Operative Techniques in Otolaryngology 30 (2019) 217–222

Operative Techniques in Otolaryngology

Transmastoid approach for surgical repair of superior canal dehiscence syndrome

Raphaelle A. Chemtob, MD, Samuel R. Barber, MD, Angela W. Zhu, MPhil, Elliott D. Kozin, MD, Daniel J. Lee, MD, FACS

From the Department of Otolaryngology, Harvard Medical School, Boston, Massachusetts

KEYWORDS
Superior canal dehiscence;
Transmastoid repair;
Middle fossa craniotomy;
Labyrinthotomy;
Third window;
Superior petrosal sinus;
Arcuate eminence

Patients with superior canal dehiscence syndrome may present with a myriad of auditory and/or vestibular complaints. Treatment of superior canal dehiscence syndrome depends on severity of symptoms and impact on quality of life. Surgery is recommended for patients with debilitating auditory and/or vestibular symptoms. The goal of surgery is to create a durable and watertight seal of the bony superior semicircular canal defect, thereby eliminating the "third window". Repair involves either resurfacing and/or plugging of the dehiscent superior canal to eliminate the third window and reduce symptoms. Surgical options include middle fossa craniotomy or transmastoid (TM) approach. The main advantages of a TM approach includes (1) avoidance of a craniotomy, (2) lower risk of CSF leak, and (3) no brain retraction. The TM approach is ideal for superior petrosal sinus superior canal dehiscence cases as the defect is found medial along the skull base and can be isolated indirectly without direct manipulation of the brain and sinus. This chapter discusses the surgical technique of TM approach.

Introduction

Superior canal dehiscence (SCD) was first characterized in 1998 as a defect of the bony covering of the superior semicircular canal (SSC)[1]. This bony defect creates a pathological "third mobile window" in the inner ear.[2-5] Patients with SCD syndrome (SCDS) may present with a myriad of auditory and/or vestibular complaints. Auditory symptoms include aural fullness, pulsatile tinnitus, autophony, hyperacusis, and hypersensitivity to bone-conducted sound, such as extraocular motion or footsteps.[3,6-8] Common vestibular symptoms include sound- (Tullio's phenomenon) and pressure- (Hennebert's sign) induced dizziness or vertigo, as well as nystagmus with pressure of the tragus or pneumotoscopy.[1,6,9,10] Estimates of SCD incidence vary from 0.5% to 1.9% of the population,[11] and over 600 cases of SCDS have been reported.[12]

The etiology of SCD may be congenital, acquired, or a combination of the two.[4,12-16] The most common subtype is a defect or thin bone of the arcuate eminence of the temporal bone overlaying the superior most aspect of the SSC (59% of patients).[17] The tegmen may thin gradually throughout life. SCD has also been associated with a prominent superior petrosal sinus resulting in a bony defect of the medial limb of the SSC proximal to the common crus (4%).[7,17] In rare cases, patients present with both defects (1%). For acquired etiologies, new onset symptoms consistent with SCDS may present after an antecedent event such as head trauma or chronic middle ear disease (Figures 1-4).

Address reprint requests and correspondence: Daniel J. Lee MD, FACS, Otology and Laryngology, Harvard Medical School, Massachusetts Eye and Ear Infirmary, 243 Charles Street, Boston, MA 02114.
E-mail address: daniel_lee@meei.harvard.edu

http://doi.org/10.1016/j.otot.2019.07.007
1043-1810/

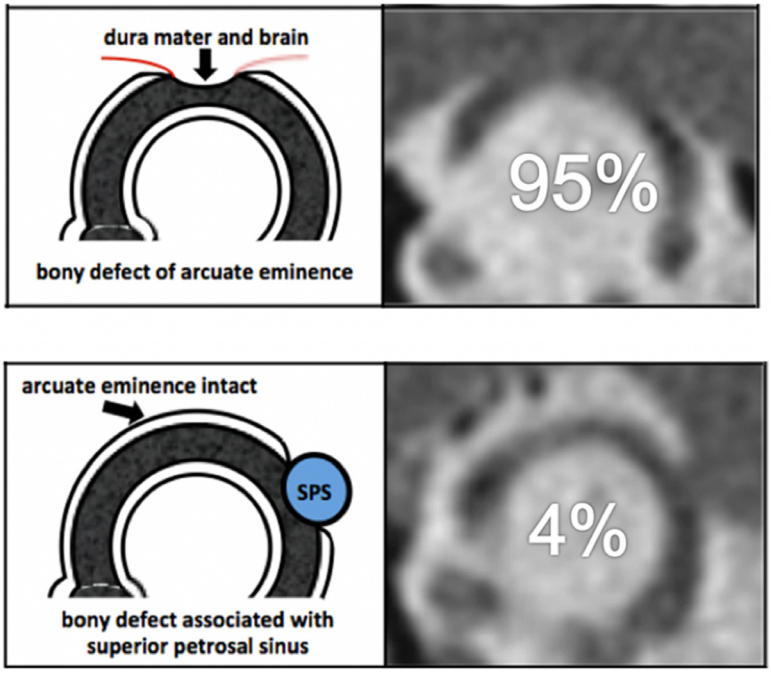

Figure 1 Location of superior canal defect influences surgical approach. The vast majority of superior canal defects are at the arcuate eminence and may readily be addressed via a middle fossa approach. Defects associate with a superior petrosal sinus (or arcuate eminence defects that fall along a severely downsloping skull base) may be better addressed by a transmastoid approach. Figure modified with permission from Lookabaugh et al.[17]

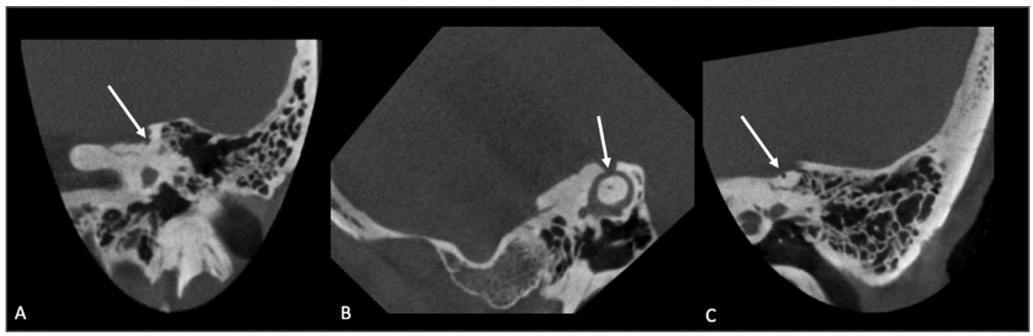

Figure 2 Superior canal dehiscence defect necessitating a transmastoid approach. (A) Coronal view, (B) Poschl view, and (C) Stenvers view. Arrow illustrates defect of the superior canal associated with the superior petrosal sinus.

Diagnostic testing and preoperative assessment

Audiologic workup will demonstrate findings that distinguish SCDS from other otologic conditions. First, pure tone audiometry will show a low frequency, conductive hearing loss on the affected side with intact acoustic reflexes.[6,11,18-20] In some cases, supranormal bone conduction is observed at −5 to −10 dB.[2,19,21,22] Secondly, cervical vestibular evoked myogenic potentials elicit responses at lower thresholds, resulting in higher amplitude cervical vestibular evoked myogenic potentials with reduced stimulation parameters.[8,23-27]

High resolution computed tomography imaging of the temporal bone requires specific parameters to identify thin or dehiscent tegmen in the area over the SSC. Image series collected in the axial plane should have thin slices with ideally 0.625 mm collimation to determine the width and length of a defect and differentiating near-dehiscence (thin bone) from a frank dehiscence when using mutiplanar reformatted views.[4,9,17,28-32] These reformatted image series include coronal, sagittal, as well as both Stenver (an oblique coronal reconstruction parallel to the petrous temporal bone, and perpendicular to the plane of the SCC), and Pöschl views (an oblique coronal reconstruction perpendicular to the petrous temporal bone, and parallel to the SSC). Radiologic classification of SCD position along the arc of the SSC and relative to the position of the superior petrosal sinus, surrounding skull base topography, and a careful assessment of tegmen and facial nerve status are crucial steps in the preoperative assessment. All patients who are surgical candidates for SCD should also undergo

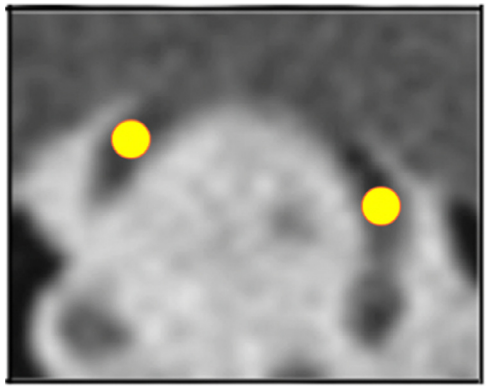

Figure 3 Pöschl view of superior canal defect. Yellow circles illustrate desired area of plugging. Care is taken not to drill too closely to the ampulla or the common crus to prevent inadvertent injury to the sensory neuroepithelium or posterior canal, respectively. Plugging may be performed by bone wax, fascia, and/or bone pâté. (Color version of figure is available online.)

high resolution MRI to rule out any associated encephaloceles, vascular anomalies, or neoplastic processes (such as meningioma) along the lateral skull base.

The diagnosis of SCDS is currently based on clinical history, a comprehensive physical examination, the presence of a confirmed thin or dehiscent tegmen over the superior canal found on imaging, and findings on audiologic workup. Novel screening tools such as power reflectance (wide band acoustic immittance testing) and laser Doppler vibrometry may be useful in the future to improve the diagnostic yield in patients in whom SCD is suspected without the need for imaging.[33-36]

Management of superior canal dehiscence syndrome

Treatment of SCDS depends on severity of symptoms and impact on quality of life. Treatment is either conservative with interval audiometric testing and avoidance of provocative stimuli or surgical repair. Patients with minimal symptoms may be followed with conservative management. Surgery is typically recommended for patients with debilitating auditory and/or vestibular symptoms impacting quality of life. The goal of surgery is to create a durable and watertight seal of the bony SCC defect, thereby eliminating the "third window". Repair involves either resurfacing and/or plugging of the dehiscent superior canal to eliminate the third window and reduce symptoms. A variety of materials are used for the repair including bone wax, fascia, bone pâté, hydroxyapatite cement, or a combination of several materials.[37-42] The 2 most common surgical procedures are via either middle fossa craniotomy or transmastoid approach. The transmastoid approach is the focus of this chapter.

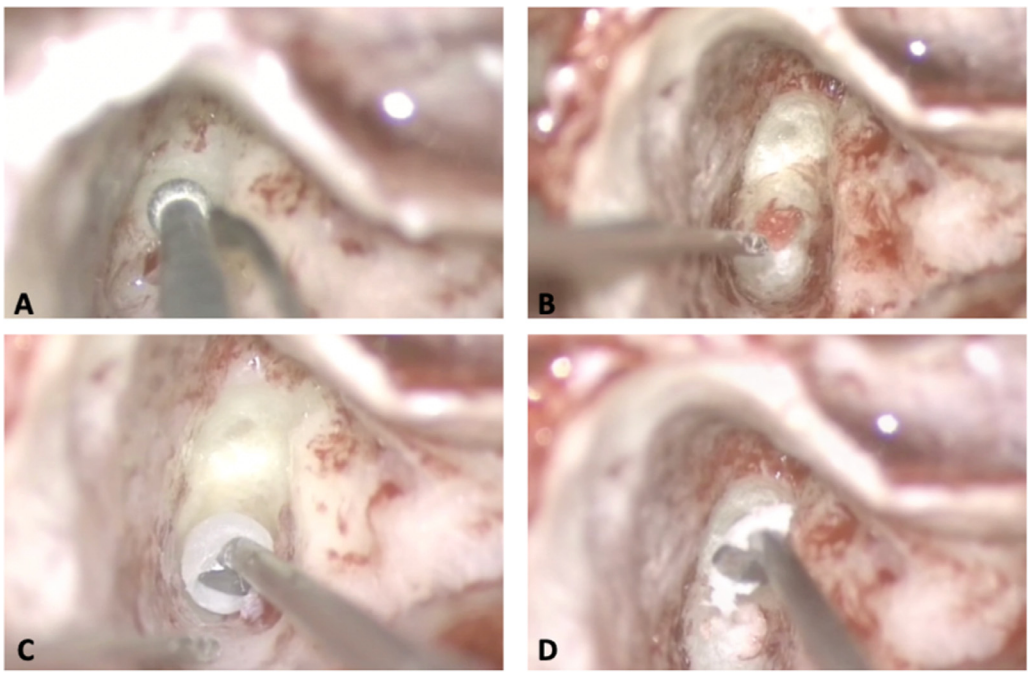

Figure 4 Transmastoid approach to superior canal dehiscence defect. (A) The drill speed is reduced to 6,000 rpm and a 1.5 mm diamond burr is used to blue line and create individual 1.0 mm labyrinthotomies on either side of the defect. Each of these labyrinthotomies are gently occluded with bone wax, allowing for "isolation" of the bony defect, without the need for direct visualization of the dehiscence and avoidance of dissection of the medial skull base and injury to the sinus. Various plugging materials may be utilized, including fascia (B), wax (C), or bone pâté (D). (Color version of figure is available online.)

Rationale for transmastoid repair of SCDS

The transmastoid approach can be used to directly or indirectly isolate a bony defect of the superior canal. The main advantages of a TM approach includes (1) avoidance of a more invasive craniotomy, (2) lower risk of CSF leak, and (3) no brain retraction. *Direct* repair of the SCD is achieved by isolating the superior canal, exposing the tegmen if necessary, drilling the lateral face of the arcuate eminence to better visualize the defect, and then either plugging or "resurfacing" the dehiscence. Based on the MEEI SCD computed tomography classification, we reserve the TM approach for *indirect* repair of medial defects of the superior canal (nonampullated limb) associated with the superior petrosal sinus, arcuate eminence defects that are located along a severely downsloping tegmen that would make even an endoscopic-assisted MFC approach difficult, as well as for revision cases in which a MFC was performed originally. The TM approach is ideal for superior petrosal sinus SCD cases as the defect is found medial along the skull base and can be isolated indirectly without direct manipulation of the brain and sinus. Based on personal experience, this unusual subtype of SCD is not typically associated with a low-lying tegmen (allowing for safe access to the superior canal using a TM approach), unlike the more common arcuate eminence defect.

Detailed description of procedure

The patient is positioned supine with the head rotated laterally away from the surgeon. The anesthesiologist is located toward the foot of the bed on the same side as the surgeon, and the scrub team located opposite the surgeon. The operating microscope base is above the head of the bed. Following intubation, needle electromyographic electrodes are placed into the orbicularis oculi and oris muscle groups for facial nerve monitoring ipsilateral to the side of surgery. A postauricular skin incision is marked approximately 5 mm posterior to the postauricular sulcus, and the patient is prepped and draped in the usual fashion. The postauricular, curvilinear incision is made through skin and areolar fascia. Temporoparietal fascia is divided with a T-shaped incision, followed by dissection of periosteum by lempert elevators. Underlying cortical mastoid bone is exposed using Weitlaner self-retaining retractors. At this time, the operating microscope is brought into the surgical field. Using a drill with a large cutting bur and suction irrigator, a cortical mastoidectomy is performed until the horizontal semicircular canal is identified. Bone situated superior to the horizontal canal and inferior to the tegmen is drilled out using a small diamond bur until exposure of the superior canal is complete.

Repair of the SCD

The drill speed is reduced to 6,000 rpm and a 1.5 mm diamond burr is used to blue line and then create individual 1.0 mm labyrinthotomies on either side of the defect (2 in total), based on imaging (or along either side of the superior petrosal sinus that can be seen during mastoid dissection). Each of these labyrinthotomies is gently occluded with bone wax, allowing for "isolation" of the bony defect, without the need for direct visualization of the dehiscence and avoidance of dissection of the medial skull base and injury to the sinus.

In cases where the dehiscence is superior at the arcuate eminence, it may not be identified. The superior canal is entered using a 1 mm diamond bur on either side of bony defect (toward either the ampullated or nonampullated ends). Care is taken not to drill too closely to the ampulla or to the common crus. Canal plugging is performed with an angled dissector (such as a round knife) at both locations, using the aforementioned materials such as 2 mm spheres of bone wax, bone pâté, or temporalis fascia is placed directly within the lumen of the canal within the opening of the labyrinthotomy. It is very important to avoid over-occlusion of the SSC as repair materials can potentially migrate to the ampulla/vestibule or common crus and be associated with postoperative vestibular hypofunction in addition to loss of SSC function. Additionally, if the dehiscence is located more medially and posteriorly, it may be repaired directly at the site. Resurfacing and/or capping may be performed, in addition to canal plugging. In either scenario, a tight seal of the labyrinthotomy is critical to reduce the risk of perilymphatic fluid loss, which can be associated with sensorineural hearing loss and global vestibular hypofunction postoperatively.[40,43]

Postoperative care

In contrast to a middle fossa craniotomy procedure, patients are expected to have faster recovery with transmastoid SCD repairs, and do not require the same inpatient level of monitoring in the immediate postoperative period. Postoperative steroids are administered variably; however; treatment courses are longer if evidence for neural deficits, such as sensorineural hearing loss, vestibular hypofunction, or facial weakness.[11] These postoperative findings typically resolve quickly and may be attributed to local changes at the surgical site from residual serosanguineous fluid or air.[40] Patients are recommended vestibular therapy in the outpatient setting. Postoperative audiologic testing allows comparison with preoperative data.

Surgical outcomes

The vast majority of patients have improvement from SCD repair. The high rate of success (92%-94%) is associated with low rate of complications that are mostly temporary during the initial recovery period (16%).[12,40,43] Multiple studies have demonstrated no difference in efficacy between MFC and transcanal approaches.[11,22,40,44] Regarding canal plugging vs resurfacing, there is not statistically

significant data from large studies to support a superior method. Studies widely report the subjective improvement of presenting symptoms. From a hearing perspective, patients consistently have decreased low frequency air-bone gaps on postoperative audiograms, which typically improve or normalize after 6 weeks as bone conduction thresholds increase or remain stable, and air conduction thresholds decrease.[6,22,38,40,44,45] Known minor complications include transient sensorineural hearing loss, benign paroxysmal positional vertigo, and vestibular hypofunction, while known major complications include permanent sensorineural hearing loss and facial nerve palsy.[40,42,43,45,46]

Limitations

A few limitations exist with a transmastoid approach for SCD repair. First, anatomic constraints can limit exposure of the superior canal when the tegmen is low. Second, a transmastoid approach can address dehiscence of the SCC, however, a MFC may be selected if multiple tegmen defects are present. Third, prolonged symptoms have been reported in patients with larger bony defects, a history of migraine headaches, and bilateral SCD.[47-49] There exists no superior approach or technique to mitigate long-term vestibular symptoms in those patients.

Disclosure

The authors reported no proprietary or commercial interest in any product mentioned or concept discussed in this article.

References

1. Minor LB, Solomon D, Zinreich JS, et al: Sound- and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal. Arch Otolaryngol Head Neck Surg 124:249–258, 1998.
2. Minor LB, Carey JP, Cremer PD, et al: Dehiscence of bone overlying the superior canal as a cause of apparent conductive hearing loss. Otol Neurotol 24:270–278, 2003.
3. Minor LB, Cremer PD, Carey JP, et al: Symptoms and signs in superior canal dehiscence syndrome. Ann N Y Acad Sci 942:259–273, 2001.
4. Carey JP, Minor LB, Nager GT: Dehiscence or thinning of bone overlying the superior semicircular canal in a temporal bone survey. Arch Otolaryngol Head Neck Surg 126:137–147, 2000.
5. Cheng YS, Kozin ED, Lee DJ: Endoscopic-assisted repair of superior canal dehiscence. Otolaryngol Clin North Am 49:1189–1204, 2016.
6. Brantberg K, Bergenius J, Mendel L, et al: Symptoms, findings and treatment in patients with dehiscence of the superior semicircular canal. Acta Otolaryngol 121:68–75, 2001.
7. McCall AA, McKenna MJ, Merchant SN, et al: Superior canal dehiscence syndrome associated with the superior petrosal sinus in pediatric and adult patients. Otol Neurotol 32:1312–1319, 2011.
8. Mikulec AA, McKenna MJ, Ramsey MJ, et al: Superior semicircular canal dehiscence presenting as conductive hearing loss without vertigo. Otol Neurotol 25:121–129, 2004.
9. Ward BK, Wenzel A, Ritzl EK, et al: Near-dehiscence: Clinical findings in patients with thin bone over the superior semicircular canal. Otol Neurotol 34(8):1421–1428, 2013.
10. Watson SR, Halmagyi GM, Colebatch JG: Vestibular hypersensitivity to sound (Tullio phenomenon): Structural and functional assessment. Neurology 54:722–728, 2000.
11. Bi WL, Brewster R, Poe D, et al: Superior semicircular canal dehiscence syndrome. J Neurosurg 127:1268–1276, 2017.
12. Ward BK, Carey JP, Minor LB: Superior canal dehiscence syndrome: Lessons from the first 20 years. Front Neurol 8:177, 2017.
13. Brandolini C, Modugno GC, Pirodda A: Dehiscence of the superior semicircular canal: A review of the literature on its possible pathogenic explanations. Eur Arch Otorhinolaryngol 271(3):435–437, 2014.
14. Hegemann SC, Carey JP: Is superior canal dehiscence congenital or acquired? A case report and review of the literature. Otolaryngol Clin North Am 44:377–382, 2011 ix.
15. Nadgir RN, Ozonoff A, Devaiah AK, et al: Superior semicircular canal dehiscence: Congenital or acquired condition? AJNR Am J Neuroradiol 32:947–949, 2011.
16. Ogutha J, Page NC, Hullar TE: Postpartum vertigo and superior semicircular canal dehiscence syndrome. Obstet Gynecol 114:434–436, 2009.
17. Lookabaugh S, Kelly HR, Carter MS, et al: Radiologic classification of superior canal dehiscence: Implications for surgical repair. Otol Neurotol 36(1):118–125, 2015.
18. Chi FL, Ren DD, Dai CF: Variety of audiologic manifestations in patients with superior semicircular canal dehiscence. Otol Neurotol 31:2–10, 2010.
19. Merchant SN, Rosowski JJ: Conductive hearing loss caused by third-window lesions of the inner ear. Otol Neurotol 29:282–289, 2008.
20. Zhou G, Gopen Q, Poe DS: Clinical and diagnostic characterization of canal dehiscence syndrome: A great otologic mimicker. Otol Neurotol 28:920–926, 2007.
21. Cox KM, Lee DJ, Carey JP, et al: Dehiscence of bone overlying the superior semicircular canal as a cause of an air-bone gap on audiometry: A case study. Am J Audiol 12:11–16, 2003.
22. Zhang L, Creighton FX Jr, Ward BK, et al: A cohort study of hearing outcomes between middle fossa craniotomy and transmastoid approach for surgical repair of superior semicircular canal dehiscence syndrome. Otol Neurotol 39:e1160–e1167, 2018.
23. Brantberg K, Lofqvist L, Fransson PA: Large vestibular evoked myogenic potentials in response to bone-conducted sounds in patients with superior canal dehiscence syndrome. Audiol Neurootol 9:173–182, 2004.
24. Brantberg K, Verrecchia L: Effectiveness of different click stimuli in diagnosing superior canal dehiscence using cervical vestibular evoked myogenic potentials. Acta Otolaryngol 132:1077–1083, 2012.
25. Milojcic R, Guinan JJ Jr, Rauch SD, et al: Vestibular evoked myogenic potentials in patients with superior semicircular canal dehiscence. Otol Neurotol 34:360–367, 2013.
26. Niesten ME, McKenna MJ, Herrmann BS, et al: Utility of cVEMPs in bilateral superior canal dehiscence syndrome. Laryngoscope 123:226–232, 2013.
27. Zuniga MG, Janky KL, Nguyen KD, et al: Ocular versus cervical VEMPs in the diagnosis of superior semicircular canal dehiscence syndrome. Otol Neurotol 34:121–126, 2013.
28. Belden CJ, Weg N, Minor LB, et al: CT evaluation of bone dehiscence of the superior semicircular canal as a cause of sound- and/or pressure-induced vertigo. Radiology 226:337–343, 2003.
29. Branstetter BFt, Harrigal C, Escott EJ, et al: Superior semicircular canal dehiscence: Oblique reformatted CT images for diagnosis. Radiology 238:938–942, 2006.
30. Ceylan N, Bayraktaroglu S, Alper H, et al: CT imaging of superior semicircular canal dehiscence: Added value of reformatted images. Acta Otolaryngol 130:996–1001, 2010.
31. Isaacson B, Vrabec JT: The radiographic prevalence of geniculate ganglion dehiscence in normal and congenitally thin temporal bones. Otol Neurotol 28:107–110, 2007.

32. Cloutier JF, Belair M, Saliba I: Superior semicircular canal dehiscence: Positive predictive value of high-resolution CT scanning. Eur Arch Otorhinolaryngol 265:1455–1460, 2008.
33. De Greef D, Aernouts J, Aerts J, et al: Viscoelastic properties of the human tympanic membrane studied with stroboscopic holography and finite element modeling. Hear Res 312:69–80, 2014.
34. Dobrev I, Furlong C, Cheng JT, et al: Full-field transient vibrometry of the human tympanic membrane by local phase correlation and high-speed holography. J Biomed Opt 19:96001, 2014.
35. Rosowski JJ, Nakajima HH, Merchant SN: Clinical utility of laser–Doppler vibrometer measurements in live normal and pathologic human ears. Ear Hear 29:3–19, 2008.
36. Rosowski JJ, Songer JE, Nakajima HH, et al: Clinical, experimental, and theoretical investigations of the effect of superior semicircular canal dehiscence on hearing mechanisms. Otol Neurotol 25:323–332, 2004.
37. Agrawal SK, Parnes LS: Transmastoid superior semicircular canal occlusion. Otol Neurotol 29:363–367, 2008.
38. Banakis Hartl RM, Cass SP: Effectiveness of transmastoid plugging for semicircular canal dehiscence syndrome. Otolaryngol Head Neck Surg 158:534–540, 2018.
39. Carey JP, Migliaccio AA, Minor LB: Semicircular canal function before and after surgery for superior canal dehiscence. Otol Neurotol 28:356–364, 2007.
40. Gioacchini FM, Alicandri-Ciufelli M, Kaleci S, et al: Outcomes and complications in superior semicircular canal dehiscence surgery: A systematic review. Laryngoscope 126:1218–1224, 2016.
41. Mikulec AA, Poe DS, McKenna MJ: Operative management of superior semicircular canal dehiscence. Laryngoscope 115:501–507, 2005.
42. Vlastarakos PV, Proikas K, Tavoulari E, et al: Efficacy assessment and complications of surgical management for superior semicircular canal dehiscence: A meta-analysis of published interventional studies. Eur Arch Otorhinolaryngol 266:177–186, 2009.
43. Xie Y, Sharon JD, Pross SE, et al: Surgical complications from superior canal dehiscence syndrome repair: Two decades of experience. Otolaryngol Head Neck Surg 157:273–280, 2017.
44. Alkhafaji MS, Varma S, Pross SE, et al: Long-term patient-reported outcomes after surgery for superior canal dehiscence syndrome. Otol Neurotol 38:1319–1326, 2017.
45. Agrawal Y, Migliaccio AA, Minor LB, et al: Vestibular hypofunction in the initial postoperative period after surgical treatment of superior semicircular canal dehiscence. Otol Neurotol 30:502–506, 2009.
46. Janky KL, Zuniga MG, Carey JP, et al: Balance dysfunction and recovery after surgery for superior canal dehiscence syndrome. Arch Otolaryngol Head Neck Surg 138:723–730, 2012.
47. Niesten ME, McKenna MJ, Grolman W, et al: Clinical factors associated with prolonged recovery after superior canal dehiscence surgery. Otol Neurotol 33:824–831, 2012.
48. Kim N, Steele CR, Puria S: Superior-semicircular-canal dehiscence: Effects of location, shape, and size on sound conduction. Hear Res 301:72–84, 2013.
49. Yuen HW, Boeddinghaus R, Eikelboom RH, et al: The relationship between the air-bone gap and the size of superior semicircular canal dehiscence. Otolaryngol Head Neck Surg 141:689–694, 2009.