Perilymphatic Fistula

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Author: Joe Walter Kutz, Jr, MD, FACS; Chief Editor: Arlen D Meyers, MD, MBA

Overview

Background

Perilymphatic or labyrinthine fistula is a condition in which an abnormal communication is present between the perilymphatic space of the inner ear and the middle ear or mastoid. The manifestations of this disease vary in severity and complexity, commonly ranging from very mild to incapacitating. Perilymphatic fistulas (PLFs) may induce hearing loss, tinnitus, aural fullness, vertigo,[1] disequilibrium, or a combination of these symptoms. The vagueness of symptoms caused by perilymphatic fistula (PLF) and the overlapping symptoms of other disease processes make the diagnosis elusive.

History of the Procedure

Sustained interest in perilymphatic fistula (PLF) began in the mid 1960s. The initial focus was on perilymphatic fistulas (PLFs) that developed following stapedectomy procedures. The recognition that loss of perilymph following stapedectomy could produce hearing loss, disequilibrium, and tinnitus led to the recognition that these same symptoms could arise from perilymph loss caused by other types of trauma. In the early 1970s, Victor Goodhill first distinguished between implosive and explosive forces as potential causes for rupture of the round or oval window membrane leading to PLF.[2]

Problem

General agreement exists that perilymphatic fistula (PLF) occurs and that leakage of perilymph from the inner ear into the middle ear can cause acute hearing loss, paroxysmal vertigo, disequilibrium, and tinnitus. Virtually no consensus exists regarding how frequently perilymphatic fistula (PLF) occurs, how it should be diagnosed, or how it should be treated when suspected. A high degree of controversy surrounds the appropriate indications for surgical exploration and operative repair of perilymphatic fistula (PLF).

Epidemiology

Frequency

The frequency of perilymphatic fistula is unknown, but it is a very rare condition.

Etiology

Perilymphatic fistula (PLF) occurs when perilymph leaks from the perilymphatic spaces of the bony labyrinth into the middle ear space. The loss of perilymph alters the balance between perilymph and endolymph within the membranous labyrinth. PLF is thus a form of inner ear fluid imbalance. Ménière disease, another form of inner ear fluid imbalance, occurs when endolymph is present in overabundance. Both disease entities alter the endolymph-to-perilymph ratio in the same direction, which may account for the difficulty involved with separating their clinical presentations.
Pathophysiology

Inner ear fluids are almost completely contained within a rigid bony framework. However, the perilymphatic space is connected to the subarachnoid space via the cochlear aqueduct. The size of the cochlear aqueduct varies dramatically between subjects. Even when it is relatively large, the cochlear aqueduct is often completely obstructed with arachnoid tissue. Evidence exists that the arachnoid becomes denser with increasing age. Thus, the patency of the cochlear aqueduct varies among persons. Establishing whether the cochlear aqueduct is patent in any particular individual is impossible.

A variety of indirect evidence suggests that pressure changes within the subarachnoid space can be communicated to the inner ear. Such pressure changes may be communicated through the cochlear aqueduct or alternatively through a patent lamina cribrosa, cochlear modiolus, or vestibular aqueduct. At least in some patients, stapes gushers appear to arise from a defective modiolus, allowing free communication between the spinal fluid and perilymphatic space.

The length of the cochlear aqueduct and the presence of arachnoid within its lumen tend to dampen the effects of sudden pressure changes in the subarachnoid space, thereby protecting the inner ear from rapid pressure changes. Simultaneous increases in pressure on the endolymphatic sac also protect the intracochlear membranes by equalizing endolymphatic and perilymphatic pressure. Even so, sudden increases in subarachnoid pressure can be transferred to the inner ear fluids.

At the round window membrane and the annulus of the oval window, the middle ear space is separated from the perilymphatic space by a soft tissue membrane. These areas permit pressure change between the middle ear and perilymphatic space. Excess pressure generated on either side of these membranes can result in a tear or rupture leading to the egress of perilymphatic fluid from the membranous labyrinth. In addition to the oval window annulus and round window membrane, which are present in every person with normal hearing, a number of other sites for perilymphatic fistula (PLF) are possible in some individuals.

- The fistula ante fenestram lies anterior to the anterior portion of the stapes footplate and is an embryonic residua formed by the resorption of precartilage in the fetus.

- Microfissures have been noted with regularity extending from the ampulla of the posterior semicircular canal to the round window.

- Congenital anomalies of the inner ear are associated with a much higher incidence of perilymphatic fistula (PLF). Incomplete partitioning of the cochlea (Mondini dysplasia) is often accompanied by incomplete formation of the stapedial footplate and a high incidence of perilymphatic fistula (PLF).

Goodhill separated the hydrodynamic forces that could potentially produce perilymphatic fistula (PLF) into implosive and explosive forces. Everyday activities, such as lifting, straining, coughing, and sneezing, are associated with increases in cerebrospinal fluid (CSF) pressure. Such pressure increases can be transmitted to the inner ear via a patent cochlear aqueduct or through the lamina cribrosa of the internal auditory canal. A precipitous increase in pressure within inner ear fluids can result in tears of the oval window annulus or round window membrane. Vulnerability to such tears may depend on the inherent strength of these tissues, which may vary among individuals. Goodhill termed tears produced by increased pressure within the inner ear fluid compartment as explosive.[2]

Conversely, pressure can increase rapidly within the middle ear space as a result of barometric pressure change, compression trauma of the ear, Valsalva maneuver, and pinched-nose sneezing. Ruptures or tears of the round window membrane, oval window annulus, or membranes protecting a fistula ante fenestram that result from increased middle ear pressure are also termed explosive forces by Goodhill.

In addition to changes related to hydrodynamic pressure, head injury may produce rupture of the membranes that seal the inner ear and prevent escape of perilymphatic fluid into the middle ear space.

The precise mechanism by which perilymph loss produces hearing loss is unclear. Decompression of the perilymphatic space may create secondary endolymphatic hydrops. Consequently, the usual symptoms of endolymphatic hydrops (Ménière disease) arise. Secondary endolymphatic hydrops has been histologically identified in experimental animals with perilymphatic fistula (PLF). Compelling indirect evidence exists that hearing loss in this circumstance is the result of loss of perilymphatic pressure. A large number of reports describe hearing loss associated with decreased subarachnoid pressure secondary to spinal anesthesia. Hardy demonstrated that such hearing loss can be reversed by injecting 20 mL of isotonic sodium chloride solution into the subarachnoid space.[3]

In 1974, Robertson demonstrated changes in the tuning curves of single spiral ganglion cells from the basal membrane after removal of perilymph from the tympanic scala of the guinea pig.[4] Flint et al showed an increase in auditory brainstem response (ABR) thresholds of 10-15 dB in guinea pigs with artificially created PLF; however, the etiology remains controversial.[5] Bohmer demonstrated no change in auditory thresholds measured in guinea pigs with simple round window membrane perforation, even though a decrease was noted in the normal positive hydrostatic pressure within the cochlea.[6]
Evidence of auditory change has also been provided by measuring electrocochleographic changes after the creation of perilymphatic fistula (PLF). Ackley demonstrated consistent increase in the ratio of the summating potential (SP) to the action potential (AP) in guinea pigs with obstructed cochlear aqueducts after the creation of PLF.[7] Others have subsequently confirmed these results.

**Presentation**

Seltzer and McCabe gathered clinical data from 91 patients with surgically confirmed perilymphatic fistula (PLF).[8] Most patients (90%) had auditory symptoms, generally hearing loss. The character of the loss varied from sudden and profound to mild and fluctuating. Tinnitus was noted in 63% of patients, and aural fullness was noted in 25%. Glasscock et al reported that 83% of their patients with traumatic PLF had complaints of sudden or fluctuating sensorineural loss.[9]

Vestibular symptoms occur in most patients. Balance disturbance can be described as true rotational vertigo, lightheadedness, disequilibrium, intolerance to motion, or any combination of these symptoms. Seltzer and McCabe reported such symptoms in 80% of patients with PLF, and Glasscock et al reported 77% incidence of true vertigo or disequilibrium.[8, 9]

In obvious cases, membrane rupture is accompanied (or followed within a few minutes) by rapid severe hearing loss, loud roaring tinnitus, and severe rotational vertigo. Vertigo is often incapacitating and accompanied by visceral symptoms (eg, sweating, pallor, nausea, vomiting). Even cursory examination demonstrates marked instability and nystagmus. Audiometric evaluation reveals sensorineural hearing loss. Platform posturography confirms disequilibrium with a vestibular pattern, and platform fistula test results are positive. Vertigo and, to a lesser degree, tinnitus and hearing loss are sometimes exacerbated by straining (the Valsalva maneuver).

Unfortunately, many perilymphatic fistulas (PLFs) do not manifest in a straightforward fashion. A combination of otologic symptoms may result, and symptoms may fluctuate in complex ways that are difficult for the patient to explain. Onset of symptoms may be delayed by several days, or the acute phase may be masked by more serious injuries in other areas. Rotational vertigo may be entirely absent, and disequilibrium may be mild, vague, and episodic. Hearing loss, tinnitus, and aural fullness may come and go unpredictably. Such elusive symptomatology is partially responsible for the controversy surrounding this disorder.

The diagnosis of perilymphatic fistula (PLF) often depends on the antecedent history of otologic surgery, trauma, diving, or congenital ear malformation. Fluctuating or sudden hearing loss with or without vestibular symptoms in a patient with a previous stapedectomy, for example, is highly suggestive of perilymphatic fistula (PLF).

Trauma is frequently cited as the cause of a perilymphatic fistula (PLF). Changes in the middle ear pressure sufficient enough to implode the round window membrane can occur during descent while flying, forced Valsalva maneuver, or suppressed sneezing. Pullen reported that 48 of 62 patients with suggested PLF following scuba diving had round window membrane rupture.[10] Pressure sufficient to rupture the round window membrane can develop at a depth of water as shallow as 4 ft. Explosive perilymphatic fistulas (PLFs) can arise from CSF pressure increases caused by physical exertion, coughing, and straining.

Klokker and Vesterhauge have reported on 4 cases of perilymph fistula that occurred in flight attendants who flew with upper respiratory infections during a 6-month period.[11] All 4 flight attendants work for a major Scandinavian airline that employs approximately 3,000 flight attendants.

Perilymphatic fistula (PLF) may be the ultimate etiology of progressive sensorineural hearing loss in some children. Its frequency remains unknown and is contested. Grundfast and Bluestone reported that 66% of 33 children with progressive sensorineural hearing loss had PLF at exploratory tympanotomy.[12] Reilly and Kenna prospectively evaluated 244 children with sensorineural hearing loss of unknown etiology; of the 54 ears that were surgically explored, 42% demonstrated active PLF.[13]

Weber et al, in a follow-up article, confirmed that surgical repair of perilymph fistula does not result in a significant risk of postoperative hearing loss and that fistula repair may prevent further hearing loss, even in patients in whom a perilymph fistula was not identified at the time of surgery.[14]

**Indications**

The indications for exploratory tympanotomy are controversial. Accurate diagnosis is difficult. If the patient’s history is suggestive of perilymphatic fistula (PLF), objective testing should be used to reinforce or reject the initial assessment. The signs and symptoms of perilymphatic fistula (PLF) are relatively nonspecific and overlap greatly with those seen in other otolaryngologic and neurologic diseases. Perilymphatic fistula (PLF) can be particularly difficult to differentiate from Ménière disease. Histologically, the two diseases are similar.
However, the final diagnosis of perilymphatic fistula (PLF) requires integration of all available information. Because perilymphatic fistulas (PLFs) can be difficult to diagnose, some surgeons believe that patients should be given the opportunity to choose surgical exploration even when perilymphatic fistula (PLF) is unlikely. Such an approach leads to many explorations with negative findings and many patients who do not benefit from the procedure. Conversely, a few patients benefit who would otherwise have had undiagnosed perilymphatic fistula (PLF).

Other surgeons prefer to use fairly rigid criteria for the diagnosis of PLF. These surgeons explore fewer individuals and have a much higher rate of positive intraoperative findings. A high percentage of their operative cases improve following surgery; however, some cases of perilymphatic fistula (PLF) are missed. These patients may have lost an opportunity for clinical improvement or cure.

Objective testing becomes especially important when the history of antecedent trauma is vague or remote. A perilymphatic fistula (PLF) test or elevated SP/AP ratio on electrocochleography (ECoG) significantly raises the likelihood of perilymphatic fistula (PLF).

Contraindications

No absolute contraindications exist to exploratory tympanotomy with round and oval window grafting in patients who can tolerate either a local or general anesthetic. A physician must exercise the usual cautions in an only-hearing ear, even though the incidence of significant hearing loss associated with this operation is low. To justify operative intervention, one must feel convinced that the likelihood of additional hearing loss is greater without surgery than with surgery.

Workup

Laboartory Studies

See the list below:

- No laboratory studies are relevant to the diagnosis of perilymphatic fistula (PLF). A possible future role for beta2-transferrin assay is discussed in Intraoperative details.

- A stable perilymph specific protein, Cochlin-tomoprotein has been characterized at the Nippon Medical School in Tokyo. Western blot assays revealed that Cochlin-tomoprotein is present in all perilymph samples and reliably absent in nonperilymph samples (specifically 98.2%).[15, 16]

Imaging Studies

See the list below:

- Obtain gadolinium-enhanced MRI scans in all individuals with unilateral otologic symptoms to exclude acoustic neuroma or other structural lesions of the cerebellar pontine angle or neuraxis.

- The incidence of perilymphatic fistula (PLF) is much higher in individuals with congenital deformities of the otic capsule. Consequently, a nonenhanced fine-cut CT scan of the temporal bones can be helpful, especially in children. A child with progressive or sudden sensorineural hearing loss in an ear that is incompletely developed has a much higher risk of perilymphatic fistula (PLF) than a child with normal inner ear radiographic morphology.

Other Tests

Audiography

Obtain an audiogram in all patients with otologic symptomatology. Sensorineural hearing loss is a regular feature of perilymphatic fistula (PLF), but any pattern of sensorineural hearing loss can result from perilymphatic fistula (PLF). Because it is otherwise relatively uncommon, low-frequency hearing loss is perhaps a bit more suggestive of perilymphatic fistula (PLF);
However, mid-frequency, high-frequency, and flat losses are also consistent with the diagnosis. Fluctuating hearing loss is common with perilymphatic fistula (PLF), as it is with Ménière’s disease.

Some authors have reported a conductive loss in patients with perilymphatic fistula (PLF), but this probably represents a small percentage of patients.

**Electrocochleography**

ECoG is a method of measuring intracochlear electrical potential changes associated with hearing. Three separate responses can be obtained, as follows:

- The cochlear microphonic is a stimulus-related alternating current (AC) potential that closely mimics the stimulus and therefore is difficult to separate from stimulus artifact. The technique used to produce standard electrocochleographic tracings eliminates the cochlear microphonic from the ECoG tracing.

- The summating potential (SP) is a stimulus-related direct current (DC) potential that reflects the time-related displacement of the cochlear partition. The SP has been shown to be sensitive to inner ear fluid imbalance, particularly in Ménière disease and perilymphatic fistula (PLF).

- The action potential is an AC potential that represents the compound action potential of the fiber's eighth nerve that discharges synchronously in response to a stimulus. The initial portion of the action potential also is known as wave I of the ABR.

The two methods of recording ECoG are transtympanic and extratympanic.

- Transtympanic: A needle electrode passed through the tympanic membrane that rests on the promontory provides the largest and most easily readable tracings because it is a near-field potential. Fewer signals need to be averaged to obtain an interpretable response, and the potentials are much longer than in other methods of ECoG. Acceptance of transtympanic ECoG has been limited because it can be painful and because a physician's presence is required to pass the electrode through the tympanic membrane.

- Extratympanic: Extratympanic ECoG is comfortable and noninvasive, and it can be performed in a nonmedical setting. However, the size of the reported potentials is significantly smaller than with transtympanic ECoG, and, consequently, the reported potentials are more difficult to identify and interpret.

ECoG is useful in the diagnosis of both Ménière disease and PLF. Both conditions produce an elevated SP/AP ratio. Increase in the SP/AP ratio appears to result because of enlargement of the SP component in patients with Ménière disease. The mechanism by which it increases in perilymphatic fistula (PLF) is controversial. Some authors believe that the increase in SP/AP ratio in patients with perilymphatic fistula (PLF) may be the consequence of a decrease in the AP.

Several guinea pig studies have shown consistent increases in the SP/AP ratio in guinea pigs with artificially induced perilymphatic fistulas (PLFs). Gibson has demonstrated an increase in the SP/AP ratio during stapedectomy.[17] The creation of a control hole in the footplate is not sufficient to produce a change in the SP/AP ratio; a change is noted only after some perilymph has been removed from the vestibule. Meyerhoff and Yellin have demonstrated that in individuals with surgically proven perilymphatic fistulas (PLFs) who have elevated SP/AP ratios preoperatively, the SP/AP ratio reliably returns to normal after surgical repair of the fistula.[18]

**Fistula test**

Fistula testing has been shown to yield positive results in patients with perilymphatic fistula (PLF). The application of positive pressure to a tympanic membrane in an ear with a fistula is known to possibly produce nystagmus. The production of nystagmus secondary to positive pressure is referred to as a positive fistula test result. The definition actually requires the presence of documentable nystagmus. The reproduction of symptomatology secondary to positive pressure may have diagnostic importance but does not constitute a positive fistula test.

An objective record of fistula testing can be made using the electronystagmogram (ENG) and the impedance bridge. To accomplish this, the emittance probe is placed into first one ear and then the other. The pressure in the external auditory canal is varied between +200 and -200 mm of mercury. The ENG has been examined for induced nystagmus. Each ear is tested separately. A positive fistula result is identified by the production of nystagmus associated with a change in pressure on the tympanic membrane. In some cases, the nystagmus can be seen to change direction as the pressure changes from positive to negative. One would expect that the patient's subjective symptoms of vertigo, with or without nausea, would be induced during the presence of nystagmoid eye movements in a positive test result. The results of the ENG fistula test then can be compared to platform fistula test results.

**Dynamic platform posturography**
This test can be used to generate a sensitive test for perilymphatic fistula (PLF). In dynamic platform posturography, pressure is applied to the external auditory canal. The increase or decrease in pressure is transmitted to the tympanic membrane middle ear space and, if a fistula is present, to the inner ear. When perilymphatic fistula (PLF) is present, abnormal sway is generated by these pressure changes. Using the acoustic impedance bridge to quantify changes in external auditory canal pressure and the dynamic platform posturography to quantify anterior, posterior, and lateral sway in response to such pressure changes, a sensitive assessment for perilymphatic fistula (PLF) can be developed. Several studies have demonstrated that patients with positive results from platform fistula testing have a high likelihood of having perilymphatic fistula (PLF).

Vestibular-evoked myogenic potential (VEMP) testing

VEMP testing is a newer diagnostic tool to evaluate patients with vestibular disorders. During VEMP testing, a suprathreshold sound is administered to the test ear. Relaxation potentials of the ipsilateral sternocleidomastoid muscle are measured and quantified. In individuals with normal hearing and vestibular function, the VEMP threshold to sounds is from 90-105 dB HL. In patients with superior canal dehiscence or perilymphatic fistula, the thresholds may be decreased to as low as 70 dB HL. Decreased VEMP thresholds with a history and symptoms suggestive of a perilymphatic fistula can provide further evidence of the presence of a fistula.[19]

Diagnostic Procedures

Poe et al performed middle ear endoscopy in 20 patients with suggested perilymphatic fistula (PLF).[20] They failed to demonstrate fistulas in any patient but placed autologous blood patches in 8 patients around the round and oval windows. No change in hearing was noted postoperatively, but 3-4 patients with preoperative vertigo had relief of symptoms, and 2-3 patients with preoperative positive fistula test results had negative test results postoperatively.

The most troublesome difficulty described by Poe was obscured vision of the oval window and round window niche, secondary to mucosal adhesions. The average human ear contains only 0.07 mL (70 mcL) of perilymph; therefore, even relatively rapid leaks are, in absolute terms, quite small. Even with magnification, leaks involving only 5-10% of the perilymph are difficult to see in an operative field because local anesthetics have been injected, irrigating fluids have been used, and a minimal amount of bleeding may be present.

Garg and Djalilian reported resolution of symptoms in two of three patients with presumed traumatic perilymphatic fistulas after intratympanic injection of blood into the middle ear space.[21]

Treatment

Medical Therapy

Some tears of the inner ear membranes probably heal without surgical intervention. Medical management includes bed rest, elevation of the head of the bed, use of stool softeners, avoidance of Valsalva maneuver, and sedation. Repeated audiometric evaluations should be performed, and medical management should be reconsidered if hearing deteriorates or balance disturbance fails to improve.

Preoperative Details

The definitive treatment of perilymphatic fistula (PLF) is surgical exploration with grafting of the fistula. Early repair of PLF offers the potential for resolution of vestibular symptoms and preservation of residual auditory function. Many authors recommend immediate surgical exploration when the likelihood of perilymphatic fistula (PLF) is high. The timing of surgical exploration in the less-defined case is controversial. Depending on strict diagnostic criteria risks underdiagnosis of the disease, whereas overzealous exploration of all patients with auditory and vestibular complaints risks performing many procedures that, in retrospect, are unnecessary. Meyerhoff and Pollock have recommended that the patient become intimately involved in the decision-making process.

Intraoperative Details
The procedure can be performed with the patient under local or general anesthesia. A standard tympanomeatal flap is designed, incised, and elevated. Generally, curetting away the posterior bony overhang (scutum) is necessary to permit adequate visualization of the round and oval window niche. These areas are then very carefully observed for the accumulation of clear fluid. However, even intraoperative observation can be inconclusive. Transudates from middle ear mucosa, irrigation, or injected anesthetic materials can collect within the dependent areas of the round window or oval window niche. The absence of detectable fluid in these areas does not exclude an intermittent or recurrent perilymphatic fistula (PLF). Use of intravenous fluorescein is not helpful because it can accumulate in the round window niches as a transudate from middle ear tissues.

Beta2-transferrin is specific for human aqueous humor, CSF, and perilymph. Western blot analysis for beta2-transferrin allows identification with very small amounts of dilute fluid. Unfortunately, the time required to perform the test prevents its use in surgical decision making. Preoperative irrigation of the middle ear via myringotomy has been suggested as a method of obtaining dilute perilymph that could be tested by Western blot analysis for the presence of beta2-transferrin, thus providing preoperative evidence of perilymphatic fistula (PLF). Intermittent or inactive perilymphatic fistula (PLF) is overlooked by this test, and its risk to the patient approaches the risk of exploratory tympanotomy. Additionally, sensitivity of Western blot analysis for beta2-transferrin has been demonstrated to be very low (29%) in one study testing specimens of known perilymph.

Provocative testing to confirm perilymphatic fistula (PLF) is advocated by some and includes intraoperative Valsalva maneuver, Trendelenburg positioning, increase in intrathoracic pressure, and compression of the internal jugular vein. Black et al observed postoperative hearing loss after intraoperative performance of provocative maneuvers in several patients; they since have discontinued this practice.[22]

Grafting is performed by removing mucosa of the round and oval window area. Autogenous tissue grafts are placed directly over the leak. If no actual leak is identified, the footplate and round window are grafted prophylactically. Adipose tissue originally was used, but its use resulted in an unacceptably high rate of recurrent fistula. Fascia or perichondrium now is used; this is reported to have decreased the incidence of recurrent fistula. Surgeons at the University of Texas Southwestern Medical Center make a small incision beneath the lobule from which parotid masseteric fascia is harvested for use as a graft. Some authors use fibrin glue; others do not.

Some surgeons do not graft unless an active leak is visualized. Other surgeons graft routinely, even if no leak can be detected by visual inspection. A poll of 167 members of the American Otologic and Neurotologic Society revealed that 78% placed grafts in all patients undergoing exploratory tympanotomy for PLF, regardless of whether a fistula could be demonstrated at operation.

**Postoperative Details**

Postoperative care is directed at maintaining the integrity of the graft. Patients are instructed to avoid heavy lifting, straining, and activities that place the head in a dependent position, all of which could lead to increasing intracranial pressure. Stool softeners are given for the first 10 postoperative days. Antinausea medications are used as necessary. Air flight is well tolerated in the immediate postoperative period because the middle ear space is filled with transudated blood, therefore providing no air-filled space upon which pressure can act.

**Follow-up**

The patient should be seen again 1-3 weeks postoperatively. A follow-up audiogram should be obtained at 6 weeks, and another follow-up audiogram should be obtained at 6 months. Beyond 6 months, follow-up care is determined by the patient's condition.

**Complications**

Few complications result from perilymphatic fistula (PLF) repair. Tympanic membrane perforations occur in 1-2% of patients. Postoperative conductive hearing loss may persist longer after round and oval window grafting than with simple exploratory tympanotomy. Approximately 5% of patients still have persistent mild (5-10 dB) postoperative hearing loss 2-3 months following surgery; however, in most patients, the loss resolves within 6 months. Some risk exists of severe-to-profound hearing loss. This is especially likely in individuals with Mondini dysplasia or other morphologic defects. These ears are unstable, and surgical manipulation can result in hearing deterioration. Conversely, additional hearing loss is almost certain in such cases, and surgical intervention with round and oval window grafting frequently is the least risky alternative. Alteration of taste as a result of chorda tympani injury occurs with some frequency. It generally resolves within a few weeks to a couple of months.
Outcome and Prognosis

Rizer and House examined the outcome of 86 patients who underwent exploratory tympanotomy for suggested perilymphatic fistula (PLF).[23] Active perilymphatic fistula (PLF) was found in 35 of the patients, and all patients had placement of oval and round window grafts. Improvement was seen in 68% of patients with surgically confirmed perilymphatic fistulas (PLFs), as opposed to 29% who did not demonstrate active perilymphatic fistula (PLF).

Similarly, a retrospective study by Prenzler et al indicated that in patients with profound sudden sensorineural hearing loss (SSNHL), exploratory tympanotomy with sealing of the round and oval windows achieves better results when perilymphatic fistula (PLF) is present than when it is not. The study involved patients with unilateral profound SSNHL who did not show clinical evidence of perilymphatic fistula (PLF), with the investigators determining that in those who were found intraoperatively to have fistula, the postoperative reduction in the percentage of hearing loss was 2.4 times greater than in those not found to have perilymphatic fistula (PLF).[24]

Hearing improvement or restoration is less likely than elimination of balance disturbance after surgical repair of perilymphatic fistula (PLF). Seltzer and McCabe reported that 49% of treated ears improved in terms of auditory function, but only 23% improved to serviceable hearing levels, and 11% had continued hearing deterioration.[8] Rizer and House noted 13.3% of their patients had hearing improvement.[23] Black et al demonstrated hearing improvement in 17% of patients, stabilization of hearing in 67%, and continued progression in 17%. [22] The same authors report improvement of vestibular systems in 83-94% of patients.

Surgical exploration is highly effective for vestibular symptomatology, but its effect on hearing loss is less predictable.

Future and Controversies

The entire area of perilymphatic fistula (PLF) is fraught with disagreement. Investigations in this area are severely constrained by the absence of any reliable tests to determine the presence of perilymphatic fistula (PLF). Because the presence or absence of the condition cannot be determined with certainty, its incidence remains unclear. Frequently, a physician cannot know whether an individual patient had a perilymphatic fistula (PLF); therefore, definitively constructing a typical clinical picture for the disorder is impossible.

A method of determining with certainty whether perilymph is within the middle ear space is needed. Perhaps a rapid assay for beta2-transferrin that could be performed intraoperatively can be developed. Until a technique is developed for unequivocal diagnosis, perilymphatic fistula (PLF) will remain mired in controversy.

Contributor Information and Disclosures

Author

Joe Walter Kutz, Jr, MD, FACS Assistant Professor, Associate Residency Director, Neurotology Fellowship Director, Department of Otolaryngology—Head and Neck Surgery, University of Texas Southwestern Medical School

Joe Walter Kutz, Jr, MD, FACS is a member of the following medical societies: Alpha Omega Alpha, American Academy of Otolaryngology-Head and Neck Surgery, American Neurotology Society, Otosclerosis Study Group, Texas Medical Association, Triological Society

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Coauthor(s)

Peter S Roland, MD Professor, Department of Neurological Surgery, Professor and Chairman, Department of Otolaryngology-Head and Neck Surgery, Director, Clinical Center for Auditory, Vestibular, and Facial Nerve Disorders, Chief of Pediatric Otology, University of Texas Southwestern Medical Center; Chief of Pediatric Otology, Children’s Medical Center of Dallas; President of Medical Staff, Parkland Memorial Hospital; Adjunct Professor of Communicative Disorders, School of Behavioral and Brain Sciences, Chief of Medical Service, Callier Center for Communicative Disorders, University of Texas School of Human Development

Peter S Roland, MD is a member of the following medical societies: Alpha Omega Alpha, American Academy of Otolaryngic Allergy, American Academy of Otolaryngology-Head and Neck Surgery, American Auditory Society, American Neurotology Society, American Otological Society, North American Skull Base Society, Society of University Otolaryngologists-Head and Neck Surgeons, The Triological Society
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Specialty Editor Board

Francisco Talavera, PharmD, PhD Adjunct Assistant Professor, University of Nebraska Medical Center College of Pharmacy; Editor-in-Chief, Medscape Drug Reference

Disclosure: Received salary from Medscape for employment for: Medscape.

Gerard J Gianoli, MD Clinical Associate Professor, Departments of Otolaryngology-Head and Neck Surgery and Pediatrics, Tulane University School of Medicine; President, The Ear and Balance Institute; Board of Directors, Ponchartrain Surgery Center

Gerard J Gianoli, MD is a member of the following medical societies: American Academy of Otolaryngology-Head and Neck Surgery, American College of Surgeons, American Neurotology Society, American Otological Society, Society of University Otolaryngologists-Head and Neck Surgeons, Triological Society

Disclosure: Nothing to disclose.

Chief Editor

Arlen D Meyers, MD, MBA Professor of Otolaryngology, Dentistry, and Engineering, University of Colorado School of Medicine

Arlen D Meyers, MD, MBA is a member of the following medical societies: American Academy of Facial Plastic and Reconstructive Surgery, American Academy of Otolaryngology-Head and Neck Surgery, American Head and Neck Society

Disclosure: Serve(d) as a director, officer, partner, employee, advisor, consultant or trustee for: Cerescan;RxRevu;Cliexa;Preacute Population Health Management;The Physicians Edge<br/>Received income in an amount equal to or greater than $250 from: The Physicians Edge, Cliexa<br/>Received stock from RxRevu; Received ownership interest from Cerescan for consulting; for: Rxblockchain;Bridge Health.

Additional Contributors

Michael E Hoffer, MD Director, Spatial Orientation Center, Department of Otolaryngology, Naval Medical Center of San Diego

Michael E Hoffer, MD is a member of the following medical societies: American Academy of Otolaryngology-Head and Neck Surgery

Disclosure: Received royalty from American biloogical group for other.

Acknowledgements

William L Meyerhoff, MD, PhD Former Chair, Former Professor, Department of Otolaryngology, University of Texas Southwestern Medical School

William L Meyerhoff, MD, PhD is a member of the following medical societies: American Academy of Facial Plastic and Reconstructive Surgery, American Academy of Otolaryngology-Head and Neck Surgery, American Auditory Society, American Bronchoesophagological Association, American College of Surgeons, American Laryngological Rhinological and Otological Society, American Medical Association, American Otological Society, Association for Research in Otolaryngology, Southern Medical Association, and Texas Medical Association

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References


