Perilymphatic fistula (PLF) has been reported as a complication of stapedectomy surgery for over three decades. However, spontaneous PLF was not reported until the late 1960s, although disease-related PLF, including sequela of cholesteatoma, appeared earlier. Other etiologies for loss of perilymph from the inner ear include head trauma/barotrauma and congenital anomalies. Surgical repair of fistula frequently stabilizes hearing, eliminates debilitating vertigo, and may even reduce tinnitus. However, data are inconsistent regarding anatomy, pathophysiology, etiology, diagnosis, and methods of repairing fistulas.

ANATOMY AND PHYSIOLOGY OF PLF ETIOLOGY

Simmons and colleagues studied surgical injuries to the round window in cats. A theory was proposed describing intralabyrinthine membrane breaks to explain sensorineural hearing loss in patients who experience popping sensation in the ear in association with hearing loss. This concept later evolved into the double-membrane break theory in which an oval or round window fistula occurs in association with an intracochlear membrane break.

Theories proposed by Goodhill and colleagues describe membrane breaks by explosive and implosive forces on the inner ear. Explosive forces enter the labyrinth by the cochlear aqueduct, lamina cribosa, or internal auditory canal. Laboratory experimental evidence proved the explosive route theory by demonstrating that cerebrospinal fluid communicated with perilymph through the cochlear aqueduct. Further, after blockage of the cochlear aqueduct inner-ear pressure remains constant despite cerebrospinal fluid (CSF) pressure changes. Additional experimental evidence showed that India ink injected into spinal cord cerebrospinal fluid (CSF) in cats was identified in high concentrations in the cochlear aqueduct and in lower concentrations in the internal auditory canal when CSF pressure was increased sufficiently to cause bulging or rupture of the round window membrane.

CSF-perilymph exchange through the cochlear aqueduct is debated in patients, however. Although the aqueduct appears to be patent in the fetus and young child, in adults it is filled with arachnoidal connective tissue that may or may not prevent free flow of CSF from the subarachnoid space into scala tympani. Anson and colleagues concluded that the tissue in the cochlear aqueduct is the type through which fluids of
the body usually pass. Other evidence indicates that arachnoidal tissue functions to plug the duct when infection is present, as demonstrated by the low incidence of labyrinthitis and deafness in cases of meningitis. Temporal bone studies show that in as many as 90% of adults the arachnoidal tissue does not interfere with the patency of the duct. More recent evidence indicates that cochlear aqueduct patency is probably age related—open in three fourths of people under age 21 and in only about one third of the elderly population.

Radioactive pharmaceuticals and dyes have been injected into the subarachnoid space to monitor CSF flow into perilymphatic spaces. Ritter and Lawrence, using such a technique, showed no evidence of CSF flow into the inner ear. However, Emmett and colleagues measured radioactivity at oval and round window fistulas and concluded that Ritter and Lawrence failed because they allowed too much time following tracer injection before their measurements were made. An alternative explanation is that cochlear aqueduct was patent in the study by Emmett and colleagues and not so in the Ritter and Lawrence study. Convincing reports by Duval and Sutherland, and Asher and Sando showed that horseradish peroxidase tracers injected into CSF of guinea pigs are transmitted to the scala tympani through the cochlear aqueduct and along cochlear and vestibular nerve fibers.

Carlborg and Farmer demonstrated that inner-ear fluid pressures were primarily affected by blockage of the cochlear aqueduct. Using a sensitive pressure transducer, they demonstrated that when cochlear aqueduct was left patent CSF pressure changes were quickly (within 5 seconds) transmitted to perilymph; however, with blocked cochlear aqueduct 10 to 20 minutes passed before pressure equilibration between these fluids was achieved. Thus, patent cochlear aqueduct in children and most adults provides a mechanism for explosive rupture of a window membrane through increased CSF pressure, accompanying physical exertion, transmitted to scala tympani.

**STAPEDECTOMY AND PLF**

In 1955, Shea performed the first stapedectomy. This surgical procedure involves opening the vestibule of the inner ear and creating a temporary perilymphatic fistula until a stapes prosthesis is placed and the oval window is closed. If the oval window is inadequately closed or later reopens, perilymph escapes into the middle ear. Report of PLF following stapedectomy described symptoms of tinnitus, vertigo, nausea, and vomiting related to oval window fistula around the stapes prosthesis. Other studies report similar symptoms associated with poststapedectomy patients, and surgical complication is reported to occur in as many as 7% of cases.

**CONGENITAL DEFECT AND PLF**

Congenital round window and oval window malformations present with or without a fistula may predispose the patient to PLF. A review of cases of meningitis related to congenital PLF was reported by Parsifer and Birken, who described several patients with repeated episodes of meningitis and with reported ear anomalies such as stapes defect around which perilymph was draining. In patients presenting with meningitis and severe sensorineural hearing loss, the possibility of PLF is to be considered. Reilly reported that 26% of children with congenital middle-ear defect had perilymph fistula. Weber and coworkers reported that 81.3% of PLF cases had congenital middle-ear defect, with malformed stapes occurring in 60% of cases.

Seltzer and McCabe concluded that congenital ear anomalies were rare (2%). It was, however, noted that some form of Mondini’s malformation was present in 8% of patients. A high rate of recurring perilymph fistulas was observed in these patients.

**STAPES GUSHER AND CONGENITAL DEFECT**

“Stapes gusher” occurs when perilymph pours into the middle ear after opening the...
vestibule surgically. The leak in these cases may occur because of CSF flow from the internal auditory canal or patent cochlear aqueduct. Glasscock in a review of stapes gusher cases determined that the internal auditory canal is the probable source of perilymphatic leaks. Shea and Goode propose that the site for stapes gushers is the cochlear aqueduct. In patients with congenital stapes fixation and elevated perilymphatic pressure, “gusher” results when the stapes is removed, causing first a conductive hearing loss that progresses to a sensorineural loss, according to Goode. He proposed that the sensorineural hearing loss may result from chronically high perilymph pressure, or perilymphatic hydrops. To prevent stapes gushers during surgery, Shea recommends making a stapes footplate hole to determine if a profuse amount of fluid will leak out.

**Spontaneous PLF and Congenital Defect**

Apparent spontaneous fistulas have been associated with exertion and trauma. In a review of PLF in children, Grundfast and Bluestone showed a predisposing congenital malformation of the oval or round window as a cause in spontaneous PLF. In children with unilateral sensorineural hearing loss of unknown cause who develop a loss in the only hearing ear, congenital malformation leading to PLF is likely. A 4-year-old child was described who bumped his head on a couch and immediately experienced buzzing tinnitus in both ears. During surgical exploration, the patient was found to have both oval and round window fistulas with a malformation of the stapes. Children who have anatomic defects may not need significant trauma for a perilymphatic fistula to develop.

**Trauma**

Seltzer and McCabe stress the need for early surgical intervention in young patients who have a known prior hearing loss, and then either a progression in the hearing loss or a new hearing loss in the better ear. This is recommended because of the high incidence of PLF observed in their series. PLF should be included in the differential diagnosis in children with unexplained unilateral sensorineural hearing loss, especially with history of head trauma, vertigo, or suspected ear deformity.

Traumatic ear fistulas can result from temporal bone injuries. Blunt head trauma may cause temporal bone fracture, but penetrating ear trauma, acoustic trauma, and barotrauma are also implicated in PLF etiology. Onset of PLF is not necessarily immediate, however. In fact, cochlear and vestibular problems are reported to represent the largest group of delayed complications of head injuries, with main causes of PLF resulting from stapedial footplate subluxation or fracture after head trauma. Patients who present with vertigo and hearing loss following head trauma should be explored for PLF.

Middle-ear explorations often fail to reveal a fistula of the window membranes; rather, fistulas of the lateral semicircular canal are reported to occur in as many as 25% of PLF cases. Blunt blows to the ear can produce PLF, such as striking the ear with hand slaps, which may cause tympanic membrane perforations as well. The mechanism causing this type of fistula is implosive.

Penetrating injuries of the temporal bone, gunshot wounds, Q-tip insertion, and surgical intervention may all potentially cause PLF. Round window fistulas may be caused by flying and diving barotrauma, and are attributed to forceful movement of the stapes while achieving middle-ear inflation through Valsalva maneuver. High-intensity acoustic trauma or blast can cause a PLF with or without a tympanic membrane perforation. Narula and Marks present two cases of PLF due to acoustic trauma. The first patient was exposed to 130 dBA through a telephone headset, while the second patient was exposed to 118 dBA of music through a loudspeaker.

PLF may be initiated by explosive forces such as sneezing, coughing, laughing, and bending over. These traumatic
events may cause a rupture of the round window membrane or the annular ligament of the stapes footplate.\textsuperscript{5,37,38} Seltzer and McCabe identified PLF of this type in 24% of their series.\textsuperscript{26} Surgical repair of traumatic PLF is recommended, with the incidence of meningitis in such cases as high as 25%.\textsuperscript{40,41}

**SYMPTOMS**

Symptoms associated with PLF include hearing loss, tinnitus, aural fullness, and vestibular symptoms ranging from debilitating vertigo to slight disequilibrium.\textsuperscript{42} Sudden or fluctuating hearing loss is reported in over 80% of patients. Vertigo or disequilibrium occurs in more than 75% of patients, and 60% of PLF patients report tinnitus.\textsuperscript{42} Hearing loss varies from sudden, profound deafness to normal pure-tone sensitivity with fluctuating speech discrimination scores.\textsuperscript{26} Patients may also present with moderate hearing loss and low speech discrimination ability, and fluctuating speech discrimination may be the only hearing-related symptom in over 20% of PLF cases.\textsuperscript{26} Normal hearing and good word discrimination ability are also reported in PLF cases.\textsuperscript{45} Such cases reportedly often present with conductive hearing loss and concomitant ossicular damage. Other conductive losses reported in PLF cases are explained by Goodhill’s theory, in which the hearing loss is caused by perilymph backsplash causing decreased energy from the middle ear to the inner ear, resulting in a conductive hearing loss.\textsuperscript{44}

Vestibular symptoms include true vertigo, disequilibrium, lightheadedness, motion intolerance, and combinations of these symptoms. Seltzer and McCabe found that in their patient series of over 100 ears some reported initial severe episodic vertigo that decreased with time.\textsuperscript{26} Other patients reported an increase in vertigo with physical exertion or straining. Persistent disequilibrium is a common complaint in PLF patients, as is intermittent dizziness. However, intermittent dizziness may be difficult to diagnose, especially in children; and, cases of bilateral disease are reportedly greater than 50% of PLF childhood cases, making this age group especially at risk for permanent inner-ear damage.\textsuperscript{45}

Vertigo is the only presenting sign of a patient with fistula reported by Koltai and Galos,\textsuperscript{46} who suggest that fistula causes a change in the endolymphatic space of the vestibular labyrinth by decompressing the perilymphatic space. When the fistula is surgically closed, a normal fluid balance between endolymph and perilymph may then exist so symptoms disappear.

Tinnitus is also a highly variable symptom in PLF patients. It is reported in PLF patients without measurable hearing loss,\textsuperscript{47} and it is perhaps the most commonly reported auditory symptom. Tinnitus descriptions range from roaring to high-pitched noise occurring intermittently to constantly in over 60% of PLF cases.\textsuperscript{26}

Although hearing loss, pressure, tinnitus, and vertigo are the symptoms most common to PLF as well as Ménière’s disease, the two diseases can be distinguished. Patients with Ménière’s disease often have fluctuating hearing, disabling vertigo, ear pressure, and distracting tinnitus, while PLF patients have stable hearing loss, nondebilitating vertigo, mild ear fullness, and tinnitus.\textsuperscript{48,49,50} However, Swift\textsuperscript{51} describes a patient with classic Ménière’s symptoms who had a PLF of the round window discovered during surgery, suggesting that many Ménière’s patients might actually be PLF cases. Swift reported the symptom of aural fullness as helpful in determining which ear to explore in 25% of cases without other localizing signs.

**DIAGNOSING PLF**

Several signs, symptoms, and tests are used in diagnosing PLF. In an attempt to diagnose a fistula, audiometric evaluation including positional audiometry, electronystagmography (ENG), ENG fistula test, Hennebert’s sign and symptom, Tullio test, glycerin testing, Quix testing, eyes-closed turning test, x-ray examination, and case history are useful.\textsuperscript{26}
Audiometric testing in PLF patients may show any type of hearing configuration including high-frequency, low-frequency, or flat audiometric loss and poor to excellent word discrimination scores. Threshold audiometry performed in various positions, including the horizontal position with the affected ear up, has been suggested. Such positioning might allow air to enter the membranous labyrinth when vertical, and interferes with stapes vibration, thus reducing thresholds, whereas in the horizontal position the cochlea refills with perilymph and hearing improves. The procedure may be sensitive to the site of the fistula because oval window fistulas correlate with 500-Hz threshold change and round window PLF correlates with 8000-Hz threshold change during positional audiometry. The procedure has particular promise when air is seen in the cochlea on x-ray. Thin sectioned laminography may show air aspirated into the cochlea or semicircular canal. In theory, air might enter the vestibule in an implosive manner, and the round window membrane would rupture, simultaneously relieving pressure and minimizing hearing loss following intracochlear membrane break.

A report by Nishioka and Yanagihara described the presence of air bubbles in experimental perilymph fistula as an explanation for inner ear morphology. Gibson described the condition of “dry cochlea” in patients with PLF as symptomatic, and other experimental studies corroborate this report.

ENG fistula testing may provide useful information if a positive result is obtained, but does not rule out a fistula if a negative result is obtained. Localization of a fistula may also be determined using the ENG fistula test as suggested by Koltai and Galos. Positive pressure applied to a patient’s right ear produced right beating nystagmus and vertigo, and round window fistula was confirmed surgically. The fistula test is also helpful in the differential diagnosis of patients who have symptoms and signs similar to Ménière’s disease but are suspected of having a fistula.

Fistula test without ENG was described by DeWeese and Saunders using a Politzer bag to increase ear canal pressure. Response of sudden vertigo frequently indicated PLF, especially in cases where cholesteatoma was suspected of eroding through the horizontal semicircular canal wall.

Hennebert testing may reveal Hennebert’s sign or symptom. This describes either a visually observed deviation of the eyes with manual application of positive or negative pressure to the external auditory canal or a subjective sense of disequilibrium. The procedure is positive in as many as 90% of PLF cases. When the procedure is positive for both ears, bilateral PLF may be confused with syphilis. An objective Hennebert’s sign was seen relatively rarely in Potter and Conner’s patients, yet when observed it was associated with oval window fistula. These investigators recommend tests for fistula and Hennebert’s sign and, when positive, exploratory tympanotomy is indicated.

Tullio phenomenon, or disequilibrium induced by sound, has been identified in some PLF patients. Test procedures vary, although presentation of intense, high- and low-frequency pure tones or narrow bands of noise for several seconds is used successfully in PLF diagnosis.

Glycerin testing is advocated because patients with PLF may have a deficiency of perilymph and a relative excess of endolymph, which would therefore dehydrate favorably and improve hearing. In patients where Ménière’s disease is not suspected and head trauma may have caused the symptoms, glycerin testing is helpful in diagnosing fistula.

Lehrer and colleagues suggest vestibular testing for PLF evaluation, including Quix test for body deviation. This is performed by having the patient stand with feet together, eyes closed, chin raised, and arms and index fingers extended. A gradual sway in a lateral direction is a positive result. Singleton devised an eyes-closed turning test in which patients attempt to walk a straight line with eyes closed, then on command turn quickly either right or left 180° and stop with subsequent instability noted. He stated that patients with a positive eyes-closed turning test will identify the ear involved by falling toward the affected side.
ENG testing may reveal positional abnormalities in one or more positions characterized by lack of latency, long duration, slow or no fatiguability, and not necessarily with the affected ear down. Spontaneous nystagmus was seen to beat to the opposite side of the affected ear in the majority of PLF cases in the patient sample described by Singleton. Bithermal caloric testing has not proven as useful in demonstrating asymmetries between vestibular labyrinths. Experimental caloric testing produces variable slow-phase velocity and prolonged duration.

Electrocochleography (ECoG) is useful in identifying endolymphatic hydrops (ELH) secondary to PLF. Summating potential is enhanced in more than 50% of active PLF cases, but findings fail to differentiate PLF cases from ELH patients with classic Ménière’s disease.

SITE OF PLF

PLF may be more common in left ears than in right ears. Goodhill attributes this finding to studies of the temporal bone where he theorizes that the patency of the cochlear aqueduct is inversely proportional to the size of the lateral sinus, and in the temporal bones studied there were smaller sinuses and larger cochlear aqueducts on the left side. Oval window PLF is more commonly reported than round window fistula or fistulas of both window membranes. More than half of PLF cases occur at the oval window and most are attributed to spontaneous and traumatic causes. The most common area of perilymph leak from the oval window region, not associated with prior ear surgery, is the fissula ante fenestram.

Round window PLF, though less common than oval window PLF, may have a higher incidence of deformity. Such defects, along with congenital tympanic cavity anomaly, are reported in over 30% of cases. Round window niche may be abnormal in over 90% of cases and the membrane is frequently in a more vertical orientation than normal. Fistulas occurred in both window membranes in 9% of the patients described by Seltzer and McCabe.

Yaniv and Traub studied head injury patients who had fistulas of the lateral semicircular canal. They reported that patients with head injuries who also experience vestibular and cochlear symptoms should be explored after 10 days for the possibility of a fistula. If a window fistula is not observed, the lateral semicircular canal should be explored. If a window fistula is repaired and symptoms persist, then exploration of the lateral semicircular is indicated. They reported the following case:

Patient was initially operated on 12 days after injury. At that stage his hearing was 30 dB. A fistula discovered in the round window was closed. Postoperatively his vertigo improved slightly, but his hearing deteriorated to 55 dB. Six weeks later he underwent surgery again. The round window was found to be intact. Surgery was performed and a fracture which was found along the lateral semicircular canal with perilymph leakage was closed. Two days postoperatively he was free of vertigo, and three weeks later his hearing improved to 30 dB, but improved no further.

These researchers emphasize that, although the incidence of lateral semicircular canal fistulas is probably higher in their series because of the severity of injuries they see, patients with vestibular and cochlear symptoms following head trauma may have fistulas of the lateral semicircular canal.

TREATMENT OF PLF

Perichondrium, subcutaneous tissue, and fat are used for grafting, while gelatin sponge is often used to support the grafts. Vein and tragal perichondral grafts have also been used. The grafting material of choice may depend on where the fistula is and whether it is necessary to remove the stapes to view the fistula. Seltzer and McCabe prefer to use subcutaneous areolar tissue because it drapes better than fascia, perichondral tissue, or other material.

Failures of patching are often attributed to fat grafts. Over half the fistulas
Results are good in controlling vertigo after fistula repair. Seltzer and McCabe report improvement in 95% of patients with vestibular symptoms by eliminating dizziness or decreasing it sufficiently to not interfere in daily living. Initial repair of PLF using fat resulted in 69% of recurrences of fistulas in the series reported by Seltzer and McCabe. Four of 10 graft sites using fat failed in the series of Potter and Conner, while vein and tragal perichondral grafts were not associated with refistulization. No patching material is seen as perfect according to Seltzer and McCabe. They report a failure rate of 15% for areolar tissue, 11% for perichondral tissue, 11% for fascia only, and 10% for prosthesis and fascia.

Preparation of the graft site may be more important than the type of seal chosen. The graft site must be exposed sufficiently and enough gelatin sponge must be placed over the grafting material to keep it in place during the postoperative period. Singleton suggests that when packing the ear tightly with Gelfoam adhesive bands may form between the eardrum and the graft material. He noted that these patients experienced vertigo with pressure movement of the tympanic membrane. The apparent sensorineural hearing loss experienced by these patients disappeared after the bands connecting the eardrum and the tissue graft were cut. To prevent formation of adhesions between the drum and graft, Singleton recommends covering the Gelfoam packing with Gelfilm sheet.

Surgical repair of PLF may have an unanticipated result. Patients who have symptoms of hydrops secondary to elevated perilymph rather than endolymph pressure may be less symptomatic because of the presence of a fistula. With spontaneous or surgical closure of the fistula, symptoms may increase and recurring PLF may develop. This might explain the rare case where surgical closure of the PLF causes symptoms to become worse. Supporting this theory, Goode suggests that the perilymph hypertension may be a cause of ear disease that presents similarly to endolymphatic hydrops. Pathologic changes show a shift of Reissner’s membrane toward the organ of Corti.

HISTORICAL PERSPECTIVE—ACKLEY ET AL.

Results are good in controlling vertigo after fistula repair. Seltzer and McCabe report improvement in 95% of patients with vestibular symptoms by eliminating dizziness or decreasing it sufficiently to not interfere in daily living. All surgically confirmed cases of PLF reported by Yaniv and Traub were without symptoms of vertigo and dizziness postsurgically. Halvey and Sade reported complete elimination of vertigo in 69% of patients, but unsteadiness persisted in 36%. Potter and Conner reported elimination of vertigo in 80%. Vertigo and disequilibrium were cured in all patients described by Lehrer and colleagues.

When hearing loss occurs following PLF, surgery provides the best chance of restoring hearing. Spontaneous PLF may heal without surgical intervention, but in many cases permanent cochlear damage may occur unless treatment is prompt. Hearing improved following surgery in 22% of patients reported by Halvey and Sade, while 56% showed no change and 22% had greater hearing loss. Althaus reported improved hearing in 34% and greater postsurgical hearing loss in 27%. Seltzer and McCabe reported improved hearing in 49% with 23% of those with a speech reception threshold (SRT) better than 35 dB and speech discrimination ability better than 80%. They found poorer postoperative hearing in 11% and stabilized hearing in 40%. As high as 80% of patients have improved hearing following closure of the PLF, although few improve to normal hearing.

A common treatment in cases of traumatic or sudden hearing loss where PLF is suspected is a trial medical management consisting of bed rest, head elevation, and avoiding straining to allow for spontaneous healing. If symptoms or hearing become worse, or if no change occurs in the first 4 weeks, then exploratory tympanotomy is advised. Emmett and Shea caution that patients with traumatic PLF may be symptomatically indistinguishable from patients with tympanic membrane perforation, who may have symptoms of hearing loss, tinnitus, unsteadiness, and vertigo. However, because a high percentage of traumatic PLF cases also involve eardrum
perforation (64% according to Emmett and Shea) surgical repair of both window membranes should always be considered when tympanic membrane perforation occurs following trauma. Althaus recommends bed rest and head elevation if the main post-traumatic symptom is sudden sensorineural hearing loss. If after 1 week there is no improvement in hearing, exploratory tympanotomy is advised.

**SUMMARY**

More than 200 medical references describe PLF in clinical cases and in laboratory animal models. Fewer than half a dozen authors deny PLF as a pathologic entity. Laboratory experimental evidence of PLF is convincing. Mystery surrounding the disorder focuses not on its existence, but rather on the challenge to clinicians to identify patients with PLF as diagnostically distinct from patients with classic Ménière’s disease.

**ACKNOWLEDGMENTS**

The authors acknowledge Cadwell Labs, Inc., Colorado Hearing Foundation, NIH (BRSG) Grant 5-35367, and R. F. Krug.

**REFERENCES**


