

PERILYMPH FISTULA: THE IOWA EXPERIENCE*†

SHARON SELTZER, MD

BRIAN F. MCCABE, MD

Iowa City, IA

ABSTRACT

Ninety-one patients with demonstrable perilymph fistulas presented with an amazing array of signs and symptoms ranging from unilateral tinnitus and aural fullness to sudden-and-profound hearing loss, roaring tinnitus, and whirling vertigo. Between 1977 and 1984, 214 exploratory tympanotomies for suspected perilymph fistula (PLF) were performed on 177 patients. One hundred six primary (initial) fistulas were found in 95 ears and 26 recurrent fistulas were identified. Eighty-two percent of the 91 patients with PLF had auditory symptoms, 8% with auditory symptoms as the sole complaint. Eighty-one percent of the patients had vestibular symptoms, 12% with vestibular symptoms alone. The type of hearing loss and the nature of the vestibular symptoms were widely variable. Of the 58 patients with preoperative auditory symptoms, 49% had improved hearing (23% improved to serviceable range), after closure of PLFs. Ninety-five percent of the patients who presented with vestibular symptoms had elimination of or decrease in their dizziness to the extent that it no longer interfered with their daily activities. The highest incidence of recurrent fistula was associated with grafts using fatty tissue. Patients with Mondini deformity were at particularly high risk for fistula recurrence.

Perilymphatic fistulas create abnormal communications between the fluid surrounding the membranous labyrinth and the middle ear space. They occur in both the round and oval windows and may be the result of stapedectomy,¹⁻³ head trauma,⁴⁻⁶ barotrauma,⁷ surgery for chronic ear disease,⁸ physical exertion,⁹ congenital anomalies,^{10,11} or idiopathic (spontaneous) causes.¹² Reports of the diagnosis and treatment of perilymph fistula (PLF) are well represented in the literature.¹³⁻¹⁶

Despite this scrutiny, however, no firm guidelines for the accurate preoperative diagnosis of PLF have been formulated. Simmons expressed the frustration of many otolaryngologists with this ambiguous state of affairs.¹⁷ The surgeon's dilemma arises because of the variety of signs and symptoms with which a patient with PLF can present and the lack of a single or narrow group of conclusive preoperative tests to support the diagnosis.

While many authors have approached the problem by trying to specify the constellation of common symptoms in cases of PLF, this discussion focuses on the differences. By highlighting the varied faces of PLF, the incidence of undiagnosed fistulas and attendant progressive hearing loss, vestibular symptoms, and possible complications may be reduced.

MATERIALS AND METHODS

Records of all patients who underwent exploratory tympanotomies from January 1977 through March 1984 were reviewed. During this period, 91 patients had surgically proven PLFs. Anatomic sites of fistulas and middle and inner ear malformations detected by mastoid tomograms were collated with detailed histories of preoperative and postoperative symptoms, predisposing factors (including previous ear operations), physical examination findings, and preoperative and postoperative audiograms.

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†From the Department of Otolaryngology-Head and Neck Surgery, University of Iowa Hospitals, Iowa City.

Send Reprint Requests to Brian F. McCabe, MD, Department of Otolaryngology-Head and Neck Surgery, University of Iowa Hospitals, Iowa City, IA 52242.

At the time of exploratory tympanotomy, obscuring mucosal sheets and tents were lysed to permit optimum visualization. The round window (RW) and oval window (OW) were each observed for precisely 5 minutes for leakage of clear fluid or a consistently shifting light reflex after repeated drying. If no change was seen initially, the patient was placed in the Trendelenburg position and (if under local anesthesia) instructed to perform a Valsalva maneuver. Patients under general anesthesia were subjected to hypoventilation to raise the blood pressure and the modified Queckenstedt maneuver.

The site of the fistula, when identified, was scarified with fine hooks and packed with fascia, subcutaneous tissue, perichondrium, or fat supported laterally by Gelfoam®. In some instances of OW fistula, stapedectomy was performed with placement of a wire-tissue prosthesis; in some patients with previous stapedectomies, the prosthesis was removed and the window grafted. These patients were kept at strict bedrest with head elevated for 24 to 48 hours postoperatively, instructed to engage in no strenuous activity, and to minimize Valsalva and other maneuvers with a similar effect for 3 weeks.

Patient Distribution. The population is skewed racially (90 of 91 patients are white and 1 is black) but is representative of the patients treated at the University of Iowa Hospitals. The youngest was 13 months old at the time of presentation and exploratory tympanotomy and the oldest was 75 years. The age and sex distribution reveal approximately equal numbers of men and women in most age groups (Table I). In the group under 19 years of age, the 2 to 1 predominance of boys over girls noted by Supance and Bluestone¹⁸ was not prevalent in this series.

RESULTS

Perilymph Fistulas. Two hundred fourteen exploratory tympanotomies for suspected PLF were performed on 177 patients. One hundred six primary (initial) fistulas were found in 95 ears. Including recurrences in this series, 132 PLFs were found in 91 patients. Of the 214 tympanotomies performed, 93

TABLE I.
Age and Sex at Time of Presentation.

Age (yr)	Male Patients	Female Patients	Total
<5	3	2	5
5-10	3	1	4
10-19	4	3	7
20-39	10	15	25
40-59	13	18	31
60-80	12	7	19
Total	45	46	91

TABLE II.

Site and Etiology of Primary (Initial) Perilymph Fistulas in 95 Ears.*

	No. of Ears	% of Site	% of Total
Oval window			
Spontaneous and traumatic	32	(61)	(34)
Following stapedectomy	16	(31)	(17)
Following ossiculoplasty	2	(04)	(02)
Following mastoidectomy	1	(02)	(01)
Following aural polypectomy	1	(02)	(01)
Subtotal	52	(100)	(55)
Round window			
Spontaneous and traumatic	31	(91)	(33)
Following stapedectomy	2	(06)	(02)
Following cochlear implant	1	(03)	(01)
Subtotal	34	(100)	(36)
Oval window and round window			
Spontaneous and traumatic	8	(89)	(08)
Following stapedectomy	1	(11)	(01)
Subtotal	9	(100)	(09)
Total	95		(100)

*Total perilymph fistulae in the series is 132 (106 nonrecurrent and 26 recurrent) in 95 ears of 91 patients.

were negative explorations (dry taps). Included in these negative explorations are seven patients who underwent reexploration for recurrent or persistent symptoms after previous patching of PLFs. Twenty patients who had a primary diagnosis of probable endolymphatic hydrops underwent an endolymphatic shunt procedure after an intraoperative negative exploration for PLF.

Slightly more than one half of identified fistulas occurred at the oval window and the preponderance were spontaneous and unrelated to previous ear operations (Tables II, III). In addition, six patients who underwent reexploration for recurrent or persistent symptoms after previous patching of PLFs had

TABLE III.

Recurrent Perilymph Fistulas.*

No. of Recurrences	Oval Window (No. of Ears)	Round Window (No. of Ears)
1	7	5†
2	3	1
3	2§	1‡
4	1	

*N=7 patients (8%) and 20 ears (21%). In three patients, no actual leak was seen at reexploration, but relief of symptoms was obtained after regrafting.

†One patient with possible Mondini deformity and initial findings of bilateral round window fistulas, bilateral oval window fistulas, and unilateral recurrence of round window fistula when reexplored following a drop in hearing. One patient with an oval window fistula 10 months following stapedectomy had fascia grafting of both windows, with temporary resolution of symptoms. Four months later, she underwent reexploration for recurrent vertigo, aural fullness, and a newly positive fistula test result. A round window fistula was found and both windows were regrafted, with resolution of her vestibular symptoms. She has had no hearing loss. A third patient had closure of an oval window fistula at another facility and on reexploration at the University of Iowa, a round window fistula was discovered and grafted.

§One patient with two recurrences at the oval window had his first two explorations and grafting of the oval window done at another medical center.

‡See case 8.

TABLE IV.

Location of Oval Window Fistula in Previously Unoperated Ears.

Site	No. of Ears*	(%)
Anterior	18	(67)
Posterior	4	(15)
Superior	3	(12)
Inferior	1	(03)
Cracked footplate	1	(03)
	27	(100)

*Of 40 previously unoperated ears, location was specified in 27.

negative tympanotomies. Congenital middle ear abnormalities were uncommon (four of 82 patients with documented PLF) and included malformations of the stapes, effaced round window, a stiff but mobile incus, and an abnormally wide and mobile stapediostapedial joint. In cases of OW fistulas not associated with previous otologic operations, PLF was most commonly noted anteriorly, at the site of the fistula ante fenestram (Table IV). Other operative findings include subluxation of the footplate, window adhesions, ossicular discontinuities, and a variety of stapes prostheses.

Predisposing Factors. The single most common predisposing factor in the occurrence of PLF was prior otologic surgery (Table V), which accounted for 24 patients (26%). Nineteen of these had PLFs following stapedectomy; 16 had OW fistulas; two, RW fistulas; and one, OW and RW fistulas. Only three were primary (early) fistulas, occurring within the first week after stapedectomy (presumably with no tissue seal of the OW ever forming) (Fig. 1). The remainder were secondary and developed 6 months to 19 years after stapedectomy. Prostheses used included a cup prosthesis with tissue graft, the House wire with tissue graft, and Teflon®-wire pistons.

Twenty-three percent of patients with PLFs had

TABLE V.

Predisposing Factors for Developing of Perilymph Fistula.

Factor	No. of Patients	(%)
Otologic surgery	24	(26)
Stapedectomy	19*	
Other	5	
External and internal trauma	21	(23)
Head trauma	10	
Barotrauma	8	
Direct trauma	2	
Acoustic trauma	1	
Exertion straining	10	(12)
Significant exertion	6	
Minimal exertion	4	
Mondini malformation	7*	(08)
Other congenital malformations	2	(02)
Upper respiratory infection	4	(04)
Congenital syphilis	1	(01)
Idiopathic factors	22	(24)
	91	(100)

*Includes one patient with associated middle ear deformity.

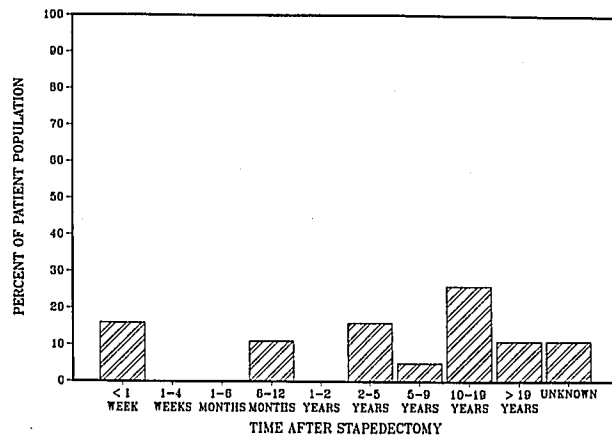


Fig. 1. Time between stapedectomy and diagnosis of PLF.

sustained direct and indirect trauma that included closed head trauma with and without documented skull or temporal bone fractures, barotrauma, direct middle ear trauma, and acoustic trauma. A history of significant exertion or straining as a precipitating factor was elicited from only six patients (7%), and an additional four noted onset of symptoms after minimal exertion, such as bending over or vacuuming (4%). Seven patients (8%) had some variant of the Mondini malformation, as shown by temporal bone tomography, while four, including one patient with associated Mondini deformity, had congenital middle ear malformations. Development of PLF was idiopathic in 24% of the cases.

Signs and Symptoms. Patients with demonstrable PLFs presented with an amazing array of signs and symptoms from sudden-and-profound hearing loss, roaring tinnitus, and whirling vertigo to only unilateral tinnitus with an isolated high frequency hearing loss or aural fullness (Tables VI, VII). Hearing loss and vestibular symptoms, if present, occurred in a variety of forms and combinations (Tables VIII, IX). Onset ranged from less than 24 hours to 23 years before diagnosis (Fig. 2). As illustrated by case 14, the apparent volume of the perilymph leak did not correlate with either the degree of hearing loss or the severity of vestibular symptoms.

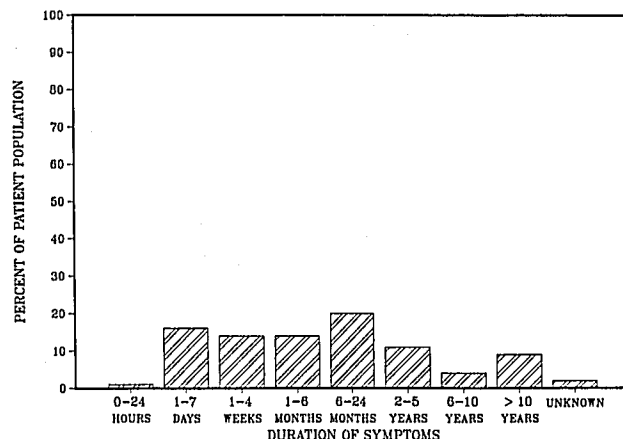


Fig. 2. Duration of symptoms prior to diagnosis of primary (initial) PLF.

TABLE VI.
Symptoms at Initial Presentation.

	No. of Patients	(%)
Hearing loss+tinnitus+vertigo	44	(48)
Hearing loss+vertigo	14	(15)
Hearing loss+tinnitus	10	(11)
Vertigo+tinnitus	4	(05)
Hearing loss	7	(08)
Vertigo	12	(13)
Tinnitus	0	(0)
	91	(100)

Hearing loss, documented in the history or the physical examination, was reported in 80 patients (84%) and included almost every kind of hearing loss from sudden-and-profound to normal pure tones with fluctuation of speech discrimination (Table IX). In some cases, a moderate hearing loss was accompanied by a disproportionately poor speech discrimination score (0% to 8%). Fluctuation of speech discrimination was a solitary feature of the hearing loss in 21% of the group; thus, fluctuating speech discrimination is an important symptom.

The equally common symptom component was vestibular, with 73 patients (80%) having a disturbance of balance at some time during the course of their illness (Table VIII). It was common for a patient to report initial severe single or multiple episodes of equilibrium disturbance which resolved partially or completely over a period of hours to months. Some patients reported occurrence or exacerbation of vertigo with straining only. Vestibular symptoms included true vertigo, a description of dysequilibrium or light-headedness, motion intolerance, and numerous combinations thereof. Two children were observed to have episodic nystagmus. It might be expected that patients with RW fistulas would be more likely to present with no vestibular symptoms. However, the absence of vestibular symptoms was not helpful in predicting site of PLFs (Table X).

Tinnitus, when present, varied from high-pitched to roaring and from constant to intermittent. It was reported in 58 (63%) patients (Table VI). In some patients it was the predominant symptom (case 8).

TABLE VII.
Additional Signs and Symptoms of Initial Presentation.

Signs and Symptoms	No. of Patients (N=91)	(%)
Aural fullness/otalgia	23	(25)
Recruitment	19	(21)
Tullio phenomenon	4	(04)
Subjective improvement in hearing or tinnitus with increased altitude	4	(04)
Positive fistula test result*	22	(24)

*Twenty-two (36%) of 61 patients tested had positive results at initial presentation; 18 of 22 were objectively positive (deviation of eyes with pneumatic otoscopy) and four were subjectively positive (sensation of dizziness with pneumatic otoscopy). Thirty-nine of 61 patients tested had negative results.

TABLE VIII.
Vestibular Symptoms.

	No. of Patients	(%)
True vertigo	13	(18)
Dysequilibrium, unsteadiness, or light-headedness	15	(21)
Motion intolerance	4	(05)
Vertigo+dysequilibrium	14	(19)
Vertigo+motion intolerance	11	(15)
Dysequilibrium+motion intolerance	5	(07)
Vertigo+dysequilibrium+motion intolerance	11	(15)
Patients with vestibular symptoms	73	(100)
Patients with no vestibular symptoms	18	
Total	91	

Other common presenting signs and symptoms recorded for a group of 61 patients included aural fullness or otalgia (25%), recruitment (21%), and a positive fistula test result (36%) (Table VII). Results of the fistula test were considered positive with deviation of the eyes or a sensation of dizziness on pneumatic otoscopy. No consistent ENG abnormality was found. Absence of any or all of these signs did not preclude the presence of a PLF, but their recurrence, persistence, or appearance in a new form was frequently associated with a persistent or recurrent PLF (Tables III and XI).

A number of patients had minimal symptoms and, in some cases, PLF was not suspected preoperatively. Several illustrative cases of patients with minimal symptoms, isolated low or high frequency hearing loss, and symptoms consistent with Meniere's disease are presented.

CASE REPORTS

Case 1

A 23-year-old woman who had a history of chronic unilateral ear disease had undergone myringotomy and tube placement at 12 years of age. An exploratory tympanotomy with removal of cholesteatoma and ossiculoplasty (autologous malleus to footplate strut) was performed at age 17. She had a resultant mild conductive hearing loss (25 dB speech recognition threshold [SRT], 100% discrimination). Five years later, on routine follow-up, she was noted to have an increase in conductive hearing loss of 5 to 15 dB at 250 through 1,000 Hz, with a decrease in SRT of 10 dB and 92% discrimination. Exploratory tympanotomy revealed an open oval window with a portion of the footplate subluxated into the vestibule. The subluxation was corrected, the

TABLE IX.
Types of Hearing Loss.

	No. of Ears	(%)
Fluctuating*	28	(35)
Severe to profound	19	(24)
High frequency	6	(07)
Low frequency	5	(06)
Other	22	(28)
	80	(100)
No hearing loss	15	
Total	95	

*Includes 14 patients with fluctuating speech discrimination as the predominant finding. Three other patients also had fluctuating speech discrimination in addition to decreased speech reception thresholds in only a portion of the frequency range.

TABLE X.
Site of Perilymph Fistulas in Patients with No Vestibular Symptoms.

	No. of Patients	(%)
Oval window	10	(56)
Round window	6	(33)
Oval and round windows	2	(11)
Total	18	(100)

OW patched with perichondrium, and total ossicular replacement prosthesis (TORP) ossiculoplasty performed. The air-bone gap was closed 5 to 20 dB in the low frequencies with a 10-dB increase in SRT and stable discrimination.

Case 2

A 42-year-old man had experienced progressive unilateral hearing loss and transient episodes of vertigo occasioned by loud noise for 3 months after an isolated incident of acoustic trauma. He was found to have a 35 dB SRT, 96% speech discrimination, and low frequency air-bone gaps ranging from 5 to 25 dB. An OW fistula was found and patched with fat, with resulting same-day audiometric improvement to 15 dB SRT. His hearing subsequently dropped to 50 dB SRT, with a primarily low frequency loss, and he continued to experience a Tullio phenomenon. Reexploration 4 months later revealed a recurrent OW fistula. He underwent stapedectomy with a placement of a fat-wire prosthesis. His hearing improved to 15 dB SRT, 92% discrimination, but he had persistent vertigo. Additional findings consisted of a positive fistula test result and positional nystagmus. At reexploration 3 months after the second procedure, he underwent grafting with fascia of a recurrent OW fistula, with stabilization of his hearing in the normal range.

Case 3

A 6-year-old girl complained of intermittent unilateral high-pitched tinnitus, being able to hear sound but not understand speech, and feeling "tippy" with imbalance to whichever side she leaned. She had been symptomatic for 5 weeks when she was first seen. At that time she had a newly documented low-to-medium frequency mixed hearing loss with 40 dB SRT, 84% discrimination. Over the course of 3 days her hearing dropped to 50 dB SRT with 24% speech discrimination (Fig. 3). She underwent exploration and patching of a RW fistula with perichondrial tissue. Her hearing returned to within normal range (15 dB SRT, 96% speech discrimination) and the vestibular symptoms subsided.

Case 4

A 38-year-old man complained of the acute onset of a constant, high-pitched, unilateral tinnitus in the right ear (AD) with occasional clanging after diving to the bottom of a pool 9 feet deep. He also noticed impairment of visual tracking to the right after the onset of tinnitus with documented abnormal saccades to the right. He had no subjective hearing loss, but the audiogram revealed an isolated high frequency hearing loss AD (Fig. 4). At exploration, a RW fistula was discovered and patched with connective tissue. Abnormal saccades resolved for 2 months, but subsequently returned. There was slight improvement in the high frequency loss, to 55 dB at 6,000 and 8,000 Hz. His tinnitus was absent only when he went into the mountains to elevations of 6,800 feet or more. He felt his symptoms did not warrant reexploration.

Case 5

A 16-year-old girl first noted the onset of constant, high-pitched tinnitus in the left ear (AS) immediately after a bout of

TABLE XI.
Adjunctive Procedures for Persistent Vestibular Symptoms.

	No.
Labyrinthectomy	4
Endolymphatic shunt	5
Middle cranial fossa vestibular nerve section	1

TABLE XII.
Serial Audiograms for Case 13.

Date	500 Hz	1,000 Hz	2,000 Hz	4,000 Hz	% Speech dB SRT Discrimination
9/78 AD	20	30	55	75	
AS	25	30	55	75	
11/80 AD	30	50	60	100	45/66%
AS	25	45	60	100	45/48%
10/83 AD	40	55	80	Not heard	50/36%
AS	50	90	110		55/40%

AD - Right ear; AS - left ear.

violent crying. She later experienced a slight hearing loss AS while playing basketball. She denied any vestibular symptoms. Her audiogram was remarkable for an isolated high frequency hearing loss AS (Fig. 5). A RW fistula AS was patched with fat. The patient was subsequently lost to follow-up.

Case 6

A 53-year-old woman noted blockage of her left ear on descent in an airplane. She was treated for middle ear effusion, without relief. She had complained of a plugged feeling and mild tinnitus AS for 2 years when she presented for treatment. She had no symptoms of dysequilibrium and had flown subsequently without additional problems. Physical examination and testing revealed normal auditory evoked potential results (ABR), objectively negative fistula results, *i.e.*, negative Hennebert's sign (no deviation of eyes on pneumatic otoscopy) and subjectively negative fistula results, *i.e.*, negative Hennebert's symptom (no subjective sensation of dizziness on pneumatic otoscopy), mild diplacusis, complete recruitment AS, and a middle and high frequency hearing loss AS (Fig. 6). An antero-inferior OW fistula was patched with fat. Initially, she had a slight decrease in hearing AS to 25 dB SRT, 90% speech discrimination. When she was seen 2 years later, she had persistent aural fullness, a constant high-pitched tinnitus, and slowly progressive hearing loss AS. In addition, she reported onset of a buzzing tinnitus AS lasting seconds and occurring after the PLF patching when she moved her head suddenly. Her hearing level and speech discrimination fluctuated over a 6-week interval with 45 dB SRT, 84% speech discrimination AS, being the poorest recorded. She had reexploration of the left ear and a fascia graft to a recurrent OW fistula and to the RW. It is too early to determine the outcome as yet.

Case 7

A 50-year-old woman complained of hearing loss, tinnitus, and aural fullness AD which started while she was swimming and diving. She experienced one 30-minute episode of light-headed dizziness. Her history included noise exposure, she had an objectively positive fistula test result, and her audiogram demonstrated a high frequency hearing loss and poor speech discrimination AD (Fig. 7). An OW fistula was repaired with fat and she had subsequent improvement in speech discrimination, but only to 76%.

Case 8

A 51-year-old woman presented with a complaint of hissing tinnitus, otalgia, and aural fullness AS for 2 weeks. She reported recurrent pressure build-up AS with a hissing, releasing sensation accompanied by light-headedness. She had no antecedent upper respiratory infection or barotrauma and denied change in

TABLE XIII.
Outcome of Perilymph Fistula Closure in Patients with Vestibular Symptoms

Vestibular Symptoms	No. of Patients	(%)
Eliminated	39	(53)
Improved	30	(41)
No change	4	(6)
Total	73	(100)

TABLE XIV.

Hearing Outcome of Perilymph Fistula Closure in Patients with Hearing Loss.

Status of Hearing	No. of Ears	(%)
Improved*	32	(49)
Improved to serviceable range†	15	(23)
Improved after ossicular reconstruction	7	(11)
Stabilized	26	(40)
Worse§	7	(11)
Subtotal	65	(100)
Incomplete documentation	15	
No hearing loss at presentation	15	
Total	95	

*Greater than 10 dB SRT or 10 dB in the affected frequencies in those patients with isolated low or high frequency hearing loss.

†SRT < 35 dB and discrimination > 80%.

§Deterioration of SRT > 10 dB or of discrimination > 10%. Five of these patients have persistent or recurrent PLF.

hearing AS. Her history included symptoms of aural fullness and hissing tinnitus AD 1 year previously for 1 week, followed by sudden complete hearing loss and severe vertigo. The balance problems resolved over a period of 4 weeks. Tomography was unremarkable and ENG showed several bouts of positional nystagmus with symmetric calorics. Fistula test results were negative (Fig. 8). At exploration, a round window fistula AS was patched with fat. She had resolution of the hissing tinnitus for 7 months. Exploration for recurrent hissing and roaring tinnitus, aural fullness, and associated light-headedness on two occasions revealed no definite fistula, but she had resolution of symptoms with repatching of the RW with tissue. Two years after the third tympanotomy, she was admitted with recurrent symptoms and scheduled for reexploration. The night before she was taken to the operating room, she experienced acute onset of vertigo with nystagmus and profound hearing loss in her only hearing ear. At tympanotomy, no fistula was seen but both windows were patched. Insufficient time has elapsed to determine the final outcome, but she can now converse over the telephone using that ear.

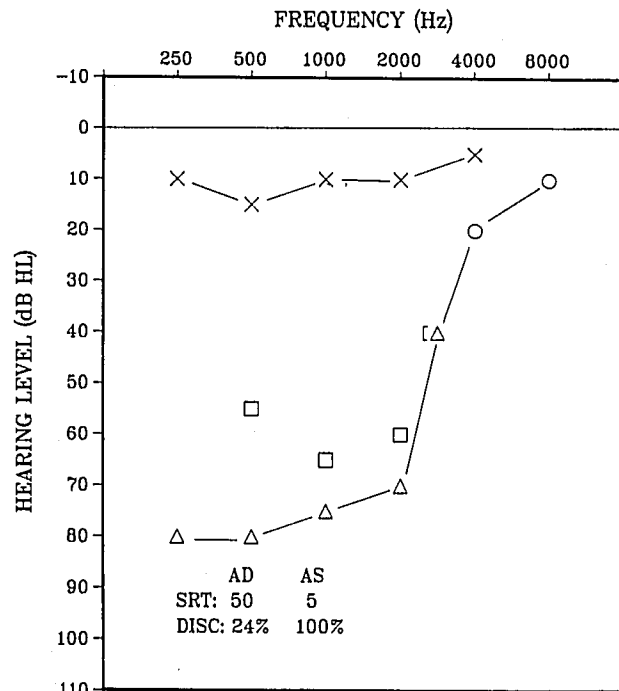


Fig. 3. (Case 3) preoperative audiogram.

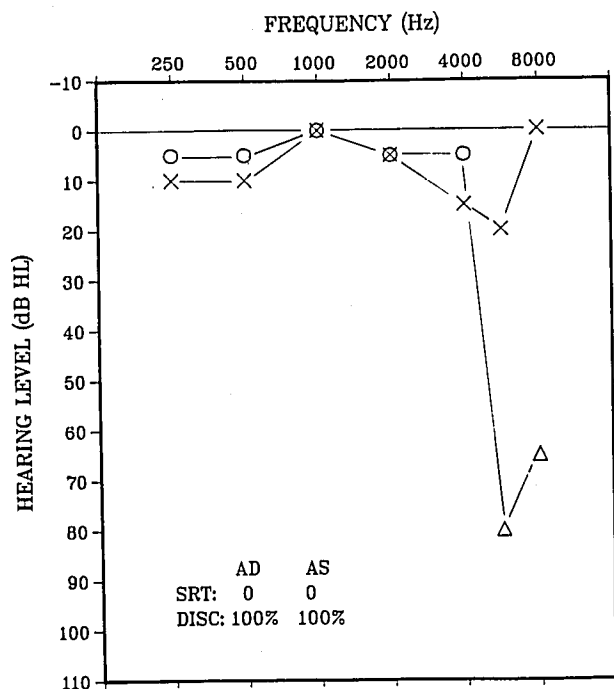


Fig. 4. (Case 4) preoperative audiogram.

Case 9

A 65-year-old man presented with a history of increasing dizziness over 10 years with nine major and 100 or more minor spells. The major spells were characterized by true vertigo lasting for 2 to 7 hours. The minor spells lasted seconds to minutes. The spells were accompanied by hearing loss and ringing tinnitus AD. He had no relief of symptoms with diuretics and a low salt diet. Fistula test results were negative and ENG showed a slow, right-beating latent nystagmus with symmetric calorics. His initial audiogram revealed a flat 70 dB sensorineural loss with 56%

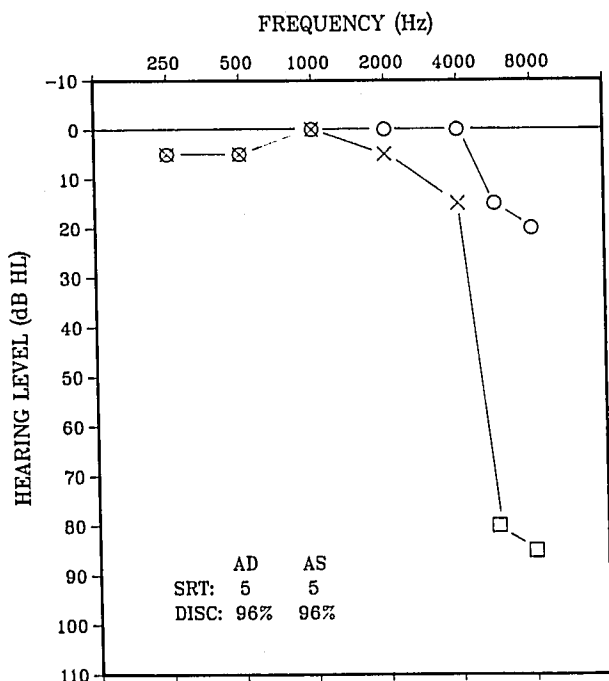


Fig. 5. (Case 5) preoperative audiogram.

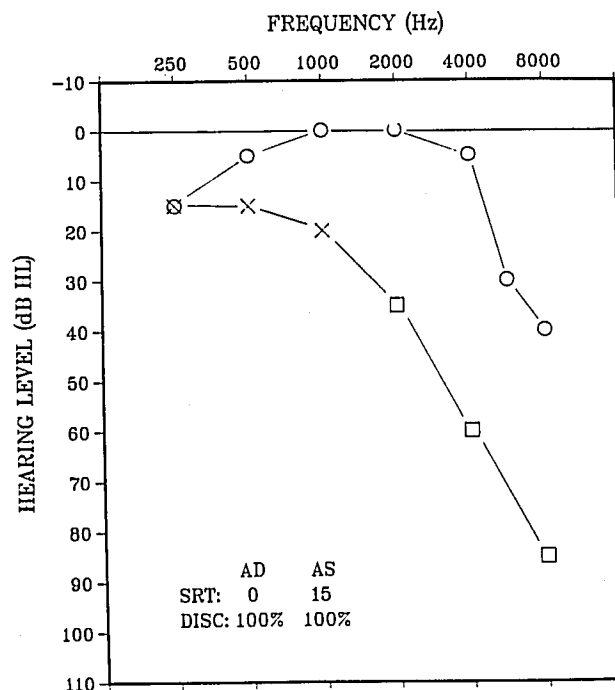


Fig. 6. (Case 6) audiogram preceding first exploratory tympanotomy.

speech discrimination AD and 25 dB SRT, 84% discrimination AS. An audiogram 2 weeks later showed a drop in discrimination AD to 28%. He underwent an endolymphatic shunt AD with the sac found in an inferior position consistent with Meniere's disease. He experienced no relief of his vestibular symptoms and complained of dizziness on swallowing. He underwent exploratory tympanotomy after reporting the new finding of a subjectively positive fistula test AD. At tympanotomy an OW fistula was identified and fat grafts were applied to both windows, with subsequent long-term relief of his spells.

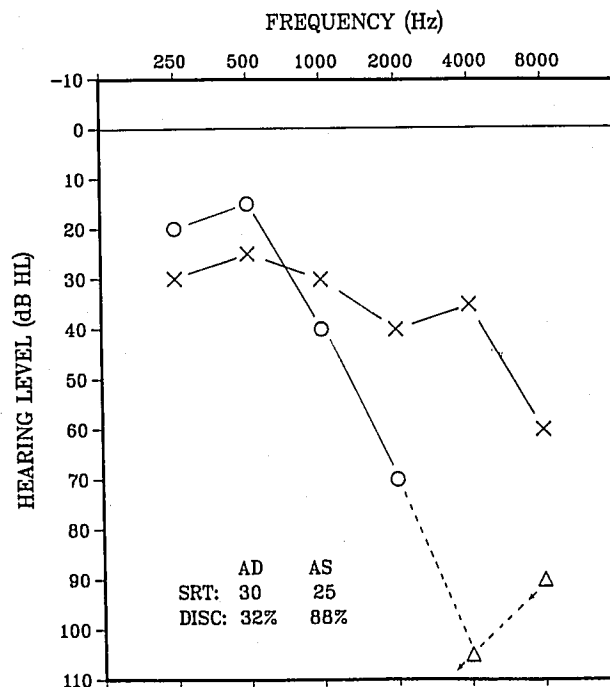


Fig. 7. (Case 7) preoperative audiogram.

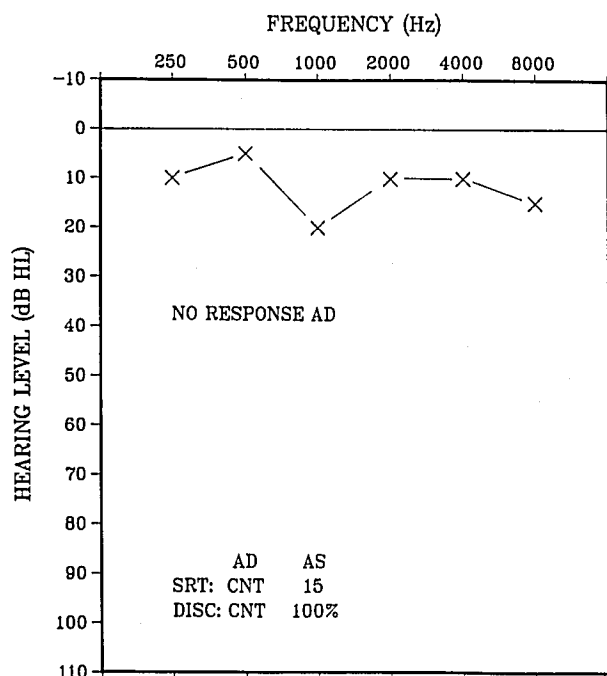


Fig. 8. (Case 8) audiogram preceding first exploratory tympanotomy.

Case 10

A 66-year-old man sustained head trauma without loss of consciousness in a motor vehicle accident. Several hours later he experienced a loss in hearing AS to unusable levels, with associated roaring tinnitus and light-headedness. He reported ataxia, motion intolerance, and episodic light-headedness over the next 18 months. Two weeks prior to presentation he awoke with no usable hearing AD; he had no associated tinnitus or vertigo. He had had gradual return of hearing AD (from 10 dB SRT, 67% to 100% speech discrimination) when first seen. At that time, in the left ear (AS), he had 70 dB SRT, 52% speech discrimination. He underwent exploratory tympanotomy AS with a fat graft to a RW fistula. Subsequently, his vestibular symptoms and subjective hearing improved, but he had a recurrence of ataxia and an SRT of 75 dB with 15% speech discrimination AS 1 month later. He presented again 5 months later with sudden hearing loss AD, which improved over the next 24 hours. At that time an audiogram showed a 45 dB SRT, 92% discrimination AD and 75 dB SRT, 20% speech discrimination AS with an improvement of 15 dB in the SRT AD after ingestion of urea. He was treated with carbogen, histamine infusion, diuretics, and Cyclospasmol® and followed with daily audiograms, which showed fluctuation in hearing level and speech discrimination in both ears (AU). He underwent exploration AD 4 days after admission with closure of an OW fistula with fascia and an endolymphatic shunt AD, with the sac found in an inferior position. Three days later, reexploration AS showed a recurrent RW fistula, which was patched with perichondrial tissue. Subsequent audiograms showed continued fluctuation of hearing AU; this patient's hearing level appeared to be very sensitive to salt ingestion. The patient had both bilateral Meniere's disease and bilateral PLFs. He had resolution of his vestibular symptoms.

Case 11

A 44-year-old man was referred for work-up of a possible acoustic neuroma. He complained of intermittent discomfort AS since a case of rheumatic fever at age 19. He developed aural fullness AS in association with an upper respiratory infection 3 months prior to presentation. He was treated with a tapering medication, possibly steroids, with no change in symptoms and referred for work-up of asymmetric hearing with reportedly abnormal ABR findings and normal mastoid x-ray films. He com-

plained of constant tinnitus AS for 2 months, and for many years had had fleeting episodes of vertigo. He had no history of ear infection, operations, or trauma. He complained of neck pain for 3 months. At presentation he had a 15 dB SRT, 80% speech discrimination, with a high frequency sensorineural hearing loss AS. A second ABR did not support the diagnosis of a compressive retrocochlear lesion. ENG caloric results were symmetrical. Posterior fossa myelogram showed symmetric filling of the internal auditory canala. Cervical spine x-ray films showed fusion of uncinata and lateral processes C7-T1 with occulta spina bifida. A cardiac evaluation revealed left atrial and ventricular enlargement. The patient was also found to be a borderline diabetic. At follow-up 1 year later, he had no additional symptoms, but a hearing loss of 45 dB, 64% speech discrimination AS. Because of the progression in hearing loss he was reexamined 6 months later. He had had several episodes of oscillopsia and 1 week of daily nonspecific dizziness in addition to progression of hearing loss to 85 dB speech detection thresholds (SDT) AS, and persistent aural fullness. A repeated ABR did not suggest a retrocochlear lesion. Head CT with contrast was unremarkable. Results of the fistula test were still negative. He underwent exploratory tympanotomy with fat patching of oval and round window fistulas AS. He had resolution of his vestibular symptoms and decrease in aural fullness, but no return of hearing. He was seen again 1½ years after the initial exploration, complained of recurrent aural fullness AS, and, for the first time, had a subjectively positive fistula test result. He underwent reexploration with a connective tissue closure of a recurrent RW fistula.

Case 12

A 14-year-old boy sustained a right temporal bone fracture at 10 months of age with self-limited bloody otorrhea and a residual hearing loss detected on a preschool audiogram. He underwent exploratory tympanotomy for conductive hearing loss AS (35 dB SRT, 96% speech discrimination) with findings of an absent incus. A homograft incus interposition from malleus to stapes was done. Follow-up audiogram demonstrated improved hearing to 9 dB SRT, 100% discrimination AD. Three years later the patient noted the sensation of air escaping AD on blowing his nose. Several days later he experienced onset of episodic vertigo, but subjectively no change in hearing. A preoperative audiogram was obtained (Fig. 9). At exploration, adhesions around the stapes

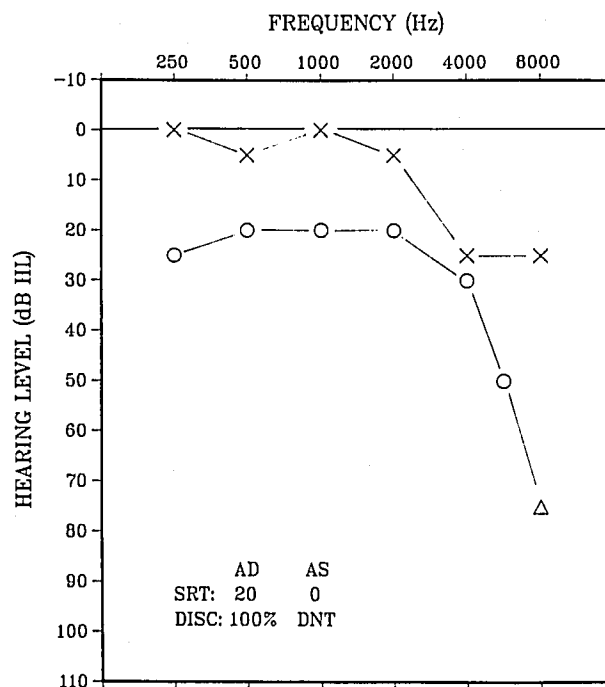


Fig. 9. (Case 12) audiogram preceding exploration for perilymph fistula.

and footplate were released and posterior OW fistula was closed with fat. Follow-up audiograms at 6 months and 1 year showed improvement of hearing to 5 dB SRT, 96% discrimination AD.

Case 13

A 9-year-old boy had a sensorineural hearing loss documented at age 2 years when an audiogram was obtained because of hearing loss found in a sibling. He had multiple ear infections treated with antibiotics, which decreased in number after age 6 to one or two infections annually. He was followed with frequent audiograms. Between the ages of 2 and 8 he had slowly progressive hearing loss AU. Over the 9 months previous to being seen, he had accelerated hearing loss AU with the more rapid drop AS. He denied vestibular symptoms, but had intermittent tinnitus AU. Outside audiograms were obtained in 1978, 1980, and 1983 (Table XII). From the time he was first seen until his ears were explored 5 weeks later, he had progressive hearing loss AD from 40 dB SRT with 56% speech discrimination to 50 dB SRT with 28% discrimination. Hearing AS was essentially stable for the period, ranging from 60 dB SRT with 16% discrimination to 55 dB SRT with 12% discrimination. Tomograms showed a possibly dilated endolymphatic aqueduct AS. On exploration, RW fistulas were found in both ears and grafted with perichondrial tissue. Six weeks postoperatively, the patient's hearing had improved somewhat, to 45 dB SRT, 52% speech discrimination AD and 55 dB SRT, 24% speech discrimination AS. When seen 3 months postoperatively, his hearing had decreased bilaterally, and it dropped further in the next weeks to 80 dB SRT, 20% AD and 95 dB SRT, 4% discrimination AS. Both ears were reexplored and bilateral OW fistulas and a recurrent RW fistula AS were found and grafted. Postoperatively, the patient had a stable hearing level but fluctuating speech discrimination.

This case suggests that the earlier a fistula is diagnosed and treated the better the prognosis (and, generally speaking, the greater the hearing loss the less the chance for significant improvement) and that fistulas have a definite predisposition for reblooming, especially in the young.

The fact that the size of the PLF may have no bearing on the extent of symptoms is illustrated by the next case.

Case 14

A 48-year-old man who worked in construction and as a pipe-fitter was referred by a neurologist. The patient had intermittent dizziness for 10 years that had become worse in recent months. He experienced sensations of floating and quivering that lasted minutes to hours, occurred up to ten times a day, and were associated with nausea. He denied any change or fluctuation in hearing, but did have aural fullness AD and intermittent buzzing tinnitus AU. The patient could initiate the symptoms by lifting heavy objects, whistling, blowing hard, or quickly turning his head. He also had a significant history of noise exposure. He reported that he had had inner ear "infections" and associated dizziness all of his life and had been treated with acetazolamide (Diamox®) and propranolol hydrochloride (Inderal®) for hypertension, angina, and migraines. Although the ENG was unremarkable, this patient said the vertigo produced during the caloric portion of the examination was dramatically different from his typical symptoms. Tympanometry performed in the course of the audiogram reproduced his typical symptoms of dizziness. His last audiogram was symmetrical, with a bilateral hearing loss at 4,000 Hz the only remarkable feature (Fig. 10). The patient underwent exploratory tympanotomy AD, and a profuse RW fistula was found. It was patched with areolar tissue and he had resolution of his episodic dizziness.

This patient was remarkable because he had no hearing loss despite the profuse nature of his perilymph leak.

OUTCOMES

Vestibular symptoms were eliminated or improved in 94% of patients after closure of the PLF (Table XIII). No patients experienced worsening of their

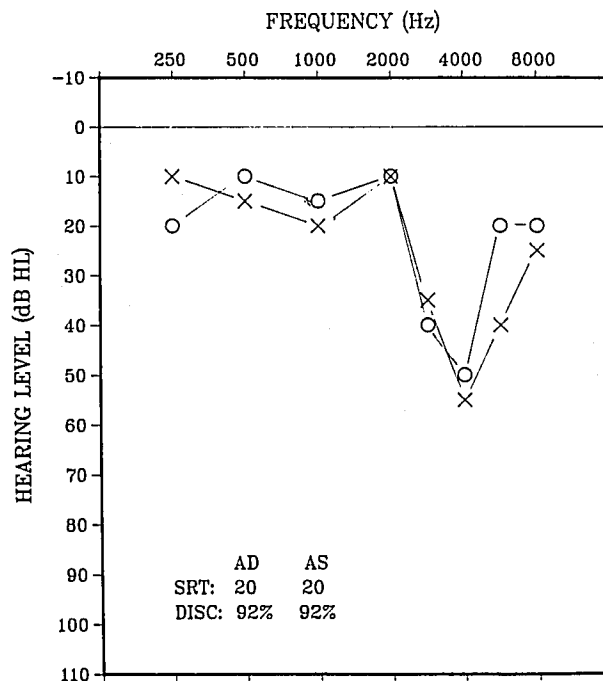


Fig. 10. (Case 14) preoperative audiogram.

vestibular symptoms. Seven patients required adjunctive procedures for persistent vertigo, with subsequent relief of their vestibular symptoms (Table XI).

Regarding hearing loss, 15 ears (17%) were omitted from the analysis because of insufficient audiologic follow-up data (Table XIV). Forty-nine percent of the ears treated showed improvement in hearing and 23% improved to serviceable levels. Serviceable hearing was considered to be SRT < 35 dB with speech discrimination > 80%. Hearing was stabilized in an additional 40%, including several stabilized at serviceable levels. Eleven percent experienced deterioration of hearing, with decreases generally 15 to 20 dB SRT with stable speech discrimination. The only two patients with significant drops in speech discrimination were subsequently reexplored and found to have recurrent or persistent PLF. New occurrence of even a minimal conductive component of hearing loss after grafting of one or both windows was found only in patients who underwent stapedectomy with tissue graft or who had a middle ear effusion at the postoperative audiometric examination.

Twenty-one percent of patients had recurrent PLF with up to four recurrences at the same site (Table III). Signs and symptoms leading to 26 positive re-explorations are noted in Table XV. The total of seven patients with recurrent fistulas does not include three patients with PLFs in the same ear, but at a different window from the initial PLF. Four of the seven patients (57%) had documented evidence of Mondini malformation. Recurrences were more likely in those ears where the PLF was initially grafted with fat (Table XVI), although no patching material was foolproof. Similar results were reported

TABLE XV.
Signs and Symptoms Leading to Reexploration* in 33 Ears.

	Incidence
Hearing loss	
Recurrent	10
Fluctuating	4
Vestibular symptoms	
Recurrent	15
Persistent	9
Tinnitus	
New	1
Recurrent	5
Aural fullness	
New	1
Persistent	1
Recurrent	4
Fistula test result positive†	
New	7
Recurrent	5

*Seven negative plus 26 positive reexplorations were done.

†One patient was explored because the fistula test result changed from subjectively to objectively positive. This patient had received grafts for an oval window fistula three times at an outside medical center. At the University of Iowa an oval window fistula was corrected by stapedectomy with placement of a fat-wire prosthesis. Vestibular symptoms resolved but the patient developed a conductive hearing loss of 10-30 dB with 96% discrimination.

by Thompson and Kohut.¹³ Subcutaneous areolar tissue is now our preferred grafting material because it drapes better than fascia, perichondrial tissue, or other materials. It is also readily available and is generally taken from a tiny incision in the postauricular crease. Patients with recurrent PLF were, of course, at increased risk for hearing loss; five patients experienced deterioration of hearing with recurrent PLF after initial stabilization or improvement of hearing following closure of the primary (initial) fistula. Only seven patients experienced deterioration in hearing, with five (71%) of these subsequently found to have persistent or recurrent PLF.

DISCUSSION

A group of patients who presented a diagnostic dilemma were those with fluctuating hearing loss and balance problems consistent with a diagnosis of Meniere's disease. In some cases, and we agonized over which ones, patients underwent exploratory tympanotomy with discovery and patching of a PLF. In case 9, the patient had a classical history for Meniere's disease; an endolymphatic shunt was

done and the endolymphatic sac was found in the inferior position typical in Meniere's disease. The patient had persistent vestibular problems and subsequently developed a subjectively positive fistula test result; an OW fistula was discovered at exploratory tympanotomy. This case raises serious questions regarding general acceptance of the (congenital) location of the endolymphatic sac inferiorly in Meniere's disease; perhaps this assumption is not warranted. Lehrer, *et al.*,¹⁹ and Simmons¹⁷ pointed out that even the glycerol test cannot provide a consistent method of differentiating cases of PLF from Meniere's disease.

Seven cases with tomographically documented Mondini deformity are included in Table XVII. Within this group, PLF recurrence was high: 57% had at least 1 recurrence, 1 had 2 recurrences, and 1 had 4 recurrences. Patient 30 underwent a subsequent endolymphatic shunt with stabilization of his hearing; patient 49 had closure of a PLF and shunting at the same procedure, and patient 53 had shunts bilaterally with exploration and closure of a PLF unilaterally. The question of indications for adjunctive endolymphatic shunt procedures in this group of patients arises. No protocols or algorithms for dealing with this set of conditions has been proposed. They are needed.

The possible additional problem of hydrops secondary to membranous labyrinthine damage caused by a PLF has been addressed by Potter and Conner.²⁰ Patients who had repair of PLF with improvement in symptoms but persistence of some dysequilibrium, as well as patients who required endolymphatic shunts for subsequent relief of their symptoms, may in fact be suffering from traumatic endolymphatic hydrops and/or from other damage to the membranous labyrinth. Development of a secondary hydrops could be due to damage to the membranous labyrinth from the initial insult causing the fistula, from the perilymph leak itself with its stresses on the membranous labyrinth or, less likely, from trauma sustained during surgical repair of the PLF.

Two groups which overlap somewhat and which deserve more detailed review are those with bilateral hearing loss, with or without documented bilateral PLF (Table XVIII), and those under 19 years of age (Table XVII). Two thirds of the patients with bilateral hearing loss were under 19 years of age, while

TABLE XVI.
Substances Used in Initial Repair of Perilymph Fistulas.

	No. of Ears	(%)	No. of Recurrences	% of Recurrences with This Tissue	% of Total Recurrences with This Tissue
Fat	41	(44)	12	(29)	(69)
Areolar tissue	20	(21)	3	(15)	(15)
Perichondrial tissue	11	(12)	2	(18)	(11)
Fascia onlay	8	(9)	2	(25)	(11)
Prosthesis and fascia	9	(10)	0	(00)	(00)
Other	5	(4)	0	(00)	(00)
Total	94	(100)	19		

TABLE XVII.
Patients Under Age 19 with Documented PLF.

Case	Age at Presentation (yr)	AD			AS			Congenital Abnormalities†	
		Hearing Loss	PLF Site	Trauma	Hearing Loss	PLF Site	Trauma	Tomograms	Exploration of Middle Ear
6	18	Stable, 20 dB	OW	Temporal bone fracture (age 10 mo) and ossiculoplasty (age 15 yrs)	None	NE	None	None	None
17	1	Fluctuating left-beating nystagmus	OW (2)	None	Fluctuating stable moderate loss	NE	None	Endolymphatic aqueduct not well seen	None
21	15	Stable, profound	NE	None	Stable	RW	Mild head trauma	None	None
23	6	Fluctuating	RW	None	None	NE	None	Normal	None
25	18	Stable, 35 dB	NE	None	Stable, 50 dB	RW	Poststapedectomy (11 yr)	None	Stapes malformation
26	16	None	NE	None	80 dB loss at 6,000-8,000 Hz	RW	Exertion	None	None
30	7	Progressive, fluctuating	RW(3), OW (1)	None	Profound	NE	None	Mondini deformity AU	None
42	4	Progressive, profound	OW, RW	None	Progressive, profound	OW	None	Mondini deformity	None
47	9	None	NE	None	None; vertigo, tinnitus	OW	Pencil in ear	None	Perforation of tympanic membrane
49	5	Progressive, profound	NE	None	Progressive, profound	RW	None	Mondini deformity AU	None
53	11	Stable, severe	NE	None	Progressive, profound	RW	None	Mondini deformity AU	None
57	9	Progressive, moderate	OW, RW	None	Progressive, severe	OW(1), OW(2)	None	Mondini deformity AS	None
68	11	Stable, mild	NE	None	Stable, moderate	OW	Mastoidectomy, ossiculoplasty	None	None
73	2½	Stable, profound	NE	None	Sudden, profound	OW (4)	None	Mondini deformity AU	None
75	1½	Stable, moderate	NE	None	Fluctuating, profound	RW	None	Mondini deformity AU	Effaced RW; stapes tilted
76	15	Progressive, fluctuating	OW	None	Progressive, fluctuating	OW	None	Normal	None
81	15	None	NE	None	None; vertigo, tinnitus	OW + RW	Barotrama	None done	None

*Demonstrated by tomography and exploration of middle ear. NE - not explored; OW - oval window; PLF - perilymph fistula; RW - round window.

only one fifth of patients were less than 19 years of age. All seven patients with Mondini deformity were under 11 years of age and had bilateral hearing loss. The high incidence of PLF in young patients with preexisting hearing loss and subsequent development of new or progressive hearing loss (Table VI) points up the need for early exploration in this group. The sudden onset of hearing loss in the unexplored ears of four of the five older patients with bilateral hearing loss suggests the possibility of undiagnosed PLF in the contralateral ears in these patients.

The percentage of ears with improved hearing after PLF closure (49%) is quite high compared with other series. Those with hearing improved to a serviceable range had been symptomatic for 3 days to 3 years. One patient was symptomatic for 23 years, yet obtained 15 to 40 dB improvement in hearing bilaterally after closure of bilateral PLFs. While improved hearing is thus possible after many years of symptoms in some patients, the need for early ex-

ploration seems clear. Case 11 is an example; the patient had 15 dB SRT, 80% speech discrimination at initial presentation, a drop to 45 dB SRT, 64% speech discrimination, and finally an 85 dB SRT at the time of exploration 1½ years after initial presentation.

Preoperative finding of an ear with no response on audiometry did not preclude a significant gain in hearing, with one patient improving to 55 dB SRT and another to 90 dB SRT with 60% discrimination. Although these are not dramatic gains, any improvement in hearing is not to be disparaged. The patient with improvement in hearing to 90 dB SRT, 60% discrimination had only 75 dB SRT, 20% speech discrimination in the opposite ear. Another patient with improvement from 75 dB, 0% discrimination to 35 dB, 60% discrimination had an audiologically dead contralateral ear. Furthermore, an early minimal improvement should not be considered final. One patient who demonstrated initial increased

TABLE XVIII.
Patients with Bilateral Hearing Loss: History and Findings.

Pt.	Age at Presentation (yr)	Site and Age at Onset of HL (yr)	Nature of Hearing Loss	SRT dB/% Discrimination	PLF Site, Adjuvant Procedure	Date of Recurrence	Symptoms Indicating Exploratory Tympanotomy				X-Ray Studies	Other History and Findings
							HL	V	T	AF		
12	66	AD 67	S	40/88%	OW, ELS	4/80	+	+	-	-		Head trauma
		AS 65	S	70/52%	RW	10/79	+	+	+	-		Sudden HL AD (9/79, 4/80)
					RW	4/80	+	-	-	-		ELS AS, sac inferior; persistent fluctuating HL AU; salt sensitive
17	1	AD 1	S	NR at 100 dB	OW, exploration for CSOM	5/81	+	+	-	-	ELA not well demonstrated	HL associated with URI
		AS 1	S (?)	NR at 100 dB	OW, NE	11/81	+	+	-	-		Localized right and left sound at 40 dB
21	15	AD 4 AS 15	S, T S	SDT 95 dB 50/88%	NE RW		+	-	+	-	None	Head trauma prior to HL AS; stabilized HL AS
30	7	AD 7	P, F	65/38%	RW, OW	10/80	+	-	+	-	Enlarged ELA	Profound HL AS associated with URI
					RW	3/81	+	-	-	-		Tinnitus and rapidly progressing HL AD
					RW	6/81	+	-	-	-		1+ reactive FTA; improved on prednisone; stabilized HL 75 dB
42	4	AD < 2	P	85 dB	Dry tap	3/83	+	+	-	-	Enlarged ELA AU; no spiral lamina apical & middle turns, right cochlea	Episodic nystagmus, ataxia, progressive HL AU
43	51	AS < 2	P, S	85 dB	OW	1/83	+	+	-	-		Hearing stabilized
		AD 50 AS 51	S None	NR 15/100%	NE RW	6/82	-	+	-	+	N	1981: AF and hissing tinnitus AD for 1 week followed by sudden profound HL AD and vertigo
						1/83	-	+	+	+		Onset (5/82) of otalgia, AF, hissing tinnitus AS with lightheadedness
					PLF not seen but symptoms relieved by patching RW	3/83	-	+	+	+		Re-exploration for recurrent symptoms
						4/84	+	+	+	+		Sudden profound HL 4/84. Can converse on phone
44	32	AD congenital	S, T	75/20%	NE							
		AS congenital	S	NR	OW	9/82	+	-	-	-	N	Possible ototoxic drug during gestation. (Sister with HL)
					OW	12/82	+	+	-	-		Result: improved hearing AS to 90 dB/56% discrimination
46	55	AD 55 AS 45	S P	55/100%	RW NE		+	+	+	-	N	Roaring tinnitus AS for 1 month with rapidly progressive to stable HL AS; hearing stabilized AD

AD - right ear; AF - aural fullness; AS - left ear; AU - both ears; CSOM - chronic secretory otitis media; ELA - endolymphatic aqueduct; ELS - endolymphatic shunt; Expl. tym. - exploratory tympanotomy; F - fluctuating; FTA - fluorescent treponemal antibody; HL - hearing loss; MEE - middle ear effusion; N - normal; NE - not explored; NR - no response; OW - oval window; P - progressive; RW - round window; S - stable; T - tinnitus; URI - upper respiratory infection; Y - vertigo.

hearing from 95 dB SRT and untestable discrimination improved to 70 dB, 20% discrimination at 10 weeks with an additional gain to 65 dB, 52% discrimination at 6 months postoperatively.

Aural fullness, reported in 25% of the patients, was often helpful in determining which ear to explore in patients with few localizing signs and symptoms.

CONCLUSIONS

Perilymph fistula is a condition which can present with a wide variety of signs and symptoms. The absence of a typical fluctuating or high frequency hearing loss, of vestibular symptoms, tinnitus, positive fistula test results, or a negative history of trauma or exertion do not eliminate the possibility of a PLF. We see no clear pattern of signs and symptoms that

TABLE XVIII (Continued).
 Patients with Bilateral Hearing Loss: History and Findings.

Pt.	Age at Presentation (yr)	Site and Age at Onset of HL (yr)	Nature of Hearing Loss	SRT dB/% Discrimination	PLF Site, Adjuvant Procedure	Date of Recurrence	Symptoms Indicating Exploratory Tympanotomy				X-Ray Studies	Other History and Findings
							HL	V	T	AF		
49	5	AD? 4	P, S, T	90-95 dB	NE						Dilated ELA AU	Result: no change; no hearing AU
		AS? 4	P, S, T	105 dB	RW, ELS		+	-	-	-		
53	11	AD < 3 AS < 3	S, T P	60/90% 95 dB	NE, ELS RW, ELS		+	+	-	-	Dilated ELA AU	Result: stabilized HL AD; improved hearing AS 65 dB/60% discrimination
57	9	AD 2	P	40/56%	RW	12/83	+	-	+	-	Possible dilated ELA AS	HL in sibling
		AS 2	P	60/16%	OW RW RW, OW	3/84 12/83 3/84	+	-	+	-		
							+	-	+	-		Initial improvement: re-explored for drop in hearing. Result: stabilized hearing AU
61	25	AD 15 AS 25	S P	NR 75%	NE OW, stapes subluxated		+	-	+	-		Profound HL AD with temporary vertigo after severe ear infection and mononucleosis (age 15 yr, 80 dB/96% discrimination). Head trauma without loss of consciousness; immediate tinnitus; rapidly progressive HL AS after 5 days. Result: improved hearing AS, 35 dB/60% discrimination
73	2½	AD > 3	Congenital (?) S, T	110 dB	NE						Dilated ELA AU. Short ELA AS, wide common crus, dilated vestibule	
		AS > 3	S, F	80 dB	OW	8/83	+	-	-	-		Myringotomy and tube AS 5/85; no MEE
					OW	12/83	+	-	-	-		Explorations for recurrent HL
					OW	3/84	+	-	-	-		
					OW	4/84	+	-	-	-		Result: improved hearing after each regrafting of PLF
75	1½	AD 1½	S, T	55 dB	NE						Enlarged ELA AU and simicircular canal AD	No speech development after 7 months
		AS 1½	Congenital (?)	85 dB	RW (also effaced RW & tilted stapes AS)		+	+	-	-		Myringotomy and tubes at 15 months; onset of episodic vertigo at 16 months. Result: stable hearing AD, improved hearing 70 dB AS
76	15	AD 7	P, F	Initial: 80/52%	OW		+	+	+	-		Progressive and fluctuating HL AU for 22 yr, episodic vertigo for 18 yr
	(Explored at 29)	AS 7	P, F	Initial: 20/100% Preop: 75/64%	OW		+	+	+	+		Subjective=fistula test AU at age 29 Result: hearing improved 15-40 dB, all frequencies AU

AD - right ear; AF - aural fullness; AS - left ear; AU - both ears; CSOM - chronic secretory otitis media; ELA - endolymphatic aqueduct; ELS - endolymphatic shunt; Expl. tym. - exploratory tympanotomy; F - fluctuating; FTA - fluorescent treponemal antibody; HL - hearing loss; MEE - middle ear effusion; N - normal; NE - not explored; NR - no response; OW - oval window; P - progressive; RW - round window; S - stable; T - tinnitus; URI - upper respiratory infection; V - vertigo.

point to exploratory tympanotomy, only a series of "flags" which, taken cumulatively in selective fashion, may be of help. Particularly notable are the negative signs, which do not necessarily eliminate the possibility of PLF:

1. A predominantly high frequency, low frequen-

cy, or flat audiometric curve.

2. Vestibular symptoms of any set nature, preponderance, or even presence (Table VII).

3. History of otologic surgery (26%), onset of external trauma such as head injury, ear injury, or barotrauma (23%) or of straining (11%).

4. An apparently clear pattern of Meniere's disease.
5. An apparently clear pattern of sudden deafness.
6. Negative fistula test results, either objectively or subjectively.
7. Very long (decades) duration of symptoms.

Although patterns in the presentation of PLF vary, the following factors are associated with a high incidence of PLF.

1. Symptoms related to hearing, which may be as minimal as fluctuation in speech discrimination alone.
2. Contralateral hearing loss, especially idiopathic contralateral hearing loss.
3. Aural fullness, especially in association with vestibular symptoms, may indicate presence of a PLF and provide useful information in determining which ear to explore.
4. Dysequilibrium with occasional spells of vertigo is the most common vestibular symptom pattern.
5. A fluctuating severe hearing loss is the single most common auditory symptom.
6. Mondini malformation.

Closure of the PLF provided elimination or improvement of vestibular symptoms in most cases and improvement or stabilization of hearing in many patients. The downside risk was low; those patients who had deterioration of hearing after exploration were few and degrees of hearing loss were generally small. The perfect material for patching PLF has not been identified, but the use of fat as the graft tissue, in our experience, has a high failure rate. We prefer subcutaneous areolar tissue for grafting because it drapes better than other tissues we have tried. Patching of both round and oval windows is recommended in all suspects because 1. even when a definite fistula cannot be confirmed, some patients obtain relief of symptoms with these measures, suggesting closure of a very small or intermittently patent PLF; 2. several patients have subsequently developed a PLF at a second site after successful closure of a fistula in the ipsilateral ear, and 3. we can identify no patients whose hearing has suffered as a result of the grafts.

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DISCUSSION

EUGENE DERLACKI, MD (Chicago, IL): I really enjoyed Doctor McCabe's beautiful presentation. The worrisome thing is that he could have just as well been discussing the signs and symptoms of Meniere's disease. There are objective signs that will help differentiate a number of these patients, but one could, on the basis of that paper (if you weren't very analytical), be exploring a lot of patients with Meniere's syndrome looking for fistulas and find that some of these negatives explorations may have been patients with pure Meniere's disorder.

BRIAN MCCABE, MD: Its very true that we frequently have a clinical presentation of perilymph fistula that looks just like that of Meniere's disorder. As a matter of fact, that's our initial diagnosis and, if the patient fails to respond to treatment, then we consider that the patient has something else. I would like to stress how we suspect the presence of perilymph fistula. We see very few gushers such as you do in Mondini's syndrome. We diagnose a fistula when we see a consistently, repeatedly shifting light reflex at one of the two windows. We suction once, and then we suction again around the window and observe. If the light reflex reappears and shifts, and does it again and again, then we accept the presence of a fistula.