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Superior semicircular canal dehiscence syndrome

CLINICAL REVIEW

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Superior semicircular canal dehiscence syndrome is a condition with troubling ear symptoms and vertigo caused by a defect in the bone between the inner ear and the middle cranial fossa. The disease is not dangerous, and for many patients it is sufficient to provide a thorough explanation for the symptoms and advice about coping strategies, balance exercises and, if necessary, use of assisted hearing devices. Surgical treatment may be appropriate for patients with severe symptoms.

Superior semicircular canal dehiscence syndrome was first described in the late 1990s (1). It was shown that a defect in the superior semicircular canal of the inner ear could cause pressure- or sound-induced vertigo, as well as hypersensitivity to bone-conducted sound. The disease is uncommon, but experience with it is increasing in Norway and internationally, and international diagnostic criteria have been published recently (2). This article is based on the authors' own clinical experiences and a non-systematic literature search in PubMed.

Prevalence

Symptomatic superior semicircular canal dehiscence is a rare condition, but its exact prevalence is still unknown. Asymptomatic dehiscence is rather more common. In a histological study, dehiscence of the superior semicircular canal was demonstrated in temporal bones from 4 out of 596 deceased adults (0.7 %) (3). In an additional 1.3 %, the bone covering was extremely thin (< 0.1 mm). On computed tomography (CT) of the temporal bone, it is difficult to distinguish between thin and entirely absent bone covering, and therefore CT alone will overestimate the prevalence of the condition (4).

Symptoms

The onset of symptoms is often acute in adulthood, and in some cases follows trauma or pressure effects (5). Hypersensitivity to bone-conducted sound means that patients hear their own voice abnormally loudly in the ear

(autophony). Other bodily sounds such as bowel sounds, heartbeat, footsteps and eye movements are also heard more loudly; this phenomenon is called conductive hyperacusis. Pulsatile tinnitus is common. Short episodes of vertigo may be induced by sound (Tullio phenomenon), particularly if the sound is loud and low frequency, or by pressure on the fold in front of the auditory canal (tragus), coughing, nose blowing or Valsalva manoeuvre. The aforementioned symptoms are considered to be typical of superior semicircular canal dehiscence syndrome. Less specific symptoms are also common, such as chronic unsteadiness and vertigo, fatigue, and difficulties with focus and concentration.

Little is currently known about the long-term course of superior semicircular canal dehiscence. The underlying bone defect does not heal spontaneously, but how patients experience symptoms and the degree of impact on everyday life may vary over time, including as a result of individual coping strategies and comorbidities.

Pathogenesis and pathophysiology

The average thickness of the bone between the superior semicircular canal and middle cranial fossa is 0.1 mm at birth, but it increases to an average of 1 mm in adulthood (3). Dehiscence in adulthood seems to be related to a lack of development of the bone in this area, and perhaps also weakening of the tough cerebral membrane (dura) (2), so that pressure can be transmitted between the labyrinth and the middle cranial fossa.

Dehiscence in the superior semicircular canal opens a *third window* into the labyrinth in addition to the round and oval windows (Figure 1). This third window lowers the total impedance of the inner ear, i.e. the ability to withstand oscillating forces from outside. Short episodes of vertigo can occur as a result of increased intracranial pressure associated with coughing, nose blowing or toilet straining because this sends a pressure pulse through the dehiscence and the superior semicircular canal's motion sensor (cupula). This can trigger a brief feeling of rotation (vertigo) and involuntary eye movements (nystagmus). The same effect can be caused by increased middle ear pressure associated with pressure equalisation (Valsalva manoeuvre).

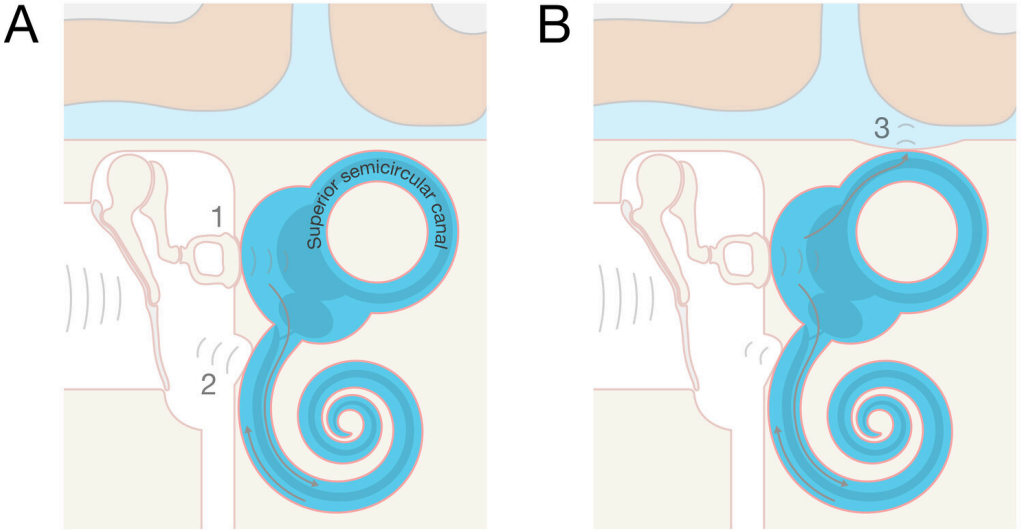

Figure 1 Pathophysiology of superior semicircular canal dehiscence. A) Normal anatomy: Two compliant areas in the labyrinth – the oval window with the stapes (point 1 in the figure) and the round window (point 2). Since fluid is incompressible, when the stapes (point 1) pushes inwards, the round window membrane (point 2) moves outwards. When sound causes the stapes to vibrate, the sound waves will travel through the cochlea causing the round window membrane to vibrate. B) Superior semicircular canal dehiscence: A third window (point 3) between the inner ear and middle cranial fossa causes part of the sound energy to follow the path of least resistance through the dehiscence and away from the cochlea. This is considered to be the probable cause of both the mechanical hearing loss and pressure- and sound-induced vertigo. At the same time, the fluid in the inner ear becomes more mobile so vibrations in the skull (including bone-conducted sound) cause abnormally intense stimulation of the cochlea (6). Illustration: Frederik Kragerud Goplen.

The abnormally loud perception of bone-conducted sound (conductive hyperacusis) is generally explained by lower cochlear impedance leading to increased sensitivity to vibrations in the skull (6).

Audiometry usually shows reduced hearing for air-conducted sound at low frequencies. This hearing loss is probably due to some of the sound energy transmitted via the auditory canal, tympanic membrane and middle ear bones to the vestibule in the inner ear following the path of least resistance away from the cochlea and to the middle cranial fossa via the dehiscence.

Investigations

If there is clinical suspicion of dehiscence, the recommendation is to refer the patient for temporal bone CT with a specific query about superior semicircular canal dehiscence. Diagnosis requires high imaging quality and radiological expertise. It is important for the CT to be taken with the thinnest possible slice thickness and with image reconstruction in the plane of and orthogonal to the superior semicircular canal.

However, symptomatic dehiscence can also occur in other areas of the labyrinth. In the event of positive results of CT scanning, further investigation is performed by an ENT specialist, and diagnosis should preferably be based on

the recently published international criteria for the condition (Box 1) (2).

Box 1 Diagnostic criteria for superior semicircular canal dehiscence syndrome (2).

Clinically definite diagnosis (criteria A–D fulfilled)

A. At least one of the following symptoms consistent with a third window in the inner ear:

1. Hypersensitivity (hyperacusis) to bone-conducted sound. This can include abnormally loud or distorted sound of one's own voice (autophony) and internal bodily sounds such as heartbeat, bowel sounds, eye movements, footsteps etc.
2. Sound-induced vertigo and/or sensation that the visual field is moving (oscillopsia) time-locked to the stimulus.
3. Pressure-induced vertigo and/or oscillopsia time-locked to the stimulus.
4. Pulsatile tinnitus.

B. At least one of the following signs or diagnostic tests indicating a third window in the inner ear:

1. Nystagmus characteristic of excitation or inhibition of the affected superior semicircular canal evoked by sound or by changes in middle ear pressure or intracranial pressure.
2. Low-frequency negative bone conduction thresholds on pure tone audiometry.
3. Enhanced vestibular-evoked myogenic potential (VEMP) responses on the affected side (low cVEMP thresholds or high oVEMP amplitudes)

C. High-resolution temporal bone CT with multiplanar reconstruction demonstrating superior semicircular canal dehiscence.

D. Not better accounted for by another vestibular disease or disorder.

It is important to test for negative bone conduction thresholds on pure tone audiometry, meaning that the patient can hear sounds that are too weak to be perceived by people with normal hearing when the sound is conducted via the skull. Bone conduction can also be tested with a tuning fork (Weber's test at 256–512 Hz), and the sound will then be heard loudly in the affected ear. Some patients hear sound when the tuning fork is placed on the ankle (malleolus). The fistula test is performed by observing eye movements, preferably using infrared video cameras in darkness (video Frenzel), while applying pressure to the tragus or with the Valsalva manoeuvre. In typical cases, pressure stimulation will induce vertigo and eye movements (vertical and rotatory nystagmus). The same can be seen with loud low-frequency sound stimulation (250–500 Hz). Measurement of cervical and ocular vestibular-evoked myogenic potentials (abbreviated as cVEMP and oVEMP respectively) has been

found to be particularly useful in diagnosing superior semicircular canal dehiscence, and demonstrates abnormal sensitivity in the affected ear with decreased cVEMP thresholds and increased oVEMP amplitudes.

Diagnostic criteria

Diagnostic criteria for superior semicircular canal dehiscence syndrome have recently been published based on an international consensus work (2). A clinically definite diagnosis requires radiological dehiscence on temporal bone CT imaging, at least one of four characteristic symptoms and at least one of three characteristic investigation findings. It is also important to identify other causes contributing to the clinical presentation. The main differential diagnoses are patulous Eustachian tube (tuba auditiva), which can cause autophony, as well as Meniere's disease and migraine, which can cause episodic vertigo and sensitivity to sound. Other common conditions are benign paroxysmal postural vertigo, neck pain (cervicogenic dizziness) and persistent postural-perceptual dizziness (7).

Treatment

The majority of patients have mild to moderate symptoms that can be managed with the provision of thorough information, if necessary in combination with adjustments in their working and personal life to reduce symptoms and discomfort. Assisted hearing devices should be considered, but experience has shown that hearing aids and tinnitus maskers are difficult to use if there is significant hyperacusis and autophony. Comorbidities are often contributory factors in patients with severe vertigo or balance disorders, and treatment with balance exercises (vestibular rehabilitation) can be helpful. It is also important to identify and optimise the treatment of any complicating conditions, such as migraine, tension headache, functional dizziness, benign paroxysmal postural vertigo and Meniere's disease.

Patients with severe symptoms are usually offered surgical treatment (2). The aim of surgery is to reduce disease-specific symptoms, and this is usually achieved by covering or occluding (plugging) the superior semicircular canal. The superior semicircular canal can be exposed via the middle fossa approach or the transmastoid approach. It is also possible to combine the transmastoid and minimal middle fossa approach (mini-craniotomy). A simpler and less invasive method is to reinforce the round window via a transcanal approach (8). There are currently few studies into the effectiveness of this method.

Surgery is reported to have a good effect, with 60–90 % of patients having improvement in disease-specific symptoms such as autophony, conductive hyperacusis, pulsatile tinnitus, and pressure- and sound-induced vertigo (9), although residual symptoms are common (10). No clear differences in the effectiveness of the aforementioned surgical methods have been documented to date (9).

Postoperative complications may include unsteadiness, vertigo, pressure sensation in the ear and headache, and more rarely significant hearing loss on the operated side or permanent facial paralysis (9). Postoperative vertigo usually subsides over time due to central vestibular compensation and can be alleviated with balance exercises (vestibular rehabilitation). The middle fossa approach requires the temporal lobe to be exposed and retracted. This leads to slightly longer postoperative hospital admission, occasionally a period of cognitive difficulties, loss of concentration and fatigue, and rarely complications such as cerebrospinal fluid leakage, epidural haematoma, stroke and epileptic seizure (11). Therefore, it is recommended that, where possible, the method with the lowest risk of complications be used (9).

Revision surgery is necessary in some cases due to lack of symptom resolution. This may be due to the dehiscence expanding or the material used to cover or plug the dehiscence having failed (12). It is important to inform patients that other symptoms, such as pressure sensation in the ear, persistent vertigo and unsteadiness, loss of hearing, headache and non-pulsatile tinnitus, may not necessarily improve with surgery (12).

User perspective

Patients may experience cognitive challenges, anxiety and worry related to the disease and any additional diagnoses. It is often necessary to make adjustments in everyday life to tolerate and manage their symptoms, which are induced by sound, pressure changes and use of vision. It is important to provide information prior to examinations that elicit the symptoms. Doctors need to be aware of the complexity of the clinical presentation, and patients need to be provided with thorough information at an early stage of the disease. Patient information about superior semicircular canal dehiscence syndrome is available at helsenorge.no.

Conclusion

Superior semicircular canal dehiscence should be suspected in patients with autophony, hypersensitivity to internal bodily sounds, and pressure- and sound-induced vertigo. Diagnosis should be based on international criteria, and most patients benefit from receiving a diagnosis and explanation for the symptoms, which can often be reduced by making small adjustments to avoid activities that trigger symptoms. Surgical treatment can be considered in patients with severe symptoms (9). There is still a need for research into the pathophysiology and effectiveness of the various treatment measures. In Norway, a project is underway that aims to coordinate and record patient trajectories and surgical outcomes. The specialist association is due to publish an update to specialist national guidelines.

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