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Review article

Superior semicircular canal dehiscence: Diagnosis and management

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ABSTRACT

The authors provide an update on the clinical manifestations, diagnosis and various approaches to the treatment of superior semicircular canal dehiscence (SSCD). SSCD is a rare condition where the bone overlying the superior semicircular canal thins or dehisces causing characteristic clinical findings. Since this was first reported in 1998 by Minor and colleagues, there has been much advancement made in terms of diagnosis and treatment. Signs and symptoms include a wide variation of both vestibular and auditory manifestations. Diagnosis made solely on clinical signs is difficult due to how varied the presentations can be and the overlap with other otologic pathologies. High-resolution CT temporal scans have been the standard in confirming superior semicircular canal dehiscence, however, MRI FIESTA scans have recently been used to image SSCD. Additionally, audiometry and vestibular evoked myogenic potential (VEMP) testing are useful screening tools. Currently, the middle fossa approach is the most common and standard surgical approach to repair SSCD. The transmastoid, endoscopic and transcanal or endaural approaches have also been recently utilized. Presently, there is no consensus as to the best approach, material or technique for repair of SSCD. As we learn more, newer and less invasive approaches and techniques are being used to treat SSCD. We present a comprehensive review of SSCD, including clinical symptoms and presentation, histopathology, diagnosis, treatment strategies and outcomes of intervention.

1. Introduction

Superior semicircular canal dehiscence (SSCD) was first described by Minor and colleagues in 1998 as the cause for sound-induced vertigo (Tullio phenomenon) not otherwise explained by infectious causes [1]. Patients experienced either vertigo or oscillopsia after exposure to certain frequencies of sounds or maneuvers which induced pressure changes within the labyrinth. The diagnosis was made radiographically by computed tomography (CT) of the temporal bones and treatment involved plugging the dehiscent area.

Dehiscence of the otic capsule overlying the superior semicircular canal leads to creation of a "mobile third window" of the inner-ear, with the first and second windows referring to the round and oval windows, respectively. When exposed to either high-intensity sounds or pressure changes such as in valsalva maneuvers, pressure changes occur across this third window. Theoretically, the pressure change generates a displacement of endolymph through positive or negative pressure in the external auditory canal towards the dehiscence in the arcuate eminence [2]. This endolymph displacement causes deflection of the cupula resulting in the wide array of symptoms associated with SSCD. The increased elasticity provided by the third window may lead to loss of acoustic energy, which would explain the manifestation of low-frequency conductive hearing loss, but also provide a mechanism for improved cochlear bone conduction by lowering the impedence on the scala vestibuli side of the cochlea [2–5].

The prevalence of SSCD is not well-defined in the literature. There have been several studies to determine its prevalence, however, the diagnosis requires both clinical symptoms and high-resolution CT temporal bone scans, which skew the population being examined. The literature reports a prevalence rate between 0.7 and 9.0% in non-pediatric populations [6–10]. In pediatric populations over 3 years of age, prevalence has been reported to be 10.7% [11]. Patients under 3 years of age are excluded from prevalence studies, because they have not fully formed the temporal bone overlying the superior semicircular canal. A recent publica-

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tion reported a correlation between patients with SSCD and higher body mass index, obstructive sleep apnea rates, and accompanying tegmen defects; suggesting a causal relationship between increased intracranial pressure and formation of SCCD [12].

2. Clinical symptoms and presentation

The range of symptoms seen in SSCD patients are all manifestations of thinning or complete dehiscence of the bone overlying the superior semicircular canal which creates a "third window". The clinical symptoms range from vertical-torsional nystagmus to autophony to bone-conduction hyperacusis (see Table 1). Vestibular manifestations include vertigo, nystagmus and oscillopsia provoked by loud sounds or pressure changes. Auditory manifestations include autophony for internally conducted sounds, bone-conduction hyperacusis on audiometry, pulsatile tinnitus, phonophobia, and aural fullness [1,13–16]. The acoustic vibrations transmitted to the oval window are partially absorbed by this third window manifesting in the distinct symptomatologies described above.

Diagnosis is often challenging based on clinical presentation alone because SSCD patients have various vestibular and auditory symptoms that may mimic other otologic pathologies [13,17,18]. Because vertigo provoked by pressure changes can be associated with other entities where there is leakage of inner ear fluid (i.e. perilymphatic fistulae), it is suspected that numerous cases of SSCD have been mistaken for other conditions.

Many patients exhibit a characteristic torsional eye movement upward and away from the affected ear. This nystagmus can be induced by sound (Tullio phenomenon) or pressure change (Hennebert's sign) when the patient is straining, as during coughing. It occurs when sound or pressure waves are transmitted over the superior canal ampulla and through the dehiscence. This stimulates the superior canal ampulla causing the characteristic nystagmus. The direction of the nystagmus is due to the direction of endolymph flow [15,17]. This nystagmus is specific to SSCD [15,16,19].

Another characteristic sign can be elicited using tuning fork testing. By placing a vibrating fork at the medial malleolus of the ankle, the patient will report being able to hear but not feel the tuning fork in the affected ear (bone-conduction hyperacusis).

A majority of patients with SSCD are acutely sensitive to internally generated or conducted sounds in the affected ear (autophony). Classic examples of autophony are an amplified heartbeat, amplified footstep, and sometimes even being able to hear eyeball movements. The dehiscence acts as a low impedance window allowing the internally generated sounds to be directed into the labyrinth.

Patients with SSCD can be exquisitely sensitive to changes in pressure of the affected ear. Patients often report finding relief from various symptoms by lying supine or sustaining a Valsalva maneuver [18]. During examination, use of the Valsalva maneuver can be exploited to illicit symptoms. The release of the Valsalva maneuver can cause a transient excitation of the superior ampulla due to a pressure change in the middle ear or the intracranial space, similar to dizziness evoked with pressure changes in the external auditory canal (Hennebert's sign).

Table 1
Table of Symptoms.

Symptoms of Superior Semicircular Canal Dehiscence [1,12,14–16]		
Category	Symptom	Description
Auditory	Autophony	Amplified heartbeat, amplified footstep, ability to hear eyeball movements
	Bone-conduction hyperacusis	Test: place vibrating force at medial malleolus of the ankle, patient will report being able to hear but not feel the tuning fork in the affected ear
	Pulsatile tinnitus	May be induced by the patients' own voice
	Low frequency hearing loss	Low frequency sounds exit preferentially through the dehiscence and are lost unless at elevated thresholds
	Phonophobia	Common and debilitating symptom of SSCD
	Aural fullness	Common symptom, but oftentimes tolerable
Vestibular	Tullio phenomenon	Characteristic torsional eye movement upward and away from the affected ear induced by *sound*
	Hennebert's sign	Characteristic torsional eye movement upward and away from the affected ear induced by *pressure changes*
	Oscillopsia	Disturbance of the visual field where objects appear to oscillate; may be induced by loud sound or pressure changes
	Vertigo	May be induced by sound or pressure changes
	Chronic disequilibrium	Thought to be related to persistent abnormal activation of vestibular receptors; most debilitating symptom

3. Histopathologic findings

Infants uniformly have thin bone over the superior semicircular canals, which contributes to the hypothesis that dehiscence may result from failure of postnatal bone development [2]. Furthermore, the thin bone may contribute to a higher chance of dehiscence following trauma and precipitate SSCD. The roof of the superior semicircular canal is composed of layers of the otic capsule, which varies from 0.5 to 0.9 mm in thickness [20]. The erosion of bone is non-focal, and spans along the length of temporal bone overlying the superior semicircular canals.

On histopathology, lamellae can be seen parallel to the surface of the superior petrosal sinus indicating that ossification is stable at one point [7]. This implies that the process leading to dehiscence could be a chronic, progressive condition. However, in 1000 specimens of temporal bones, there were no local bone changes, osteoporotic changes or underlying bone diseases found in any specimens to explain the thinning. Furthermore, the prevalence of otosclerosis is not significantly different in cases of SSCD [7].

4. Diagnosis

The diagnosis of SSCD can be elusive, requiring the clinician to consider all of the following: auditory and vestibular symptoms, audiogram and vestibular evoked myogenic potential (VEMP) testing results, and CT temporal bone scans findings. Although the diagnosis cannot be based on symptoms alone, the combination of autophony for internally-conducted sounds, hyperacusis, and imbalance or vertigo provoked with loud sounds or pressure changes should raise suspicion for SSCD [5]. Similarly, while a wide range of audiogram results can be found in SSCD cases, a low-frequency conductive hearing loss with bone conduction hyperacusis, combined with preserved acoustic reflexes should prompt suspicion. Thus, the diagnosis of SSCD can be further elucidated by the use of audiometry testing.

Conductive hearing loss can be measured by air-bone gaps, which is calculated by the difference between air and bone conduction thresholds on audiograms. The low frequency conductive hearing loss may be explained by the preferential loss of these frequencies through the dehiscence [3–5,17]. Sounds are only able to reach the cochlea at elevated thresholds. However, this low frequency hearing loss is not specific to SSCD and cannot be used as

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