A perilymphatic fistula is defined as a leak of perilymph, the fluid contained within the bony capsule of the inner ear, into surrounding structures, either the mastoid air spaces or the middle ear. While the labyrinthine capsule consists of extremely dense bone, abnormal openings between the middle ear structures and the fluids of the inner ear can be created by congenital or acquired defects. The flow of fluid stimulates vestibular receptors and produces episodes of vertigo, often associated with a sensorineural fluctuation in hearing. Hearing loss may progress and result in total sensorineural loss and the vertigo may be persistent. Symptomatology, however, is highly varied and difficult at times to correlate with objective auditory and vestibular tests. One of the most common complications of cholesteatoma involving the middle ear and mastoid is erosion of the horizontal semicircular canal, creating a direct contact between the keratoma, its associated infection and the perilymph of the labyrinth. When the cholesteatoma mass becomes large, the patient may induce nystagmus with finger pressure in the external auditory canal, a sure sign of fistulization.

Mastoid surgery for chronic otitis media and cholesteatoma performed in the pre-microsurgery era as a life-saving measure occasionally resulted in surgical creation of a fistula by the surgical drill or inadvertent dislodging of the stapes. The resultant opening between the middle and inner ear sometimes results in a non-hearing ear, postoperatively.

"Symptomatology, however, is highly varied and difficult at times to correlate with objective auditory and vestibular tests."

By way of historical interest, the surgical microscope was first used by a Swedish ear surgeon in 1922 for creating a surgical fistula from the mastoid to the inner ear for hearing restoration in otosclerosis. The progression of advances in optics from monocular to binocular scopes changed the entire field of otologic surgery and has made inadvertent fistulization a rare surgical complication.

In 1955, surgical attention for hearing improvement in otosclerosis was shifted from the bypass fenestration operation to stapes removal. In the next 15 years, numerous reports occurred in the literature regarding oval window fistulae occurring after stapes surgery. Most of these related to the types of prosthesis and materials used to seal the oval window niche. With the greater adoption of connective tissue seals, the incidence of fistulae dropped markedly. However, recognition of the complex of symptoms typical of perilymphatic
fistulae became recognizable under other circumstances. These were predominately cases of head trauma, Barotrauma, or physical exertion producing elevation of intracranial pressure. Abnormal persistent communications between the inner ear and middle ear became suspect in patients with head and neck injuries, excessive coughing and sneezing, changes in barometric pressure (either in aircraft or scuba diving), exposure to high sound pressure levels of noise, and congenital auricular deformities.

Pathophysiology Of Fistulae: Simmons, in 1968, reviewed 15 patients who suffered sudden unilateral hearing loss in whom he felt increased intracranial pressure produced intracochlear membrane damage. He cited a case of increased intracranial pressure associated with alcohol intake and rupture occurring with yelling at a football game. Temporal bone studies in 1981 by Gussen confirmed the existence of intracochlear membrane rupture and sudden hearing loss. Numerous other reports have indicated that not only may intracochlear membrane ruptures occur, but breaks through the oval and round windows are more common. Goodhill described both explosive and implosive pressures producing fistulae. Pressures reached the ear in the explosive route by way of the cochlear aqueduct and the internal auditory canal. Physical exertion or coughing and sneezing may produce fistulae by this method.

Possible extreme Valsalva maneuvers or other changes in barometric pressure may produce inward pressure on the suspensory ligaments of the stapes in the oval window or on the round window membrane and a consequent fistula by the implosive method.

Anatomic Variables: The normal inflow tract for the entrance of fluids into the cochlea extending from the subarachnoid space to the scala tympani is called the cochlear aqueduct. In congenitally malformed ears, this may be shorter and wider than normal and result in high intracochlear perilymphatic pressure. Grundfast and Bluestone, reporting on perilymphatic fistula in children, also have noted a higher incidence of spontaneous fistulae occurring in those with congenitally deformed ears. There may be associated conductive hearing loss due to congenital fixation of the stapes footplate. For this reason, this group of individuals with either conductive or sensorineural hearing loss is carefully followed for development of any symptoms of fistula. Surgical closure of the fistula is felt to be helpful in preventing sudden hearing losses in these individuals. Very early on in the history of stapedectomy, it was learned that this procedure in such ears would often result in a “perilymph gusher” in which perilymph literally erupted from the inner ear upon removal of the stapes. Quite frequently, these ears ended up as total sensorineural losses following surgery.

The outflow tract from the inner ear is the vestibular aqueduct which contains the endolymphatic duct and sac. Changes in patency of this system are felt to be influential in causation of some types of fistulae.

The fistula ante-fenestrum, a defect in the bony labyrinthine capsule filled with connective tissue, is felt to be a site of predi-

“There are few otologic surgeons who feel that perilymphatic fistulae may be due to minor cases of head trauma . . .”

lection for spontaneous fistulae. A challenging study was recently performed by Kohut, RI, of the University of Chicago, of temporal bone collection in which microscopic temporal bone findings were used to predict a history of dizziness likely to be of perilymphatic fistula origin in the clinical records of those whose temporal bones were studied. The percent of correlation was high, offering supportive histopathologic evidence of perilymphatic fistulae.

Finally, anatomic variations in minor as well as major congenital abnormalities may be associated with the presence of either increased fluid pressure within the inner ear from a more widely patent cochlear aqueduct or patency of the cribiform area of the internal auditory canal. This provides invasive opportunities for bacterial meningitis, secondary to otitis media. Cases of otorrhea, that is, spontaneous cerebrospinal fluid leaks, are also more common in congenitally deformed ears. Surgical correction of these variant
channels is often critical in prevention of recurring otogenic bouts of meningitis.

The Fistula Problem In Trauma: In spite of extensive literature discussing the presence of perilymphatic fistula in congenital ears, post-stapedectomy cases, mastoid and neurotologic procedures, and severe cases of Barotrauma or head trauma, the presence or absence of fistulae in the instance of more minor degrees of trauma remains controversial. There are a few otologic surgeons who feel that perilymphatic fistulae may be due to minor cases of head trauma, often occurring years before the onset of vertiginous symptoms. Surgical closure of perilymphatic fistulae is recommended quite frequently by these individuals. There are those at the opposite end of the scale who feel that the presence of inner to middle ear fistulas is rare in their practice. In a questionnaire to active surgeons in the American Otologic and Neurotologic Societies, Gordon B. Hughes, M.D., et al, recently reported the results of the survey. One hundred sixty seven surgeons performed an average of 197 otologic procedures per year (median 161; range 10 1500). Middle ear explorations were performed on an average of 4.6 fistula explorations per year per surgeon with a median of 3 and a range of 0 - 50. Most surgeons agreed that spontaneous healing of fistulas does occur and this has been demonstrated in animal studies. Many of these are presumed to be minor fistulae, such as that presumed to occur with severe, airtight nose blowing. Ninety two percent of the surgeons agreed that in patients with sudden or fluctuating hearing loss who either had a stapedectomy or penetrating trauma should be explored without delay. With its protean manifestations, it becomes clear that diagnostic studies are critical in the approach to the patient with possible fistula.

Diagnosis: The dramatic case of recurring meningitis occurring in otherwise healthy individuals is rare compared to the vast numbers of individuals who have a sensorineural or conductive hearing loss that fluctuates and may or may not be associated with a sense of pressure in the ear, tinnitus and/or vertigo. In classic severe cases, all three signs occur together and certainly mimic those of a Meniere’s disorder. While fluctuating hearing loss is probably the most common auditory symptom, others have reported sudden losses to a severe degree or mild or even vague hearing changes. Conductive losses are those most often seen with post-stapedectomy patients. The number of these, as indicated above, has greatly diminished following the use of a live tissue seal of the oval window at the initial stapedectomy.

Vestibular symptoms may also vary a great deal from true vertigo to motion intolerance. Positional nystagmus has also been reported. The incidence of positional nystagmus however, is controversial. Objective testing for the presence of a fistula through vestibular means varies from clinical testing to various laboratory procedures. A positive fistula test is one in which air pressure against the eardrum produces nystagmus (Hennebert’s Sign). If nystagmus is not visible but the patient experiences a sense of dizziness or motion, this is termed Hennebert’s Symptom. According to Hennebert’s original description, the presence of a positive fistula sign was supposed to be indicative of syphilis involving the temporal bone. Tullio’s Phenomenon, loud sound producing dizziness, also maybe symptomatic of a fistula.

Semicircular canals
Anterior
Posterior
Lateral
Ampullae of
semicircular canals
Oval window
Vestibule
Cochlea
Malleus
Incus
Stapes
Round window
Tympanic membrane

Details of the middle ear and bony labyrinth of the internal ear.

continued
Use of glycerol intravenously to dehydrate the inner ear has been considered helpful by some when combined with electrochleography with the demonstration of an increased ratio of summating potential to action potential. Unfortunately, an active Meniere’s disorder will produce similar findings.

If a presumptive diagnosis is made, the consideration for surgery should usually be tempered with a period of observation.

Electronystagmography is helpful in detecting the presence of nystagmus in response to pressure changes in the external auditory canal when such changes may not be visible. However, subjective sensations of dizziness experienced by the patient in response to this test may not be noted because of broad somatosensory input relative to being in a prone position on the table. When electronystagmography is not used, the patient opens his eyes for visualization and visual suppression may occur, eliminating potential observation of nystagmus. A new testing method, dynamic posturography, eliminates these pitfalls.

Dynamic Posturography: Dynamic posturography is used to evaluate both motor and sensory aspects of balance. The classic Romberg test does not enable identification of sensory versus motor deficits in evaluation of a dizzy patient. Motor testing in this phase of posturography enables assessment of the automatic reflex balance responses which are not tested in ENG and standard caloric testing. In addition, sensory input from visual and somatosensory receptors may be altered by sway referencing, the visual surroundings and the platform on which the patient stands. In sway referencing, visual input is altered by the simultaneous moving of the visual surroundings to coincide with the patient’s body sway. Similarly, coincident motion of the platform via computer control alters proprioceptive input from the soles of the feet and the ankle joints. These tests permit evaluation of visual, vestibular and somatosensory inputs into the vestibular system and aid in the evaluation of the dizzy patient.

In testing for the presence of perilymphatic fistula, the inner ear is isolated by removing visual input through eye closure and by altering somatosensory input by platform sway referencing. The patient then depends largely on inner ear mechanisms for the main avenue of balance. Alternating pressures are placed in the external auditory canal, varying from +300 millimeters of water to -300 millimeters of water and the patient’s change in body sway is recorded. If a timely increase in sway is noted or if the patient falls (caught by a support harness), a fistula may be present. Posturography appears to offer significant advantages in the diagnosis of perilymph fistulas, but much more correlated studies with surgical results must be obtained to determine the ultimate predictive value of this test.

Currently, the overall clinical judgement of the surgeon involved seems to be the most important factor in making a final decision as to surgical exploration of the middle ear. Head trauma or significant Barotrauma are important points in consideration for surgery, especially in incidences of sudden hearing loss.

In the overall medical management of possible fistulae, if a presumptive diagnosis is made, the consideration for surgery should usually be tempered with a period of observation. While longstanding bedrest is inimical to health, a limited program of bedrest and avoidance of activities which produce increased intracranial pressure is important. Minor types of changes in intracranial pressure, such as sneezing or nose blowing which may produce microfistulas, should always be observed before considering surgery. The presence of temporal bone abnormalities increases the possibility of a fistula. Children with such problems should be closely monitored for possible fistula formation. Stability versus progression of symptoms may be influential in deciding whether or not surgery is indicated. Progression of hearing loss or vertigo would be favored by most surgeons for surgical closure.

A positive fistula test is one in which air pressure against the ear drum produces nystagmus (Hennebert’s Sign).
Resolving hearing loss or dizziness in patients with potential fistulae usually results in continued medical observation. The risk of meningitis stemming from the ear with a fistula is minimal in those absent any anatomic variation. There should be continued observation in those in whom threatening hearing loss is not a problem but dizziness persists. This type of individualized conservative approach to each patient seems most appropriate when all factors are considered.

Surgical Management:
Surgical approach to the ear for closure of a fistula involves middle ear exploration through a tympanotomy approach. This is best done under local anesthesia without extrusion of the anesthetic materials into the middle ear. Each normal communication between the inner and middle ear (oval and round windows), is carefully observed for evidence of clear excess fluid discharge. Care must be taken to ensure adequate visualization by curettage of the bony posterior canal wall overhang which may obscure the oval window, including visualization around the chordatypani nerve which carries taste to the anterior 2/3 of the tongue. Round window fistulas may require removal of thin bony structures obscuring the round window membrane. Use of steep Trendelenberg position, as well as jugular vein compression, may aid in identification of perilymph leaks.

Different types of connective tissue have been used to close abnormal communications between the inner and middle ear. Pericondrium from the tracheal cartilage seems to be preferred by many.

The goals of surgery are to control vestibular symptoms and to promote improvement in hearing loss. The results are generally good. However, there is considerable variation in reported long-term results, with revision surgery necessary at times in variable numbers of cases.

CONCLUSIONS:
1. Diagnosis of perilymphatic fistula is still a clinical diagnosis, based on the total clinical picture. Laboratory testing, particularly dynamic posturography with pressure tests, appears to be promising as an aid to diagnosis. History and physical examination still form the cornerstone of diagnosis, however.

2. Post-stapedectomy fistulae, now increasingly rare, and penetrating trauma fistulae nearly always require surgical intervention.

3. If hearing is stable and dizziness is improving, surgery is usually unnecessary. This is also true in cases of sudden hearing loss without a history of trauma.

4. Temporal bone anomalies or progressive hearing loss in childhood and cases of severe Barotrauma frequently requires surgery, although long-term results are not yet totally understood.

5. The average inner ear exploration rate of 4.6 for 197 active otologic surgeons seems acceptable.

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