Canal dehiscence
Wade W. Chien, John P. Carey and Lloyd B. Minor

Department of Otolaryngology–Head & Neck Surgery, The Johns Hopkins University School of Medicine, Baltimore, Maryland, USA

Correspondence to Wade W. Chien, Department of Otolaryngology–Head & Neck Surgery, The Johns Hopkins University School of Medicine, 601 N. Caroline Street, 6th floor, Baltimore, MD 21287, USA
Tel: +1 410 955 7808; fax: +1 410 614 8610; e-mail: wchien1@jhmi.edu

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Purpose of review
The aim is to review canal dehiscence involving the superior, lateral, and posterior semicircular canals. The main focus will be on superior semicircular canal dehiscence.

Recent findings
Canal dehiscence involving the superior, lateral, and posterior semicircular canals can have different etiologies, including developmental abnormality, congenital defect, chronic otitis media with cholesteatoma, and high-riding jugular bulb. However, their clinical presentation can be very similar, with patients complaining of vertigo, oscillopsia, and sometimes hearing loss. Canal dehiscence causes an abnormal communication between the inner ear and the surrounding structures. This creates a third mobile window within the inner ear, disrupting its normal mechanics and causing symptoms.

Summary
Superior semicircular canal dehiscence is now a well-established entity in the medical literature. Surgical repair is effective at relieving patients’ vestibular symptoms. Lateral semicircular canal dehiscence is usually associated with chronic otitis media. Posterior semicircular canal dehiscence is a rare entity, with similar clinical presentations and treatment options as the other canal dehiscences.

Keywords
cholesteatoma, chronic otitis media, hearing loss, Hennebert sign, labyrinthine fistula, perilymphatic fistula, posterior semicircular canal dehiscence, superior semicircular canal dehiscence syndrome, Tullio phenomenon, vertigo

Introduction
Canal dehiscence causes an abnormal communication between the inner ear and the surrounding structures. Under normal circumstances, the inner ear is fluid-filled and is encased by the dense otic capsule bone with two mobile windows: the oval window and the round window. Semicircular canal dehiscence results in a third mobile window in the inner ear, which can lead to areas of increased compliance with symptoms and signs that can be understood based on the abnormal pressure transmission in the system. Semicircular canal dehiscence involving the superior, lateral, and posterior canals can have different etiologies, but they may present similarly. This review article will focus primarily on superior semicircular canal dehiscence, which is the most common clinical presentation of semicircular canal dehiscence, but lateral canal and posterior canal dehiscences will also be discussed.

Superior semicircular canal dehiscence syndrome
A syndrome of vertigo and oscillopsia was recently identified [1–4]. The etiology of this syndrome was localized to a dehiscence of the bone covering the superior semicircular canal. The abnormal communication between the superior semicircular canal and the brain can result in vertigo and oscillopsia induced by loud sounds (Tullio phenomenon), by changes in pressure in the ear canal that are transmitted to the middle ear (Hennebert sign), or by Valsalva maneuvers [1,2,4]. Some patients with superior semicircular canal dehiscence syndrome were previously explored for possible perilymphatic fistula [2] with no improvement of their symptoms postoperatively. The key to the identification of this syndrome was the analysis of the evoked eye movements in the sound- or pressure-induced symptoms. These eye movements evoked by the sound and pressure stimuli were noted to align with the plane of the dehiscent superior semicircular canal, although larger dehiscences can result in alignment of the evoked eye movements in other planes [3]. Figure 1 shows an example of the evoked nystagmus in a patient with bilateral superior canal dehiscence syndrome when loud tones are presented to each ear independently. The scleral search coil technique was used to make three-dimensional recording of eye movements (horizontal, vertical, and torsional components) from both eyes but tones were presented to the right ear (upper traces) or left ear (lower traces). Figure 2 shows...
Figure 1 Horizontal (H), vertical (V), and torsional (T) eye positions recorded with scleral search coils and plotted against time for the left eye (thick traces) and right eye (thin traces) in a patient with bilateral superior canal dehiscence syndrome.

![Figure 1](image-url)

The thick black line (bottom) indicates the duration of a 3-kHz tone that was presented at 110 dB hearing level in darkness to the right ear (top traces) and to the left ear (bottom traces). The scale markers indicate a 5° eye rotation and a 5-s time interval. Positive direction for the horizontal, vertical, and torsional axis is defined as left, down, and clockwise (rotation of the superior pole of the patient’s eye toward his right side). In response to a tone presented to the right ear, the patient developed nystagmus with upward, counterclockwise slow phases, consistent with excitation of the right superior semicircular canal. In response to a tone presented to the left ear, the patient developed nystagmus with upward, clockwise slow phases, consistent with excitation of the left superior semicircular canal. The nystagmus was sustained for the duration of the tone and was conjugate, without evidence of vertical (skew) disconjugacy. The median slow-phase nystagmus speed was 3°/s. Reproduced with permission from [3].

The symptoms and signs in superior semicircular canal dehiscence syndrome can be understood from the effect of the dehiscence in creation of a third mobile window in the inner ear [1]. The direction of the evoked nystagmus (either excitation or inhibition) can be predicted based upon the direction of endolymph flow within the superior canal (either ampullofugal or ampullopetal) in response to Valsalva maneuvers or pressure in the ear canal. A Valsalva maneuver against pinched nostrils (forcing air into the middle ear through the Eustachian tube) results in inward displacement of the stapes and ampullofugal (excitatory) motion of the superior canal ampulla. Conversely, a Valsalva maneuver against a closed glottis (taking a deep breath and bearing down) results in increased intrathoracic pressure, decreased jugular venous return, and increased intracranial pressure. The membranous superior canal in the area of the dehiscence is compressed by the increased intracranial pressure and ampullopetal (inhibitory) motion of the superior canal ampulla is induced. Positive pressure in the external canal causes inward motion of the tympanic membrane and stapes footplate with ampullofugal motion of the superior canal ampulla. Oppositely directed motion is induced by negative pressure in the external canal, resulting in ampullopetal motion of the superior canal ampulla.

Confirmation of the bone dehiscence overlying the superior canal in patients with the syndrome has been obtained with high-resolution computed tomography (CT) scans of the temporal bones [7]. Conventional temporal bone CT scans are performed with 1.0-mm collimation, and images are displayed in the axial and coronal planes. These scans have a relatively low specificity (high number of false positives) in the identification of superior canal dehiscence because of the effects of partial volume averaging. The specificity and positive predictive value of these scans is improved when 0.5-mm-collimated multislice CT scans are performed with reformation of the images in the plane of the superior canal [7]. Figure 3 shows CT images with reformation in the plane and orthogonal to the plane of each superior semicircular canal in a patient with superior canal dehiscence syndrome. CT studies have also shown that the thickness of bone overlying the intact superior canal in a patient with unilateral dehiscence can be significantly thinner than in patients without superior canal dehiscence [8]. This finding and observations from a review of 1000 histologically processed temporal bones sectioned in a plane parallel to the petrous ridge [9], suggest an underlying developmental or congenital abnormality that leads to the development of the syndrome. The bone overlying the superior canal continues to develop after birth through 3 years of age. A premature arrest in the development of this bone may be the cause or predisposing factor for superior canal dehiscence syndrome. However, it should be emphasized that the bony defect is a necessary but not sufficient condition to produce the clinical syndrome. Pressure transmission through the dehiscence must also be present, and this may be

Key points
- Dehiscence of the bone covering the semicircular canals can cause vertigo and/or hearing loss.
- The evoked eye movements provide important clues as to the location of the abnormality.
- Surgical repair of the bony dehiscence is effective at alleviating patient symptoms.

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symptoms may still not develop until the elasticity of the dura is sufficient to transmit pressure between the middle fossa and inner ear. This may occur with a sudden change in intracranial pressure, as might occur with a vigorous Valsalva maneuver or with a shear stress across the skull base associated with a closed head injury. In our initial experience, 59% of patients reported a history of one of these inciting factors [2].

Vestibular-evoked myogenic potentials (VEMPs, cervical and ocular) have been useful in assessing for the presence of superior canal dehiscence syndrome [10–13]. In cervical VEMPs, short-latency relaxation potentials evoked by clicks or by tone bursts are recorded from surface electromyographic electrodes placed on the skin over the ipsilateral sternocleidomastoid muscle while that muscle is tonically contracted [14,15]. Ocular VEMPs are excitatory responses recorded from surface electrodes directly below the eye in response to the same stimuli. Previous studies have shown that these responses arise from the vestibular end organs (utricle and saccule), and not from the cochlea [14,16,17,18]. The threshold for eliciting the VEMP response is lower in an ear affected with superior canal dehiscence than in a normal ear [10–12,19]. This is true for both air-conduction and bone-conduction stimuli [13]. The mechanism for this is still under investigation but is postulated to be the fact that the sacculle lies directly in the pathway between the stapes footplate and the mobile third window (the dehiscence), causing it to be more sensitive than usual to sound stimuli. The abnormal VEMP threshold can be normalized by surgical repair of the dehiscence [13] (Fig. 4).

Some patients with the vestibular symptoms and signs indicative of superior canal dehiscence syndrome have also been noted to have auditory manifestations of the disorder. The Weber tuning fork test typically lateralizes to the affected ear, and patients may also hear a tuning fork placed on the lateral malleolus of the foot [5,10]. Bone conduction thresholds on audiometry can be less than 0 dB normal hearing level. Therefore, an air–bone gap can exist even when air conduction thresholds are normal [12,20].

Autophony is a common complaint in patients with superior canal dehiscence. Patients can also complain of seemingly bizarre symptoms such as hearing their eye movements or hearing their pulse. In a study examining the otologic manifestations of superior canal dehiscence, Zhou et al. [21] found that the signs and symptoms of superior canal dehiscence can be very similar to other otologic conditions, including patulous Eustachian tube. The authors found that patients with patulous Eustachian tube tend to experience autophony of their voice and breathing with equal loudness whereas patients with superior canal dehiscence tend to complain of autophony of their voice and other facilitated bone-conducted sounds but not breathing. Interestingly, the signs and

Figure 2 The axis of slow-phase eye velocity corresponding to the data shown in Figure 1
symptoms of both conditions can improve with the patient in the supine position.

Patients with superior canal dehiscence can present predominantly with auditory signs and symptoms [20,22]. In a study on auditory manifestations of superior canal dehiscence, four patients with dehiscence of bone overlying the superior canal were found to have air–bone gaps in the affected ears that were greatest at lower frequencies and averaged $24 \pm 7$ dB over the frequency range 250–4000 Hz [20]. Three of these patients had undergone stapedectomy before identification of superior canal dehiscence. The air–bone gap was unchanged postoperatively. Each patient had an intact VEMP response from the affected ear, a finding that would not have been expected based upon a middle ear cause of conductive hearing loss. One patient underwent resurfacing of the superior canal through a middle fossa approach. Postoperatively, his vestibular symptoms were relieved, and his air conduction thresholds were improved by 20 dB. Thus, patients with superior canal dehiscence can have an apparent conductive hearing loss, because the third window formed by the dehiscence causes a dissipation of acoustic energy transmitted through air conduction mechanisms [23]. The clinical manifestations of superior canal dehiscence include vestibular signs and symptoms in some patients, vestibular and auditory abnormalities in others, and exclusively auditory findings in others. A recent study found that patients with larger dehiscences ($\geq 2.5$ mm) presented predominantly with both vestibular and auditory abnormalities, whereas patients with smaller dehiscences present with either auditory or vestibular dysfunction [24*]. The study also found that the location of the dehiscence did not have any influence on the
clinical manifestations in these patients. The dehiscence size in this study was estimated using temporal bone CT scans, which may not be as accurate as intra-operative measurement of dehiscence size.

Patients who experience only mild effects of the symptoms and signs associated with superior canal dehiscence may not require any specific treatment other than avoidance of the stimuli that evoke these effects. Placement of a tympanostomy tube may be beneficial in some patients with principally pressure-induced symptoms. For patients who are debilitated by symptoms associated with the syndrome, surgical repair of the superior canal through a middle cranial fossa approach can be effective in alleviating or attenuating the vestibular signs and symptoms [1,2,11,25]. It has been found that plugging the canal dehiscence provided better outcomes than resurfacing [26]. Surgical repair of SCD is effective at alleviating both vestibular and auditory symptoms [20,26,27]. More recently, a transmastoid approach for superior canal plugging has been described [28]. The advantages for this technique include the lack of craniotomy and temporal lobe retraction. However, this technique may not be feasible when the dura is low hanging or when there is extensive cranial base dehiscence requiring reconstruction. The transmastoid approach also does not allow for visual confirmation of the dehiscence and confirmation that the plugs are placed proximal and distal to the full extent of the dehiscence.

**Lateral semicircular canal dehiscence caused by chronic otitis media**

Chronic otitis media can result in erosion of the dense otic capsule bone surrounding the structures of the labyrinth, leading to fistula formation. The most common site of labyrinthine fistula in the setting of chronic otitis media is the lateral semicircular canal [29]. Figure 5 shows CT
images of the right temporal bone in a patient with an erosion of the right lateral semicircular canal.

In a temporal bone histopathology study, Jang and Merchant [30] found that labyrinthine fistula can be caused by chronic otitis media with cholesteatoma, or chronic granulomatous otitis media without cholesteatoma. The proposed mechanisms for bony erosion were chronic infection and inflammation leading to resorption osteitis from osteoclastic activation and osteolysis resulting from either direct pressure by cholesteatoma or by the production of osteolytic compounds by the cholesteatoma matrix. Hakuba et al. [29] reviewed 375 revision surgeries performed for recurrent chronic otitis media during a 22-year period at their clinic. Labyrinthine fistulae were recognized at revision surgery in 29 ears. The previous primary surgery in these cases had been intact canal wall mastoidectomy in four patients and canal wall down mastoidectomy in 25 patients. Overall, the incidence of fistula was 2% in the revisions of previous intact canal wall procedures and 13% in revisions of previous canal wall down procedures. This is most likely due to the extensive nature of the disease process in cases which required canal wall down surgery. In those patients with canal wall down procedures, all 25 patients experienced vertigo, 13 experienced otorrhea, hearing loss was noted in three, headache in one, and facial palsy in one. Spontaneous nystagmus was noted in six cases: toward the side of the involved ear in three cases and toward the contralateral side in three. A fistula test conducted with a Politzer bulb was positive in 14 ears and negative in five ears. In the remaining six ears, pressure in the ear canal induced a sensation of vertigo without accompanying nystagmus.

The management of lateral canal fistula is controversial. Some advocate complete removal of cholesteatoma matrix and repair of canal fistula. Surgical repair can be accomplished using conchal cartilage, bone paste (bone dust mixed with fibrin glue), or temporalis fascia after the inflammatory granulation tissue has been removed.

Others advocate leaving a thin layer of cholesteatoma matrix intact in order to prevent labyrinthine exposure and potential deafness associated with it. In a review article by Copeland and Buchman [31], they found that studies that left the cholesteatoma matrix intact had a hearing preservation rate of 83% postoperatively, similar to the hearing preservation rate of 84% in studies that removed the cholesteatoma matrix. More recently, Chen et al. [32] described using canal occlusion technique to treat lateral canal fistula after cholesteatoma has been completely removed, in a manner similar to that used in the transmastoid superior canal dehiscence repair. They found that only two out of 22 patients in their study had further hearing loss after surgical repair (5–15 dB decrease in postoperative hearing), and the remaining 20 patients had stable or improved hearing after surgical repair.

**Posterior semicircular canal dehiscence**

Posterior semicircular canal dehiscence is a rare clinical entity (Fig. 6). It has been described in association with high-riding jugular bulb [33] and fibrous dysplasia [34]. In a case series of 12 patients with posterior semicircular canal dehiscence, Gopen et al. [35] found that these patients can present with auditory as well as vestibular complaints. All patients in this case series had hearing loss, and two patients had suprathreshold bone conduction. VEMP testing showed decreased threshold in all patients. One of the patients presented with conductive hearing loss and underwent an unsuccessful stapedectomy procedure. It is interesting that two patients in this series had evidence of superior canal dehiscence as well, indicating that there may be an underlying abnormality in bone development which could account for canal dehiscence in general.

Figure 6 Computed tomography of the temporal bone demonstrating a posterior semicircular canal dehiscence into the jugular fossa in both axial and coronal views
Conclusion
Dehiscence of the bone covering the semicircular canals can be caused by various pathologic processes, including trauma, infection, cholesteatoma, and congenital developmental abnormalities. This results in a disruption of the normal mechanics of the inner ear, which leads to vertigo and/or hearing loss. Patients may experience Tullio phenomenon (eye movements and vertigo evoked by loud noises) as well as Hennébert sign (eye movements and vertigo induced by pressure in the external auditory canal). The direction of these evoked eye movements provides important insight as to the location of the abnormality. High-resolution CT scans can be beneficial in evaluation of the underlying abnormality. Surgical repair of the dehiscent canal is effective at alleviating patient symptoms.

References and recommended reading
Papers of particular interest, published within the annual period of review, have been highlighted as:
• of special interest
•• of outstanding interest

Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 90).

5 Ewald JR. Physiologic investigation of the end-organs of the eighth nerve [in German]. Germany: Bergmann; 1892.
This article summarizes the current evidence on VEMP.
This article examines the correlation between dehiscence size and clinical findings.
This article is the first case series of 12 patients with posterior semicircular canal dehiscence.