Chapter 171: Perilymphatic Fistula

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The term *perilymphatic fistula (PLF)* defines a communication of the perilymphatic fluid space, usually with the middle ear cavity. Because a natural communication between the perilymphatic space and the middle ear does not occur, such communication occurs as a result of an injury that disrupts the bony or soft tissue barriers between perilymph and middle ear space (Fee, 1968). A common example of PLF in clinical experience occurs as a result of subluxation/fracture of the stapes footplate secondary to direct trauma of the ossicular chain (Emmett and Shea, 1980; Silverstein, 1973). The usual clinical course after such injury is one of vestibular (ataxia) and auditory (sensorineural hearing loss) symptoms, which may progress to a profound loss of hearing unless there is early and effective repair of the communication (Arragg and Paparella, 1964). The current controversy surrounding PLF concerns patients with auditory and vestibular symptoms occurring spontaneously or as a result of indirect trauma, such as sudden pressure changes in the middle ear and/or intracranial space. It should be pointed out that patients with serous middle ear effusion and/or meningitis as a result of a congenital aplasia of the labyrinth are not included in this discussion of PLF because they concern cerebrospinal fluid leaks rather than perilymphatic fluid leaks.

This chapter offers a reasonable approach to the controversy of PLF and its role in patients with auditory and/or vestibular symptoms. The approach is based on basic and clinical observations and principles.

Experimental Observations

Several experimental observations bear on the subject. First, the round window membrane can be displaced and even ruptured in the experimental animal (cat) by increasing the cerebrospinal fluid pressure after experimental cannulation of the subarachnoid space (Harker et al, 1974). The pathway for this increase in perilymphatic fluid pressure occurs through the cochlear aqueduct, which is short and wide in the cat as compared to humans where it is long and narrow. The human cochlear aqueduct is usually filled with periotic duct tissue and does not readily permit the flow of cerebrospinal fluid into the perilymphatic fluid compartment (scala tympani). However, in cases of subarachnoid hemorrhage, temporal bone studies have demonstrated that increased cerebrospinal fluid pressure may permit the flow of cerebrospinal fluid and blood contents into the perilymphatic fluid spaces if the cochlear aqueduct is wide (Holden and Schuknecht, 1968). In temporal bones where the cochlear aqueduct is narrow, blood is not found in the perilymph, even with high subarachnoid pressure. In humans, therefore, subarachnoid pressure may or may not be transmitted to the scala tympani, depending on the anatomy of the cochlear aqueduct. Pertinent to these observations is the fact that in laboratory animals such as the cat, guinea pig, or chinchilla, the round window membrane is very thin (Richardson et al, 1971) and susceptible to ruptures with markedly increased perilymphatic fluid pressure. In primates, the round window membrane is a much thicker structure (Kawabata and Paparella, 1971) and resistant to such increased pressure.

The second experimental observation is that the round window membrane heals spontaneously in the experimental animal (Simmons et al, 1962). Presumably spontaneous

repair occurs in other connective tissue structures, such as the annular ligament, which surround the perilymphatic space. Healing of even large round window membrane tears takes place within 7 to 10 days in the laboratory animal.

Finally when round window membrane rupture has been produced experimentally, no significant permanent change in cochlear potentials has been demonstrated (Simmons et al, 1962; Weisskopf et al, 1978). These potentials are preserved as the ruptured membrane heals spontaneously. If intracochlear membrane ruptures (for example, Reissner's membrane) are associated with round window membrane defects, however, then significant pathologic changes in the cochlea are manifested by decreased cochlear potentials and a permanent sensorineural hearing deficit (Oshiro et al, 1989). Presumably the reason for this effect is an increased longitudinal flow of toxic endolymph in the perilymphatic compartment produced by the round window membrane rupture along with an intracochlear membrane break. This observation has added support to the theory of double membrane breaks, which are thought to be responsible for sensorineural hearing loss after traumatic insults to the labyrinth (Simmons, 1979).

Clinical Observations

Clinical observations are also pertinent to a discussion of this topic.

First, a persistent perilymph to middle ear communication usually leads to a permanent and profound sensorineural hearing loss from serofibrinous labyrinthitis and reduced vestibular symptoms because of compensation for the vestibular deficit. This observation suggests that the fistula between the perilymphatic space and the middle ear is causally related to symptoms of serous labyrinthitis, that is, sensorineural hearing loss and disequilibrium.

Second, clinical experience with patients suffering direct trauma to the ossicular chain indicates that the injury is usually in the oval window region rather than the round window membrane (Emmett and Shea, 1980; Silverstein, 1973). Indirect trauma such as from head injury or from sudden middle ear pressure changes may be transmitted more effectively to the oval window than the round window membrane (that is, the force concentrated at the oval window footplate is greater than that transmitted to the round window membrane). Moreover, the relatively thick round window membrane is more capable of withstanding sudden displacement than the annular ligament. This suggests that the likelihood of fistulization is greater at the oval window than at the round window level. The higher incidence of PLF reported at the oval window supports these observations (Singleton et al, 1978).

Third, although clinical signs associated with PLF vary, both sensorineural hearing loss and vestibular symptoms usually occur in varying degrees. Therefore, selective auditory or vestibular symptoms are not characteristic of PLF. Clinical test findings are variable and not pathognomonic of PLF (Singleton et al, 1978). Even a positive fistula test (nystagmus in response to pneumatic otoscopy) may be associated with negative exploration for PLF. The vestibular response as measured by caloric stimulation may be normal or decreased (Love and Waguespack, 1981; Singleton et al, 1978). This variable response results because vestibular sensitivity associated with a serous labyrinthitis may be normal or reduced.

Diagnostic Evaluation

Clinical history is probably the most important diagnostic information of PLF (Hughes et al, 1990). A history of trauma, whether it be *direct* to the tympanic membrane and ossicular chain or *indirect* as a result of head injury or barotrauma, has both experimental and clinical supporting evidence of injury to the window regions. A history of sudden auditory and vestibular symptoms after such trauma is strongly suggestive of a traumatic injury to the labyrinth. Typical auditory symptoms are a threshold elevation with poor word discrimination scores, whereas vestibular symptoms may include both ataxia as well as positional vertigo and nystagmus (Healy et al, 1976; Simmons, 1982; Singleton et al, 1978). The presence of both auditory and vestibular symptoms suggests a serous labyrinthitis as a result of either PLF or intracochlear membrane breaks. Supporting signs, such as a positive fistula response, fluctuating sensorineural hearing loss, and either spontaneous nystagmus or nystagmus induced by positional testing, are indicators of labyrinth injury.

With a history of trauma followed by auditory and vestibular symptoms in a previously normally functioning ear, the diagnosis of PLF through the oval or round windows should be entertained. Because considerable clinical and experimental evidence predicts spontaneous healing of defects in the round window membrane or annular ligament, a period of bed rest, with ancillary measures such as elevation of the head and avoidance of physical exertion, should be administered for a period of 10 to 14 days. If this conservative program fails to bring improvement in auditory or vestibular symptoms, then urgent exploration of the middle ear is recommended.

Management

Surgical exploration of the middle ear for PLF should be performed under local anesthesia, not only to reduce the amount of bleeding and fluid accumulation in the middle ear, but also to allow a patient's response with Valsalva maneuver to enhance an area of perilymph leakage. Complete control of bleeding is necessary before accurate examination of the middle ear space can be performed. Adequate curettage of the posterior superior canal wall is usually required to fully visualize the oval and round window regions. It is preferable to expose the oval window and the round window niches without manipulating any mucosal folds. The oval window niche is carefully examined first with a No. 24 aspirator tip to determine whether fluid has accumulated. If no clear fluid has accumulated, it is advisable to repeatedly (two to three times) depress the ossicular chain and/or to have the patient perform a Valsalva maneuver. This maneuver may help to identify the presence of a small PLF. If no fluid has accumulated, then the round window niche is examined in a similar fashion, without instrumentation, and after a Valsalva maneuver and/or ossicular manipulation. If no fluid has accumulated in either the oval or round window niches, no surgical repair with soft tissue is carried out. Some authors have recommended the placement of soft tissue in the windows believing that a small unrecognized fistula may be present. If such repair is performed without visible evidence of PLF, the treatment cannot be assessed objectively.

If fluid has been detected in either the oval or round window niches, preparation of the area in question should be carried out and sealed with an adipose tissue graft. This preparation includes the elevation of mucosa around the area of leakage with exposure of bone and with the placement of small tissue (fat) grafts, approximately 2 to 3 mm in diameter

to adequately obliterate the defect. Gelfoam is used to stabilize the placement of the adipose tissue grafts. If fluid is detected in the round window niche, removal of the promontory overhand enhances examination of the round window membrane. Elevation of mucosal folds in the niche and the round window membrane should precede the placement of soft tissue grafts. Postoperatively, the patient's activity should be restricted for at least 1 week to 10 days to permit the fibrous tissue repair of the defect. Recovery of hearing, if forthcoming, should be evident in approximately 4 to 6 weeks; therefore, evaluation of auditory function should be performed no sooner than 6 to 8 weeks postoperatively.

In clinical practice, the incidence of PLF secondary to indirect trauma is very low. In a study of 167 surgeons (Hughes et al, 1990) most (60%) performed three or fewer explorations for PLF per year, and the average number of PLF explorations for the total group was less than five per year. Because the positive identification of PLF at surgery is probably lower than the number of explorations, the actual number of PLFs is very small. I have performed twelve middle ear explorations for PLF over a 25-year period. Only four definite PLFs in the oval window were identified and repaired in this group; no PLFs were identified in the round window region. Hearing recovery and improved vestibular symptoms were achieved in these four patients.

Summary

One can say with confidence that PLFs do occur, but the incidence is extremely low. The recommendations to explore surgically for PLF should be based on a history of trauma (direct or indirect), auditory and vestibular symptoms and signs, and failure to improve over a period (10 to 14 days) of conservative management. Recognition and repair of PLF at surgery should be based on careful objective examination of the oval and round window regions.