



PERILYMPH FISTULA

This handout is intended as a general introduction to the topic. As each person is affected differently, speak with your health care professional for individual advice.



Key points

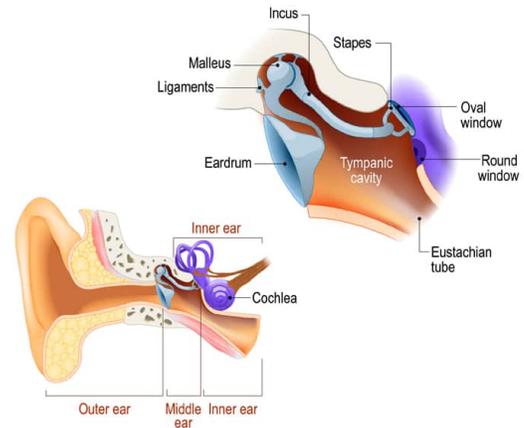
- An abnormal opening or tear in one or both of the membranes separating the middle and inner ear.
- Can cause sudden or progressive hearing loss and episodes of vertigo (spinning sensation).
- Rare and controversial condition – not everyone agrees on the definition, diagnosis, or treatment.
- May be caused by disease, a pressure injury from an external or internal source, or no obvious reason.
- Treatment and management may include avoiding triggers, treating other medical conditions, or surgery.

What is perilymph fistula?

A fistula is an abnormal connection between two parts of the body. Perilymph is the fluid that fills the bony labyrinth in the inner ear.

A perilymph fistula is an opening or tear in one or both of the membranes separating the middle and inner ear. This opening lets perilymph leak out of the inner ear and into a nearby area like the middle ear. It may appear in the oval or round window, in one of the semicircular canals (similar to a semicircular canal dehiscence) or somewhere else.

MIDDLE EAR



Perilymph fistula can cause sudden or progressive hearing loss and episodes of vertigo.

Perilymph fistula is a controversial condition. Doctors and researchers do not all agree on how it should be defined, when it should be diagnosed or how it should be treated. There is no specific diagnostic test for it, and the definition of perilymph fistula has changed over the years. Also, the symptoms are similar to those of many other balance problems. But the diagnosis is becoming more accepted.

Perilymph fistula is rare. One group of researchers has estimated that about 1.5 in 100,000 adults has perilymph fistula. It seems to be more common among children with sensorineural hearing loss that does not have an obvious cause (idiopathic).

What are the causes?

Many different things can cause perilymph fistula. Recently, the following four groups of causes have been identified:

- Disease (such as meningitis), trauma (such as head injury), surgery or other treatment
- Pressure injury (barotrauma) from an external source such as flying or diving

- Pressure injury from an internal source such as straining, sneezing or coughing
- No apparent cause (idiopathic)

What are the symptoms?

Perilymph fistula can cause both balance and hearing symptoms.

Balance symptoms of perilymph fistula may include:

- a feeling of spinning or movement (vertigo)
- feeling off-balance or wobbly (disequilibrium)
- discomfort with motion
- nausea or vomiting

Hearing symptoms of perilymph fistula may include:

- sudden or progressive sensorineural hearing loss that sometimes gets better or worse (fluctuates)
- tinnitus (ringing, roaring or whooshing in the ears)
- sensitivity to everyday sounds (hyperacusis)
- a feeling of fullness or running water in the ear

Some people with perilymph fistula may have trouble making sense of complex surroundings such as crowds or traffic, or they may have trouble concentrating or remembering things.

Some people with perilymph fistula find that their symptoms are triggered by loud noises. The symptoms may also get worse with activities that cause pressure changes inside or outside the body, including:

- coughing
- sneezing
- nose-blowing
- straining
- exercise
- flying

How is it diagnosed?

Perilymph fistula may be diagnosed by a primary care doctor or an emergency doctor, but it is more often diagnosed by a specialist, such as an otolaryngologist or an otologist.

The doctor will ask about your symptoms. Try to be as specific as possible about the symptoms, when they started, and when they get better or worse.

The doctor will also ask about your medical history, including injuries, illnesses, any medications you are taking or recently stopped taking, any conditions you have been diagnosed with in the past and any recent activities that cause pressure changes, like flying or SCUBA diving. The doctor will also do a thorough physical and neurological exam.

You will probably have some of the following diagnostic tests:

- hearing and vestibular function tests
- an imaging test such as magnetic resonance imaging (MRI) or computed tomography (CT); this can help to rule out conditions such as a semicircular canal dehiscence

Many other conditions cause similar symptoms, so the doctor will need to rule out conditions like Ménière's disease or migraine before making a diagnosis of perilymph fistula.

How is it treated and managed?

Different doctors and researchers have different opinions about the best way to treat perilymph fistula. Some doctors believe that fast treatment is necessary for a good outcome. Others have found that patients may benefit from treatment even if years have passed since the symptoms started.

Avoiding triggers

Some doctors prefer to wait a few months and see if the fistula will heal on its own. During this time, you should avoid lifting, flying, playing wind instruments, diving, or anything else that could raise the pressure inside your body. Your doctor may even suggest a period of bed rest, although this is an

older treatment and many doctors today do not believe it is necessary.

Treating other possible conditions

Your doctor may suggest trying some treatments for Ménière's disease or migraine especially if the diagnosis of perilymph fistula is not definite.

Surgery

If your perilymph fistula can be seen with imaging, or if you definitely had an injury or another event that could have caused the fistula, your doctor may suggest surgery to seal the hole. Surgery for treating perilymph fistula may involve surgical glue, tissue grafts and/or laser surgery. It may be a simple procedure in your doctor's office, or it may be a more complicated operation in a hospital. This will partly depend on where the fistula is located.

Another surgical option for treating perilymph fistula is a pressure-equalizing tube (PE tube or tympanostomy tube). The surgeon makes a tiny hole in the eardrum on the same side as the fistula and then places a small, soft tube in the hole. This helps to balance the pressure inside and outside the middle ear. This can help to protect the ear against external sources of pressure, like flying.

In extremely rare cases when nothing else works and the symptoms are unbearable, the doctor may suggest a vestibular nerve section as a very last resort.

A less invasive option is to inject the ear with gentamicin, an antibiotic that damages the cells of the ear and destroys the part of the ear that controls balance. In most cases, patients who have this treatment no longer have vertigo. But there is a risk of hearing loss with this treatment.

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What to expect in the future

New tests for perilymph fistula are being developed. These tests look for a specific molecule (or biomarker) that is only found in perilymph, known as Cochlin-tomoprotein or CTP. To do the test, a doctor takes a sample of fluid from the middle ear during exploratory surgery. A lab then tests the sample. If the test shows CTP in the fluid, it means that perilymph is leaking into the middle ear. However, this test may only find about 1 in 5 cases of perilymph fistula. At present, the test is only available in Japan, but it may be approved in other countries eventually.

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