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Clinical Forum

A Critical Appraisal of Spontaneous Perilymphatic Fistulas of the Inner Ear

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Objective: This article provides an overview of relevant data supporting and refuting the existence of spontaneous perilymph fistula, as well as critically reviewing the literature pertaining to their evaluation and management.
Data Sources: Sources used were relevant English language clinical and basic science publications.
Study Selection: A Medline search dating back to 1966 for articles concerning perilymphatic fistula, including both human and animal data, was performed. Articles were included if they contained relevant data or were significant reviews of the subject. A traditional bibliography search was then completed to acquire articles missed by the computerized search, including works published before 1966.
Data Extraction: The data from each publication were critically reviewed. Emphasis on understanding the clinical features of surgically created perilymph fistulas was used to more objectively assess the data regarding spontaneous perilymph fistulas.
Data Synthesis: The data were not amenable to formal meta-analysis or valid data summarization; however, when possible trends and contrasting data were emphasized.
Conclusions: Spontaneous perilymph fistulas are very rare occurrences and the majority are likely incited by a pressure-altering event. Current methodologies do not provide sufficient specificity and sensitivity to accurately diagnose perilymph fistulas. The results of endoscopic studies of the middle ear in the evaluation of perilymphatic fistula suggest a low incidence compared with the large number of fistulas reported in the literature. A high index of suspicion must be maintained, and appropriate preoperative counseling should reflect the current controversies. Questions must continue to be asked and further research pursued to help distinguish reality from myth.
Key Words: Perilymph fistula—Histopathology—Diagnosis—Auditory—Vestibular.
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A perilymph fistula is an abnormal connection between the fluid-filled inner ear and the gas-filled middle ear and mastoid cavity. Although this clinical entity was proposed more than a century ago, the topic of spontaneous perilymph fistula continues to stimulate great controversy among otologists. In this forum, we will explore the evidence supporting and refuting the existence of spontaneous perilymph fistula and critically review the literature pertaining to their evaluation and management.

HISTORY

Prosper Menière initiated what may be the first modern clinical controversy regarding labyrinthine pathology (Fig. 1). In the year before his death, Menière presented a treatise to the Imperial Academy of Medicine on a specific constellation of symptoms involving vertigo, tinnitus, and hearing loss (1). His autopsy findings of isolated labyrinthine pathology refuted the prevailing concept of "apoplectiform cerebral congestion" and suggested a relationship between the inner ear and vestibulocochlear dysfunction. Controversy ensued, sparked by Trousseau, Professor-in-Chief of the Hotel Dieu in Paris and proponent of the generalized and widely encompassing diagnosis of cerebral congestion. Menière's rebuttals, published the same year (1861), provided several case reports detail-

FIG. 1. Prosper Menière (1799–1862). Controversy exists regarding the placement of the accent in his last name. Old French records show a single stress, but current custom is to place an accent over both syllables.

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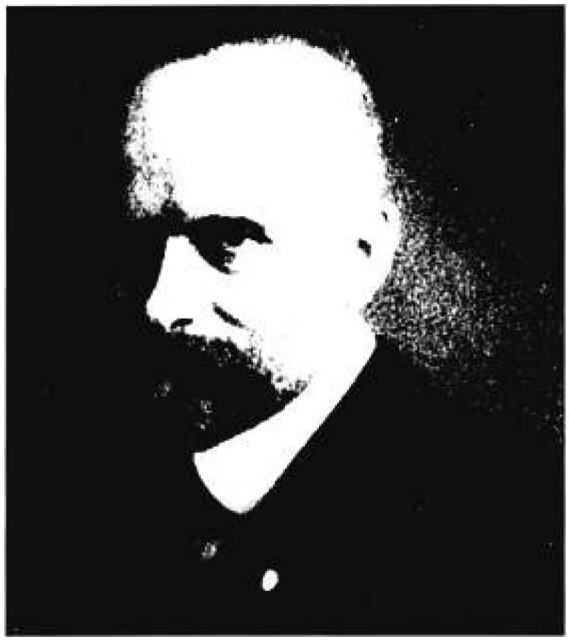

FIG. 2. Robert Bárány (1876–1936). Bárány was awarded the Nobel Prize for Medicine and Physiology in 1915 (one of two otolaryngologists ever to receive this honor). At the time, he was a Russian prisoner of war, and he was exchanged for a Russian officer by the Red Cross so he could deliver his Nobel address. Later, he was a Professor of Otolaryngology at Uppsala University in Sweden.

ing the specific time course and symptoms of the disease bearing his eponym in an effort to distinguish this entity from the broader diagnosis. It is ironic that current supporters of the diagnosis of spontaneous perilymph fistula are similarly challenged by their detractors to provide specific criteria to distinguish it from the constellation of symptoms comprising Menière's disease.

The late nineteenth century and early twentieth century saw several reports confirming the association between the vestibular labyrinth and disorders of balance. Gellè, in 1883, was able to associate vertigo of the type found in Menière's disease with mobility of the stapes and pathology of the oval and round windows (2). Barnick, in 1897, reported vestibulocochlear symptomology after perilymph leak from the round window after head trauma (3). Hennebert, in 1905, demonstrated oculo-vestibular disturbance with pressure changes in the external canal of otherwise normal appearing ears (4). These findings led many to propose the existence of labyrinthine capsule fistulas allowing communication between the middle ear and inner ear fluid (5). However, surgical explorations, albeit without the aid of an operating microscope, failed to demonstrate such fistulas. Further, histologic examination of the temporal bone found no evidence of otic capsule pathology (5). Such findings led to disenchantment with the concept of inner ear fistulas and the conclusion by Bárány that Hennebert's sign reflected increased stapes footplate mobility (Fig. 2) (6).

The diagnosis of inner ear disease remained largely unchanged during the early 1900s. Vertigo, and some cases of hearing loss, were often ascribed to Meniere's disease. Variants of Meniere's disease were also described to explain additional or absent symptoms (7,8). Also, abundant infectious processes (*e.g.*, mumps, syphilis, influenza, measles, scarlet fever) were responsible for and able to explain many additional cases of hearing loss (9). Starting in the 1920s, and particularly in the 1940s and 1950s, case reports appeared in the literature describing rapidly progressive and sudden hearing losses with unclear etiologies. Many of these cases were associated with vestibular dysfunction but did not adhere to the patterns of presentation outlined by Menière (10).

In 1949, Simpson reviewed 100 cases of hearing loss with normal otoscopic exams (9). Although he defined approximately 20 etiologic categories of hearing loss, he concluded that 71% of these cases reflected vascular compromise. He included instances of apparent noise-induced hearing loss, reasoning that the explosions caused vasoconstriction of the cochlear end-arteries. He notes, however, that the vascular theory "is based on clinical grounds and theoretical speculation" with "no direct experimental evidence to support it."

Others also accepted vascular compromise as a cause of vestibulocochlear dysfunction. Hallberg, in 1956, attributed 50% of 178 cases to vascular accidents (11). He noted that these events cause sudden, unilateral deafness and tinnitus followed by vertigo of variable duration. Hallberg found it difficult to explain, however, that more than 30% of his patients were between 20 and 39 years old, younger than the normal age for onset of intimal vascular disease. Bolognesi reported five patients with similar presentation and age distribution (12). Three of his patients, interestingly, showed improvement after anticoagulation.

FIG. 3. Samuel Rosen (1894–1981). Rosen reintroduced the stapes mobilization procedure, which led to the surgical creation of a perilymphatic fistula during stapedectomy.

FIG. 4. Victor Goodhill (1911–1994). Goodhill performed stapes surgery from the front of the patient. In this manner, the oval window was in a nondependent position. He contributed the concepts of implosive and explosive mechanics of perilymphatic fistula formation.

Not all physicians were willing to accept vascular insufficiency as an explanation for sudden vestibulocochlear compromise. Lindsay and Zuidema reported 12 cases of unexplained sudden deafness, many of these associated with vertigo and imbalance (13). They acknowledged the common assumption that these may be vascular in origin, but refused to accept that as a viable explanation. Similarly, Brown, in 1957, reviewed 34 patient cases of rapid onset deafness, the majority associated with vestibular symptoms and tinnitus (14). He distinguished these from Meniere's disease by history and also felt the etiology to be unknown.

At about the same time, Rosen (Fig. 3) reintroduced stapes mobilization as a treatment for otosclerosis (15). As oval window surgeries became increasingly prevalent, so did reports of postoperative complications. Bergstrîm and Ivstam reported two cases of postoperative vertigo—one of delayed onset and another of prolonged duration (16). They attributed the former to coincidental vascular accident, and the latter remained unexplained. They also noted a personal communication from Rosen regarding postsurgical episodes of sudden deafness.

In 1963, Steffen and colleagues reported on the findings at exploration of patients exhibiting poststapedectomy hearing loss, tinnitus, and vertigo (17). The onset was weeks to years after surgery. Approximately half of the patients had gross evidence of perilymph egress from the oval window. It was believed that the prosthesis thinned the oval window, permitting fistula formation and escape of perilymph. The same year reports of poststapedectomy meningitis were also published (18,19). Meningitis occurred 2 months, 18 months, and 2 years after initial surgery. One patient died, and temporal bone histopathology showed a labyrinthine fistula in the stapes footplate (18).

By 1967, the term perilymph fistula was widely used, and it was a recognized complication of stapes surgery. All reports agreed that diagnosis was difficult and symptomatology was often consistent with that found in Meniere's disease. For example, Hemenway and coworkers noted tinnitus, fluctuating hearing loss, fullness, and vertigo (20). Patients less frequently described "popping or gurgling" noises and positional or motion-associated dizziness. They indicated that many patients presenting years after surgery were treated for Meniere's disease. Goodhill found similar presentations, adding that the onset of symptoms sometimes appeared spontaneously (Fig. 4) (21). Harrison and colleagues observed postsurgical fistulas presenting "so similar to...labyrinthine hydrops that it makes the differential diagnosis very difficult, if not impossible (22)." House found hearing loss and/or vertigo to be present in all cases, but in varying degrees (23,24). He felt that the symptomatology was often identical to that in Meniere's disease and even diagnosed Meniere's disease in a patient showing no improvement after fistula repair. Further, he noted that no test other than exploration was able to accurately diagnose a perilymph fistula.

Before 1968, the occurrence of perilymph fistulas exclusive of poststapes surgery had not been reported. Fee then reported three patients with known or suspected head trauma who presented with vertigo, fluctuating hearing loss, and tinnitus (25). Tympanotomy was performed, and all three were found to have leak of perilymph from around the oval

FIG. 5. F. Blair Simmons (1930–1998). Simmons was an otologic pioneer who, among his basic and clinical contributions, made important observations regarding the clinical manifestations of perilymphatic fistula.

window. The leaks were patched, and all patients experienced significant improvement in their symptoms. Like House, Fee concluded that diagnosis relied upon surgical exploration. Two years later, Stroud and Calcaterra reported the first cases of spontaneous perilymph fistulas (26). They postulated that an increased intracranial pressure was the etiologic event producing the fistula. They also noted the difficulty of diagnosis and the similarity between the clinical presentations and Meniere's disease. Age of onset and associated physical activity prompted exploration, at which perilymph fistula was confirmed. In neither of these reports was the previously popular concept of vascular compromise considered.

In the early 1970s, Goodhill proposed his theory on implosive and explosive forces leading to membranous rupture and the formation of perilymph fistulas (26). This provided the matrix for multiple pathophysiologic mechanisms that would explain the development of these lesions. As was seen in the 1950s with diagnoses of vascular accidents, awareness of fistulas led to increased identification, and many reports appeared through the 1970s and 1980s. Case reports of sudden deafness, isolated vestibular symptoms, and vestibulocochlear dysfunction in children were now explained by perilymph fistula (28–31). By the late 1980s and early 1990s, enough cases were evaluated that large series of explorations were reported (32–35).

Although early reports often demonstrated confirmation of fistulas at exploration, many of the later series had low rates of positive exploration. Further, repair of identifiable fistulas often had variable rates of success with many surgeons "repairing" negative explorations as well. Shelton and Simmons (Fig. 5) reported a 51% positive exploration rate (33). Major symptoms resolved in 64% of these patients and in 44% of those with negative explorations. Rizer and House had only a 41% positive exploration rate, with 68% of these patients improving (34). Of their patients with negative explorations, 29% noted improvement. Such reports led to critical evaluation of diagnostic measures to evaluate for perilymph fistula and of the surgeries performed (36). At the extreme, several otologists have contested the existence of spontaneous fistulas and the motives of those performing repairs (37–39).

We are now at a stage of rational evaluation where the initial excitement of a new diagnostic entity and the backlash against such fervor has calmed. Focus is now on the anatomic bases of spontaneous perilymph fistulas, understanding associated symptoms, improving both preoperative and intraoperative diagnostic testing, and increasing outcome success rates.

ANATOMIC BASES FOR PERILYMPH FISTULAS

At the turn of the century, acceptance of perilymph fistulas lost favor after direct observation and histologic evaluation failed to confirm fistulas in the labyrinthine capsule (40). Occasional, and usually informal, reports appeared around the 1920s of natural dehiscences in the otic capsule. For example, decades after the fact, Holmgren recalled having seen a perilymph leak from the fissula ante fenestra (41). Crowe, in 1928, presented a histopathologic study of temporal bones from patients having died of acute meningitis (42). He described the fissula ante fenestrum as a natural potential portal between middle and inner ears. Dench, in the presentation's accompanying transcript, recalled observing dehiscences over the promontory near the oval and round windows.

In the 1930s, formal studies of the fissula ante fenestrum and otic capsule microfissures appeared (43–47). Microfissures were consistently observed in several regions of the temporal bone, particularly between the round window niche and posterior canal ampulla. Other reports suggested that many of these fissures were artifact, and further investigation soon waned (48–50). Studies on the fissula ante fenestrum were likewise contradictory with little agreement as to the histologic composition of the region.

Early histologic examination of the fissula ante fenestra described the region as being cartilaginous, of dense fibrous tissue, or of loose fibrous tissue (51). In an attempt to clarify the issue, Kohut and colleagues examined the temporal bones of five patients with purported perilymph leaks (51,52). They found a loose fibrous composition in the fissula ante fenestra of all bones. The cores extended from inner to middle ear and showed no evidence of dense impermeable material. As these patients had surgically confirmed leaks, Kohut and colleagues concluded that this histologic pattern was consistent with potential, and likely, permeability to inner ear fluid.

Other investigators focused on the early reports of microfissures and began to look for natural weaknesses in the otic capsule that may be prone to "rupture." Okano and coworkers investigated the development of the round window niche with attention to the microfissures previously described in the 1930s (53). They found a fissure in all their temporal bone specimens of patients 6 years and older. Further, they described a distinct area of ossification correlating anatomically with the fissure connecting the round window niche and the ampulla of the posterior canal. In a later investigation, the same fissure was found in all temporal bones of patients older than 8 years (54). It was postulated that such a fissure may permit the leak of perilymph into the middle ear.

Harada and colleagues, in 1981, investigated a microfissure around the oval window; this was also described years earlier (54). They found an oval window microfissure in 25% of 331 temporal bones examined (n = 84). Further, the incidence of the microfissure increased with increasing age. They interpreted the frequency and similarity of their findings as being inconsistent with that of a processing artifact. Harada and colleagues felt that repetitive or continuous stress on the otic capsule may predispose a person to such fissures. In addition, they speculated on the possible relationship between such a fissure and perilymph fistulas.

Sato and associates studied microfissures by computer reconstruction of histologic sections (55). They found a round window fissure in 21 of 24 (87%) of cases and a microfissure at the oval window in 7 of 24 (29%) of those cases. The length of the fissures was found to increase

with age. They believed that this might reflect accumulation of stress on "weak points" of the otic capsule. The reconstruction of the oval window showed predominance of fissures posterosuperiorly in contrast to the clinical findings of anterior perilymph leak. They thus concluded that spontaneous leak around the oval window was more consistent with a patent fissula ante fenestrum than a patent microfissure.

Kamerer and coworkers further supported the relationship between microfissures and perilymph fistulas (56). They reported four cases of patients clinically suspected of having perilymph fistulas. Upon exploration, these patients were found to have perilymph leak from microfissures around the oval and round windows. The report presented histologic *examples* demonstrating the type of fissures likely had by these patients. Kamerer and colleagues speculated on the correlation between location of a microfissure (and thus site of resultant perilymph leak) and the predominant symptoms at presentation. There was no direct corroboration, however, between the clinical and histopathologic cases.

Kohut and colleagues examined temporal bones of normal patients and those of patients with a diagnosis of labyrinthine hydrops (40). In all patients who did not have labyrinthine hydrops, they found the fissula ante fenestrum closed by cartilage or bone. They similarly found a round window fissure sealed by dense collagen or new bone. Of the 11 patients with labyrinthine hydrops, two had a history suggestive of perilymph fistula. It was only in these two patients that a patent fissula ante fenestrum and incompletely obstructed microfissure was found.

Kohut followed the above study with a report of a case of surgically proven perilymph fistula and histopathologic confirmation (57). At exploration the patient was found to have two areas of perilymph leak around the round and oval windows. After patching the leaks, the patient's symptoms markedly improved. He died several years later of an unrelated cause and his temporal bones were examined. The fissula ante fenestrum was found to be free from bony and cartilaginous obstruction. The entire length was filled with loose connective tissue. A round window fissure was likewise free from obstruction and composed of a fibrinous core. The middle ear openings of these "pathways" were sealed with the well adherent graft. Kohut and colleagues surmised that the fibrinous cores are permeable and accounted for the leaks observed at exploration. It should be noted that the histology showed no evidence of endolymphatic hydrops to explain his vestibular dysfunction.

Kohut and coworkers went further and designed a blinded study to correlate clinical findings of perilymph fistula with histologic findings (58). One investigator reviewed medical charts and selected patient histories suspicious for having a perilymph leak. Another reviewer examined temporal bone sections and listed those patients with patent fissula ante fenestra or round window microfissures. Upon comparison they found they had a sensitivity of 59% and a specificity of 91%. That is, clinically they were "underdiagnosing" the proposed anatomic basis for perilymph fistula (only 59% selection). However, clinically they were very accurate in predicting those patients with sealed or obstructed fissures (91% selection rate).

In contrast, no relationship between the presence of histologically patent fissures and clinical presentations of sudden sensorineural hearing loss was found by El Shazly and Linthicum (59). They concluded that "microfissures...are a common histopathologic finding and have no clinical significance." Their study, however, provided no data as to the vestibular symptoms of their patients and compared patients with sudden hearing loss with patients with nonsudden loss without analyzing unaffected individuals. The Kohut group of patients, in contrast, was selected for presence of fistula based on specific audiologic and vestibular symptoms.

In addition to examining the otic capsule for "natural" weaknesses, other anatomical bases for perilymph fistulas have been proposed. Goodhill, as previously noted, devised the concept of an "explosive" etiology underlying some perilymph fistulas (27). He postulated an increased cerebrospinal fluid (CSF) pressure transmitted to the inner ear by the cochlear aqueduct or internal auditory canal. Indeed, Glasscock, in 1973, described three cases of perilymph gusher controlled only by occluding the internal auditory canal (60). Most investigations, however, have implicated the cochlear aqueduct as the principle conduit between CSF and perilymph.

Ahlèn, in 1947, using a rabbit model, provided evidence of change in cochlear aqueduct perilymph flow with respiration, increased abdominal and intracranial pressure, and changes in regional hemodynamics (61). In 1963, Kerth and Allen performed simultaneous measurements of CSF pressure and perilymph pressure in the cat (62). They found immediate and approximately equal rise in perilymph pressure with increase in CSF pressure. Further, the perilymph pressure change was prevented by obliteration of the cochlear aqueduct. Studies in other experimental animals produced a similar relationship (63). The guinea pig, cat, and rhesus monkey all showed rise in perilymph pressure with CSF pressure increase. The more rapid response in the cat was attributed to a more patent cochlear aqueduct.

Beentjes measured endolabyrinthine pressures in the cat and found that an abrupt change in CSF pressure resulted in an equal, but attenuated, change in perilymph pressure (64). He postulated that the cochlear aqueduct and the round window membrane might play a buffering role against sudden CSF pressure changes. Allen, using a biophysical model, showed that small changes in the diameter of the cochlear aqueduct would profoundly affect flow rate from the CSF to perilymph (65). Further, based upon physical parameters, he noted that small fistulas might not allow egress of perilymph unless CSF pressure was increased. Nomura measured the pressure changes in CSF associated with inner ear damage (66). He showed that infusion of fluid into the subarachnoid space to a pressure of approximately 400 mmHg could rupture the guinea pig round and oval windows.

The clinical significance of the relationship between CSF and perilymph pressure was studied in 1994 by Sakikawa and colleagues (67). They measured the CSF

pressure of patients undergoing diagnostic lumbar punctures during various activities. These included nose blowing, breath holding, and sniffing. Nose blowing and breath holding were found to result in marked increases in CSF pressure. The CSF pressure was found to increase in some patients to more than 500 mmHg. Furthermore, three patients with suspected perilymph fistula (one confirmed surgically) demonstrated an average response greater than patients without suspicion of fistula. However, there was no statistical analysis of these differences. Sakikawa and colleagues postulated that patients with perilymph fistula might have a predisposition to dramatic increase in CSF pressure with physical strain. Such increase would then be transmitted to the inner ear by the cochlear aqueduct and lead to the explosive formation of a perilymph fistula.

Weider and colleagues reported on a patient with fluctuating hearing noted on exploration to have a perilymph leak from the round and oval windows (68). The leaks were patched and the patient's hearing stabilized. Several months later, the patient's symptoms recurred and a computed tomographic (CT) scan noted an enlarged cochlear aqueduct infundibulum. Upon exploration, the duct was grossly enlarged, easily allowing passage of a 4 mm right angle pick. The cochlear aqueduct was obliterated, and postoperatively the patient's hearing fluctuation improved.

Several studies have contested the clinical significance of an abnormally wide cochlear aqueduct. Carlborg and colleagues, in 1982, demonstrated in the cat model that closure (*i.e.* narrowing) of the cochlear aqueduct was actually more significant for pressure modulation than wide patency (69). Closure of the cochlear aqueduct left the inner ear without protection from middle ear pressure changes and thus susceptible to *implosive* fistula formation. Jackler and Hwang examined CT scans of 100 temporal bones and established a normogram for cochlear aqueduct width (70). They found that although there is variation in the medial orifice, the lateral conduit is fairly consistent and relatively narrow. They noted limited ability of CT to adequately resolve the narrowest portion of the conduit and thus, at present, a lack of evidence that widened cochlear aqueducts exist.

PATIENT PRESENTATIONS

The presentation of patient's with perilymphatic fistula varies. Often, associated antecedent events such as surgery or trauma serve to increase the clinician's suspicion of a perilymph leak. Cases of supposed spontaneous perilymph fistula lack a readily apparent inciting incident and make the diagnosis more difficult. The otologic complaints seen with inner ear fistulas are generally nonspecific, and patients are often initially diagnosed with other disease processes, most notably Meniere's disease (71). Reports from large series of patients, however, have attempted to define criteria that may increase a clinician's suspicion of perilymph fistula.

Seltzer and McCabe, in 1986, reported on the Iowa experience with perilymph fistulas by analyzing the chief complaints of patients with confirmed leaks (32). Based on history, 24% of patients had no antecedent event associated with the onset of symptoms. The most common presentation was the triad of hearing loss, vertigo, and tinnitus. By comparison, it was less common to have only two of these symptoms and rare to have just one. In fact, not one of 91 patients with a surgically confirmed perilymph fistula presented solely with the complaint of tinnitus. More than 80% of patients noted some change in hearing, with the most salient clinical feature being fluctuation in speech discrimination. Approximately 80% of patients also had vestibular complaints that included vertigo, dysequilibrium, light-headedness, and motion intolerance. The most common vestibular disturbance associated with perilymph fistula consisted of dysequilibrium with occasional episodes of vertigo. Of the associated signs and symptoms, 25% experienced aural fullness and 21% noted recruitment. Duration of symptoms in this series was between 1 day and 23 years. Seltzer and McCabe noted "no clear pattern of signs and symptoms" that would mandate surgical exploration, yet remarked on the above patterns of presentation that should raise one's suspicion. A follow-up report of 115 confirmed perilymph leaks stressed again the importance of fluctuating hearing loss and dysequilibrium punctuated by vertigo (72). These studies analyzed the presentations of only those patients with positive explorations and did not refer to those patients with negative explorations.

Weider and Johnson, in 1988, noted similar presentation in New Hampshire (73). The majority of their patients had some combination of vertigo, hearing loss, and tinnitus. They remarked on several historical events that may increase suspicion of perilymph fistula. Agreeing with the Iowa study, they included prolonged dysequilibrium and aural fullness. Unlike Seltzer and McCabe, they had a very high rate of positive explorations. Further, they believed that those patients without visible leaks who improved postoperatively likely had true fistulas missed on exploration.

In 1988, Shelton and Simmons reported on the Stanford experience with perilymph fistulas (33). Overall, they confirmed a perilymph leak in 51% of the 65 patients explored. A leak was found in 15 of 33 patients (45%) with supposed spontaneous fistulas as compared with 56% of patients with antecedent events. Patient symptoms ranged from 1 day to 28 years, with a mean of approximately 4 years. Hearing loss was the chief complaint of 28% of patients, yet 54% had abnormal audiograms. No distinguishing pattern of audiogram was noted, although 50% had downsloping losses. The most common symptom was of vestibular dysfunction; this was present in 73% of patients. Although the majority had episodic symptoms, no form of balance disturbance was significantly associated with a perilymph fistula. There was no mention of the incidence of complaint of tinnitus.

Rizer and House, in 1991, presented the House Ear Clinic experience of 86 fistula explorations and 35 confirmed perilymph leaks (34). Approximately 23% of patients with a confirmed fistula had no known event associated with the onset of symptoms. Average duration of symptoms was

approximately 3 years. Of those with a fistula, the primary symptoms were hearing loss (54%) and dizziness (46%). Similar to the Iowa study, no patient noted tinnitus as the chief complaint. Of patients with a surgically confirmed fistula, 74% had a sudden onset of symptoms compared with 65% in those with a negative exploration. There was no statistically significant association between patient symptoms and the findings at surgical exploration.

Other series have reported similar findings at presentation. Black and colleagues presented the Portland experience and noted dysequilibrium as the most common presenting complaint (74). Most patients noted a precipitating event, although 20% were considered spontaneous. As in other studies, symptoms ranged from days to decades. Interestingly, they had an 88% positive exploration rate with a high incidence of bilateral leaks.

The most recent perilymph fistula "experience series" comes out of Washington, DC (75). These researchers analyzed data from 197 surgical repairs of suspected perilymph fistulas. Inciting events were noted in 65% of patients, with 35% considered of undetermined etiology. Duration of symptoms ranged from 1 day to 40 years, with a mean of 3.1 years. Vestibular complaints were present in 87% of patients, and 35% of patients reported hearing fluctuation or loss. Tinnitus was found in 25% of patients, but no report was made of its association with other symptoms. Similar to the Iowa experience, significance was given to concurrent aural fullness. Contrary to most studies, but similar to the New Hampshire report, a clinical improvement after patching was taken as confirmation of a fistula regardless of the results of direct observation at exploration. Thus, direct comparison with other series is difficult. Their data analysis failed to demonstrate a pattern of symptoms having a higher correlation with actual perilymph fistula.

It may be concluded from these studies that a wide variety of presentations may be associated with perilymph fistulas. The difficulty in establishing a diagnosis is supported by the consistent finding that patients are not explored for approximately 3 to 4 years and many go decades before surgical exploration. Many presentations mimic the pattern found in Meniere's disease; however, some findings have been established in these series that may increase the likelihood of finding a perilymph fistula upon exploration.

In summary, patterns of hearing disturbance more associated with perilymph fistula are sudden loss or rapidly worsening progression (76). Additionally, hearing symptoms are usually fluctuant and attention must be paid specifically to speech discrimination (32). Vestibular disturbances appear to be the most prevalent complaint of patients with perilymph fistulas (29,74,77). Although all forms of imbalance have been described, continuous dysequilibrium punctuated by episodic vertigo and positional vertigo are of note (32,76). Most patients will have a combination of symptoms, because isolated dysfunction is rare (32,78–80). Tinnitus appears to be nonspecific, yet one should note that isolated tinnitus was not seen in association with a perilymph fistula (32,34). Aural fullness was a complaint in approximately 20% of patients with perilymph fistulas (32,75).

DIAGNOSTIC TESTS FOR PERILYMPH FISTULAS

Many otologic pathologies can be diagnosed by history and basic physical exam; however, the nonspecific presentation of patients with idiopathic or spontaneous perilymph fistulas demands further diagnostic evaluation. Specifically, the absence of a positive fistula test or highly suspicious history may make the clinician and patient uneasy about preceding with formal middle ear exploration. Thus, some form of preoperative noninvasive testing that would raise the clinical suspicion of a perilymph leak is desirable. The predominance of complaints with respect to the vestibular and auditory systems have thus focused attention on vestibular function testing and audiologic evaluation.

As noted previously, patients with perilymph fistulas often present with complaints of hearing loss. In fact, Shelton and Simmons found abnormal audiograms in almost twice as many patients as complained of hearing changes (33). Attempts at finding specific patterns on audiograms that would indicate a perilymph leak have generally failed (32,81,82). However, fluctuation in pure tone averages and in discrimination has frequently been reported (25,29,72,76,82).

There have been reports of conductive hearing losses in perilymph fistula patients (21,83). This is presumably secondary to air entry into the labyrinth at the fistula site. Hazell and colleagues, using this premise, performed audiograms with the patient's affected ear facing up (83). They found no prognostic value, except in cases of large oval window leaks. As these patients are usually poststapedectomy, the history may actually provide stronger diagnostic criteria than the testing.

The lack of specificity and sensitivity of standard audiograms has led some to investigate the use of electrocochleography (ECOG) in diagnosing perilymph fistulas. Arenberg and coworkers found abnormal ECOGs (*i.e.*, summation potential/action potential ≥ 0.5) in 52% of patients with surgically confirmed perilymph leaks (79). In contrast, abnormal speech reception thresholds (*i.e.*, >25dB) were found in only 22% of patients. However, they were unable to distinguish perilymph fistula patients from those with Meniere's disease.

Campbell and colleagues created perilymph fistulas in guinea pigs and correlated ECOG with acute and healed states (84). They found significant change in the SP/AP ratio only when the fistulas were acute, with reversion to normal when it was sealed. It was not differentiated whether a patent but nonleaking fistula could be identified.

Aso and Gibson, in 1994, performed intraoperative ECOGs on patients without visible leaks of perilymph (85). Initially the ECOGs were normal, but upon suctioning the round window niches significant change was noted. Such change did not occur upon suctioning the margins of the oval window. Only the round windows were patched, and postoperatively the patients noted relief of vertiginous symptoms. It would thus appear that acute perilymph flow is necessary for a change in the ECOG.

The vestibular system has also been investigated as an adjunct to perilymph fistula diagnosis. For example, Wall and colleagues have developed the chinchilla as an animal model to study the diagnostic efficacy of the fistula test (86,87). In their system, a computer controls the application of pressure to the external canal while ocular motion is monitored by electrooculography. They have found 80% sensitivity for the detection of surgically created fistulas.

Clinically, most series have reported on the fistula test or the elicitation of Hennebert's sign. These terms have essentially been used synonymously and involve applying positive and negative pressure to an intact tympanic membrane with a pneumatic otoscope. The subject is then observed for the elicitation of nystagmus or questioned regarding the onset of dysequilibrium. Some authors refer to induced nystagmus as an objective sign and dysequilibrium as a subjective symptom (32). The fistula test has been performed with various modifications to increase sensitivity in detecting a perilymph leak. Kohut and Cole, for example, administer the test with the patient standing and eyes closed (76,78). Additionally, the test has frequently been administered with electronystagmography (ENG) monitoring (34,88–90). Black and colleagues perform the test on a moving posture platform to isolate what they believe is the more sensitive vestibulo-spinal system (80).

Reports of association between a positive fistula test and documented perilymph fistulas vary. Rizer and House performed a fistula test with and without ENG enhancement (34). There was no significant difference between those with confirmed fistulas and those without fistulas; 58% versus 56% (by non-ENG) and 33% versus 26% (by ENG enhancement). Healy and colleagues reviewed the literature and their own cases and noted a positive fistula test in only 25% of patients with perilymph leak (29). Seltzer and McCabe noted almost identical results, with only 24% of patients with surgically confirmed leaks demonstrating a positive fistula test (32).

In contrast, Podoshin and associates reported a positive fistula test in 77% of those confirmed with a fistula (90). Vartiainen and coworkers found 33% of those with a fistula had a positive fistula test, which was statistically significant when compared with the 8% positive response in those without a fistula (89). This study, however, found no statistical significance when performing the fistula test with ENG enhancement. Of interest is a recent report by Ostrowski and colleagues who surgically explored three patients with a positive fistula test (91). In two of three patients fistulas were noted, and in all patients the round and oval windows were patched. On postoperative testing, however, all three patients had persistent positive fistula tests. Interestingly, they had ocular torsion and not directional nystagmus, which may indicate otolith and not cupula stimulation. This may indicate an alternate means of stimulation (e.g., hypermobile footplate) and not have any relation to perilymph leak from the inner ear.

Black and colleagues, using the moving platform fistula test, had a positive result in 97% of tested ears but 70% of patients (80). Upon exploration, they confirmed a fistula in 73 ears: a 97% positive exploration rate. They calculated 97% sensitivity for their modified fistula test. Meyerhoff noted an abnormal platform fistula test in only 9 of 27 patients, yet also had a very high positive exploration rate (35).

Sporadic results of other vestibular tests have been reported with regard to the presence of perilymph fistulas. Shelton and Simmons noted 8 of 10 patients with hypoactive calorics had confirmed fistulas on exploration (33). Vartiainen and associates found no significance using calorics between patients with confirmed fistulas and those with negative explorations (89). Black and colleagues found 50% of patients had abnormal calorics compared with 30% with a positive fistula test (80). Shelton and Simmons noted 18 patients with positional nystagmus of which 10 had confirmed fistulas (33). Healy and coworkers, reviewing 47 cases of perilymph fistula, noted 17% with spontaneous nystagmus, 30% with positive calorics, and 43% with positive positional testing (29). They also noted in their own series that 9 of 19 patients had a positive Romberg sign. The results of vestibular testing appear as varied as the presentations of the patients and no single test is able to distinguish perilymph fistulas from Meniere's disease or to clearly indicate the presence of a perilymph leak.

CONFIRMATION OF PERILYMPH FISTULAS

In the absence of significant inciting events, the clinical presentation of perilymph fistula can be indistinguishable from Meniere's disease (71). As noted by House, even in patients after stapedectomy, "there is no positive method available to diagnose a fistula accurately without exploration (23)." Although direct observation would seem to be capable of providing definitive proof of a fistula, it is often difficult to see a perilymph leak or to prove that fluid is indeed of inner ear origin. For example, it has been reported that injected local anesthetic may pass into the middle ear and mimic perilymph pooling (92).

The volume of perilymph is reported to be approximately 75 μl (76), and determination of a leak would thus necessitate visualizing small amounts pooling in the middle ear. Rarely have cases been reported of a dramatic perilymph leak (60), which may actually represent significant CSF loss as well. Because of the small amount of perilymph, many authors advocate prolonged microscopic examination of suspicious regions or confirmation of the leak by at least two surgical personnel (33). Seltzer and McCabe suggest looking for a shift in the light reflex as an indication of fluid egress (32). Todd and Jackson proposed using a drop of mineral oil to trap and better resolve minute amounts of fluid (93). Others have used the Trendelenburg position, Valsalva maneuver, or internal jugular compression (90,94).

The majority of reports on perilymph fistulas have used the operative microscope to confirm perilymph leak. From these reports, a positive exploration rate can be extrapolated. The national perilymphatic fistula survey of 1991 found approximately a 50% rate of surgical validation of fistulas among individual practitioners and those within institutions (36). This group, however, included approximately 10% of respondents who have never found a fistula and 10% who

have always identified a fistula. Black and colleagues reviewed the results of exploration from 1968 to 1990 (77). They found a positive exploration rate of approximately 70%. This survey, however, included early studies that only reported confirmed fistulas and studies examining primarily traumatic and congenital etiologies. A compilation of recent explorations for perilymph fistula of mixed etiology shows a positive exploration rate of 55%, similar to the survey of 1991 (see Table 1).

The low rate of positive findings by operative microscope has led many to investigate other forms of detecting perilymph fistulas. For example, to improve resolution, attention has turned to the use of endoscopes for visualizing fistulas. The first use of the endoscope in middle ear evaluation was reported in 1967 by Mer and colleagues who performed the procedure through existing tympanic membrane perforations (95). Reduction in size and improvements in resolution of endoscopes have allowed further evaluation of the middle ear and associated structures (96–98). Recently, the use of rigid and flexible endoscopes as an in-office substitute for middle ear exploration was evaluated (99). The procedure was found to be well tolerated and to provide excellent visualization and resolution of middle ear structures.

Poe and colleagues reported on the use of endoscopes for the detection of perilymph fistulas (99). They performed in-office endoscopy on 20 patients with a clinical history suspicious for perilymph fistula including 6 with positive fistula tests. No active leaks were observed, and there were no reported anatomic anomalies. However, in their positive control studies in which they created fistulas in 10 cats, they were able to visualize clearly the defect in all animals. They did not follow-up their negative office findings with formal middle ear explorations. In discussing the 100% negative exploration rate, they noted that they did not use injected local anesthetic that had been shown to pool in the middle ear (92).

Ogawa and co-workers, in 1994, used flexible and rigid endoscopy to examine the middle ears of eight patients with suspected perilymph fistula (100). Using a rigid endoscope, they observed a leak in two patients, which was confirmed on formal middle ear exploration. One additional patient was found with a fistula at exploration that was not visualized by endoscopy. They concluded that endoscopy is useful for diagnosis and follow-up in patients with perilymph fistula.

Rosenberg and colleagues employed rigid endoscopy in various neurotologic procedures (101). In 13 patients with suspected perilymph fistula, in-office endoscopy found no evidence of leak. They reported excellent visualization of the oval and round windows. Injectable local anesthetic was avoided in this study, as well, to avoid possible transudation of fluid into the middle ear. There was no report of results from formal middle ear exploration.

Poe and Bottrill reported on three patients with suspected perilymph fistula on whom they performed minimally invasive endoscopy followed by formal tympanotomy (102). On endoscopy, no perilymph fistula was observed in any patient, despite excellent visualization of the oval and round windows. The canal was then injected with lidocaine and epinephrine, after which a tympanomeatal flap was raised. In all cases fluid was seen pooling in the region of the oval and round windows. They noted, anecdotally, that in 48 consecutive endoscopic explorations for perilymph fistula, they have found no leaks. In interpreting the results, they reassessed the incidence of perilymph fistula and warned the reader again about contamination by local anesthetic and transudation of fluid. They noted that endoscopy without injection of local anesthetic may be more sensitive than observing accumulation of fluid for the diagnosis of perilymph fistula.

Pyykkö and colleagues, in 1995, evaluated patients for perilymph fistula with a battery of tests including in-office endoscopy by a myringotomy after topical anesthesia (103). They used a 5° and 25° endoscope, but they were able to visualize at least part of the round window in only 35% of patients. Further, in 61 endoscopies, they noted one membrane covered traumatic fistula of the round window. They identified four additional cases with abnormal mucosa that were considered "possible candidates" for perilymph fistula. No correlations with formal exploration and no treatment outcomes were reported.

The variability in observing perilymph fistulas, whether because of observer bias, mistaken identification of fluids, or the possible fluctuant nature of perilymph leaks, has focused attention on developing objective tests to detect perilymph in the middle ear. In fact, as early as 1968, Fee noted the need for identifying fluid as being of inner ear origin and speculated about labeling perilymph with isotopes (25). In 1970, Stroud and Calcaterra attempted to

TABLE 1. *Results of exploratory tympanostomy (1986-1997)*

Study	Positive ears	Total ears	Percentage observed leaks (%)
Seltzer and McCabe, 1986	121	214	57
Weider and Johnson, 1988	31	35	89
Shelton and Simmons, 1988	33	78	51
Rizer and House, 1991	35	86	41
Vartiainen et al., 1991	26	51	51
Black et al., 1991	79	90	88
Meyerhoff, 1993	26	30	87
Podoshin et al., 1994	40	53	75
Cole, 1995	31	40	76
Fitzgerald et al., 1997	55	197	28
Ostrowski et al., 1997	2	3	66
Totals/average	479	877	55

label albumin with technetium, but failed to get migration of labeled albumin into the perilymph (26). Fluorescein had earlier been shown to apparently enter the perilymph by the cochlear aqueduct from labeled CSF (104,105). Clinically, fluorescein was even used to label CSF and detect occult leaks causing rhinorrhea or otorrhea (106). This strategy was adopted by Rauch and used to detect perilymphatic fistula after intrathecal injection of fluorescein (107). The use of intrathecal fluorescein is, in fact, a non-FDA approved application, and neurologic complications have been observed in up to 25% of patients (108).

Because intrathecal injection of fluorescein is not without risk, alternative perilymph labeling methods have been pursued. Syms and associates noted the ability to detect perilymph leak after intravenous administration of fluorescein (109). The basis for this was, in part, established by Applebaum, who characterized the pharmacokinetics of intravenous fluorescein injection (110). Applebaum noted fluorescence of the perilymph within 2 minutes of injection. Further, by comparing CSF labeling kinetics, he concluded that perilymph labeling arose by transudation across cochlear vasculature and not by diffusion from the cochlear aqueduct.

The assumption of perilymph labeling, either by CSF diffusion or cochlear transudation was challenged in 1993. Poe and colleagues observed pooling of label around the oval and round windows after intravenous fluorescein injection (111). Upon mechanical rupture of the membrane, the mixture of perilymph and blood fluoresced. However, when rupturing the membrane hemostatically, no fluorescent label was detected in the perilymph. They concluded that fluorescein was in the blood vessels and not in the inner ear fluid. Likewise, Bojrab and Bhansali showed fluorescence in vessels and mucosa, but not in the perilymph itself (112).

In an interesting use of fluorescein, Arenberg and Wu labeled their local anesthetic with fluorescein (113). Therefore, fluid in the middle ear could be shown to be anesthetic by virtue of positive fluorescence. Nonfluorescent fluid in the middle ear would increase the suspicion of an inner ear origin. In 10 patients, all showed seepage of fluorescein-positive fluid into dependent portions of the middle ear after elevation of the tympanomeatal flap. By using the endoscope through a myringotomy incision before raising the flap, they observed a "dry" ear in four patients.

The conflicting reports and uncertainty of perilymph labeling led to further attempts at definitively distinguishing perilymph from other fluids. In 1991, Paugh and colleagues subjected perilymph, CSF, and serum to two-dimensional gel electrophoresis (114). By separating proteins based on charge and molecular weight, they identified several spots found only, but consistently, in perilymph. They postulated the ability to use these markers to confirm the presence of perilymph in the middle ear. Drawbacks to this process are the need for specialized laboratory equipment and personnel, as well as a 48-hour protocol and thus delay in results.

Thalmann and colleagues similarly examined the use of two-dimensional gel electrophoresis for the identification of perilymph (115,116). Although they found several candidate proteins to distinguish perilymph from serum or CSF (*e.g.*, apo D, apo J, beta-2 transferrin), they noted limitations of the applicability of such a technique. Namely, samples needed to be relatively pure and concentrated, thus becoming unreliable in the presence of a local transudate or dilution from local anesthetic.

Silverstein reported the identification of perilymph by protein concentration in a rapid, simple office test (117). Using protein paper that generated differing shades of green based on protein concentration, he performed office myringotomy and collection of middle ear fluid in microcapillary tubes. This test was based on different protein concentrations between perilymph (200 mg%), CSF (20 mg%), and serum (7000 mg%). He noted the possibility of erroneous results secondary to anesthetic dilution of middle ear transudate.

As noted, several proteins can distinguish perilymph from other body fluids. The most actively studied has been beta-2 transferrin. This protein can be studied by simple one-dimensional gel electrophoresis and is found in perilymph and CSF, but not in serum (118,119).

Investigators at Pittsburgh critically evaluated the use of beta-2 transferrin for detection of perilymph and CSF leaks (120,121). They cited collection methods, laboratory protocol, concurrent patient disease, and physician acceptance as potential sources of error in evaluating perilymph for evidence of fistula and leak. With these factors in mind, they performed a prospective single-blinded study of the ability to detect perilymphatic fistula by beta-2 transferrin testing (122). Gelfoam was placed on the oval and round windows in patients with suspected fistulas and in patients undergoing other otologic procedures. The pledgets were blindly tested for beta-2 transferrin. Of 10 patients who were not suspected of having fistula, no fistulas were observed and no samples were positive for beta-2 transferrin. Of 10 patients who were suspected of having perilymph fistulas, 9 were observed on tympanostomy and 66% of these were positive for the protein. They concluded that beta-2 transferrin testing is a better predictor of perilymph fistula than direct observation. However, the evaluation of sensitivity and specificity of the test was predicated on surgical observation as the gold standard.

A more extensive study was later produced by the Pittsburgh group, examining 43 patients with suspected perilymph fistula (123). In this study, they had a 54% observation of fistula in clinically suspected cases. Of the 23 surgically "confirmed" leaks, only 26% were positive for beta-2 transferrin. Further, of 20 patients suspected of fistula, in which no fistula was observed, 2 were positive for beta-2 transferrin. Rather than assign two cases of false-positive to the beta-2 transferrin test, the authors suspected a false-negative exploration. The authors concluded that beta-2 transferrin is at least as accurate as surgical exploration. This study was well designed, but, as in the previous study, and indeed with any test for perilymph fistula, lacked an adequate standard (87).

Further investigation of the efficacy of beta-2 transferrin testing was performed by Levenson and colleagues

(124). They collected samples of perilymph and/or CSF from patients undergoing various neurotologic procedures. Although 100% of the CSF samples were positive, only 2 of 13 perilymph samples demonstrated beta-2 transferrin. The authors concluded that beta-2 transferrin testing for the identification of perilymph is unreliable. They noted that negative results may be secondary to inadequate collection methods or sample volumes. Further, they noted that a positive test might result from CSF "contamination" of perilymph through congenital anomalies of the IAC or cochlear aqueduct.

The unreliability of beta-2 transferrin testing has been recently reiterated by Buchman and colleagues (125). In a blinded and controlled study, they identified only 5% of known perilymph samples by beta-2-transferrin immunoelectrophoresis. They concluded that such testing may be unreliable by the currently employed collection and analysis protocols. Telian and associates have also noted the unreliability of beta-2 transferrin testing and have thus investigated the efficacy of apolipoprotein D as a marker for perilymph (126). Their initial findings are encouraging, as 75% of perilymph samples were accurately identified and no false positive test results were noted.

Visualization of perilymph leaks by noninvasive methods has been attempted. These have primarily depended upon high resolution CT scans of the temporal bones. In 1986, Zalzal and colleagues retrospectively reviewed their experience with CT evaluation of 124 children with various otologic complaints (127). They noted that although CT scan may be useful for many congenital and acquired anatomic abnormalities, it was particularly inadequate in evaluating the oval and round window regions. Of three children operatively found to have perilymph fistula, none were diagnosed by CT scan.

Reilly, in 1989, performed a prospective study of children with sensorineural hearing loss and/or vertigo (128). Of 234 CT scans, 11% had abnormal morphology of the cochlea, internal auditory canal, and vestibular aqueduct. There was no report of increased suspicion of a perilymph fistula based upon the CT scans. Of 137 conventional polytomograms, 7 had distortion of the oval and round windows. Upon exploration, 15 children were found with congenital perilymph fistulas. Of these 15, only 3 were noted to have had abnormal radiographic findings. Thus, it appears that clinical history is more sensitive for the presence of a perilymph fistula than is radiographic assessment.

Shusterman and coworkers reported similar findings with CT evaluation (129). Of seven patients (eight ears) with suspected perilymph fistula, six were confirmed to have perilymph fistula at surgical exploration. However, five of these patients had CT scans interpreted as normal, and the other two demonstrated abnormalities, not of the oval or round windows, but reportedly of the endolymphatic ducts. The authors indicated a high false-negative rate of CT evaluation and noted that exploration for perilymph fistula should be based upon history and clinical exam and not on radiography.

A better correlation between CT interpretation and operative findings was reported by Weissman and colleagues (130). They again reviewed the CT scans of 10 children with surgically confirmed congenital perilymph fistulas. On direct exam, they found that the most common middle ear abnormalities associated with a fistula were of the stapes and round window. Only 33% of abnormal stapes could be identified on CT scan, and none of 3 abnormal round windows was detected preoperatively. However, more than 50% of patients with a perilymph leak had an abnormal CT scan of the inner or middle ear. They concluded that a CT scan could increase the suspicion of a perilymph fistula by virtue of identifying associated middle and inner ear pathology.

Lövblad and colleagues presented an alternative approach to the use of CT scans (131). They performed intrathecal injection of iodinated contrast in a child with sensorineural hearing loss and clear otorrhea. Computed tomographic scan demonstrated an abnormal stapes and contrast material within the tympanic cavity suggesting a perilymph leak. Surgical exploration confirmed the abnormal stapes and demonstrated a perforated oval window with perilymph egress. They note that, although invasive, intrathecal contrast is efficacious in demonstrating continuity of the inner and middle ears. Further, they suggested that magnetic resonance imaging (MRI) may provide the higher resolution necessary to localize the fistula rather than to just suggest the existence of a leak.

Morris and associates attempted to assess the feasibility of MRI in perilymph fistula detection (132). They created round window fistulas unilaterally in experimental cats. Precontrast and postcontrast MRI images were acquired using intrathecal gadolinium enhancement. They found passage of contrast from CSF to the perilymph and then pooling of enhanced fluid in the mastoid bulla. They concluded that MRI could detect communication between the inner and middle ears but a noninvasive, more cost-effective modality would be preferred.

TREAMENT OF PERILYMPH FISTULAS

The difficulty in visualizing perilymph leaks makes precise and directed therapeutic intervention challenging. Antecedent surgery, congenital anomalies or erosive disease can lead an otologist to the site of leak and helps in directing fistula repair. Cases of idiopathic or spontaneous fistula, however, provide both a diagnostic and therapeutic challenge. Further, evaluation of outcomes is difficult, as there is no definitive way to confirm that a fistula was or was not present. Nevertheless, numerous series have been published presenting methods of repair as well as surgical outcomes.

Conservative or medical therapy for suspected perilymph leaks is rarely reported, as many consider fistulas a surgically correctable problem. Davis, reporting the University of North Carolina approach to fistulas, noted that patients believed to have spontaneous leaks are prescribed bedrest, head elevation to 30°, and avoidance of lifting or pressure increasing activities (94). Similarly, Nomura noted that if a fistula is considered idiopathic, a 1-week trial of strict bedrest may allow spontaneous healing (133). There have

been no reports of the percentage of patients that respond to such therapy.

The principle approach to repairing perilymph fistulas is covering the leak site with some form of autologous tissue. The difficulty in visualizing a fistula, however, presents a dilemma as to further management in the face of a negative exploration. Additionally, if a leak is seen at one window, it is unclear if concurrent but inactive fistula is present at other sites. Hughes and colleagues reported on practices of otologists in the United States and found that 78% place patches on both windows regardless of findings at direct observation (37).

Various tissues have been used to patch the oval and round windows. These include fat, fascia, perichondrium, and areolar tissue (29,34,57,94,133). Seltzer and McCabe have used several materials and have found that subcutaneous areolar tissue conforms best to the oval and round window regions (32). Further, they reported a slightly higher incidence of recurrence with the use of fat. Davis likewise found perichondrium to be superior to fat (94). Vartiainen and colleagues used fascia, but noted that in two reexplorations, the fascia had disappeared (89). Kohut and coworkers reported good adherence to bone of an areolar tissue and fibrin glue patch upon histologic examination 4 years after surgery (57). Black and associates used the Argon laser to clean the mucosa surrounding the oval and round window and placed a fascia, fibrinogen, and thrombin graft (77). Further, they buttressed the graft with Avitene and laser coagulated the edges to promote a seal.

Outcome measures for perilymph fistulas are difficult to analyze because of the range of complaints at initial presentation. Most series report a subjective improvement rate stratified by the cochlear and vestibular systems. There have been some postoperative objective test reports, usually audiograms and less frequently vestibular tests. Occasionally, overall response to therapy is reported, and these have generated significant controversy. For example, Shelton and Simmons noted that 64% of patients with a confirmed fistula felt improvement (33). However, 44% of patients with negative exploration also noted improvement. If only spontaneous fistulas were examined, response rates were essentially the same in both groups. Rizer and House had similar results in which 68% of confirmed fistula patients improved and 29% of nonfistula patients also improved (34). This has led some to doubt a negative exploration and consider a fistula present in any patient that responds to patching (73,75). Furthermore, the inability to distinguish between true cure and a placebo effect has generated criticism of perilymph fistula surgery (33,38). The placebo effect of surgery has been shown to be quite strong with sham operations demonstrating positive outcomes in 35% to 65% of cases (134,135).

Vestibular complaints with perilymph fistulas have been found to be the most common and have likewise been found to be the most treatable. Seltzer and McCabe found improvement in 94% of patients with primary vestibular complaints (32). Similarly, Black and colleagues showed improvement in 84% to 89% of their patients (74,77); with Healy and coworkers, 100% of their patients improved (29); with House and associates, 72% to 78% of their patients improved (34); and with Fitzgerald and colleagues, 87% of their patients had improvement in vestibular symptoms (75). Using objective testing, Black and coworkers found no significant change in preoperative and postoperative ENGs or rotational testing (74). They did demonstrate significant improvement in dynamic posturography, with 12 of 32 patients having normal tests after fistula closure.

Hearing loss is less common than vestibular disturbance and has proved more frustrating in providing meaningful intervention. Seltzer and McCabe were able to improve hearing in 49% of patients with confirmed fistula, but only 23% achieved speech reception thresholds > 35dB or discriminations > 80% (32). In 40% of patients, hearing remained stable. Fitzgerald and colleagues had similar results, with 40% of patients presenting with sensorineural hearing loss improving (75). Shelton and Simmons noted less success and stabilized hearing in 50%, but had significant improvement in only two patients (33). Black and colleagues had improvement in 15% of patients and no further hearing loss in 67% (74,77). Healy and coworkers noted 9 of 20 patients had better hearing, but 4 had further hearing loss (29). Rizer and House noted only 13% to 18% of their patients had improved hearing (34). Meyerhoff noted 24 of 27 patients with abnormal ECoGs preoperatively and only 3 abnormal tests postoperatively (35). However, hearing thresholds were unchanged in 18 of the 27 patients.

AUTHORS' COMMENTS

Even the staunchest detractors of the occurrence of spontaneous perilymph fistulas agree that fistulas of defined etiology do exist (38). The most common occurrence of perilymph fistulas is postsurgical at the site of the oval window. Although such patients may present with signs and symptoms similar to Meniere's disease, they are generally explored based upon a highly suspicious antecedent event. Less accepted, but not as strongly disputed, are those patients presenting with similar complaints after trauma. Many of these patients are also explored, and patching of the oval and round windows is common. Strongest debate centers on those patients, likewise with similar complaints, in whom the inciting incident is a common event, like sneezing or lifting, or in whom no event is recalled at all. Herein lies the major controversies surrounding perilymph fistulas: can perilymph fistulas occur "spontaneously?"; what symptoms are specifically associated with such fistulas?; what tests can diagnose a fistula?; what is the "gold standard" for confirming the presence of a fistula?; how should a fistula be treated?

Evidence has been presented demonstrating developmental centers of the otic capsule that correlate with reported sites of perilymph leak. Further studies have shown apparently age-related, acquired fissures in the otic capsule. Additionally, postmortem temporal bone analyses have correlated clinical findings of fistulas with histopathologic studies. These findings appear to provide strong evidence

for the generation of leaks at these sites, yet controversy persists. Critics have raised issues of processing artifact, yet the consistency of these findings makes such a case unlikely. What needs to be answered, however, is if fissures are acquired, why is the incidence of perilymph fistula not age-related? Further, there has been no definitive proof that the core of these fissures is truly permeable. Additionally, it must be explained why such weaknesses would suddenly permit the flow of perilymph.

There has been evidence presented that CSF pressure is closely tied to perilymph pressure and may underlie inner ear membrane ruptures. The presumed mechanism is transmission by means of a widened cochlear aqueduct. Indeed, studies have shown that in some people CSF pressure rises dramatically with sneezing or the Valsalva maneuver. However, another study disputed the presence of widened cochlear aqueducts. In the opposite extreme, it was calculated that an extremely narrow or closed aqueduct may make the inner ear susceptible to implosive etiologies, such as potential increases in middle ear pressure, also during sneezing or the Valsalva maneuver. This issue clearly has yet to be resolved.

There have been many studies investigating the relationship of presenting symptoms with the presence of a perilymph fistula. Although claims have been made that specific patterns of hearing loss or vestibular disturbance should increase one's suspicion of a perilymph leak, they all suffer from lack of a standard for significant data analysis. The standard in most cases has been the finding of fluid around the oval or round windows at surgical exploration. Fitzgerald and colleagues, conversely, used improvement after patching as their standard regardless of the finding of an active perilymph leak (75). Until a definitive means of confirming the presence of a perilymph leak is developed, studies analyzing patterns of symptoms will be inherently flawed.

The decision to explore the middle ear is still based primarily upon a highly suspicious history. Exploration in the absence of inciting events, as in spontaneous fistulas, appears to be based on a constellation of signs and symptoms. As described above, none of these provides a high rate of success for detecting a fistula, but may be all the clinician has upon which to base a differential. Exploration in many cases appears to be a last resort and may do nothing more than - reassure the physician and patient that all possibilities have been entertained.

The results of endoscopic studies of the middle ear to establish definitive data for the evaluation of perilymph fistulas are particularly disturbing, in light of the large number of fistulas reported in the literature. The incidence of a positive exploration with endoscopes is exceedingly low. In fact, Poe and colleagues in several reports have yet to find an active leak (99,102). Even more troubling is their visualization of fluid, similar to those reported as positive explorations, only after anesthetic injection or tympanomeatal flap elevation. The fluid may indeed be anesthetic or perhaps raising of the tympanomeatal flap provokes a transudate from the local tissue. Alternatively, middle ear pressure gradients may be such that perilymph egress in the recumbent stable patient is only possible after the flap is raised. This would indicate that some of these explorations may be positive, although there would still be a high rate of false positives. Current methodologies do not provide sufficient specificity and sensitivity to accurately diagnose perilymph fistulas.

The otologist must decide the best therapeutic option in a patient suspected of having a perilymph fistula. Improvements in symptoms after patching the oval and round windows are about equal whether a fistula is visualized or not. Controlled studies are lacking, as few physicians are willing to do nothing when faced with a negative exploration. If the suspicion to explore were high, it would seem reasonable to place patches in areas of the most common leaks regardless of observation. The morbidity from such procedures is very low and with some negative explorations improvement has been noted in almost 50% of patients. Whether this is a placebo effect or closure of nonvisualized fistulas is not known, but helping 50% of patients with such a challenging presentation can be seductive.

It is the authors' opinion that perilymph fistulas do exist. They most certainly occur after surgery and posttrauma. Spontaneous fistulas are very rare occurrences and the majority are likely incited by a pressure-altering event. A high index of suspicion should provide the impetus to explore and, regardless of the findings, patches should be placed. The patient must be informed preoperatively of issues related to middle ear exploration and fistula treatment and the controversies regarding these entities. The patient and physician should be prepared for no response to treatment, particularly regarding hearing improvement. Alternative diagnoses need to be entertained and pursued where warranted. Frustration must be recognized by both physician and patient, and compassion for a patient with real but nonspecific symptoms maintained. Above all, questions must continue to be asked and further research pursued to help distinguish reality from myth.

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INVITED COMMENTS

This article is a very well written, thorough, and "critical" examination of the important literature concerning perilymphatic fistulas. The controversies are quite fairly presented.

The authors have very correctly noted that the fundamental problem with any discussion on perilymphatic fistulas is that there is no objective measurement on which a diagnosis may be made. Exceptions may include very obvious wide open defects found largely in postsurgical or occasionally in head trauma cases. The majority of reports use a clinical judgment on the presence of clear fluid welling up repeatedly in the oval or round window niche areas observed during surgical exploration as their "gold standard." The presence of fluid may be difficult to ascertain or, if present, is impossible to visibly distinguish from injected anesthetic or surgical transudate artifacts.

While performing previous work with fluorescein intravenous injections in an attempt to examine for its presence in perilymph, I observed fluorescein-labeled fluid pooling continuously in the dependent oval and round window niches of cat and dog models before any manipulation of the labyrinth. The fluid was presumably transudate from the surgical exposure and was impossible to completely eliminate. We concluded that it was likely to be equally impossible to visually differentiate between artifacts and perilymph by inspection alone.

All of the literature presented in this review must be reinterpreted in the light that visual inspection for fistulas is probably highly inaccurate. The poor correlation of signs, symptoms, and physiologic testing with surgical observation of fistulas is not surprising given the artifact problems. Observation alone should no longer be held as an adequate standard, and newer standards must be sought through protein analysis, imaging, or safer intrathecal tracers.

Endoscopic examinations with only topical anesthetic reduce the artifact problem and may be a more reliable means for observation. The authors noted the paucity of identified fistulas reported endoscopically. Pyykko in more than 200 cases found 2 fistulas by office endoscopy.

I have explored 13 patients to date with an intraoperative endoscopy followed by injection of 1 ml 1% Lidocaine and epinephrine 1:100,000, and conventional open exposure. Six cases showed no fistula by endoscopy, but appeared positive on open exposure. These were suspected to be surgical contamination by artifact and false positives. Five cases were equivocal because of bleeding from adhesions caused by previous surgery. Two cases showed true oval window fistulas at the anterior rim in both endoscopy and open exams and both were poststapedectomy cases.

I have now found a single case, out of 81 office endoscopies, of a noniatrogenic fistula in a scuba diver. The patient was treated with a transtympanic injection of autologous venous blood as suggested by Dr. Jean Bernard Causse for poststapedectomy-suspected fistulas, who found it successful and lasting in one third of the patients. The diver had an immediate, impressive relief of symptoms that has lasted longer than 1 year. Blood patch techniques should be further studied as a possible treatment. If significant relief is gained but symptoms return within 1 to 2 weeks as the blood disappears, it may warrant a definitive fistula repair. It should be noted that any manipulation of the ear may favorably influence Meniere's disease as well; therefore, a positive response cannot be equated with definitive identification of a fistula.

The authors noted that the fistula may be a problem of air gaining access to the labyrinth and not because of fluid leaking out. Many surgeons have found unsuspected asymptomatic fistulas after stapes surgery, and patients under local anesthesia do not generally report vertigo with an open leaking vestibule unless the surgeon aspirates the perilymph. Electrocochleographic changes during aspiration of the vestibule has been reported, and pneumolabyrinth has been documented in a fistula case by computed tomography scan. Improvements in imaging resolution will be necessary before minute amounts of air will be detectable on a routine basis. Blood patches may be useful for temporary occlusion of a suspected fistula and pneumolabyrinth.

The histologic identification of microfissures filled with loose fibrous tissue that widen with age is difficult to accept as a true source for leakage. Thin fibrous adhesions are usually grossly effective fluid barriers and even Kohut used loose areolar tissue to patch possible fistulas. The authors note that if these fissures were indeed responsible for leaks, then it should follow that the incidence should rise with age as the fissures widen.

It is my opinion that noniatrogenic fistulas are rare and that "spontaneous fistulas" are even more rare unless a significant congenital defect is present. For previously nonoperated patients, I now recommend fistula exploration for fluctuating or progressive sensorineural hearing loss and/or vertigo associated with straining or valsalva if they have a positive objective or subjective fistula test response, and they must have a history for a significant direct trauma or barotrauma. Individual exceptions are made case by case for children. When an exploration is negative, I do not recommend patching the windows, because it is likely they may have developing hydrops. Patients who later develop

Meniere's disease may benefit from intratympanic injections of medications, which may not be effective if the windows have been patched. If there is uncertainty about the exam and treatment is desired, a blood patch may be tried.

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In this well-documented article, the authors make a critical analysis of numerous publications concerning perilymphatic fistulas. Through a complete historical background, they underline:

- the similarity of symptoms with numerous vestibular pathologies and especially the Meniere's disease.
- the absence of an objective specific test.
- the importance of middle ear exploration to see this fistula.

The importance of middle ear exploration to see this fistula

Among controversial elements, let's note the difficulty in history taking for the search for antecedents, in the event of possible spontaneous fistulas.

The lack of specificity of the fistula test, the difficulties in confirming the presence of a fistula during surgery (B2 transferine and fluoreceine remain controversial), the variability of fistula closing techniques.

The authors scarcely mention the symptom production mechanism on the physiopathologic level.

In our experiment with a set of 86 explorations for fistulas,let's cite:

- the interest of Tullio's sign (vertigo or nystagmus after a very high-level sound stimulation at low and very high audio frequencies).
- the interest of computed tomographic scan when searching for luxation of the ossicular chain, pneumolabyrinth, inner ear malformation, large vestibular aqueduct, otologic surgical antecedents, and declive fluid in the tympani sinus in a positional scanner.
- the interest of intraoperative provocative tests.

Indeed, in this set, 62% of fistulas were visible, only 55% of which were after intraoperative provocative tests.

I believe the satisfactory results of this surgery, the risk of spontaneous evolution, and the simplicity of the surgical investigation seem to justify an operative indication for any symptomatology suspected by clinical and paraclinical investigations.

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The authors have done a credible job of summarizing the historical and recent literature on perilymph fistulas (PLF). However, the vast majority of the article discusses the complex problem of PLFs in general (mostly without regard to cause or rigorous discussion of pathophysiology), rather than focusing on spontaneous PLFs (SPLFs), which by definition have no cause. In fact, the authors don't mention SPLFs until near the end of the article. Bottom line: The content of the article is not consistent with the title.

Among the best ways to develop or perseverate a "controversy" is to obfuscate the problem. Do we really think that PLFs simply happen for no reason, analogous to prevailing opinion on "spontaneous combustion" before the age of enlightenment? Based upon the preponderance of literature to date, it is far more likely that an "event" (most likely a sudden, transient pressure differential across the oval or round window) causes PLFs, with the patient (and/or their physician) simply unaware of the event or unable to link a precipitating event with the subsequent emergence of PLF symptoms. Had the authors presented a definition of "spontaneous" PLF, then critically reviewed the relevant literature, a fruitful discussion of the issues would be possible.

Throughout the paper, there is a "soft thread" in which the authors seem to want to link SPLFs with vague symptoms, subclinical findings, and/or negative surgical explorations. I don't believe the authors were successful in doing this, but it may be interpreted in that way by some readers. More importantly, the authors seem to confuse complications of PLFs (especially secondary endolymphatic hydrops and hearing loss) with the basic pathophysiology of PLFs. Defects in the otic capsule, oval windows, or round windows, including PLFs, usually do not cause hair cell or neural damage (the usual cause of permanent hearing loss in PLF patients) or secondary hydrops unless the defect results from extreme pressure changes or unless the PLF persists. "Gold standard" example: stapes surgery. An oval window PLF must be created for a successful stapedotomy or stapedectomy, but most patients do not develop permanent hearing loss or secondary hydrops (well documented complications of persistent poststapedectomy PLFs) before the iatrogenic fistula heals.

A fundamental flaw of this review is represented in the statement: "Many otologic pathologies can be diagnosed by history and basic physical examination; however, the nonspecific presentation with idiopathic or spontaneous perilymph fistulas demands further diagnostic evaluation." The authors need to defend this statement with factual data. I know of no inner ear condition that can be "diagnosed" by history and basic physical examination. A differential diagnosis with a high probability of accuracy sometimes can be developed with an accurate history and a physical. The "gold standard" example is otosclerosis. Histopathologic confirmation is required to establish the diagnoses objectively. All conductive hearing losses are not caused by otosclerosis and all fluctuating auditory and vestibular symptoms are not PLFs. As for PLFs, inappropriate indications for exploration will lead to high false negative rates for the "diagnosis" of otosclerosis at tympanotomy. Perhaps it also should be stressed that there are no tests of auditory or vestibular function that are "diagnostic" of PLFs or any other histopathologic diagnosis.

The authors' comparison of Meniere's disease and PLFs seems weak. It is true that some PLF patients with a secondary hydrops will present with symptoms similar to idiopathic endolymphatic hydrops. However, by definition, a "diagnosis" of Meniere's disease means that the endolymphatic hydrops is idiopathic, i.e., no cause can be demon-

strated. Although this may, on the surface, seem a reasonable rationale to compare the pathology between the two conditions, it should be recalled that a PLF is basically a mechanical defect between the perilymph fluid space and the air space of the middle ear. Consequently, any damage to the hair cells, the neural transduction systems, and/or the fluid control systems of the inner ear are complications of a PLF, not a primary pathophysiology as presumably occurs in Meniere's disease. In fact, the confusion between the two conditions was highlighted when an observant Dr. Fee (a Canadian otolaryngologist) explored the middle ears of head trauma patients who were thought to have Meniere's disease. The American Academy of Otolaryngology criteria for Meniere's disease is quite different from the basic presenting complaints of PLF patients. This differentiation was not adequately discussed and is critical for a productive exchange of ideas and perspective on PLFs. A review and critical assessment of the diagnostic criteria presented in the literature would likely produce a more productive approach to addressing and resolving any "controversy" about PLFs.

The discussion of sudden hearing loss and PLFs is also disturbing, because the relationship between round window PLFs and sudden deafness secondary to barotrauma has been well established. To confuse this cause of PLFs with other pathologies of the ear is inexcusable and seems to result from speculation when the diagnosis is unclear. Why not simply state the obvious (i.e., that the diagnosis is uncertain or unknown) and avoid tagging a patient with an incorrect diagnosis? Would this not alter the level of controversy to a focus on data as opposed to unsubstantiated opinion? In fairness to the authors, the basic problem here is the lack of adequate pathologic data. Causes of PLFs, sudden deafness, and most other inner ear diseases do not usually kill patients. Thus, the availability of timely pathologic examination is impossible. Until a generally accepted method of diagnosis is developed and an adequate pathologic basis of inner ear disorders is established, this lack of clinical–pathologic correlation has the inevitable effect of fueling "controversy" for the diagnosis and management of many otologic conditions.

The section on diagnostic techniques (endoscopic versus tympanotomy) for identification and the discussion of local versus perilymph fluid provides a reasonable overview of the current status: unsatisfactory. The main problem with endoscopic examination is the monocular view and the inability to fully view the oval and round windows, especially in patients with traumatic fistulas who almost invariably have membranous obstruction of these critical areas.

The issue of local anesthetic versus perilymph identification at tympanotomy is easily resolved. The surgeon may simply dry out the mucosa overlying the area of interest with suction. Perilymph leaks appear in the most dependent operative area, not from the external canal direction. It also helps to avoid injecting too much local anesthetic.

The authors do not seem to realize that an otic capsule defect (fistula) without perilymph leakage can yield symptoms identical with those arising from an active perilymph leak. At least two "gold standards" (one for perilymph in the middle ear and one for otic capsule defects) are therefore needed for the diagnosis of "PLFs."

The final paragraph tries to summarize the "state of the art," but the statements made don't go together. For example, the authors say that SPLFs are "rare occurrences" and that the majority are probably incited by a "pressure altering event." However, this conclusion was not presented or defended by data in the text. All the other statements in the concluding paragraph are devoted to general statements about PLFs. In my opinion, this serves only to guarantee continued obfuscation of the issues.

The authors should be complimented upon the enormous effort put forth. I appreciate the opportunity to comment on this article.

Bernard Fraysse
Toulouse, France

This article is well written and a complete review of previously published material on perilymph fistulas. The authors have tried to be objective, but at times their prejudices have slanted the material. The title refers to "spontaneous fistula," but much of the material refers to acquired fistulas from trauma or after surgery. In general, this article is an excellent review of the literature without any original material.

Early work with stapedectomy surgery was associated with the occurrence of perilymph fistulas. The authors do a good job reviewing this topic, but fail to recognize that fistulas rarely or never occur today in stapes surgery. We were seeing fistulas when the technique was to remove the entire stapes and stapes footplate, replacing them with a single loop wire prosthesis over a pad of gelfoam. The gelfoam created a very thin membrane, which could rupture or leak. In addition, the wire tended to migrate to the side of the oval window, further increasing the chances of a fistula. In the past, we have had the opportunity to revise cases because of a recurrence of the conductive hearing loss to find the wire in an entirely open oval window without any membrane covering. These patients did not have vertigo or dizziness. Today, we are using a small fenestra technique. A 0.7 mm fenestra is created in the center of the footplate and a 0.6 mm piston placed. This is sealed with a blood patch. Since we have been using this technique, we have not seen any perilymph fistulas.

Stapes surgery creates a temporary perilymph fistula, but the patient rarely has vertigo or dizziness after the surgery. If perilymph fistulas cause so many problems, why do we not see this during or shortly after stapes surgery or cochlear implant surgery? It is true that patients with postoperative stapes fistula do, at times, complain of fullness in the ear, and in a few patients dizziness occurs with changes in altitude. I have also seen similar symptoms in patients who have a long prosthesis. By changing the prosthesis to a shorter one, the symptoms are relieved.

The authors refer to our paper (Rizer and House) on several occasions. The research for the paper was motivated by an experience I had reviewing a group of more than 80 patients from another institution who had undergone fistula surgery over a 2-year period. Two other nationally

prominent surgeons and I were asked to look at these cases. We were to give our opinion whether surgery was indicated and if the findings at surgery and the results justified the surgery. We all agreed that in our opinion in most cases there was not an indication and the results did not substantiate the surgery. This prompted me to look at our experience at the House Ear Clinic over 12 years with 10 surgeons. We found 86 procedures in these 12 years and, as has been pointed out, fistulas were found in only 41% of the cases. If there was a fistula, 68% improved, but if no fistula, 29% improved. What the authors failed to point out was that the majority of these positive explorations were in postoperative stapes patients who had previously undergone a stapes over gelfoam procedure.

The authors spend a great deal of time on the anatomical justification for fistulas. They base much of this on Kohut's work. Kohut found "loose fibrous composition in the fissula ante fenestra" in bones of patients who supposedly had perilymph fistulas. As has been pointed out by many others, these fissures are common in all temporal bones and don't prove the existence of active perilymph leaks. It is doubtful that there would be symptoms even if a small amount of perilymph could travel through these paths.

The authors refer to work by El Shazly and Linthicum at the House Ear Institute. They were looking at patients who had sudden sensorineural hearing loss. They found that the "microfissures" are common and have no clinical significance. This study was prompted by Goodhill's claim that the sudden sensorineural loss was caused by spontaneous perilymph fistulas. Shortly after Goodhill proposed this, the physicians of the House Ear Clinic began to explore ears in patients with sudden hearing loss. After approximately 100 explorations, they did not find any fistulas. For this reason, we do not explore sudden hearing loss cases in search of fistulas.

Diagnosing perilymph fistulas preoperatively is virtually impossible. This is well pointed out and covered in this article. Results of the surgery are also difficult to gage. Most of those who report improvement are referring to dizziness, not hearing gain. This has also been our experience. Hearing rarely improves after repair of the fistula. Most series, including ours, show a substantial number of patients reporting improvement of symptoms with both positive and negative explorations. This would make one consider the placebo effect of surgery. In fact, we (Rizer & House) found in negative explorations a significant number of patients improved when a prophylactic repair was not performed.

The authors state "Conservative or medical therapy for suspected perilymph leaks is rarely reported, as many consider fistulas a surgically correctable problem." I would not agree with this statement. When we see a patient with sudden sensorineural hearing loss or sudden onset of vertigo, we do not think of a fistula, whereas many others would. We treat these medically, with reasonably good results. If in fact these are fistulas, this is an example of medical treatment. I would like to also point out that many fistulas close spontaneously. The best example of this is in postoperative stapes and cochlear implants. We have had good results with scuba divers who have suffered sudden hearing loss. In our fistula study, we surgically explored five such cases early with good hearing return. A large number of similar cases were not explored, with similar good hearing results.

In conclusion, this is an excellent and complete review of the literature. In the authors' comments, several questions are asked:

Can perilymph fistulas occur "spontaneously"? Doubtful or extremely rare.

What symptoms are specifically associated with such fistulas? I don't know, because we rarely diagnosis spontaneous fistulas.

What tests can diagnose a fistula? None.

What is the "gold standard" for confirming the presence of a fistula? Surgery.

How should a fistula be treated? Initially with steroids and rest for a week or two. If not, better to perform surgery.

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