Superior semicircular canal dehiscence syndrome

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Introduction

This article includes discussion of superior semicircular canal dehiscence syndrome and SSCD. The foregoing terms may include synonyms, similar disorders, variations in usage, and abbreviations.

Overview

The author explains the clinical presentation, pathophysiology, diagnostic work-up, and management of superior semicircular canal dehiscence syndrome.

Key points

• The Tullio phenomenon is sound-induced vertigo, nystagmus, or both.

• Hennebert sign is pressure-induced vertigo, nystagmus, or both, elicited by insufflation of the external auditory canal.

• Typically, in affected patients with superior semicircular canal dehiscence syndrome, there is a several-year history of symptoms that may include sound-induced vertigo (Tullio phenomenon), pressure-induced vertigo (Hennebert sign), or oscillopsia as well as chronic disequilibrium.

• Characteristic signs include sound-induced eye movement and head tilt, Valsalva-induced eye movement, Hennebert sign, and postural sway induced by external auditory canal pressure. Other signs can include pulse-synchronous rotatory nystagmus, hyperacusis to bone-conducted sounds, and conductive hearing loss.

• Patients with bilateral superior semicircular canal dehiscence may have vertical oscillopsia and impaired vision during locomotion, disequilibrium, spontaneous pulse-synchronous vertical pendular nystagmus, and Valsalva-induced upbeat jerk nystagmus.

• Superior semicircular canal dehiscence syndrome is caused by missing bone over the superior aspect of a semicircular canal, creating a third "mobile window" in the bony labyrinth. This may be a developmental abnormality.

• High-resolution computed tomography of the temporal bones is usually considered the definitive test for superior semicircular canal dehiscence.

• Patients also have lowered vestibular-evoked myogenic potential thresholds and larger vestibular-evoked myogenic potential amplitudes; vestibular-evoked myogenic potential studies are highly sensitive and specific for superior canal dehiscence (although rare patients with posterior canal dehiscence will also have abnormal vestibular-evoked myogenic potentials).

• Patients with superior semicircular canal dehiscence and disabling disequilibrium have benefited from surgically plugging or patching (ie, resurfacing or "reroofing") the dehiscent superior semicircular canal through a middle cranial fossa approach or, more recently, with a less complicated and potentially safer transmastoid approach.

Historical note and terminology

The Tullio phenomenon is sound-induced vertigo, nystagmus, or both. In experiments conducted during the first half of the 20th century, Italian biologist and physiologist Pietro Tullio (1881–1941), Dutch otolaryngologist Eelco Huizinga (1893–1976), AJH van Eunen, and colleagues established that (1) fenestration of individual semicircular canals made pigeons susceptible to sound-evoked eye and head movements in the plane of the fenestrated canals and (2) these responses were transiently abolished by applying cocaine to the ampulla of the fenestrated canal (Tullio 1929; Huizinga 1934;1935; Van Eunen et al 1943; Addams-Williams et al 2014).

Hennebert sign is pressure-induced vertigo, nystagmus, or both elicited by insufflation of the external auditory canal.
Belgian otolaryngologist Camille Hennebert (1867–1954) originally reported this phenomenon in syphilitic patients (Hennebert 1911).

Both the Tullio phenomenon and Hennebert sign can occur together in patients (Cawthorne 1956; Erlich and Lawson 1980; Minor et al 1998), presumably because of disruption of the bony labyrinth (Cawthorne 1949; Cawthorne 1956). Examples include erosion of bone of the horizontal semicircular canal by cholesteatoma and surgical fenestration of the horizontal semicircular canal as a treatment for hearing loss in otosclerosis (Minor et al 1998).

Superior semicircular canal dehiscence syndrome is a newly recognized vestibular disorder, first reported by American otolaryngologist Lloyd Minor and colleagues in 1998. Characteristic clinical features of the disorder include both Tullio phenomenon and Hennebert sign (Minor et al 1998; Adams and Levine 2011; Basura et al 2014).

**Clinical manifestations**

**Presentation and course**

Superior semicircular canal dehiscence has been reported predominantly in adults, with roughly equal numbers of men and women. Typically, affected patients have a several year history of audio-vestibular symptoms that may include sound-induced vertigo (Tullio phenomenon), pressure-induced vertigo (Hennebert sign), or oscillopsia as well as chronic disequilibrium (Minor et al 1998; Minor et al 2001; Brantberg et al 1999; Watson et al 2000; Hillman et al 2006; Wilkinson et al 2007; Chi et al 2010; Kaski et al 2012; Shuman et al 2012; Basura et al 2014). Patients may report symptoms with loud noises, humming, coughing, sneezing, or straining (Minor et al 1998; Minor et al 2001; Brantberg et al 1999; Strupp et al 2000; Hillman et al 2006; Wilkinson et al 2007; Zhou et al 2007; Basura et al 2014). Other symptoms can include autophony of voice, aural fullness, pulsatile tinnitus, head rotation, induced tinnitus, transient unsteadiness and gaze instability after rapid head movements, positional vertigo, and drop attacks (Brantberg et al 1999; Brantberg et al 2001; Brantberg et al 2005; Mong et al 1999; Smullen et al 1999; Minor et al 2001; Hillman et al 2006; Wilkinson et al 2007; Zhou et al 2007; Matteo and Hudgins 2008; Puwanarajah et al 2008; Chi et al 2010; Nam et al 2010; Basura et al 2014). Symptoms are often increased during upper respiratory infections (Brantberg et al 1999; Brantberg et al 2001). Some cases may be bilateral (Albuquerque and Bronstein 2004; Deutschlander et al 2004; Tilikete et al 2004; Kanaan et al 2011). Superior semicircular canal dehiscence may also be asymptomatic (Brantberg et al 1999; Albuquerque and Bronstein 2004).

Characteristic signs include sound-induced eye movement and head tilt, Valsalva-induced eye movement, Hennebert sign, and postural sway induced by external auditory canal pressure (Minor et al 1998; Hillman et al 2006; Chi et al 2010; Basura et al 2014). When Tullio phenomenon exists in the superior semicircular canal dehiscence syndrome, loud sound induces a mixed vertical-torsional nystagmus, in which the slow phase rotates up and away from the affected ear, ie, in the plane of the dehiscent semicircular canal (Basura et al 2014). Other signs can include pulse-synchronous rotatory nystagmus (Younge et al 2003). Occasional patients may have a positive Dix-Hallpike test (Smullen et al 1999), although it is not entirely clear that this is not coincidental. Some patients may have hyperacusis to bone-conducted sounds (Minor et al 2001; Cox et al 2003; Halmagyi et al 2003a; Rosowski et al 2004), conductive hearing loss (Cox et al 2003; Minor et al 2003; Mikulec et al 2004; Rosowskii et al 2004; Hillman et al 2006; Wilkinson et al 2007; Martin et al 2009; Chi et al 2010), profound sensorineural hearing loss (Chi et al 2010), mixed hearing loss (Martin et al 2009; Chi et al 2010), and normal hearing (Chi et al 2010). Conductive hearing loss tends to be in the low frequencies, whereas sensorineural loss generally affects the high frequencies (McEvoy et al 2013).

Different investigators have reported conflicting results concerning whether there is a relationship between the size of dehiscence and the severity of vestibular symptoms or the degree of hearing loss, with some reporting no association (Chi et al 2010) and others reporting that larger superior canal dehiscences show significantly more vestibulocochlear symptoms or signs, lower vestibular evoked myogenic potential thresholds, and objective vestibular findings compared with smaller ones (Pfammatter et al 2010). Furthermore, an air-bone gap is consistently shown at the low frequency when the dehiscence is larger than 3 mm; the size of the average air-bone gap correlates with the size of the dehiscence (Yuen et al 2009).

Patients with bilateral superior semicircular canal dehiscence may have vertical oscillopsia and impaired vision during locomotion, disequilibrium, spontaneous pulse-synchronous vertical pendular nystagmus, and Valsalva-induced upbeat jerk nystagmus (Deutschlander et al 2004; Tilikete et al 2004).
A similar clinical disorder may occur with dehiscence of bone over the posterior semicircular canal (Krombach et al 2003; Mikulec and Poe 2006; Zhou et al 2007; Paladin et al 2008; Gopen et al 2010; Peress et al 2015). As with dehiscence of the superior semicircular canal, associated hearing loss may be mixed, conductive, or sensorineural, with downward-sloping configuration being the most common (Gopen et al 2010). Vestibular symptoms are more common in the adult patients with dehiscence of the posterior semicircular canal than in the pediatric patients, with chronic disequilibrium the most common complaint (Gopen et al 2010; Lee et al 2011). An anecdotal report indicates that posterior canal dehiscence may present clinically as Meniere disease (Peress et al 2015).

Prognosis and complications

About half of the reported patients have developed chronic disequilibrium, which in some cases is severe and debilitating (Minor et al 1998). Few longitudinal data are available on untreated patients. Although few longitudinal data are available on untreated patients, it appears that progression of symptoms can be associated with a wider air-bone gap on audiometry, lower cVEMP thresholds, and a larger bony defect (Lookabaugh et al 2016).

Clinical vignette

At 22 years of age, a woman noted vertical oscillopsia induced by loud noises in the left ear (Minor et al 1998). She experienced vertigo with Valsalva maneuver or pressure in the left external auditory canal. A presumptive diagnosis of perilymph fistula prompted exploratory surgery of the middle ear, but the ossicular chain was normal and no leakage of perilymph was identified at surgery. Similarly, medical treatment with a low-salt diet, diuretics, and vestibular suppressants (apparently for a presumptive diagnosis of Ménière syndrome) was not beneficial in relieving her symptoms.

Sound-induced symptoms increased over the 2-year period following her initial surgery, and she developed persistent disequilibrium that was exacerbated by head movements. Examination showed normal eye movements with no spontaneous, gaze-evoked, or post-head-shaking nystagmus. Pure tones from 500 to 1500 Hz at an intensity of 100 dB in the left ear elicited upward, clockwise movement of the eyes (from the patient's perspective). Either positive pressure insufflation of the left external auditory canal or Valsalva maneuver against pinched nostrils elicited a vertical-torsional nystagmus, with upward, clockwise slow phases (from the patient's perspective). Release of Valsalva, Valsalva maneuver against a closed glottis, jugular venous compression, or negative pressure insufflation of the left external auditory canal produced an oppositely directed nystagmus. Neither pure tones nor insufflation of the right ear produced eye movement. Audiometry, electronystagmography, MRI, syphilis serologies, and Lyme titers were normal. Coronal computed tomography of the temporal bones demonstrated dehiscence of the bone overlying the left superior semicircular canal.

She was treated with plugging of the superior semicircular canal through a middle cranial fossa approach. At surgery, there was an absence of bone over the superior portion of the superior semicircular duct in the area of the arcuate eminence. The canal was packed with fascia, bone dust, and fibrin glue. Cortical bone was replaced over the plug. Postoperatively her hearing was unchanged. Her sound-induced and pressure-induced symptoms in the right ear and her disequilibrium resolved completely, although she continued to have nystagmus evoked by Valsalva maneuvers.

Biological basis

Etiology and pathogenesis

Superior semicircular canal dehiscence syndrome is caused by missing bone over the superior aspect of a semicircular canal, creating a third “mobile window” in the bony labyrinth. It is likely that there are congenital and acquired forms of the disorder.

The disorder may be a developmental anomaly (Minor et al 1998; Brantberg et al 1999; Tsunoda and Terasaki 2002; Hirvonen et al 2003; Kuhn and Clenney 2010; Hegemann and Carey 2011; Fraile Rodrigo et al 2016; Sugihara et al 2016). At least 4 families have been reported in which 2 first-degree relatives had superior semicircular canal dehiscence syndrome, suggesting that genetic factors may contribute to the disorder (Brantberg et al 1999; Niesten et al 2014); nevertheless, most cases have been sporadic (Minor et al 1998; Brantberg et al 1999; Mong et al 1999; Smullen et al 1999; Cremer et al 2000; Minor 2000). Findings of multiple malformations of temporal bone structures in anecdotal cases with superior semicircular canal dehiscence syndrome lend support to the theory that semicircular canal dehiscence may have an underlying developmental or congenital etiology (Wang and Parnes 2010; Brandolini
and Modugno 2011); indeed, semicircular canal anomalies are common in large vestibular aqueduct syndrome, most commonly dehiscence of the superior or posterior semicircular canals (Ma et al 2009; Brando­lini and Modugno 2011). Radiographic superior semicircular canal dehiscence is more prevalent in younger children, especially infants younger than 12 to 24 months, indicating that the bony covering of the superior semicircular canal develops normally at a young age (Jackson et al 2015; Sugihara et al 2016).

On the other hand, the increased radiologic prevalence of superior semicircular canal dehiscence among older individuals suggests that some cases are acquired (Nadgir et al 2011). Only a few patients have been reported with a history of head trauma or a history supporting possible changes in middle ear or intracranial pressure prior to symptom onset (Minor et al 1998). There has been no documented history of recent otologic infection or evidence of infection at surgery (Minor et al 1998). Rare cases may be caused by other local pathology that results in erosion of the canal, such as an enlarged superior petrosal sinus draining a large cerebellar developmental venous anomaly (Puwanarajah et al 2008; McCall et al 2011). Temporal bone histopathology in some cases is consistent with the idea that superior canal dehiscence may result from failure of postnatal bone development, and that minor trauma may disrupt the thin bone or dura overlying the superior canal (Teixido et al 2012). Patients with superior semicircular canal dehiscence have a higher prevalence of obesity, higher rates of obstructive sleep apnea, and are more likely to have accompanying tegmental defects, but the causal or non-causal nature of these associations is unclear (Schutt et al 2015).

Horizontal canal dehiscence is typically, but not exclusively, associated with chronic otitis media (Chien et al 2011; Zhang et al 2011).

Posterior semicircular canal dehiscence is relatively rare, but may occur as a developmental anomaly and may be associated with other developmental abnormalities (Chien et al 2011). The prevalence of Chiari type 1 malformations is elevated in patients with semicircular canal dehiscence, especially among patients with posterior canal dehiscence (Kuhn and Clenney 2010). Most patients with posterior semicircular canal dehiscence have dehiscence into a high-riding jugular bulb, but some may have otic capsule dysplasias, eg, enlarged vestibular aqueduct with a Mondini malformation (Gopen et al 2010).

Under normal physiologic conditions, sound is transmitted from the ossicular chain in the middle ear, through the oval window, to the cochlea where it stimulates hair cells on the basilar membrane to produce the sensation of sound. The cochlear round window allows the sound pressure to dissipate from the cochlea back to the middle ear. Normally, the vestibular apparatus does not have a mobile window, so pressure remains relatively constant and the vestibular end organs are not stimulated by sound, by pressure applied to the external auditory canal, or by Valsalva-induced changes in middle ear or intracranial pressure.

A defect in the bony labyrinth can function as a “third mobile window” (ie, in addition to the oval and round windows), allowing transmission of sound and pressure to the vestibular apparatus (Cawthorne 1956; Minor et al 1998; Hirvonen et al 2001). The resulting clinical manifestations are the Tullio phenomenon and Hennebert sign.

With superior semicircular canal dehiscence specifically, the bony defect allows sound or pressure to initiate endolymph flow in the superior semicircular canal. Sound, positive pressure in the external auditory canal, and Valsalva maneuver against pinched nostrils (ie, inflating the Eustachian tubes and middle ears) all increase middle ear pressure, which is in turn transmitted to the vestibule, causing both excitatory (ie, ampullofugal) deflection of the cupula of the superior semicircular canal and an outwardly directed pressure gradient in the area of the bony defect (Minor et al 1998; Hirvonen et al 2001). Stimulation of the superior semicircular canal ampullary nerve causes upward and torsional movements of both eyes in the plane of the stimulated canal (ie, intorsion in the ipsilateral eye and extorsion in the contralateral eye) (Cremer et al 2000; Ostrowski et al 2001). A high-magnitude, low-threshold vestibulo-ocular reflex that aligns with the superior canal can be evoked by air-conducted clicks, suggesting that the superior canal is hypersensitive to sound (Aw et al 2006). In contrast, negative pressure in the external auditory canal, jugular venous compression, and Valsalva maneuver against a closed glottis all produce inhibitory (ie, ampullopetal) deflection of the cupula of the superior semicircular canal and an inwardly directed pressure gradient in the area of the bony defect (Minor et al 1998). Jugular venous compression and Valsalva maneuver against a closed glottis both increase intracranial pressure (because of impaired venous outflow), which in the presence of superior semicircular canal dehiscence is transmitted directly to the apex of the superior semicircular canal, producing ampullopetal endolymphatic flow.
Some patients may present with an air-bone gap on audiometry and apparent conductive hearing loss (Cox et al 2003; Halmagyi et al 2003a; Minor et al 2003; Merchant and Rosowski 2008; Luers et al 2015). The length of dehiscence measured intraoperatively correlates with the maximum air-bone gap (Chien et al 2012). Minor proposed that the third mobile window caused by the superior semicircular canal dehiscence produces dissipation of acoustic energy and a conductive hearing loss (Minor et al 2003). More specifically, the pathological third window allows air-conducted sound energy entering the vestibule through the stapes to be shunted away from the cochlea, particularly at low frequencies (Merchant and Rosowski 2008). Above-normal thresholds for bone-conducted sound may also occur due to a lowering of the impedance on the scala vestibule side of the cochlear partition. Animal studies support Minor's proposed mechanism: in chinchillas, surgically induced superior canal dehiscence will produce a decrease in cochlear potentials in response to low-frequency sound stimuli that reverses when the dehiscence is patched with cyanoacrylate glue (Songer and Rosowski 2005; Songer and Rosowski 2007). However, in some cases, superior semicircular canal dehiscence can produce a conductive hearing gain, causing diagnostic confusion and potentially inappropriate management, including surgery (Halmagyi et al 2003a). In addition, studies of the effects of a superior semicircular canal dehiscence in fresh cadaveric human temporal bones found a much smaller conductive abnormality than that reported in some clinical in vivo studies, suggesting that other mechanisms may be involved in cases with large conductive losses (Luers et al 2015).

Anterior canal hypofunction may result from dural tissue plugging the membranous canal through the dehiscent bone, particularly with dehiscences larger than 5 mm (Cremer et al 2000; Deutschlander et al 2004). When bilateral, symptoms suggestive of bilateral vestibular failure may result (Deutschlander et al 2004).

**Epidemiology**

Limited epidemiologic information on superior semicircular canal dehiscence is available. Superior semicircular canal dehiscence syndrome is probably not rare and has undoubtedly been under-recognized for several reasons: (1) some cases are asymptomatic (Smullen et al 1999); (2) many of the diagnosed cases were initially diagnosed as other conditions (Minor et al 1998; Brantberg et al 1999; Smullen et al 1999; Halmagyi et al 2003a); (3) superior semicircular canal dehiscence is present in temporal bone specimens from 0.6% to 0.7% of individuals, and the bone overlying the superior semicircular canal is markedly thinned in an additional 1.3% of individuals (Carey et al 2000; Crovetto et al 2010); and (4) CT scanning gives a higher prevalence of superior semicircular canal dehiscence than anatomical studies, with estimates as high as 3.6% (Crovetto et al 2010; Masaki 2011), although higher rates are seen when studies are restricted to children (Sugihara et al 2016).

In a CT study of 503 children less than 18 years of age, all of whom had undergone imaging including the temporal bones, the incidence of superior semicircular canal dehiscence was 6.2% (Sugihara et al 2016). The prevalence of superior semicircular canal dehiscence is highest under age 2 years and then progressively decreases with age: less than 2 years, 36.7%; 2 to 8 years, 5.6%; and 9 to 18 years, 3.2% (Sugihara et al 2016). This supports a developmental basis for superior semicircular canal dehiscence, with overlying bone maturation occurring normally during early childhood (Sugihara et al 2016).

Thinning or dehiscence of the superior semicircular canal may occur on the middle cranial fossa floor or adjacent to the superior petrosal sinus (Koo et al 2010). A deep groove of the superior petrosal sinus may cause superior canal dehiscence close to the common crus, and costimulation of the superior and posterior canals in this circumstance may explain the mainly torsional nystagmus induced by sound and vibration stimuli (Koo et al 2010).

CT studies have generally given estimates of the prevalence of posterior canal dehiscence from 0.6% to 1.2% (Crovetto et al 2010; Erdogan et al 2011).

**Differential diagnosis**

The key symptoms of superior semicircular canal dehiscence are sound-induced and pressure-induced vestibular symptoms and chronic disequilibrium (Minor et al 1998), which are findings that have been reported in a number of other conditions. The Tullio phenomenon (Tullio 1929) has been associated with syphilitic labyrinthitis, perilymphatic fistulas (Fox et al 1988), Ménière disease, congenital deafness (Kwee 1976), and other conditions (Nields and Kueton 1991). Hennebert sign has been associated with syphilitic labyrinthitis (Hennebert 1911), perilymph fistula (Kohut et al 1979), and Ménière syndrome (Nadol 1977).
Several patients with superior semicircular canal dehiscence were initially thought to have perilymph fistulas and underwent middle ear exploratory surgery without benefit (Minor et al 1998; Mong et al 1999). Another patient was treated surgically for middle ear and mastoid disease, which delayed the diagnosis of superior semicircular canal dehiscence (Ramsey et al 2004). In addition, apparently some patients were initially considered to have Ménière syndrome and were treated with a low-salt diet, diuretics (eg, triamterene, hydrochlorothiazide, acetazolamide), benzodiazepines (eg, lorazepam, clonazepam), and endolymphatic sac decompression and vestibular nerve section without benefit (Minor et al 1998; Mong et al 1999; Smullen et al 1999). Superior semicircular canal dehiscence syndrome may also be confused with otosclerosis (Cox et al 2003; Halmagyi et al 2003a; Minor et al 2003; Mikulec et al 2004; Zhou et al 2007; Van Rompaey et al 2011), benign paroxysmal vertigo (Brantberg et al 1999) or psychosomatic illness (Minor et al 1998). In all reported cases, syphilis serologies and Lyme titers have been normal.

Superior canal dehiscence can be confused with dehiscence of bone over the posterior semicircular canal (Krombach et al 2003; Mikulec and Poe 2006; Zhou et al 2007). In a case of suspected superior canal dehiscence based on high-resolution CT with an abnormal vestibular evoked myogenic potential study, surgical exploration failed to identify a superior canal dehiscence and instead found an unexpected posterior canal dehiscence (Zhou et al 2007).

Some patients may present with an air-bone gap on audiometry and apparent conductive hearing loss, which may be confused with otosclerosis or other disorders of the middle ear (Cox et al 2003; Halmagyi et al 2003a; Minor et al 2003; Mikulec et al 2004; Zhou et al 2007; Van Rompaey et al 2011).

One patient has been reported with a superior canal dehiscence close to the common crus related to a venous malformation (Brantberg et al 2004a). Superior canal dehiscence syndrome should be considered in the differential diagnosis of patients with persistent audiovestibular symptoms after stapes surgery (Hope and Fagan 2010); high-resolution computed tomography of the temporal bone and vestibular evoked myogenic potential testing, if available, have the greatest utility in confirming the diagnosis.

Bilateral cases can resemble bilateral vestibular failure clinically, although caloric testing is normal (Deutschlander et al 2004).

**Diagnostic workup**

Clinical examination should include assessment for sound-, pressure-, and vibration-induced vertical-torsional nystagmus. Effects of pure tones can be assessed with standard earphones used in audiometric testing. Effects of pressure can be assessed by positive- and negative-pressure insufflation of the external auditory canal; by manually occluding the external canal with the tragus and then pushing, with a Valsalva maneuver, against pinched nostrils or a closed glottis; and by jugular venous compression (Minor et al 1998). Effects of vibration can be assessed using a 100 Hz vibrator for 10 to 15 seconds applied to the vertex, bilateral mastoid, or bilateral occipital cranial areas of patients in the seated position, although the vertical-torsional nystagmus is usually best observed with suboccipital vibration applied on the side of dehiscence (White et al 2007; Manzari et al 2008). The nystagmus is suppressed with fixation; therefore, examination should be performed with Frenzel lenses (Minor et al 1998; Cremer et al 2000).

Loud sounds with pure tones (100 dB, 110 dB nHL) can be used to stimulate both ears for 5 seconds at frequencies of 500, 1000, and 2000 Hz. In preliminary studies, such loud sounds elicit vertigo in approximately 80% of cases, with characteristic associated eye movements (rotational with superior canal dehiscence and horizontal with lateral canal dehiscence) (Yu et al 2011).

High-resolution computed tomography of the temporal bones is usually considered the definitive test. High-resolution computed tomography of the temporal bones demonstrates dehiscence of bone overlying the affected semicircular canal (Minor et al 1998; Mong et al 1999; Smullen et al 1999; Cremer 2000; Minor 2000; Williamson et al 2003; Wilkinson et al 2007; Manzari et al 2008; Chi et al 2010). Some studies have suggested a low specificity of CT (Williamson et al 2003; Re et al 2013), so it is important to correlate CT findings suggestive of superior canal dehiscence with clinical symptoms. The diagnostic utility of computed tomography reportedly improves with 0.5 mm-collimated helical CT and reformation in the plane of the superior semicircular canal (Belden et al 2003), although another study suggests that coronal reformation from multi-detector row CT of the temporal bone are sufficient for the evaluation of superior semicircular canal dehiscence (Branstetter et al 2006; Thabet et al 2012). In some cases,
the defect may be bilateral (Smullen et al 1999; Thabet et al 2012). In contrast, routine cerebral magnetic resonance imaging, with and without gadolinium, is often normal (Minor et al 1998; Mong et al 1999; Smullen et al 1999; Cremer 2000; Minor 2000). Other studies have also been normal or have shown mild, inconsistent, and nonspecific abnormalities (Minor et al 1998; Mong et al 1999; Smullen et al 1999; Cremer 2000; Minor 2000). Some steady-state free precession gradient-echo pulse sequences have demonstrated high sensitivity and high negative predictive value so that negative findings on MRI using FIESTA (Fast Imaging Employing Steady-state Acquisition) sequence effectively eliminate the need to use CT to detect semicircular canal dehiscence; only patients with positive findings on MRI then require CT (Browaeys et al 2013).

Weber's test may lateralize to the symptomatic ear (Brantberg et al 2001). Audiometry generally shows normal pure tone thresholds, normal speech discrimination, normal otocoustic emissions, normal tympanometry (type A tympanograms), and normal acoustic reflexes (Minor et al 1998; Mong et al 1999; Smullen et al 1999; Cremer 2000; Minor 2000; Banerjee et al 2005; Zhou et al 2007). Some patients may have mild to moderate low-frequency conductive, sensorineural, or mixed hearing loss in the affected ear or in both ears (Minor et al 1998; Brantberg et al 1999; Mikulec et al 2004; Rosowskii et al 2004; Banerjee et al 2005; Zhou et al 2007). Electronystagmography generally shows no localizing abnormalities, including no caloric weakness (Minor et al 1998), although occasional atypical cases may have caloric weakness (Smullen et al 1999). Rotary chair testing is normal or in a minority of patients shows gain asymmetries consistent with vestibular hypofunction in the affected ear (Minor et al 1998; Smullen et al 1999).

Patients also have lowered vestibular evoked myogenic potential thresholds and larger vestibular evoked myogenic potential amplitudes (Brantberg et al 1999; Brantberg et al 2004b; Minor et al 2001; Streubel et al 2001; Halmagyi et al 2003b; Zhou et al 2007; Teixido et al 2008; Welgampola et al 2008; Gopen et al 2010; Manzari et al 2011); vestibular evoked myogenic potential studies are highly sensitive and specific for superior canal dehiscence (although rare patients with posterior canal dehiscence will also have abnormal vestibular evoked myogenic potentials) (Zhou et al 2007). Enhanced sound- and vibration-induced vestibular evoked myogenic potentials and their lower threshold in patients with superior canal dehiscence have been interpreted as being due to the dehiscence, allowing sound and vibration to activate the receptors of the dehiscent semicircular canal. However, a patient with bilateral superior canal dehiscence and bilaterally decreased superior canal function also showed enhanced vestibular evoked myogenic potentials with reduced thresholds, suggesting that enhanced otolithic stimulation by sound and vibration after dehiscence may play a role in producing the characteristic changes in vestibular evoked myogenic potentials with superior canal dehiscence (Manzari et al 2011). Ocular and cervical vestibular evoked myogenic potentials evoked by air-conducted sound are equally useful in diagnosis and follow-up of superior canal dehiscence, whereas thresholds for bone vibration are less useful (Welgampola et al 2008). The n10 component of the ocular vestibular evoked myogenic potential in response to a very high frequency stimulus (4000 Hz), either air-conducted sound or bone conducted vibration, is a fast (“single trial”) indicator of probable superior canal dehiscence (Manzari et al 2013). In contrast, standard techniques demonstrating enhanced amplitude and reduced thresholds of vestibular-evoked myogenic potentials require multiple trials that can be both time consuming and tiring for patients (Manzari et al 2013).

Syphilis serologies, Lyme titers, complete blood count, serum electrolytes, erythrocyte sedimentation rate, rheumatoid factor, antinuclear antibodies, and complement levels have been normal (Minor et al 1998; Brantberg et al 1999; Mong et al 1999).

If a patient has an air-bone gap on audiometry with preserved acoustic reflexes, the patient should be assessed for sound-, pressure-, and vibration-induced nystagmus and should have vestibular evoked myogenic potential testing if available (Cox et al 2003; Halmagyi et al 2003a; Minor et al 2003; Brantberg et al 2004b; Manzari et al 2008). If the patient has either sound-induced or pressure-induced nystagmus with preserved vestibular evoked myogenic potentials, high-resolution spiral CT should be obtained (Halmagyi et al 2003a).

**Management**

In most cases, recognition of the cause of a patient's symptoms allows them to avoid the troublesome stimuli, and no further treatment is required (Minor et al 1998).

Surgical repair of dehiscence can relieve symptoms with low overall morbidity, although patients may have persistent or recurrent symptoms despite surgery, and there is also a risk of hearing loss and vestibulopathy (Chi et al 2010; Kozin et al 2015). Patients with superior semicircular canal dehiscence and disabling disequilibrium have benefited
from surgically plugging, capping (with hydroxyapatite cement), patching (ie, resurfacing or “reroofing”), and plugging with resurfacing the dehiscent superior semicircular canal (Minor et al 1998; Brantberg et al 1999; Brantberg et al 2001; Smullen et al 1999; Minor 2000; Martin et al 2004; Banerjee et al 2005; Mikulec et al 2005; Carey et al 2007; Wilkinson et al 2007; Goddard and Wilkinson 2014; Mueller et al 2014; Gioacchini et al 2016). Surgical treatment is effective for the vestibular symptoms associated with superior canal dehiscence, but there is no evidence for improvement of hearing loss after surgical treatment (Ziylan et al 2017). So far there have been relatively few patients treated surgically, and different authorities disagree as to the relative strengths and weaknesses of the different surgical techniques (Gioacchini et al 2016; Ziylan et al 2017), with some reporting that the different techniques do not significantly differ in terms of either success rate or surgical complications (Gioacchini et al 2016), whereas others report that plugging using transmastoid approach had a lower complication rate, lower revision rate, and a shorter hospital stay (Ziylan et al 2017).

Surgical repair of a dehiscent superior semicircular canal has most commonly been accomplished through a middle cranial fossa approach. Middle fossa repair of superior semicircular canal dehiscence is generally safe and effective with excellent sensorineural hearing preservation (Phillips et al 2010; Chung et al 2016). A transmastoid approach to plugging and resurfacing the dehiscent superior semicircular canal has been developed that does not require a craniotomy and, therefore, also does not involve temporal lobe retraction (and potential damage from this) (Mikulec and Poe 2006; Agrawal and Parnes 2008; Crovetto et al 2008; Teixido et al 2008; Teixido et al 2011; Deschenes et al 2009; Chi et al 2010; Fiorino et al 2010; Amoodi et al 2011; Beyea et al 2012). The transmastoid approach generally requires only an overnight hospital stay (Amoodi et al 2011).

Clinical benefits of canal plugging and reproofing can include resolution of episodic vertigo, mixed hearing loss (Wilkinson et al 2007; Agrawal and Parnes 2008; Goddard and Wilkinson 2014), and reduced dizziness handicap (Crane et al 2008). Sound-induced and pressure-induced symptoms as well as the associated mixed hearing loss may resolve completely with surgery (Minor et al 1998; Wilkinson et al 2007). Vestibular evoked myogenic potential thresholds normalize with canal plugging for superior canal dehiscence (Welgampola et al 2008; Goddard and Wilkinson 2014).

Hypofunction of the operated superior canal is intended, but the function of the other ipsilateral semicircular canals is typically preserved, although vestibular hypofunction of all ipsilateral canals can occasionally occur postoperatively (Carey et al 2007; Agrawal et al 2009; Mantokoudis et al 2016). Immediate postoperative vestibular hypofunction affecting the horizontal canal (as assessed either with bedside horizontal head thrust testing or quantitative video head impulse testing) is common, particularly with larger dehiscences, but typically resolves within 6 weeks postoperatively (Agrawal et al 2009). Postoperative vestibular hypofunction may be due to postoperative labyrinthine inflammation, or to intraoperative loss of perilymph, which may be more likely with larger dehiscences (Agrawal et al 2009; Mantokoudis et al 2016). Full recovery of vestibulo-ocular response gain is typical for the horizontal canal but not always for the posterior canal (Mantokoudis et al 2016).

Endolymphatic hydrops, sensorineural hearing loss, ocular torsion without perceptual tilt, signs of vestibular hypofunction without loss of hearing, transient facial weakness, and benign paroxysmal positioning vertigo are reported complications of surgery (Minor et al 1998; Minor 2000; Strupp et al 2003; Mikulec et al 2005; Ward et al 2012; Goddard and Wilkinson 2014; Barber et al 2016). Surgical plugging via a middle cranial fossa approach is associated with mild high-frequency sensorineural hearing loss that persists in 25%, whereas speech discrimination is generally unchanged (Ward et al 2012). In some cases repair may be possible without plugging (Smullen et al 1999; Minor 2000; Banerjee et al 2005; Mikulec et al 2005).

Poor surgical outcomes may occur from inadequate repair of the dehiscence or from mechanical damage to the membranous labyrinth (Kozin et al 2015). Recurrent or persistent symptoms may necessitate additional plugging procedures (Minor et al 1998). Revision surgery can be curative in carefully selected patients, but there may be a higher failure rate than with the primary surgery (Sharon et al 2016).

Second-side surgery may be prompted by sound- and pressure-induced vertigo in patients with bilateral superior canal dehiscence syndrome (Agrawal et al 2012). Although second-side surgery may be associated with some increased oscillopsia, patients generally find this preferable to the other symptoms of the condition (Agrawal et al 2012).

A novel approach in development to address current operative limitations and improve surgical outcomes is the creation of customized, fixed-length prostheses using 3-dimensional printing technology (Kozin et al 2015).
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**References especially recommended by the author or editor for general reading.

ICD and OMIM codes

ICD codes

ICD-9:
Labyrinthitis: 386.30
Labyrinthitis, syphilitic: 095.8
Vertigo: 780.4
Vertigo, Meniere: 386.00

ICD-10:
Labyrinthitis: H83.0
Labyrinthitis, syphilitic: A52.7
Vertigo NOS: R42
Ménière or vertigo: H81.0

Profile
Age range of presentation
19-44 years
45-64 years
65+ years

Sex preponderance
male=female

Family history
family history may be obtained

Heredity
heredity may be a factor

Population groups selectively affected
none selectively affected

Occupation groups selectively affected
none selectively affected

Differential diagnosis list
syphilitic labyrinthitis
perilymphatic fistulas
Ménière disease
congenital deafness
benign paroxysmal vertigo
psychosomatic illness
otosclerosis
disorders of the middle ear
posterior superior semicircular canal dehiscence

Associated disorders
Meniere syndrome
Perilymph fistula
Syphilitic labyrinthitis

Other topics to consider
Dizziness
Otic capsule dysplasia
Positional vertigo