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# Perilymph fistula

## Four year experience with a new audiometric test

*by*

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The phenomenon of a position-dependent fluctuation of hearing loss, in patients subsequently found to have perilymphatic fistula, is reported. Eleven patients with rupture of the oval or round windows of the labyrinth showed a lowering of thresholds on audiometry after lying horizontally for 30 minutes with the affected ear uppermost. It is suggested that air had entered the membranous labyrinth, interfered with transmission of footplate vibration to the perilymph column, and was displaced by this manoeuvre to produce a small, but measurable, hearing gain. The diagnostic and prognostic significance of this finding is discussed.

### Introduction

Since our first report of a position-dependent fluctuation in hearing loss in four cases of perilymph fistula (Fraser and Flood, 1982), we have gained greater experience in its use as a diagnostic test and recent publications have provided supportive evidence to our original suggestion of the underlying mechanism. It was suggested that perilymph leakage into the middle ear might be associated with a retrograde passage of air into the labyrinth, altering the hydraulic efficiency of the stapedial footplate. This air might be expelled by appropriate positioning of the patient, thus eliminating some of the conductive hearing loss.

### Method

The audiometric test is performed as follows:

(a) A pure tone audiogram, measuring right and left air conduction thresholds, and a speech audiogram are obtained.
(b) The subject lies horizontally for half an hour with the affected ear uppermost.
(c) The pure tone and speech audiograms are then repeated with the patient remaining in this position.

A minimum hearing gain of 10 dB in at least two frequencies on pure tone audiometry is termed a positive result. Insufficient data are available, as yet, to determine suitable values for alterations in the speech audiogram. An artefactual learning-factor may apply in the latter testing. Care must be taken that pressure on the undermost, non-test ear does not kink the meatus and so alter the level of masking provided. Over 100 such tests have been performed in the four years 1979–1982. The most marked fluctuation associated with a perilymph fistula, which also provided the best hearing result with closure, is described.

### Case history

A 45-year-old man presented with a 10-year history of progressive right-sided hearing loss with minimal vertigo. This was of gradual onset, with no history of precipitating trauma. Otoscopy showed no abnormality, but tym-



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panometry showed increased compliance on the right (1.6 cc of air) with normal compliance on the left. Pure tone audiometry (Fig. 1) showed a mixed conductive/sensorineural deafness. Nevertheless, stapedial reflexes were obtained ipsilaterally at 100–115 dB thresholds. It is notable that these reflexes were reversed, i.e., an acoustic stimulus produced increased compliance. Tomography showed symmetrical widening of both internal auditory meati but was felt to be within normal limits.

Vestibular examination showed no abnormality but for a spontaneous first degree nystagmus to the left on E.N.G. testing. There was no positional nystagmus and the fistula test was negative.

Positional audiometry showed an improvement of up to 20 dB at low frequencies on pure tone audiometry (Fig. 2) and an increase in maximal speech discrimination score from 60 per cent to 95 per cent (Fig. 3).

At tympanotomy there was a large defect in the round window membrane with a moderate leak of perilymph. The oval window was normal and the ossicular chain intact and mobile. A fat graft was applied to plug the round window niche and, by three months after the operation, there was no vertigo and the air-bone gap had closed to within 10 dB (Fig. 4).

## Results

A perilymph fistula was found at tympanotomy in 14 patients who had undergone postural audiometry (Table I). Eleven patients had given a positive test result (true positive) and 3 a negative result (false negative). Case 15 developed a sudden sensorineural deafness of some 70 dB with vertigo during ascent from a dive. One week later he gave a positive test result and fluid meniscus was observed on otoscopy. Unfortunately he declined tympanotomy and is lost to follow-up. Eight patients giving a negative test result showed no perilymph leak on exploration (true negative). No false positive results have been obtained on pure tone audiometry (Table II).

Of the 11 true positive tests, 10 showed an improved pure tone audiogram; in 6 this was accompanied by increased speech discrimination. Improved thresholds tended to occur in the low frequencies (below 2 kHz), and most marked in those with a flat audiogram and a notable conductive component. One patient showed a subjective improvement in speech

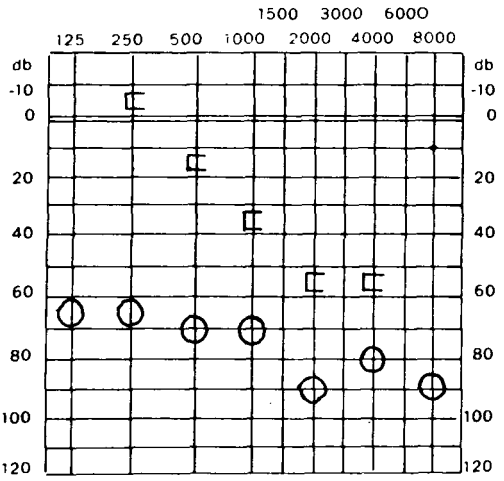



FIG. 1

Pre-operative pure tone audiogram
○ masked air conduction
⊏ masked bone conduction

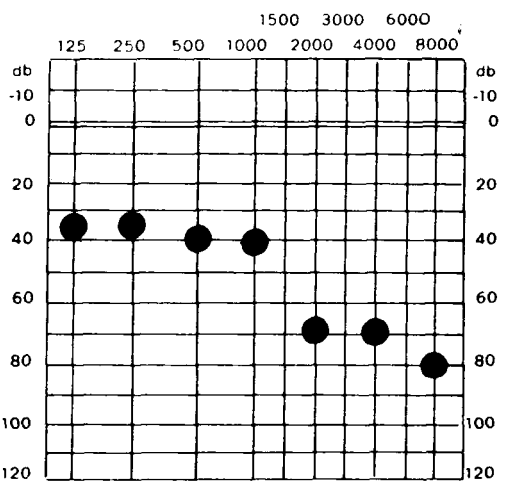



FIG. 2

Positional pure tone audiogram
● masked air conduction after 30 minutes lying with right ear uppermost.

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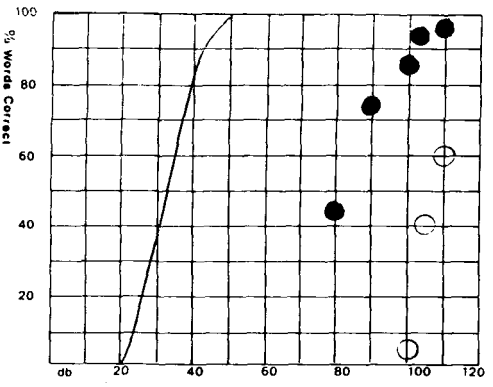



FIG. 3.
Positional speech audiogram, masked
○ pre-positional
● post positional

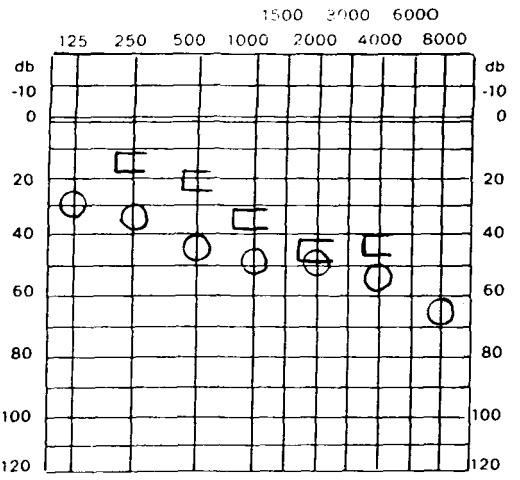



FIG. 4.
Pure tone audiogram 3 months after closure of perilymph leak
○ masked air conduction
⊏ masked bone conduction

TABLE I
AUDIOMETRIC RESULTS IN PATIENTS WITH CONFIRMED PERILYMPH FISTULA

| Case | Precipitating factor | Duration history | PTA test | Speech audio | Site of leak | Hearing after closure |
|---|---|---|---|---|---|---|
| 1 | Stapedectomy | 12 years | + | − | OW | Deteriorated |
| 2 | Idiopathic | 6 years | + | − | RW | Improved |
| 3 | Stapedectomy | 2 years | + | + | RW | Improved |
| 4 | Lifting | 2 months | − | + | OW | Unchanged |
| 5 | Idiopathic (sudden) | 2 months | + | + | RW | Unchanged |
| 6 | Idiopathic (sudden) | 2 years | + | + | RW | Unchanged |
| 7 | Idiopathic | 2 years | + | − | RW | Unchanged |
| 8 | Barotrauma | 9 years | + | − | RW | Improved |
| 9 | Idiopathic (sudden) | 1 year | + | + | RW | Unchanged |
| 10 | Idiopathic (sudden) | 2 years | + | + | RW | Unchanged |
| 11 | Idiopathic | 3 years | + | + | RW | Improved |
| 12 | Idiopathic | 8 years | − | − | RW | Unchanged |
| 13 | Syringe | 2 years | − | − | RW | Deteriorated |
| 14 | Head injury | 15 years | − | − | RW (bilaterally) | Unchanged |
| 15 | Diving | 1 week | + | − | No tympanotomy | — |

intelligibility and increased speech discrimination scores, without alterations in pure tone audiometry. The false negative tests occurred in two patients with islands of hearing preservation only and one with a sloping high frequency loss.

Of the 14 patients with proven perilymph fistula, 11 had troublesome vertigo associated with their hearing loss. Vestibular abnormalities were found in 12. Spontaneous nystagmus on E.N.G. was detected in 10; and postural nystagmus with the affect ear uppermost, in 4.

With closure of the leak there was subjective improvement in vertigo in all the patients affected. Only 4 showed a gain in hearing, however, all in the true positive group. Further deterioration of hearing occurred in

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TABLE II
CORRELATION BETWEEN AUDIOMETRIC TEST RESULTS AND TYMPANOTOMY FINDINGS.

| Number of Tympanotomies | Findings | Test Result |
|---|---|---|
| 22 | 14 Leaks | 11 positive<br>3 negative |
| | 8 no leaks | 0 positive<br>8 negative |

2. Case 13 lost all hearing in the only hearing ear and is currently being evaluated for insertion of a multichannel intra-cochlear electrode.

## Discussion

We have previously suggested (Fraser and Flood, 1982) that this fluctuation of hearing is related to displacement of air from the cochlea and elimination of the 'cochlear-conductive' loss produced.

There is evidence for entry of air into the vestibule in labyrinthine window rupture. Pashley (1982) reported radiological demonstration of air in the labyrinth in a 4-year-old boy with simultaneous oval and round window ruptures. Tomography, in the face-down position, showed an air-filled posterior semicircular canal. This was associated with diurnal and even momentary fluctuations in auditory thresholds, too rapid to be ascribed to alterations in cochlear hair cell physiology.

Increased compliance was noted in two cases in this series and this may reflect the effect of an air bubble deep to the stapes footplate producing increased mobility. Markedly increased compliance has previously been described in post-stapedectomy fistulae (Moon and Hahn, 1978) and spontaneous labyrinthine window ruptures (Goodman and Morioka, 1978). The air-filled or 'empty' cochlea is sometimes found at tympanotomy, or bubbles may be seen in the perilymph (Grundfast and Bluestone, 1978).

Entry of air into the vestibule may affect hearing. In 1929 Meurman suggested that an air bubble in the perilymph was responsible for the nystagmus which he observed during experiments on artificial membrane ruptures in rabbits. He proposed that it might be displacing the macula of the utricle. Air in the vestibule will hamper transmission of ossicular vibration to the perilymph, producing inefficient stapedial excursion. Goodhill (1967) described a conductive hearing loss as a manifestation of a post-stapedectomy fistula. He suggested that leakage or backsplash of perilymph interfered with the transducer mechanism of the piston.

This effect would be even greater where the footplate is no longer in contact with a relatively incompressible liquid but must transmit its vibration through a highly compressible gaseous medium.

Most descriptions of a rupture of the oval or round window membrane convey an image of a steady trickle of perilymph into the middle ear, constantly replenished from the C.S.F., and this is certainly the picture seen in the recumbent patient during stapedectomy or exploratory tympanotomy. However, should middle ear pressure exceed that of the perilymph, air will tend to enter in a retrograde fashion. Indeed, this is the mechanism of the 'aerodynamic implosive fistula mechanism' described by Goodhill (1981) as producing membrane rupture. Air in the cerebral ventricles is a common radiological finding in cases of C.S.F. rhinorrhoea or otorrhoea.

The recumbent cerebrospinal fluid pressure is 80–150 mmHg, yet perilymph loss during stapedectomy is usually insignificant. This is because pressure equalization through the cochlear aqueduct and along the meningeal sheaths in the internal auditory meatus is usually slow (Weisskopf *et al.* 1978). Myers (1973) has demonstrated that compression of both jugular veins in monkeys produces a rise in perilymph pressure only after a 20 second interval and notes that, when 'gushers' are encountered, elevating the head reduces perilymph flow. The erect posture thus favours air entry into the cochlea. In such a position, the pressure of the cerebrospinal fluid may be as low as 85 mm C.S.F. at the level of the cisterna magna (Langfitt, 1975). The transmission of alterations in C.S.F. pressure to the perilymph becomes slower with the progressive obliteration of the lumen of the cochlear

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aqueduct with age. Patency can be demonstrated in 82 per cent of children under 16 years of age but in only 30 per cent of the over 60s (Wlodyka, 1978). Certainly spontaneous perilymph fistula seems to be an affliction of the younger patient. Singleton *et al.* (1978) found the average age to be 46.6 years in 34 fistula cases.

Despite much research, diagnosis of perilymph fistula remains difficult. In our 12 patients with a positive test result, a history of antecedent trauma or surgery was found in less than half. Four patients had suffered a sudden spontaneous hearing loss, whilst three showed a progressive hearing loss of between 2–6 years duration. Blair-Simmons (1978) could find no precipitating factors in one-third of his cases. We did not demonstrate the predominance of left ear fistula previously reported (Goodhill *et al.*, 1973). In this series 7 right, 7 left and 1 bilateral fistulae occurred.

Vestibular abnormalities were evident in all but three of our patients. Postional nystagmus was found in 26 of 34 fistula cases studied by Singleton *et al.* (1978). This was characterized by a short latency and long duration occuring with the affected ear undermost. Spontaneous nystagmus on electronystagmography was a more common finding in our series. The E.N.G. fistula test uses the impedance bridge to alter external canal pressure and electronystagmography to detect nystagmus. Of 74 patients tested by Daspit *et al.* (1980), 6 underwent tympanotomy after giving a positive result and 5 proved to have a fistula.

Postitional fluctuation did not indicate a better prognosis for hearing gain with closure. Only 4 of the 11 true positive cases maintained an improvement in the audiogram after surgery. In isolation a tear in the round window produces little hearing loss. Thus removal of the round window membrane in the guinea pig produces a maximum 10 dB elevation of threshold estimated with the cochlear microphonic (Weisskopf *et al.* 1978). The greater hearing loss usually encountered must be the result of further cochlear damage. The pressure producing labyrinthine window rupture may be associated with inner membrane tears allowing a toxic mixture of endolymph and perilymph. Such defects or 'holes in the cochlea' may be demonstrated as islands of hearing loss, if pure tone audiometry is performed in 100 Hz increments at low frequencies (Blair-Simmons 1978).

It has been suggested (Pashley, 1982) that, in those subjects who have an anatomical abnormality predisposing to fistula, little force is needed to produce a labyrinthine window rupture, there are fewer vestibular signs and the prognosis for hearing is better. Though it has been our impression that early closure improves prognosis (Fraser and Harborow, 1975), hearing gains were achieved in patients with histories of between 2–9 years in this series.

Tympanotomy remains the mainstay of diagnosis. Pitfalls may still occur, however. Mucous membrane folds may be found in the round window niche in 55 per cent of temporal bones (Stewart and Belal, 1981), often producing a well-defined curtain lateral to the membrane and confused for it. Disturbance of these folds to visualize the membrane may disrupt it, as connecting strands commonly occur. Iatrogenic fistula may also be produced during drilling of the superior and anterior walls of the niche to gain access to the membrane. Local anaesthetic infiltration should not be mistaken for perilymph.

In over 100 tests there have been no false positive results but three false negatives using the criteria described, and this procedure should find a place in the investigation of patients suspected of having perilymphatic fistula. The phenomenon of cochlear air entry may yet provide the solution to a reliable non-invasive diagnosis of labyrinthine window defects. Could the presence of air in the labyrinth be demonstrated radiologically, e.g. with high resolution computerized tomography, it would be pathognomonic of a fistula.

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