Superior semicircular canal dehiscence syndrome

Wenya Linda Bi, MD, PhD,1 Ryan Brewster, BA,1 Dennis Poe, MD,2,3 David Vernick, MD,3 Daniel J. Lee, MD,3–5 C. Eduardo Corrales, MD,3,5 and Ian F. Dunn, MD1

1Center for Skull Base and Pituitary Surgery, Department of Neurosurgery, Brigham and Women’s Hospital; 2Department of Otolaryngology and Communication Enhancement, Boston Children’s Hospital; 3Department of Otolaryngology, Harvard Medical School; 4Department of Otolaryngology, Massachusetts Eye and Ear Infirmary; and 5Division of Otolaryngology, Brigham and Women’s Hospital, Boston, Massachusetts

Superior semicircular canal dehiscence (SSCD) syndrome is an increasingly recognized cause of vestibular and/or auditory symptoms in both adults and children. These symptoms are believed to result from the presence of a pathological mobile “third window” into the labyrinth due to deficiency in the osseous shell, leading to inadvertent hydroacoustic transmissions through the cochlea and labyrinth. The most common bony defect of the superior canal is found over the arcuate eminence, with rare cases involving the posteromedial limb of the superior canal associated with the superior petrosal sinus. Operative intervention is indicated for intractable or debilitating symptoms that persist despite conservative management and vestibular sedation. Surgical repair can be accomplished by reconstruction or plugging of the bony defect or reinforcement of the round window through a variety of operative approaches. The authors review the etiology, pathophysiology, presentation, diagnosis, surgical options, and outcomes in the treatment of this entity, with a focus on potential pitfalls that may be encountered during clinical management.

https://thejns.org/doi/abs/10.3171/2016.9.JNS16503

KEY WORDS superior semicircular canal dehiscence syndrome; superior semicircular canal; hearing loss; dizziness; transtemporal repair; transmastoid approach; skull base

Superior semicircular canal dehiscence syndrome has been an increasingly recognized cause of vestibular and/or auditory symptoms in both adults and children since its initial description less than 2 decades ago. Patients commonly present with vertigo, disequilibrium, conductive hearing loss, pulsatile tinnitus, or aural fullness. Dizziness can be associated with sound-induced (Tullio’s phenomenon) or pressure-induced (Hennebert’s sign) triggers. These symptoms are believed to result from the presence of a pathological mobile “third window” into the labyrinth due to deficiency in the osseous shell, leading to inadvertent hydroacoustic transmissions through the cochlea and labyrinth. The most common bony defect of the superior canal is found over the arcuate eminence, with rare cases involving the posteromedial limb of the superior canal associated with the superior petrosal sinus. Operative intervention is indicated for intractable or debilitating symptoms that persist despite conservative management and vestibular sedation. Surgical repair can be accomplished by reconstruction or plugging of the bony defect or reinforcement of the round window through a variety of operative approaches. The authors review the etiology, pathophysiology, presentation, diagnosis, surgical options, and outcomes in the treatment of this entity, with a focus on potential pitfalls that may be encountered during clinical management.

Epidemiology

The roof of the SSC is dehiscent in 0.5% and nearly dehiscent in another 1.4% of cadaveric temporal bone specimens, localized to either the floor of the middle fossa or between the SSC and the superior petrosal sinus.18,62 Near dehiscence, established at a bone thickness of 0.1 mm or...
less in 1 study, compares to an average roof depth of 1 mm in control adult specimens. Series in which HRCT was used have estimated an incidence of SSCD of 3%–10% in both adults and children, which may be an overestimate given that even HRCT slices are thicker than the thickness of some middle fossa floors. Superior semicircular canal dehiscence on one side is also associated with contralateral thinning or dehiscence in one-third to half of all cases on both cadaveric and radiological studies.

**Etiology**

The SSC resides under the floor of the middle cranial fossa, and is often denoted by the presence of a prominence protruding into the floor known as the arcuate eminence. In up to 15% of the population, no arcuate eminence is discernible. In other patients, the arcuate eminence may be rotated posteriorly from the position of the SSC. Both congenital and acquired causes of SSCD have been proposed.

During development, the covering of the SSC grows from a diaphanous state to a robust trilaminar capsule between birth and 3 years of age. The SSC also protrudes prominently into the middle fossa during gestation, compared with a reduced prominent temporal bone location in adults, which may lead to adhesion of the membranous labyrinth to dura mater before complete ossification of the bony labyrinth. A computer simulation model of inner-ear ontogeny has attributed SSCD to a malpositioned primitive otocyst and impaired migration of mesenchymal cells to form the apical cap of the canal. Failure or inadequate osseous development may then manifest in overt clinical symptoms following a second hit, such as minor trauma, infection, inflammation, or chronic pressure from CSF pulsations or the overlying temporal lobe.

The presence of SSCD in young children supports a congenital etiology. An increasing prevalence of canal dehiscence with age in temporal bone imaging series suggests additional acquired factors. Frequent co-occurrence with tegmen mastoideum and tympanic erosion and the incidence of bilateral SSCD can support either acquired or congenital etiologies. Dehiscence in the posterior semicircular canal and other locations has been reported, but is much more rare.

Chronic elevated intracranial pressure (ICP) has been hypothesized to underlie co-occurrence of tegmen defects, patulous internal auditory canal, and SSCD, because idiopathic intracranial hypertension is associated with tegmen defects and CSF leakage. Indeed, up to 15% of patients undergoing middle fossa craniotomy for repair of spontaneous CSF otorrhea are noted to have dehiscence of the SSC apex—higher than the reported prevalence in the general population. Some patients with SSCD are also observed to have higher rates of obstructive sleep apnea and a higher body mass index, suggestive of a correlation between elevated ICP and development of dehiscence.

Several families with multiple members who have been treated for SSCD have been reported. However, the contribution of genes implicated in inner-ear development and familial deafness syndromes, such as the homeobox genes $HMX3$ and $POU3F4$, $Netrin1$, cochlin (COCH), and the solute carrier $SLC26A4$, to SSCD remains unclear.

**Pathophysiology**

Superior semicircular canal dehiscence is hypothesized to cause symptoms by creation of a mobile “third window.” In normal conditions, the oval window, to which the stapes footplate attaches, regulates sound input into the inner ear, whereas the round window regulates sound and mechanical energy release from the scala tympani of the inner ear, among other functions. These 2 windows typically direct forces associated with sound waves along the spiral basilar membrane. In contrast, the membranous vestibular labyrinth is insulated from sound or pressure changes in the middle ear and CSF by a bony capsule, but becomes sensitive to such changes if an osseous defect occurs (Fig. 1A and B). Hence, addition of a nonphysiological third window at the roof of the SSC results in preferential hydroacoustic shunting toward the pathologic bony defect and away from the cochlear partition, to produce symptoms of vertigo, conductive hyperacusis, or conductive hearing loss. Transmission of ICP through the vestibular system from the SSCD to the round window also produces increased compliance of the inner ear, with Hennet’s sign, conductive hearing loss, and the perception of pulsatile tinnitus.

The concept of a third window has been conjectured to result in stimuli-induced vertigo in chronic adhesive otitis by disrupting the bony labyrinth. Similar phenomena have been observed in dehiscence of posterior and horizontal semicircular canals from various pathologies such as cholesteatoma. Animal models, in which the bony apex of the SSC is drilled while preserving the membranous labyrinth, corroborate this hypothesis. Fenetration of the bone overlying selected semicircular canals evokes eye and head movements in the plane of the canal with loud noises, producing a sound-induced vestibular activation known as the Tullio phenomenon. Removal of the bony cap of the SSC in rodents also produces a significant air-bone gap in response to clicks and a pattern of auditory brain responses, consistent with SSCD findings in patients. This underlying pathophysiology of an altered pressure gradient between mobile window outlets has prompted targeted treatment strategies, as discussed below.

**Clinical Presentation**

Patients can present with vertigo, disequilibrium, oscillopsia, conductive hyperacusis, autophony, conductive hearing loss, hearing distortion, gaze-evoked tinnitus, and aural fullness or blockage. A quarter of patients report an auditory sensation, described as a “swishing,” associated with motion of the eyes or body, particularly toward the affected ear. There is an enhanced perception of bone-conducted sounds producing autophony of one’s own voice, neck creaking, chewing, and footsteps hitting the ground. As opposed to the autophony of patulous eustachian tube, there is little or no autophony of one’s breathing sounds. Less commonly, dysautonomia, such as bradycardia and hypotension, may be triggered by
sound or pressure alteration of vestibulosympathetic reflexes. Unprovoked drop attacks have also been reported in isolated cases, with amelioration following SSCD repair. Symptoms vary widely from patient to patient. Precipitating factors for the vertigo or oscillopsia include loud noise, or activities that produce changes in the middle ear or ICP, such as lifting, straining, nose blowing, Valsalva maneuvers, or pressure on the ear. Exercise and exertion-induced vestibular symptoms have been associated with SSCD adjacent to the superior petrosal sinus in particular. Antecedent head trauma or barotrauma may be elicited in the history.

**Diagnostic Evaluation**

Differential diagnoses that often mimic SSCD syndrome include eustachian tube dilatory dysfunction, patulous eustachian tube, otosclerosis, chronic otitis media, perilymph fistula, labyrinthitis, Ménière’s disease, migraine-associated vertigo, and vestibular schwannoma, establishing it as a great otological mimicker. Accurate diagnosis relies on consistent symptoms, signs, audiometric and vestibular results, and imaging confirmation. In particular, HRCT is increasingly found to overestimate the true incidence of the clinical syndrome, because the presence of bony dehiscence alone does not necessarily indicate a pathological condition. Fortunately, the direction-specific physiology of semicircular canal biomechanics produces several localizing signs that aid diagnosis. Adjunctive testing, including provocative testing, audiometry, electrocochleography (ECoG), and sound-induced cervical VEMPs are necessary to confirm the clinical significance of the radiology findings.

**Examination and Provocative Testing**

Signs of SSCD syndrome can be induced with tones at escalating frequencies and intensities, compression of the tragus to increase external auditory canal pressure, jugular venous compression, and Valsalva maneuvers. Vertical torsional eye movements produced by these stimuli and observed with the patient wearing Frenzel lenses are pathognomonic for SSCD syndrome. Nystagmus or vestibular activation elicited by pressure changes at the external auditory canal, known as Hennebert’s sign, is also classically associated with canal dehiscence but is not diagnostic of it. Weber’s test typically lateralizes to the symptomatic ear; this is attributed to a conductive hearing loss or intralabyrinthine hearing loss.

**Audiometry Testing**

Hearing loss may be conductive, sensorineural, or mixed. Bone conduction is decreased and air conduction is increased due to the presence of a mobile third window, thereby producing the air-bone gap. Normal symmetrical hearing helps eliminate retrocochlear pathologies as
Superior semicircular canal dehiscence

Superior semicircular canal dehiscence (SSCD) is a condition where the bone that forms the roof of the superior semicircular canal is thinner than normal, allowing for a direct pathway between the canal and the middle cranial fossa. This condition is often associated with vertigo and hearing loss. The stapedial (or acoustic) reflex is typically preserved in SSCD, in contrast to other causes of hearing loss such as otosclerosis, where it is characteristically absent. Patients with predominantly vestibular rather than auditory symptoms may still exhibit some degree of hearing loss. Therefore, formal pure-tone audiometry is frequently useful to establish a baseline, especially before planned operative interventions. After operative repair of SSCD, the air-bone gap may partially or completely resolve in long-term follow-up.

Electrocochleography Testing

Electrocochleography offers an alternative perioperative testing modality to assess SSCD pathological features and repair. ECoG measurements reveal a significantly higher cochlear summating potential/action potential ratio in ears affected by SSCD, with improvement observed following operative repair. The ECoG measurement is not specific to canal dehiscence because abnormal values are also observed in other inner-ear conditions such as Ménière’s disease and perilymphatic fistula. Isolated observation of intraoperative ECoG has also noted normalization of the summating potential/action potential ratio immediately following canal occlusion.

Vestibular Evoked Myogenic Potential Testing

The VEMP assesses the response of postural muscles that are under tonic control of the medial vestibulospinal tract to vestibular activation. Auditory stimuli, such as loud clicks, usually induce relaxation of the ipsilateral sternocleidomastoid VEMP or orbicularis oculi VEMP, which is measurable on electromyography. The presence of SSCD renders patients more sensitive to the auditory stimulus and produces a vestibulocollic response, in which the sternocleidomastoid muscle relaxes to vestibular stimulation, at a lower (<65 dB) threshold than in control subjects on the affected side. The VEMP can also distinguish between patients with near dehiscence, or thin bone, versus those with true dehiscence over the SSC, both of whom can present with similar symptoms. Following successful repair of the dehiscence, the VEMP normalizes.

Imaging

A thin-cut HRCT scan, particularly in the coronal plane, or an oblique cut directed to the plane of the semicircular canal, is confirmatory of the clinical diagnosis. However, even HRCT can produce false-positive results if the canal roof is thinner than the image resolution slice thickness, as might be expected given an average SSC roof thickness of 1 mm in cadaveric studies. Osteoporosis can also lead to a false-positive reading because of lack of calcium in the imaged bone. Reformatted images parallel (Pöschl’s plane) or perpendicular (Stenver’s plane) to the SSC may aid visualization of a dehiscence, as do novel 3D reconstruction techniques. An MRI study with a fast imaging employing steady-state acquisition (FIESTA) sequence demonstrates 100% sensitivity and negative predictive value, but 96.5% specificity and 61% positive predictive value compared with CT in evaluating SSCD, suggesting that MRI might be adequate in excluding the diagnosis. Of note, near dehiscence with extremely thin bone can produce similar signs and symptoms as a fully dehiscent apical cap, and may benefit from operative intervention.

Management

Management after clinical diagnosis begins with avoidance of provocative stimuli and vestibular sedation. It is important to note that a majority of patients identified with SSCD syndrome in the literature have not undergone
surgical repair. Operative intervention is indicated for intractable or debilitating symptoms. Surgical repair can be accomplished through resurfacing of the SSC roof, plugging of the osseous defect, or reinforcement of the round or oval window to diminish the third window effect (Fig. IC and D).

Canal Plugging

The original report of SSCD operative repair described a middle fossa subtemporal extradural approach to expose the roof of the SSC and plug the defect with fascia, which has been subsequently expanded to muscle, bone wax, bone pâté, and bone pâté. Given the risk of false-positive CT diagnoses, the middle fossa approach allows for face visualization and confirmation of the defect prior to operative repair (either occlusion or resurfacing), and can be further augmented by endoscopy in select cases.

As an alternative, transmastoid occlusion of the SSC appeals to otologists due to their ability to access large areas of dehiscence via a familiar approach, the ability to occlude the canal without manipulating the defect, and the relatively lower invasiveness of a mastoidectomy compared with a craniotomy. This can be achieved with bone pâté or temporalis fascia placed within the SSC, with > 90% postoperative improvement in vestibular and hearing symptoms. Some prefer the transmastoid approach for repair of medially located SSCD adjacent to the superior petrosal sinus. This surgical approach, however, offers more limited visualization of the dehiscence and may be less favorable in the setting of a poorly pneumatized temporal bone, low-lying temporal fossa, or extensive concurrent tegmen defects. Concurrent plugging of the defect, along with reconstruction or resurfacing of the floor, is achievable through either a middle fossa or a transmastoid approach.

The choice of reconstructive material may also influence long-term outcome, although experience remains limited. In an animal model of semicircular canal occlusion, comparison of bone dust, bone wax, or muscle revealed that the bone dust group had the best hearing outcomes, with periosteal osteoneogenesis; the bone wax group had the poorest hearing outcomes, with periosteal osteoneogenesis observed but with the presence of perilymphatic inflammation.

Canal Roof Resurfacing and Capping

Reconstruction or resurfacing of the dehiscent middle fossa floor with fascia, perichondrium, cartilage, split-thickness cortical bone, bone wax, bone pâté or calcium phosphate, synthetic elastomer, or a combination thereof, is a common alternative to canal plugging. The use of hydroxyapatite cement for reconstructing the apical defect has also been termed capping. Notably, resurfacing allows for reconstruction of multiple middle fossa floor defects, which is regularly encountered in the setting of SSCD.

As with canal plugging, resurfacing can be accomplished via either a middle fossa or a transmastoid approach, with their respective strengths and limitations in terms of ease of visualization, available surface area for reconstruction, and surgical corridor. Endoscopic assistance provides another adjunct, based on surgeon preference.

Analysis of select cases of recurrent SSCD symptoms after use of an autologous temporal bone split-thickness calvarial bone graft to resurface the middle fossa has revealed resorption of the bone graft; this prompted the conclusion that the entire temporal bone is abnormal in patients with SSCD and that resurfacing using a calvarial bone graft carries a higher risk of resorption. Concern for graft resorption or migration as a mechanism of resurfacing failure over time prompts some surgeons to favor bone source substitute or silicone elastomer over autologous grafts. Securing of the fascia-bone graft to the calvaria with a small plate has also been performed, with satisfactory results. Evolving appreciation of the biological sequelae of reconstruction suggests that experience from earlier resurfacing series may not be comparable to contemporary results because of modifications to the technique.

Round Window Reinforcement

Several groups empirically investigated tissue graft reinforcement of the round window, having observed that patients who had undergone procedures to repair a presumed perilymph fistula prior to the initial report of SSCD, in retrospect may have had SSCD and may have experienced significant benefit. The mechanism for why it may help is uncertain, but it has been postulated that it may be influencing the impedance of the inner-ear system, perhaps reducing global hypercompliance and consequent compression-related endolymph displacement. The round window is directly accessible through a tympanic membrane opening and can be reinforced with soft tissue such as fascia, perichondrium, or muscle, gently filling the round window niche after excoriating the mucosa, but carefully preserving the membrane. Gelfoam or an autologous blood patch placed through a myringotomy can be done as a trial in an office setting. All of these can also be removed in the event of symptom worsening. Over-stiffening of the round window with bone wax or cartilage may divert additional acoustic energy into the dehiscence and cause a worsening of symptoms that can be reversed by removal of the material. It appears that gentle reinforcement of the round window may be beneficial—for reasons to be determined—as opposed to severe dampening of the round window, which exacerbates the condition, as may be predicted from the “third window” model.

There is a paucity of clinical data examining outcomes following round window reinforcement for SSCD syndrome. Postoperative improvement in the full gamut of SSCD-related symptoms has been reported, with the exception that conductive hearing loss does not improve. Although some advocate that round window occlusion should be performed in patients whose primary symptom is conductive hyperacusis, others suggest that both oval and round window reinforcement may be beneficial for that symptom.

A concern is that the clinical outcome following round window reinforcement may not be as durable as superior canal plugging or resurfacing techniques; symptoms may
Superior semicircular canal dehiscence
J Neurosurg Volume 127 • December 2017

Considerations in Pediatric Patients

Experience with SSCD diagnosis and management in children remains limited,47,52,53 although imaging series suggest a 3%–10% incidence.21,73 Children present with a varied constellation of hearing loss, vertigo, and disequilibrium, with auditory symptoms typically occurring first. Diagnosis of vestibular symptoms is more challenging in younger children, and may manifest as delayed onset ofWalking.47 The radiographic appearance of bilateral SSCD is frequently detected and does not warrant intervention in the absence of symptoms. Repair of SSCD on one side can provoke or be followed by development of contralateral symptoms attributable to the second canal dehiscence, and patients must be counseled appropriately.4 Subsequent plugging of the second side is associated with oscillopsia in half of the patients, but still provides satisfactory relief of the primary debilitating symptoms. Bilateral canal dehiscence is also associated with a prolonged course of symptom recovery following unilateral canal plugging, for which patients should be counseled.66

Perioperative Considerations

Perioperative steroids are given variably across reports, often for longer durations if signs of sensorineural hearing loss, labyrinthine hypofunction, or facial paresis are detected postoperatively.56,85 Early postoperative persistence or slight decline in auditory and vestibular function may be due to fluid, blood, or air at the surgical site, and may be monitored. Preoperative and postoperative audiometry and vestibular testing allow objective tracking of symptoms and outcomes, especially as experience with this disease entity accrues.

Outcomes

The majority of patients report improvement in presenting symptoms following operative repair of SSCD, regardless of the technique.5,45,52,57,61,62,66 Many studies cite a higher success rate with canal plugging compared with resurfacing,57 especially as a means to treat vestibular imbalance; however, canal plugging demonstrates efficacy equivalent to capping with hydroxyapatite, which is in essence a reconstruction operation with nonmigratory material.84 In a meta-analysis, all operative modalities achieved equal success if auditory symptomatology was the primary indication for intervention. In addition to symptom resolution, improvements in the VEMP threshold and air-conduction gap are frequently observed following successful repair of canal dehiscence.

Vestibular hypofunction1,50 and hearing loss60 are the most feared complications following SSCD repair. Impairment of the SSC function in isolation is common after plugging.17 More global vestibular hypofunction is observed in approximately one-third of patients in the early postoperative period following middle fossa plugging, with higher risk associated with an increased length of the dehiscent area.3 This may result from intraoperative loss or displacement of perilymph and redistribution of endolymph into the horizontal and posterior canals by the plug or during dural elevation over the dehiscent canal, during which extra caution should be exerted. These symptoms typically resolve over time as perioperative fluid resolves and physiological vestibular compensation is attained,43 with 11% of patients demonstrating vestibular impairment at 6 weeks postoperatively.3 Because of the concern for compromise of SSC function, some surgeons favor resurfacing over plugging strategies; capping with hydroxyapatite may offer a suitable option of minimizing postoperative vestibular side effects while providing high rates of relief from preoperative vestibular handicap.

Significant hearing loss is uncommon after SSCD repair, and does not correlate with either the plugging or resurfacing technique.46 In comparison, mild high-frequency sensorineural hearing loss, without change in speech discrimination, has been observed in 25% of patients after canal plugging via a middle fossa approach.85 Early methods emphasized full occlusion of the membranous superior canal during plugging, which might incur a higher risk of postoperative hearing loss.60 Use of bone pâté for plugging may be associated with a lower risk of hearing impairment compared with other occlusive materials.25 Not surprisingly, the incidence of hearing loss rises in revision procedures for SSCD, presumably due to adhesions at the surgical site and injury to the membranous labyrinth while separating the dura. Prior surgery on the cochlea or stapes also increases the risk for hearing loss following SSCD repair.60

Patients with bilateral SSCD, larger defects, and a history of migraines are more likely to experience a prolonged duration of symptoms following operative repair.66 Facial nerve dysfunction, CSF leakage, and other neurological morbidities are rare after SSCD repair. On the whole, quality of life measures and outcomes remain to be clarified for patients with SSCD.

Conclusions

Over the past 2 decades, SSCD has been increasingly recognized as a treatable cause of disequilibrium and
hearing loss. Its distinctive pathophysiology, arising from the introduction of a mobile “third window” formed by an unprotected membranous labyrinth at the apex of the superior canal, accounts for characteristic symptoms, diagnostic signs, and treatment options. Regardless of the approach, most patients experience alleviation of presenting symptoms following operative repair of the SSCD, although risk of vestibular hypofunction and hearing loss remain avenues for future improvement. The choice of operative strategy ultimately depends on the surgeon’s experience and preference. A multidisciplinary approach to the management of SSCD will continue to optimize outcomes for this newly recognized disease entity.

References
Superior semicircular canal dehiscence


44. Kartush JM, Kemink JL, Graham MD: The arcuate eminence. *Presse Med Belg Brux* 45:151, 1911


74. Shia WT, Kartush JM: Superior canal dehiscence.


83. Tullio P: Das Ohr und die Entstehung der Sprache und Schrift. Berlin: Urban & Schwarzenberg, 1929


Disclosures
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author Contributions
Conception and design: Bi. Acquisition of data: Bi, Brewster. Drafting the article: Bi. Critically revising the article: Dunn, Bi, Poe, Vernick, Lee, Corrales. Reviewed submitted version of manuscript: Dunn, Bi, Brewster, Corrales. Approved the final version of the manuscript on behalf of all authors: Dunn. Study supervision: Dunn, Bi.

Correspondence
Ian F. Dunn, Department of Neurosurgery, Brigham and Women’s Hospital, 15 Francis St., Boston, MA 02115. email: idunn@partners.org.