beltramello A. A dehiscent superior semicircular canal may be plugged and resurfaced via the transmastoid route. *Otol Neurotol.* 2010; 31:136–9. [PubMed: 19707169]
10. Agrawal SK, Parnes LS. Transmastoid superior semicircular canal occlusion. *Otol Neurotol.* 2008; 29:363–7. [PubMed: 18180691]
11. Sass K. Sensitivity and specificity of transtympanic electrocochleography in Ménière's disease. *Acta Otolaryngol.* 1998; 118:150–6. [PubMed: 9583780]
12. Ferraro J, Best LG, Arenberg IK. The use of electrocochleography in the diagnosis, assessment, and monitoring of endolymphatic hydrops. *Otolaryngol Clin North Am.* 1983; 16:69–82. [PubMed: 6856313]
13. Pou AM, Hirsch BE, Durrant JD, Gold SR, Kamerer DB. The efficacy of tympanic electrocochleography in the diagnosis of endolymphatic hydrops. *Am J Otol.* 1996; 17:607–11. [PubMed: 8841707]
14. Ikino CM, de Almeida ER. Summating potential-action potential waveform amplitude and width in the diagnosis of Ménière's disease. *Laryngoscope.* 2006; 116:1766–9. [PubMed: 17003737]
15. Arts HA, Adams ME, Telian SA, El-Kashlan H, Kileny PR. Reversible electrocochleographic abnormalities in superior canal dehiscence. *Otol Neurotol.* 2009; 30:79–86. [PubMed: 19092559]
16. Adams ME, Kileny PR, Telian SA, et al. Electrocochleography as a diagnostic and intraoperative adjunct in superior semicircular canal dehiscence syndrome. *Otol Neurotol.* 2011; 32:1506–12. [PubMed: 22072263]
17. Gouveris H, Mann W. Association between surgical steps and intraoperative auditory brainstem response and electrocochleography waveforms during hearing preservation vestibular schwannoma surgery. *Eur Arch Otorhinolaryngol.* 2009; 266:225–9. [PubMed: 18553092]
18. Simon MV. Neurophysiologic intraoperative monitoring of the vestibulocochlear nerve. *J Clin Neurophysiol.* 2011; 28:566–81. [PubMed: 22146352]
19. Colletti V, Fiorino FG, Mocella S, Policante Z. ECochG, CNAP and ABR monitoring during vestibular Schwannoma surgery. *Audiology.* 1998; 37:27–37. [PubMed: 9474437]
20. Brackmann DE, Owens RM, Friedman RA, et al. Prognostic factors for hearing preservation in vestibular schwannoma surgery. *Am J Otol.* 2000; 21:417–24. [PubMed: 10821558]
21. Schlake HP, Milewski C, Goldbrunner RH, et al. Combined intra-operative monitoring of hearing by means of auditory brainstem responses (ABR) and transtympanic electrocochleography (ECochG) during surgery of intra- and extrameatal acoustic neurinomas. *Acta Neurochirurgica.* 2001; 143:985–95. [PubMed: 11685605]
22. Nadol JB Jr, Chiong CM, Ojemann RG, et al. Preservation of hearing and facial nerve function in resection of acoustic neuroma. *Laryngoscope.* 1992; 102:1153–8. [PubMed: 1405966]
23. Belden CJ, Weg N, Minor LB, Zinreich SJ. CT evaluation of bone dehiscence of the superior semicircular canal as a cause of sound-and/or pressure-induced vertigo. *Radiology.* 2003; 226:337–43. [PubMed: 12563123]
24. Hirvonen TP, Weg N, Zinreich SJ, Minor LB. High-resolution CT findings suggest a developmental abnormality underlying superior canal dehiscence syndrome. *Acta Otolaryngologica.* 2003; 123:477–81.
25. Minor LB. Clinical manifestations of superior semicircular canal dehiscence. *Laryngoscope.* 2005; 115:1717–27. [PubMed: 16222184]
26. Chien WW, Janky K, Minor LB, Carey JP. Superior canal dehiscence size: multivariate assessment of clinical impact. *Otol Neurotol.* 2012; 33:810–5. [PubMed: 22664896]

27. Margolis RH, Rieks D, Fournier EM, Levine SE. Tympanic electrocochleography for diagnosis of Ménière's disease. *Arch Otolaryngol Head Neck Surg.* 1995; 121:44–55. [PubMed: 7803022]
28. El Hadi T, Sorrentino T, Calmels MN, Fraysse B, Deguine O, Marx M. Spontaneous tegmen defect and semicircular canal dehiscence: same etiopathogenic entity? *Otol Neurotol.* 2012; 33:591–5. [PubMed: 22429945]
29. Ferraro JA, Tibbils RP. SP/AP area ratio in the diagnosis of Ménière's disease. *Am J Audiol.* 1999; 8:21–8. [PubMed: 10499115]
30. Andrews JC, Bohmer A, Hoffman LF. The measurement and manipulation of intralabyrinthine pressure in experimental endolymphatic hydrops. *Laryngoscope.* 1991; 101(6 Pt 1):661–8. [PubMed: 2041448]
31. Youssef AS, Downes AE. Intraoperative neurophysiological monitoring in vestibular schwannoma surgery: advances and clinical implications. *Neurosurg Focus.* 2009; 27:E9. [PubMed: 19795957]
32. Curthoys IS, Vulovic V, Sokolic L, Pogson J, Burgess AM. Irregular primary otolith afferents from the guinea pig utricular and saccular maculae respond to both bone conducted vibration and to air conducted sound. *Brain Res Bull.* 2012; 89:16–21. [PubMed: 22814095]
33. Colebatch JG, Halmagyi GM, Skuse NF. Myogenic potentials generated by a click-evoked vestibulocollic reflex. *J Neurol Neurosurg Psychiatry.* 1994; 57:190–7. [PubMed: 8126503]
34. Welgampola MS, Myrie OA, Minor LB, Carey JP. Vestibular-evoked myogenic potential thresholds normalize on plugging superior canal dehiscence. *Neurology.* 2008; 70:464–72. [PubMed: 18250291]

Fig. 1.

A, Preoperative and postoperative audiograms of an example patient. Air-bone gap closed postoperatively. Intraoperative ECoG recordings are shown in (B–D). B, Baseline recording (C) after plugging (D) closing recording.

Table 1
Characteristics of the study participants

Characteristics	Total N = 34 ears
Sex, female, n (%)	24 (71)
Affected ears, left, n (%)	16 (47)
Patient symptoms, n (%)	
Autophony	31 (91)
Pulsatile tinnitus	28 (82)
Sound-induced vertigo	27 (79)
Pressure-induced vertigo	23 (68)
Fullness, affected ear	17 (50)
Chronic disequilibrium	17 (50)
Conductive hearing loss ^a	28 (82)
Sound/pressure-induced nystagmus	20 (59)
Sound-induced head movements	2 (6)

^aConductive hearing loss defined as at least 15 dB air-bone gap at 250, 500, 1,000, or 2,000 Hz.

Table 2

Intraoperative electrocochleography results

N = 34 ears	SP/AP ratio		SP amplitude (µV)		AP amplitude (µV)	
	Affected side	Contralateral	Affected side	Contralateral	Affected side	Contralateral
Preplugging	0.62 (0.45–0.74)	0.33 (0.25–0.41)	0.24 (0.09–0.34)	0.10 (0.06–0.14)	0.12 (0.06–0.20)	0.28 (0.15–0.34)
Postplugging	0.42 (0.29–0.52) ^a	0.30 (0.25–0.35)	0.12 (0.06–0.22) ^a	0.08 (0.05–0.15)	0.17 (0.09–0.32)	0.20 (0.11–0.29)

Values are medians (interquartile ranges).

^aStatistical significant difference of pairwise comparisons from preplugging to postplugging ($p < 0.05$). Pairwise comparisons were only performed if Kruskal-Wallis test was significant at $p < 0.05$.

SP indicates summing potential; AP, action potential; IQR, interquartile range; µV, microvolts.

Table 3
Intraoperative auditory brainstem response recordings in affected ears of patients with SCD and controls

	Baseline recording		Closing recording	
	SCD (n = 19)	Control (n = 22)	SCD (n = 19)	Control (n = 22)
Wave I latency (ms)	2.47 (2.31–2.81)	2.47 (2.03–2.69)	2.44 (2.09–2.78)	2.47 (2.20–2.69)
Wave III latency (ms)	4.94 (4.75–5.19)	5.00 (4.75–5.31)	4.84 (4.47–5.25)	4.88 (4.63–5.19)
Wave V latency (ms)	7.03 (6.25–7.59)	7.06 (6.81–7.34)	6.75 (6.44–7.41)	7.00 (6.47–7.31)
Interwave latency I–III (ms)	2.31 (2.19–2.53)	2.69 (2.13–3.03)	2.19 (2.06–2.63)	2.38 (2.03–2.78)
Interwave latency III–V (ms)	1.94 (1.44–2.62)	2.06 (1.81–2.38)	1.97 (1.81–2.47)	2.00 (1.69–2.44)
Wave I amplitude (μV)	2.43 (1.57–3.06)	1.83 (1.20–2.67)	2.20 (1.57–3.29)	1.80 (1.25–2.67)
Wave III amplitude (μV)	3.20 (2.00–4.00)	2.75 (1.63–3.57)	2.10 (1.30–3.20)	2.84 (1.75–4.27)

SCD indicates superior canal dehiscence; ms, milliseconds; μV, microvolts.

Table 4
Preoperative and postoperative hearing outcomes for affected and contralateral ears

Outcomes (N = 34 ears)	Affected ear		Contralateral ear	
	Preoperative	Postoperative	Preoperative	Postoperative
Four-frequency PTA, dB HL, bone conduction	9.8 (10.8)	18.0 (14.1) ^a	13.3 (12.1)	11.3 (13.9)
Four-frequency PTA, dB HL, air conduction	18.5 (9.8)	24.2 (15.9) ^a	15.6 (11.8)	15.5 (12.7)
Speech Discrimination Score (%)	98.3 (4.3)	96.8 (4.8)	98.3 (5.3)	99.2 (1.9)
Average low-frequency ABG	13.5 (7.8)	7.2 (7.1) ^a	4.4 (5.3)	6.0 (4.4)

Values are means (SD).

^aStatistical significant difference from preoperative audiometry of affected ear at $p < 0.05$.

ABG indicates air-bone gap (average of 0.25, 0.5, 1, 2 kHz); PTA, pure-tone average (0.5, 1, 2, 4 kHz).