

DISORDERS

TEAR OR DEFECT

An abnormal connection in the membranes separating the inner and middle ear can result in severe symptoms.

ARTICLE

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Perilymph Fistula

By The Vestibular Disorders Association, with edits by Dr. Dennis Fitzgerald, MD

OVERVIEW

A *perilymph* fistula (PLF) is an abnormal connection (a tear or defect) in one or both of the small, thin membranes (the oval window and the round window) that separate the air filled middle ear and the fluid filled perilymphatic space of the inner ear. This small opening allows perilymph (fluid) to leak into the middle ear.

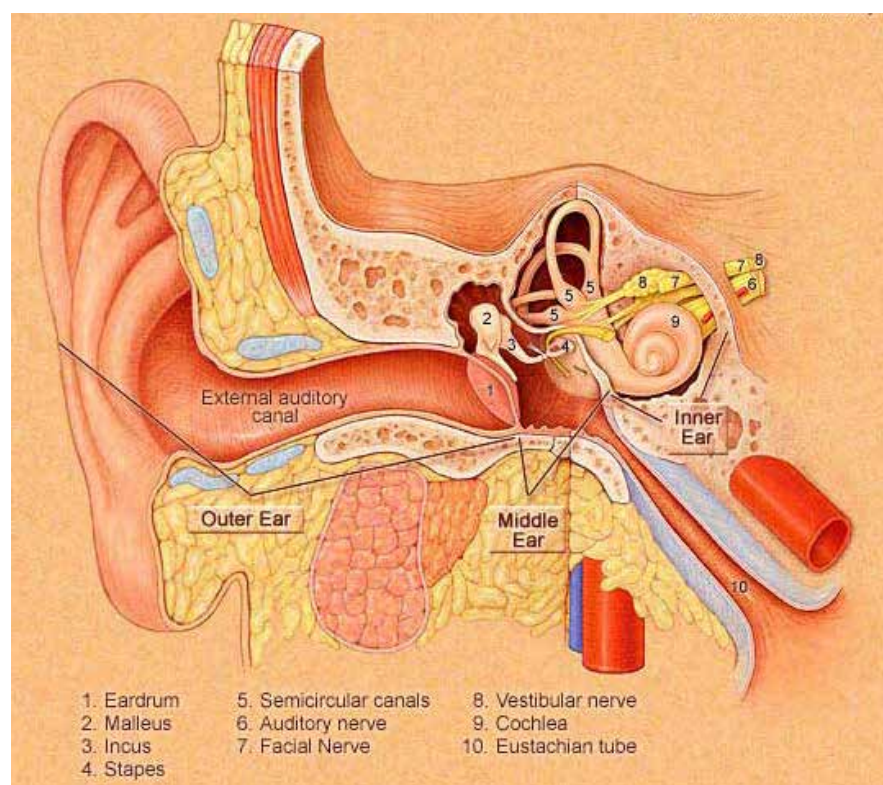


Fig. 1: Anatomy of the inner and middle ear. A fistula is an abnormal connection between the air-filled middle ear and the fluid filled inner ear. The two weakest points are membranes located at the stapes foot plate (the "oval window"), #4 here, and just below, a small niche called the "round window". There can also be fistulae at other points, such as the superior semicircular canal, as well as other semicircular canals, but most require erosion of bone, and can be seen on a temporal bone CT scan. There can also be openings in the bone that cannot be seen on CT scan (although visible on autopsy). © Vestibular Disorders Association. Image used with permission from T. C. Hain.



Changes in air pressure that occur in the middle ear (for example, when your ears “pop” in an airplane) normally do not affect your inner ear. However, when a fistula is present, changes in middle ear pressure will directly affect the inner ear, stimulating the balance and/or hearing structures within and causing PLF symptoms.

The perilymphatic space of the inner ear is connected to the cerebrospinal fluid (CSF) that surrounds the brain. Perilymphatic fluid, which is high in sodium (Na⁺), is similar in composition to CSF. When an abnormal connection between the membranes between the middle and inner ear exists, perilymph in the inner ear escapes, driven by the hydrostatic pressure of the CSF, and is replaced by CSF. This can also result in lower than normal levels of CSF fluid around the brain and spinal cord, which may result in symptoms such as mild headache.

Patients with PLF often feel frustrated and depressed because, while they don’t feel well, they look fine to others. PLF patients specifically and vestibular patients in general often have a challenging time explaining to friends and family what they are going through. Sometimes it is enough to ask your support network for patience and understanding while you explore diagnosis and treatment options and learn to cope with the symptoms brought on by persistent dizziness.

HISTORY

The small amount of fluid leaking from the inner ear to the middle ear is not detectable by the patient and is not generally visible to the surgeon who sets out to patch the leak. 40 years ago, when PLF first became an item of concern, the presumption was that there was a tear in the round window or the ligamentous attachment of the footplate of the stapes to the edge of the oval window. A novel idea was put forth by Dr. Robert Kohut, based on post-mortem examination of temporal bones in patients who had suffered sudden hearing loss. His pioneering work indicated that the leak sites could be microfissures in the area just in front of the oval window or in the floor of the round window niche.¹

SYMPTOMS

The symptoms of a PLF most commonly include ear fullness, fluctuating or “sensitive” hearing, dizziness without true vertigo (spinning), and motion intolerance. Vertigo or sudden hearing loss can occur from a PLF. Most people with fistulas find that

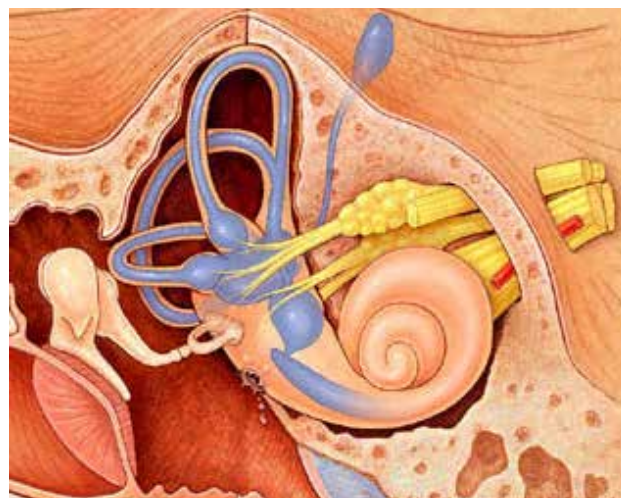


Fig. 2: Round window fistula. An opening in the round window allows perilymph to leak out into the middle ear. In this artist's depiction, for clarity, bone is not shown between the middle and inner ears. While it is difficult to be sure, it seems likely that in most cases there is only a small oozing of fluid between the perilymphatic space and the air-filled middle ear. © *Vestibular Disorders Association*. Image used with permission from T. C. Hain.

their symptoms get worse with changes in altitude (fast elevators, airplanes, and travel over mountain passes) or increased CSF pressure resulting from heavy lifting, bending over, and coughing or sneezing.

CAUSES

Head trauma is the most common cause of fistulas, usually involving a direct blow to the head or in some cases a “whiplash” injury. Other common causes include ear trauma, objects perforating the eardrum, or “ear block” on descent of an airplane or SCUBA diving. Fistulas may also develop after rapid increases in intracranial pressure, such as may occur with weightlifting or childbirth.

Fistulas are infrequently present from birth. A long-running controversy has surrounded the idea of a “spontaneous PLF.” Instead, what may occur is that a patient has a causative event but does not see an ear specialist right away. The passage of time blurs the memory of such an event so that the PLF might seem to have been spontaneous. Rarely, PLF’s occur in both ears, and only after a severe head injury.

DIAGNOSIS

There is no positive way to diagnosis a PLF. For many years it was thought that it could be confirmed by performing a tympanotomy (surgical exploration of the middle ear) and directly viewing



the area of the suspected fistula to detect a fluid leak. However, since the leak would be only a few microliters of clear fluid, visual detection has been found to be virtually impossible. Larger amounts of fluid leakage may indicate a CSF leak due to a congenital defect in the inner ear.

A physician can arrive at a presumptive diagnosis through a thorough probing for events close in time to the onset of symptoms, along with a variety of tests. These tests can include hearing tests (audiogram, ECOG), balance tests (VNG, VEMP) and some form of a "fistula test."

Historically, a platform pressure test developed by Dr. F. Owen Black was seen to be the most reliable test to determine if a PLF was present. However, this equipment is no longer in production, and only a small number still exist today.

In the end, a physician has to present the possibility of a PLF to the patient based on history, test results, and the lack of spontaneous resolution of symptoms. Together the physician and patient (or guardian) must decide whether to undertake an operation to patch the oval and round window areas. Immediately following surgery there is a period of bed rest, followed by a period of restricted activity. Four to six weeks later a reassessment of the patient's symptoms is done to determine if the patching successfully corrected the PLF.

TREATMENT

When a traumatic event results in sudden onset of hearing loss or dizziness, the patient is advised to severely restrict physical activity for 7-14 days. If the symptoms do not improve or they plateau, testing is ordered. If the tests are compatible with the diagnosis of PLF, a surgical intervention may be considered. Persons with diagnosed fistulas who are awaiting surgery should avoid lifting, straining and bending over as these activities can cause a worsening of the symptoms.

A PLF repair involves an operation, often under general anesthesia, working through the ear canal. The eardrum is lifted up and minute soft tissue grafts are placed around the base of the stapes (stirrup) and in the round window niche. The operation usually takes about 45-60 minutes to complete. There is very little, if any, pain. Some patients are kept overnight to restrict activity. Once discharged the patient is advised to spend three days at home with limited activity. After three days the patient may return to sedentary work activities. The patient is advised to avoid lifting more than

10 lbs. for one month and avoid sporting activities. After one month there are additional restrictions suggested on activities such as contact sports, diving, weight lifting, and roller coasters. All of these activities have resulted in recurrent PLF's after an initial successful repair.

REFERENCES

1. Kohut RI, Hinojosa R, Ryu JH. Perilymphatic fistula: a single-blind clinical; histopathologic study. *Adv. Otorhinolaryngol* 1988; 42:148.

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