LET’S TALK ABOUT . . .

SEMICIRCULAR CANAL DEHISCENCE (SCD)

Key points

- Semicircular canal dehiscence (SCD) is a rare condition caused by a hole in one of the bony canals in the inner ear.
- SCD can cause both hearing and balance symptoms.
- People with SCD often hear internal sounds – their own voice, heartbeat, joints cracking or even their eyeballs moving.
- Surgery is suggested for people whose symptoms are severe.

What is SCD?

Semicircular canal dehiscence syndrome (SCD) is a collection of hearing and balance symptoms that are caused by a tiny hole (called a dehiscence, pronounced “dee-HISS-ence”) in one or more of the semicircular canals in the inner ear. Usually, SCD affects the superior semicircular canal, but sometimes there is a hole in the posterior semicircular canal. SCD may affect one or both ears.

SCD was first described in 1998. It is not clear how many people have SCD. It seems to be more common in older adults, but young children can also have SCD. Some people have a hole in the semicircular canal, but do not have symptoms.

It is not clear what causes these holes. In some cases, they may happen as part of fetal development. In other cases, they may be caused by injury or thinning bone later in life. Scientists are still researching this question.

What causes SCD?

The inner ear has two connected parts: the semicircular canals, which help you balance, and the cochlea, which lets you hear. These structures are surrounded by bone and filled with a fluid called endolymph.

There are three semicircular canals in each ear, arranged at right angles to each other. They sense when your head tilts or turns. For example, if you nod your head yes, the endolymph in the superior semicircular canal tries to stay still while the canal moves. (Imagine you are holding a loop of clear garden hose, with one end in each hand, and that it is partly filled with water. If you raise one end of the hose, the water will move toward the other end, because it is staying in one place while the hose is moving.) This makes the endolymph push against specialized hair cells, which send a signal to your brain that your head is moving. This is an important part of the brain’s balance system.

The cochlea is a tube that is rolled up so it looks like a snail shell. It turns sound waves into nerve signals that travel to your brain so you can hear. Sound waves enter the ear canal and make the eardrum vibrate, and these vibrations are sent to the stapes (stirrup) bone.

This tiny bone pushes on a structure called the oval window, which is at one end of the cochlea, and this causes pressure waves in the fluid (endolymph) inside the cochlea. A structure called the organ of Corti turns these pressure waves into nerve signals.

Finally, the pressure waves leave the cochlea through another structure called the round window. The round window acts like a release valve for the cochlea: it moves out when the oval window moves in, and vice versa.

The oval window and the round window are membranes. The rest of the inner ear is rigid, because it is encased in bone. Normally, this means that the round window and the oval window
are the only parts of the inner ear that move in response to sound waves, so the sound waves stay in the cochlea.

But if there is a hole in the bone that covers one of the semicircular canals, there is now a “third window” in the inner ear. This means that instead of staying in the cochlea, some of the sound waves can leak into the semicircular canal.

This leads to several problems:

- The pressure of sound waves in the cochlea is less, so they do not all get turned into nerve impulses. This means that you may not hear some sounds well.
- Sound waves can push against the hair cells in the semicircular canals, so your brain may think your head is moving when it is not.
- Sound waves from inside the body can enter the semicircular canals and then the cochlea, so those internal sounds can be distorted or louder than normal.

Sometimes the hole is up to two mm across. But the hole can be as small as a pinhole and still cause symptoms.

Symptoms of SCD

SCD can cause both hearing and balance symptoms.

Balance symptoms of SCD are often caused by loud sounds or pressure changes (either inside the body, such as when you are coughing, or outside the body, such as when you are going up in an airplane). They may include:

- Vertigo (a spinning sensation), dizziness (for instance, a sensation of a jolt or being pushed or pulled) and nystagmus (rapid, involuntary eye movements) in response to loud sounds or pressure changes; this is called the Tullio phenomenon when it is caused by sounds and Hennebert’s sign when it is caused by pressure changes
- Feeling unsteady or as though you are going to fall (disequilibrium)
- A vision problem called oscillopsia, where what you see blurs or seems to jump around in response to loud sounds or pressure changes

- A feeling of fullness in the ear

Hearing symptoms of SCD may include:

- A form of hearing loss called low-frequency pseudo-conductive hearing loss. “Low-frequency” means it is harder to hear sounds with a lower pitch. “Pseudo-conductive” means that even though some of the hearing test results are similar to conductive hearing loss, your middle ear function is normal.
- Hearing abnormally loud sounds from inside your own body (bone conduction hyperacusis), such as your voice (autophony), your heartbeat, your joints cracking or even your eyeballs moving
- Hearing rhythmic or whooshing noises that don’t appear to have an outside source (pulsatile tinnitus)
- Sounds from outside the body, or your own voice, are distorted or hard to hear
- Sensitivity to loud sounds (phonophobia), as they may provoke Tullio phenomenon

How SCD is diagnosed

SCD is usually diagnosed by a specialist, such as an otolaryngologist (an ear, nose and throat or ENT doctor) or a neurologist.

Your doctor will ask about your symptoms and your medical history. Your doctor will also do a thorough physical and neurological exam, including an ear exam.

You will likely have the following diagnostic tests:

- hearing and balance tests, including audiometry, vestibular evoked myogenic potential (VEMP) and videonystagmography (VNG)
- imaging (high-resolution CT scan)
Treatment and management of SCD

At present, the only treatment that addresses the cause of SCD is surgery. But if your symptoms are mild, the drawbacks of surgery may outweigh the benefits. Your doctor may suggest starting with less invasive ways to manage your symptoms. Some people find that just knowing what is causing their symptoms is helpful. Other people find their symptoms improve with various techniques, including:

- avoiding triggers such as loud sounds and pressure changes; earplugs may help to make sounds quieter
- tinnitus retraining therapy
- using a hearing aid
- treating migraines, if you have them

Your doctor will want to see you regularly to follow up on how you are doing and see if your symptoms are better or worse.

Surgery

Surgery for SCD is usually suggested for people whose symptoms are severe and who find that other approaches do not help.

There are several different surgical options, including:

- canal plugging to block the semicircular canal
- canal capping or surfacing to close the hole
- reinforcement of the round or oval window

Which procedure is used will depend on various factors, including:

- where the hole is and how big it is
- the shapes of your ear and skull bones
- your state of health
- how experienced your surgeon is with each procedure

Some techniques have better results than others, and some have a higher risk of hearing loss. Your surgeon will discuss the risks and benefits with you, including potential side effects and how long it usually takes to recover from the procedure.

Some of the symptoms of migraine are similar to those of SCD, and SCD can trigger migraines. If you have migraines, it is important to have them under control before surgery.

Similarly, head injury sometimes causes SCD, but the symptoms of concussion and SCD can be similar or overlap. It is important to be sure that SCD is causing your symptoms before moving ahead with surgery.

Most people who have surgery for SCD have good outcomes, with fewer symptoms and a better quality of life. The most common side effects of surgery include:

- Reduced function of the affected semicircular canal and/or another nearby semicircular canal
- Reduced function of one or both of the otoliths (the utricle and saccule), which are part of the balance system and help to sense head movements
- Problems with vestibular function right after surgery; these usually improve in the weeks after surgery
- Benign paroxysmal positional vertigo (BPPV)
- High-frequency sensorineural hearing loss

People tend to take longer to recover after surgery if they have:

- holes on both sides
- larger holes
- migraines

What to expect in the future

Researchers are studying the causes of SCD, as well as ways to prevent, diagnose and treat it. Surgeons are also developing new methods and improving existing methods so that they work better and take less time to recover from. Some researchers believe that one day it may be possible to use custom 3-D printing to cover the hole.
More resources and sources

View more SCD resources as well as sources used for this handout: https://bit.ly/2NSRKlr

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If you find the information in this handout valuable, we ask you for your help. The cause of supporting those affected by balance and dizziness disorders with up-to-date, evidence-based information written for Canadians, needs you. Will you consider becoming its champion by making a gift online or by mail?

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