# Transmastoid Repair of Superior Semicircular Canal Dehiscence Associated with the Superior Petrosal Sinus: Presentation of Two Cases and Review of the Literature

## Abstract

**Educational Objective:** At the conclusion of this presentation, the participants should be able to describe the anatomy, etiology, and presenting symptoms of superior semicircular canal dehiscence syndrome, understand the relevant anatomy and unique considerations for dehiscences associated with the superior petrosal sinus, and be familiar with the transmastoid approach for occlusion of the superior semicircular canal.

**Objectives:**
1. Describe the presentation, evaluation, treatment, and outcomes of two patients with superior semicircular canal dehiscence (SCD) associated with the superior petrosal sinus (SPS).
2. Discuss the rationale for transmastoid occlusion of the superior semicircular canal for SCD when it is associated with the SPS.

**Study Design:** Case series with literature review.

**Methods:** The records of 2 patients who presented to our institution with SCD associated with the SPS were reviewed.

**Results:** Both patients presented with autophony, conductive hyperacusis, disequilibrium, and pulsatile tinnitus. Low VEMP thresholds on the left and CT scans suggested left SCD involving the abutting SPS. Both subjects underwent successful and uncomplicated transmastoid plugging of the left superior semicircular canal with significant improvement in presenting symptoms.

**Conclusions:** The understanding and treatment of SCDs continues to evolve. Dehiscence can be associated with either the middle cranial fossa floor or the SPS. Dehiscences associated with the SPS are more medially located and visualization may require additional temporal lobe retraction when repaired via middle cranial fossa approach. Additionally, risk of intraoperative bleeding is likely increased. In contrast, transmastoid access may afford excellent visualization of relevant structures without brain retraction and minimal risk of bleeding. Our findings demonstrate that the transmastoid approach may be a useful option for the management of SCD associated with the SPS.

## Case Narratives

### Case 1

This 60-year-old woman presented with left-sided autophony, conductive hyperacusis, pulsatile tinnitus, and imbalance without true vertigo. Examination was notable for a positive Hennebert’s sign. CT scan of the temporal bones (Figures 1-2) was notable for dehiscence of the left superior semicircular canal (SSC) at the interface with the superior petrosal sinus (SPS). Vestibular testing revealed abnormally low ocular vestibular evoked myogenic potential (OcVEMP) threshold on the left. On MRI/MRA, a non-filling left transverse sinus was noted. She ultimately underwent transmastoid plugging of the left SSC. Postoperatively, she noted complete resolution of autophony and pulsatile tinnitus. Some persistent mild imbalance and motion-induced vertigo was noted.

### Case 2

This 40-year-old woman presented with left-sided autophony, conductive hyperacusis, ringing and pulsatile tinnitus, constant generalized imbalance, and rare 15-20 minute episodes of vertigo. Physical exam was normal. CT scan of the temporal bones revealed left-sided SCD with the SPS (Figures 3-4). Vestibular testing demonstrated decreased cervical VEMP thresholds on the left. She underwent transmastoid plugging of the left SCC (Figures 5-6) with complete resolution of her autophony and near-complete resolution of her imbalance and tinnitus.

## Intraoperative Photographs

![Figure 1: Exposure of the left superior petrosal sinus (faint blue structure, white arrow) and superior semicircular canal (top of image is anterior).](image1)

![Figure 2: Floor of middle cranial fossa.](image2)

![Figure 3: Unroofing the SSC, exposing both superior and inferior area of superior semicircular canal.](image3)

![Figure 4: Transmastoid middle fossa craniotomy repair of superior semicircular canal dehiscence using a soft tissue graft.](image4)

![Figure 5: Delivery of middle fossa dura.](image5)

![Figure 6: The dural remnant was then sutured to the pericranium with 4-0 Vicryl.](image6)

## Discussion

- Superior semicircular canal dehiscence syndrome (SCDS), first described in 1998 by Minor, et al., most commonly presents with symptoms of sound/pressure induced vertigo, autophony, conductive hearing loss, and conductive hyperacusis.
- SCDS is caused by a bony dehiscence over the SSC, creating a “third window” that allows abnormal movement of perilymph, particularly with increased middle ear pressures.
- This bony dehiscence can communicate with the middle fossa or (perhaps less commonly) the superior petrosal sinus (SPS).
- Cadaveric temporal bone studies have suggested a higher rate of SCD with the SPS than is seen in the symptomatic population, suggesting this subset is less often symptomatic.
- First described in North America in 2008, the transmastoid approach for treatment of SCDS is an alternative to the more common middle fossa (MF) approach.
- Repair of SCD with the SPS via MF approach entails increased need for temporal lobe retraction for visualization medial to the arculate eminence and need to dissect the SPS free from the area of dehiscence, increasing risk of bleeding.
- Teixido, et al. (2011) describe a transmastoid middle fossa resurfacing (as opposed to plugging) technique that produced similarly excellent results in 3 patients with SCD and the SPS.

## References