Noise-induced perilymph fistula

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Abstract
Perilymph fistulae are difficult to diagnose because they present with a wide variety of signs and symptoms, they are associated with many etiologies, and they often mimic other conditions. In this article, we describe a case of perilymph fistula that featured one of its more rare causes: acoustic trauma—specifically, damage from a loud blast from the siren of a fire engine. We also review the literature and discuss the difficulties of diagnosis and treatment and the possible mechanisms by which acoustic trauma and other etiologies cause perilymph fistulae.

Introduction
A perilymph fistula is an abnormal communication that allows perilymph to escape from the inner ear to the middle ear. The abnormal flow of perilymph can result in continuous or fluctuating sensorineural hearing loss, tinnitus, and vertigo. A wide variety of signs, symptoms, and etiologies are associated with perilymph fistulae, but these same features are seen with many other conditions as well, which makes diagnosis and management both difficult and controversial.

During the past 30 years, very few reports of perilymph fistulae caused by exposure to excessive noise have been reported. Those that have been reported include cases associated with exposure to gunfire, exploding fire-crackers, and loudspeakers. Until now, the most recent report of a case of a noise-induced perilymph fistula was published more than 15 years ago. In this article, we describe a new case of perilymph fistula that was caused by exposure to noise.

Case report
A 27-year-old volunteer fireman presented 3 weeks after he had experienced an acoustic trauma as the result of a blast from the siren of a fire engine. When the trauma occurred, the patient’s right ear was approximately 18 inches from the siren. This type of siren (Q2B; Federal Signal Corp.; University Park, Ill.) can produce 140 dB at a distance of 18 inches and 136 dB at 30 inches.

Immediately after the incident, the patient noted a sensation of lancinating right ear pain, extremely loud tinnitus, and mild dizziness. The tinnitus persisted at a high intensity for approximately 6 hours after the incident. Thereafter, the intensity diminished and the patient noticed a right-sided hearing loss. The dizziness persisted as an intermittent, daily sensation of unsteadiness without rotary vertigo; this sensation was exacerbated by head motion and relieved by avoidance of head motion.

Examination revealed that the right tympanic membrane was mobile and that it had a slightly prominent vascular strip. There was no spontaneous nystagmus, and on positional testing the patient experienced feelings of unsteadiness without nystagmus or a sensation of rotary vertigo. Findings on the tandem Romberg’s test were equivocal, with the patient falling backward with his eyes open. With a 512-Hz tuning fork, Rinne’s test was positive bilaterally, and Weber’s test revealed that the sound was louder on the left side, indicating a right-sided sensorineural hearing loss. Pneumatic otoscopy of the right ear elicited Hennebert’s sign.

Pure-tone audiometry revealed a 30-dB threshold at 6,000 Hz. Thresholds in the left ear were within normal limits, with the exception of a 30-dB threshold at 6,000 Hz. The patient had no air-bone gap, his speech reception thresholds were consistent with the findings on the pure-tone audiogram, and his speech discrimination scores were normal.

Electronystagmography (ENG) detected a mild left-beating nystagmus on positional testing with the right ear down, without a sensation of vertigo. A fistula test in the right ear was positive, with both a mild right-beating nystagmus and a sensation of vertigo.

A follow-up audiogram obtained 2 weeks later showed little change in air conduction, bone conduction, and speech reception thresholds, but the speech discrimination score had fallen to 88% at 65 dB (it subsequently improved to 96% at 65 dB on the next audiogram prior to middle ear exploration). Findings on magnetic resonance imaging
of the brain and internal auditory canals were normal. Laboratory testing revealed normal values for the complete blood count, thyroid-stimulating hormone, Lyme titer, fluorescent treponemal antibody, antinuclear antibody, Rh factor, erythrocyte sedimentation rate, and antibodies to inner ear antigens. The lipid profile showed elevated levels of triglycerides, total cholesterol, and low-density lipoprotein cholesterol.

Over the next few weeks, the patient's hearing loss and tinnitus had stabilized but still persisted, and his periods of dizziness became more severe. He was scheduled for surgery. In the operating room, he received local anesthesia and sedation. The malleus and incus were found to be intact and mobile, and there was no evidence of a fistula of the round window during Valsalva's maneuver. However, some filmy adhesions were noted to be draping the stapes superstructure over the promontory, the facial nerve, and the oval window; a fluid collection in the footplate area was also obvious. The adhesions were lysed, the mucosa was scraped off the footplate, and the area of the annulus was denuded. A piece of loose areolar tissue was harvested through a postauricular incision and used to seal the oval window. Gel foam was applied to fill the middle ear and stabilize the graft.

Postoperatively, the patient's dizziness disappeared for a brief period, but his hearing loss gradually became worse. Eventually, the dizziness also returned, and his clinical course became complicated. He was taken back to the operating room three more times to revise his perilymph fistula repair. During this period, he also developed a right-sided facial paralysis secondary to Ramsay Hunt syndrome, and he underwent total facial nerve decompression to fully restore his facial nerve function.

Over time, the patient's hearing improved but never fully recovered. His balance improved and stabilized, but he continued to experience bouts of dizziness. Vestibular nerve section was offered to him several times throughout the course of treatment, but he repeatedly declined. His management was also rendered more complex by his early decision to seek a permanent disability designation through legal action and by his addiction to prescription narcotics obtained from other physicians.

**Discussion**

The diagnosis of a perilymph fistula can be difficult because of its many possible causes and the highly variable signs and symptoms that have been associated with the condition. The preferred method of establishing a diagnosis is visual inspection during exploratory tympanotomy, even though findings are often equivocal. During the past few years, efforts have been undertaken to establish more objective diagnostic criteria, but controversy persists.

Perilymph fistula first became recognized as a legitimate clinical diagnosis around the mid-1960s. The most common cause is otologic surgery, particularly stapes surgery. The first report of a perilymph fistula that was not caused iatrogenically was published by Fee in 1968. He described the cases of 3 patients who each experienced tinnitus, vertigo, and fluctuating hearing loss following head trauma; each was found to have a leak of perilymph from the oval window area during exploratory tympanotomy. In 1970, Stroud and Calcaterra reported the first cases of spontaneous oval window perilymph fistulae and hypothesized that the inciting events were episodes of increased intracranial pressure.

In 1971, Goodhill published his well-known theory on the pathophysiology of spontaneous perilymph fistulae. H proposed that fistulae occur via two possible mechanisms. The first is the hydrodynamic explosive route, in which increased intracranial pressure is transmitted to the perilymphatic space through a patent cochlear aqueduct internal auditory canal, causing a rupture at either the oval or round window. The second is the aerodynamic implosive route, in which a sudden increase in middle ear pressure from either the external auditory canal or the eustachian tube causes a rupture of the oval or round window.

Since then, various anatomic studies have been performed on temporal bones, and theories have arisen about the significance of microfissures in the otic capsule in the area of the oval window, the round window niche, and the posterior canal ampulla. Moreover, investigators have studied the importance of pathways between the intracranial cavity and the inner ear, particularly the cochlear aqueduct. Yet despite all this additional work, not many major alterations have been made to Goodhill's theory, and it is still widely cited today.

One of the most important components in the diagnosis of a perilymph fistula is the patient's history. Knowledge of previous otologic diseases and surgery, head trauma, barotrauma, congenital malformations and syndromes, bouts of recurrent meningitis, and other infectious diseases (e.g., syphilis) is key to the evaluation of a possible perilymph fistula. The patient's symptoms are important as well, but they are not always diagnostic. Because the otologic symptoms of a perilymph fistula are so variable, patients are often misdiagnosed as having some other otologic condition, usually Ménière's disease.

The variability of signs and symptoms in patients with perilymph fistula has also been widely studied, and the data have been used to help develop diagnostic criteria. In 1986, Seltzer and McCabe published the first large series of patients with perilymph fistula. They found that 83% of patients with surgically confirmed fistulae had experienced a hearing loss; the most common type

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of hearing loss (36% of patients) involved fluctuations in both pure-tone threshold and speech discrimination. Some 80% of patients had vestibular complaints, the most common being disequilibrium with occasional episodes of vertigo. Approximately 53% of patients reported tinnitus, 25% had aural fullness, and 21% had recruitment. The most common presentation overall was the triad of hearing loss, vertigo, and tinnitus. Twenty-four percent of these patients had no known precipitating event. Other large studies were reported by Weider and Johnson in 1988, Shelton and Simmons in 1988, and Rizer and House in 1991.

Likewise, physical examination findings also tend to be variable and inconsistent. Often, results are completely normal. Nevertheless, a careful otologic examination is important to look for other possibly related pathology, such as congenital, infectious, inflammatory, autoimmune, and other causes. Over the years, the most frequently studied physical examination finding has been the fistula test for Hennebert’s sign. As might be expected, the results of these studies have been variable. Seltzer and McCabe found that only about 25% of patients with surgically confirmed fistulae had had a positive fistula test, while other authors reported positive fistula tests in as many as 77% of patients. Rizer and House found that the percentage of positive tests in patients with surgically confirmed fistulae was not significantly different from the percentage of positive tests in patients without surgically confirmed fistulae.

Audiologic and ENG findings also tend to be variable. Patients may present with almost any pattern of hearing loss, although fluctuating sensorineural hearing loss with fluctuating speech discrimination is most common. ENG may detect directional preponderance, reduced caloric response, spontaneous nystagmus, positional nystagmus, or no abnormalities at all. An ENG fistula test may be useful as a supplement to the clinical fistula test. Electrocochleography may show an increased ratio of summing potential to action potential in patients with perilymph fistulae but, overall, findings can be similar to those in patients with Ménière’s disease.

Ultimately, a diagnosis of perilymph fistula must be confirmed by direct visualization. However, even surgical confirmation can be controversial because it is sometimes difficult to see the leak of perilymph and to prove that any visualized fluid is actually emanating from the inner ear.

Various methods have been proposed to help identify perilymph fistulae. The most widely used method is exploratory tympanotomy under microscopic visualization with intraoperative measures such as Valsalva’s maneuver, bilateral jugular vein compression, and Trendelenburg’s positioning; endoscopes, labeling methods, and rapid fluid analysis have all been studied and, again, results have been variable.

Some controversy also attends to the treatment of perilymph fistulae. The goal of surgery is to stabilize hearing and relieve vestibular symptoms. For patients with idiopathic perilymph fistulae, some groups recommend conservative management first—bed rest, head elevation, and avoidance of strenuous activity. However, most otologists recommend early surgical exploration and repair with various materials, such as subcutaneous areolar tissue, fat, perichondrium, fascia, fibrin glue, and microfibrillar collagen.

Despite the paucity of reports on noise-induced perilymph fistulae, we do know that it is possible for a sufficiently loud acoustic stimulus to cause enough movement of the oval window to rupture it. In their 1985 case report on perilymph fistula caused by loudspeaker noise, Narula and Marks mentioned that Dahlmann reported in 1954 that the minimum pressure on the tympanic membrane necessary to cause visible movement of the stapes at the oval window is 60 mm Hg, which corresponds to a sound level of 84 dB. The patient in that case report had been exposed to a sound level of 130 dB, and our patient had been exposed to approximately 140 dB.

It is not inconceivable that our patient’s ongoing symptoms might have been attributable to posttraumatic endolymphatic hydrops rather than to his initial perilymph fistula. As mentioned, the signs and symptoms of perilymph fistulae and the findings on diagnostic tests all tend to mimic those seen in patients with Ménière’s disease, and our case was no exception. A perilymph leak was obvious at the time of our patient’s first and second surgical procedures, but a fistula could not be confirmed during the subsequent operations. Studies by DiBiase and Arriaga, McGill and Schuknecht, and Kemink and Graham provided histopathologic evidence of endolymphatic hydrops in patients with delayed-onset vertigo following noise-induced hearing loss. In a study of soldiers exposed to noise from firearms (155 to 190 dB), Ylikoski reported that all of the soldiers who had experienced balance problems following exposure to noise did so on a delayed basis (from 6 to 29 yr) and that more than half of these patients reported attacks of vertigo accompanied by tinnitus. Finally, trauma that is severe enough to cause a perilymph fistula may be severe enough to also cause delayed hydrops that can be very difficult to differentiate from a recurrent fistula.

In conclusion, it is important to recognize perilymph fistula as a cause of hearing loss and dizziness because it is potentially curable, despite the controversies over diagnosis and treatment. Additional clinical and basic research should be encouraged to help answer the remaining questions about this complex entity.
References