Endoscopic-Assisted Repair of Superior Canal Dehiscence

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KEYWORDS

• Endoscopic ear surgery • Superior canal dehiscence
• Superior canal dehiscence syndrome • Lateral skull base • Middle fossa craniotomy
• Endoscope • Otology

KEY POINTS

- Middle fossa craniotomy (MFC) and transmastoid approaches are the most common surgical techniques to repair superior canal dehiscence (SCD). The advantage of the MFC approach is direct visualization and surgical access to the SCD without the need for drilling the skull base in most cases.
- Endoscopes provide a superior view (compared with binocular microscopy) of arcuate eminence defects using a MFC approach, especially medial dehiscences that are found along the downsloping tegmen.
- Improved visualization of arcuate eminence defect using the endoscope is achieved with a minimally invasive skin, soft tissue and craniotomy approach, with reduced temporal lobe retraction, and avoidance of drilling overlying bony ridges that obscure the line of sight.
- Endoscopes provide superior transillumination of the skull base and localization of blue-lined superior canals, which is important when repairing symptomatic "near dehiscence" SCD.
- Massachusetts Eye and Ear radiologic classification of SCD and surrounding skull base topography is helpful in preoperative planning and can anticipate the need for an endoscopic-guided repair.

INTRODUCTION

First characterized by Minor and colleagues[1] in 1998, superior canal dehiscence (SCD) is a bony defect of the superior canal that is associated with vestibular and/or auditory dysfunction. Classic symptoms associated with SCD syndrome (SCDS) include aural

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fullness, pulsating tinnitus, autophony, and conductive hyperacusis (hearing one's own voice, footsteps, and/or eye movements in the affected ear) and dizziness or vertigo evoked by loud sounds (Tullio phenomenon) or Valsalva maneuver (Hennebert sign).[2] Symptoms can vary widely among patients with SCDS, and not all patients with these defects have symptoms.

The concept of a pathologic mobile "third window" of the inner ear (the oval and round windows being the first two) is widely accepted as the explanation for SCDS pathophysiology.[3–6] Based on this concept, it is hypothesized that a third window allows for transmission of fluid pressure between the intralabyrinthine space and the cranial vault.[4,7–10] With this alternative path of low impedance, acoustic flow entering from the oval window is shunted toward the dehiscence causing hearing loss and deflection of the superior canal cupula away from the ampulla (ampullofugal) resulting in the stimulation of the affected superior canal.[3] Conversely, straining or Valsalva against closed glottis can lead to labyrinthine fluid flow away from the dehiscence and toward the ampulla (ampullopetal) resulting in inhibition of the superior canal ampulla.[11] Dizziness and vertigo can arise from either condition. Hypersensitivity to sounds and vibrations conducted through the body is a common feature of SCDS and is thought to be part of the third window phenomenon, but the underlying mechanism is still a subject of ongoing research.

Conservative management is adequate for most patients with SCDS, but for patients with severe symptoms, surgical repair is a feasible option.[12,13] Microscopic-assisted middle fossa craniotomy (MFC) is a widely used technique to repair SCDs because it is well-established and safe.[14,15] Alternatively, some surgeons favor a less invasive transmastoid approach to either directly repair the dehiscence (through a tegmen defect created from the mastoid area)[16–20] or indirectly isolating the dehiscence (by creating labyrinthotomies on either side of the SCD and then plugging the canal).[21–23] At the Massachusetts Eye and Ear Infirmary (MEEI) we believe that the transmastoid approach is ideal for SCDs associated with the superior petrosal sinus (**Fig. 1**E)[24] and revision cases, where the MFC approach is often complicated by prior attempts to repair the defect directly.

The MFC approach with binocular microscope provides excellent visualization of the lateral skull base and direct surgical access to the SCD, which is essential to ensuring adequate repair of the entire defect. However, a subset of patients with SCDS has unfavorable skull base topography and visualizing the defect under the microscopic is challenging (**Fig. 2**). Rigid endoscopes provide a superior view of the medial skull base and defects "hidden" from the microscopic view (**Fig. 3**). We now favor a small craniotomy approach for SCD repairs using endoscopes as an adjunct when necessary. This article discusses (1) the diagnosis of SCD, (2) the preoperative evaluation of SCD with an emphasis on patient selection and radiologic classification, (3) the MEEI endoscopic-assisted surgical technique to repair SCD via MFC, and (4) pearls and pitfalls with this approach.

DIAGNOSIS OF SUPERIOR CANAL DEHISCENCE

The diagnosis of SCDS encompasses a detailed clinical history, comprehensive head and neck examination, audiometric testing (with stapedius reflex), vestibular evoked myogenic potential (VEMP) testing, and high-resolution temporal bone computed tomography (CT).[9,25] Differential diagnoses to consider include otitis media with effusion, vestibular migraines, Meniere disease, benign paroxysmal positional vertigo, and patulous eustachian tube dysfunction.[26] If the patient has unilateral SCDS (or a less symptomatic contralateral ear), 512-Hz tuning fork testing usually lateralizes to