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Perilymph fistula—a diagnostic dilemma

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Abstract
A retrospective series is presented of 51 cases operated on for suspected perilymph fistula. In 26 ears a fistula was identified at surgery. A positive fistula test was found to strongly indicate a perilymph fistula but was more often negative than positive in surgically demonstrated fistula ears. Other vestibular tests were found to be of little value in the pre-operative diagnosis. Ears with a surgically demonstrated fistula and sensorineural hearing loss had either flat or downward-sloping audiograms. Difficulties in diagnosing a perilymph fistula at tympanotomy are discussed. At follow-up, vestibular symptoms were found to be eliminated or improved in 96 per cent of cases with surgically demonstrated fistulae and in 68 per cent of cases in which no fistula was detected at tympanotomy but hearing improved significantly in only one ear (4 per cent) of the former group and in five ears (20 per cent) of the latter group.

Introduction

Since Fee's (1968) and Simmon's (1968) first descriptions of window fistulae unrelated to stapedectomy, numerous reports have been published concerning this entity. Despite these, diagnosis of perilymph fistulae continue to present great problems to the practicing otologist. Even at surgery, identification of a perilymph fistula is often very difficult and misdiagnosis is possible.

We present our experience during a 12-year period with special emphasis on pre-operative diagnosis, operative and follow-up findings.

Patients and methods

Medical records of all patients who had undergone tympanotomy for suspected perilymph fistulae between 1978 and 1989 were studied. During this period, 54 patients were operated on in the Department of Otolaryngology, University Hospital of Kuopio and two in Central Hospital, Mikkeli. Data from two patients were missing and one patient had failed to attend the follow-up examinations; these three patients were excluded. Thus, the material studied consists of 51 patients, each having a follow-up of at least six months. The mean follow-up period was 1.8 years.

The decision to carry out an exploratory tympanotomy was based on history, clinical, audiological and vestibular evaluation. Results of the fistula test were considered positive with deviation of the eyes on pneumatic otoscopy. In Hallpike caloric tests, a side difference of 25 per cent or more was regarded as indicating a canal paresis. A pneumatic otoscope was used for testing Hennebert's sign both with positive and negative pressure changes. The eye movements were recorded in a dark room with electronystagmography (ENG) and the electrodes were placed for recording horizontal nystagmus. The patients were asked to keep the eyes closed. The test was considered positive if nystagmic beats were seen with pressure changes. When a positive test was observed, the test was also performed on the other ear.

At surgery, performed in most cases under general anaesthesia, the site of the fistula, when identified, was carefully scarified and packed with temporalis fascia held in position by Gelfoam.® If no fistula was identified, both the oval and round windows were packed with fascia, after the bony rims of the windows were first meticulously de-epithelized. In three cases, however, only the round window was sealed.

Hearing results were judged from the audiogram obtained one day before surgery and the most recent post-operative audiogram. Hearing was scored as 'improved' if there was at least a 20 dB improvement in the 500 to 2,000 Hz average.

The post-operative balance results were recorded as 'cured', 'improved', or 'no change'. The descriptor 'improved' was used for patients who post-operatively were significantly better but still had minor degrees of spatial disorientation but no longer affected their life-styles.

The chi-square test was used for the statistical analysis of the results.

Results

Age and sex distribution of the patients is given in Table I. The youngest patient was seven and the oldest 66 years old.

The possible aetiological factors are presented in

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TABLE I

Age (years)	No. of patients Male	Female	Total
<10	–	1	1
10–19	2	–	2
20–39	8	7	15
40–59	14	11	25
>60	4	4	8
Total	28	23	51

Table II. Head trauma was the most common factor. A history of significant exertion or straining as a precipitating factor was elicited from only seven patients (14 per cent). The time interval between the onset of symptoms and surgery varied from two days to more than 20 years. All post-stapedectomy fistulae were secondary, developing three months to 15 years after the primary operation.

At surgery, a perilymph fistula was found in 26 ears (51 per cent). There were 18 fistulae in the oval window, five in the round window, one in the fissula ante fenestram, and two ears with fistulae in both oval and round windows. Further in this paper the patients with surgically demonstrated fistula are named as Group A and those with negative finding in tympanotomy as Group B.

Audiological findings

Pre-operative hearing levels are shown in Table III. The number of totally deaf ears was relatively large (18, or 35 per cent). Patients in Group A with sensorineural hearing loss had either high frequency loss or flat audiogram curve (Table IV). In Group B there were four ears with low frequency loss.

Most ears (86 per cent) did not undergo any change post-operatively (Table V). Surprisingly, there were in Group B five ears and in Group A only one ear, in which hearing improved significantly (20 dB or more). In only four ears (8 per cent)—one in Group A and three in Group B—hearing had improved to a socially acceptable level (air conduction thresholds 30 dB or better at frequencies 0.5, 1, and 2 kHz). On the other hand, hearing became worse in only one ear many years after surgery. This patient had been operated on because of round window fistula and had a 65 dB sensorineural hearing loss in the affected ear both pre-operatively and immediately after surgery. She returned after six years suffering from severe vertigo and a 95 dB sensorineural loss in the same ear. At this time, labyrinth destruction was performed and vertigo was eliminated.

TABLE II
POSSIBLE AETIOLOGICAL FACTORS

	No.	(%)
Otological surgery		
Stapedectomy	6	12
Mastoidectomy	1	2
Head injury		
with fracture of the temporal bone	12	24
without fracture	4	8
direct (penetrating)	1	2
Barotrauma	2	4
Exertion or straining	7	14
No obvious factor	18	35
Total	51	

Vestibular findings

At presentation 48 patients (94 per cent) had vertigo or dizziness. Vestibular symptoms included true vertigo, a description of dysequilibrium or lightheadedness, motion intolerance, and numerous combinations of these. Pre-operative vestibular examination showed that the fistula test was significantly more often positive in Group A than in Group B (Table VI). Investigation for spontaneous nystagmus and Hennebert's sign, and the results of bithermal caloric tests revealed no significant differences between the two groups.

At follow-up (Table VII) vestibular symptoms were found to be eliminated or improved in 96 per cent of patients who had a surgically proved fistula (Group A) and in 68 per cent of patients with no apparent fistula (Group B); the difference was significant ($p<0.01$). No patients experienced worsening of their vestibular symptoms.

Of the patients who did not benefit from surgery, one was a 62-year-old man who had sustained a fracture of the temporal bone. At tympanotomy, an oval window fistula was packed. Four years post-operatively he still has motion intolerance. Two other patients had both suffered from sudden sensorineural hearing loss with vertigo. At tympanotomy no fistula was detected but both windows were sealed. These two patients have still intermittent vertigo, and despite repeated neurootological examinations their definite diagnosis has remained obscure. The remaining five patients suffered from sensorineural hearing loss and episodic vertigo. In two patients Hennebert's sign and in one patient fistula test was positive and in the remaining two patients vertigo was provoked by physical exercise or straining. Audiologically, the lesion was cochlear and in vestibular studies canal paresis was seen. Glycerol tests performed twice in four and once in one patient were negative.

Re-exploration

Seven patients (13.7 per cent)—all in Group A—required a second tympanotomy, the interval between the two procedures ranging from one month to six years. The most common indication (six cases) was vertigo; three patients had persistent vertigo after the first operation and three developed a recurrence after six months. One patient was re-operated because of poor hearing after repair of post-stapedectomy oval window fistula.

Of the cases requiring re-exploration, five were oval

TABLE III
HEARING PRE-OPERATIVELY

Hearing loss	Group A	Group B	Total
Conductive or combined	4	–	4
Sensorineural			
<30 dB	3	3	6
30–60 dB	2	4	6
>60 dB	6	11	17
Totally deaf ear	11	7	18
Total	26	25	51

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TABLE IV
CONFIGURATION OF THRESHOLD AUDIOGRAMS IN 47 PATIENTS WITH SENSORINEURAL HEARING LOSS

	Group A	Group B	Total
Downward	3	4	7
Flat	7	9	16
Upward	–	4	4
Severe to profound	1	1	2
Deaf ear	11	7	18
Total	22	25	47

window and two round window fistulae. There was a recurrent perilymph leak in one of the re-explored ears.

All six patients with vertigo who underwent re-exploration had their symptoms either cured (4) or improved (2).

Discussion

We have found the diagnosis of perilymph fistula to be a very difficult task. A positive fistula test strongly indicates a fistula but in our patients with apparent fistulae it was more frequently negative than positive; Weider and Johnson (1988) have reported the same observation. Ruben and Yankelowitz (1989) reported that in children the fistula test had a sensitivity of 80 per cent and a specificity of 38 per cent. In this study the Hennebert's sign was helpful in some cases, but it seems to give both false negative and false positive results. Some authors (*e.g.* Glasscock *et al.* 1987; Singleton *et al.*, 1978) have reported that a reduced vestibular response in bithermal caloric testing was the most reproducible finding on ENG. We found a canal paresis in nearly equal numbers of our patients with an apparent fistula (39 per cent) and in patients without (33 per cent). Hence, we agree with Weider and Johnson (1988) that ENG is not particularly helpful in diagnosing a perilymph fistula.

Our patients with surgically documented perilymph fistulae and a sensorineural hearing loss had either flat or downward sloping audiograms but none had a pure low frequence loss. The number of these cases is, however, so limited (10) that no conclusion can be drawn. Seltzer and McCabe (1986), Glasscock *et al.* (1987) and Muntarbhorn and Webber (1987) have found that neither the severity nor the pattern of hearing loss predict which patients will have a perilymph fistula.

So, there is a great demand for a reliable method with high sensitivity and high specificity to diagnose a perilymph fistula. The decision to operate should be based on adequate historic data combined with clinical, audiological and vestibular findings instead of a single test.

The differential diagnosis between early stage Menière's disease and perilymph fistula is sometimes difficult. We have packed the round and oval windows with unsuccessful outcome in five patients and successfully in one patient who had symptoms related to Menière's syndrome. According to Weider and Johnson (1988), 'what appears to be a straightforward history of Menière's disease may be Menière's disease, Menière's disease plus a perilymph fistula, or a perilymph fistula only'. A tympanotomy is a short operation with little morbidity, and, in our opinion, it is better to operate on some cases unnecessarily than leave to a patient without care, because early surgery of a perilymph fistula may save the hearing (Glasscock *et al.*, 1987).

TABLE V
HEARING OUTCOME AT THE END OF FOLLOW-UP

	Group A	Group B	Total
Improved	1 (4%)	5 (20%)	6 (12%)
Unchanged	24 (92%)	20 (80%)	44 (86%)
Worse	1 (4%)	–	1 (2%)
Total	26	25	51

Identification of a perilymph fistula even at tympanotomy can be very difficult. Naturally, a post-stapedectomy oval window fistula and a fracture in the footplate are often easily recognizable but especially a fistula in the round window is not usually visible because the round window membrane is directed postero-inferiorly in most cases. In our early cases we drilled the bony overhang of the round window but often failed to visualize the membrane. Later we have abandoned this drilling technique for fear of inner ear damage. Most often the diagnosis is based on the accumulation of clear fluid after repeated suctions in the window niche. This finding may sometimes lead to a false diagnosis because the fluid can be local anaesthetic or serous tissue fluid when adhesions are lysed. On the other hand, when no perilymph fistula or leakage can be seen, many authors recommend increasing the intracranial pressure by jugular vein compression or Valsalva manoeuvre. Early in this series we used these methods in some doubtful cases but they gave no help.

The present series shows that at tympanotomy overdiagnosing is not so frequent as underdiagnosing. According to Seltzer and McCabe (1986) and Shelton and Simmons (1988), a sizeable number of perilymph fistulae are intermittent and this intermittence may explain the improvement seen after treating ears in which no perilymph leak was seen.

At tympanotomy, a most meticulous exploration of the whole tympanic cavity is essential, as illustrated in one of our cases, in which a perilymph leak from the fisula ante fenestram was found. Earlier, Williams and Keene (1986) have reported a similar case. In addition, according to Kamerer *et al.* (1987), microfissures in the vicinity of the round window niche and the oval window niche may be a source of perilymph leakage.

We have used fascia to seal the perilymph fistula. In one case at two re-explorations the fascia graft was

TABLE VI
RESULTS OF PRE-OPERATIVE VESTIBULAR EXAMINATION

	Group A (n = 26)	Group B (n = 25)	Significance of difference between groups
Spontaneous nystagmus			
—present	10/23 (43%)	5/25 (20%)	NS
—not detected	13/23 (56%)	20/25 (80%)	
Fistula test			
—positive	7/21 (33%)	2/25 (8%)	P<0.05
—negative	14/21 (67%)	23/25 (92%)	
Hennebert's sign			
—positive	2/7 (29%)	3/12 (25%)	NS
—negative	5/7 (71%)	9/12 (75%)	
Caloric tests			
—canal paresis	7/18 (39%)	6/18 (33%)	NS
—symmetric	11/18 (61%)	12/18 (67%)	

NS = not significant.

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TABLE VII
OUTCOME OF 48 patients with vestibular symptoms

	Group A No.(%)	Group B No.(%)	Total No.(%)
Cured	14 (54)	10 (45)	24 (50)
Improved	11 (42)	5 (23)	16 (33)
Unchanged	1 (4)	7 (32)	8 (17)
Total	26	22	48

found to have totally disappeared. Other material, such as fat, vein graft, perichondrium, and subcutaneous areolar tissue, have been used by many authors but usually with a rather high rate of recurrence (Thompson and Kohut, 1979; Potter and Conner, 1983; Seltzer and McCabe, 1986; Muntarbhorn and Webber, 1987). Probably careful elevation of the mucosa around the fistula site prior to graft insertion is the most important step in this operation. The use of cryoprecipitate glue, as suggested by Weider and Johnson (1988), may be helpful in stabilizing the graft tissue.

Controversy exists among otologists concerning the treatment of cases in which no fistula can be detected at tympanotomy. Most authors recommend to seal both windows. Pappas and Schneiderman (1989) and House and Rizer (1989) disagree, the former authors because they have observed that packing of autogenous tissue into the area of the ear windows may further dampen hearing and the latter authors consider it useless. We have found no complications related to surgery, and because of a good outcome in some patients with no apparent fistula, we think it is wise to patch the oval and round windows in patients undergoing tympanotomy for a suspected perilymph fistula, even if no fistula is found.

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