The Postural and Cognitive Disabilites of Chronic Perilymph Fistula (PLF) after Mild Head Trauma

Jeremy Hornibrook1,2,3*
1Department of Otolaryngology–Head and Neck Surgery, Christchurch Hospital, New Zealand
2University of Canterbury, Christchurch, New Zealand
3University of Otago Christchurch School of Medicine, New Zealand

Abstract

Perilymph Fistula (PLF) is a leak of perilymph at the oval or round window. It was first recognized in the early days of stapedectomy surgery as causing balance problems before sealing of the prosthesis with natural tissue became routine. It then became apparent that head trauma and barotraumatic trauma from flying or diving could be a cause of PLF. Large institutional published case series and descriptions of “spontaneous” PLF with no trauma history led to skepticism and a bitter debate between “believers” and “non-believers”. The main criticisms were a lack of reliable symptoms and diagnostic tests and operative traps in distinguishing leaking perilymph from local anaesthetic. A close examination of the literature reveals a disparity of definitions and assumptions on symptoms, particularly vestibular. Acute PLF is diagnosis that will easily come to mind for a patient presenting with aural barotrauma from diving or flying, an airbag explosion, or a direct blow to the ear canal. But it may not be considered in a patient with subtle symptoms (particularly balance) after mild head trauma or whiplash that may have occurred months or years prior, where the symptoms have been blamed on traumatic brain injury. There is increasing evidence from animal and human studies that fixed peripheral vestibular damage can cause cognitive disabilities. A chronic PLF is a rare example of an unstable peripheral abnormality that can cause a demonstrable chronic unilateral postural instability and cognitive difficulties. These symptoms are likely due to chronic otoolith organ dysfunction. The video component of this article documents these in a middle aged male after a mild head injury and their complete resolution following PLF repair.

Keywords: Perilymph fistula; Trauma; Traumatic brain injury; Postural instability; Sideways stepping test; Cognitive disability; Tympanotomy

Introduction

Perilymph Fistula (PLF) has been a controversial issue in otolaryngology for over fifty years [1]. Many hold strong views on its existence or otherwise, the symptoms it might cause, the tests which might predict it, the reliability of what is described at exploration, and the effect of repair on symptoms. PLF is a true fistula with a leak of perilymph at the oval or round window, and currently excludes other conditions with “fistula” tests due to a dehiscent horizontal canal from cholesteatoma and the superior canal dehiscence syndrome. It was first recognized in the early days of stapedectomy as causing disequilibrium and balance problems when sealing of the stapedotomy was with blood clot or gelatine sponge. It was solved by routine sealing with natural tissue [2]. In 1968 it was reported that a PLF may follow a head injury [3]. The term “spontaneous PLF” based on 4 patients with no apparent trauma history [4] coincided with a proliferation of large institutional PLF series, skepticism of it and a vigorous debate between “believers” and “non-believers” [5]. In a survey of active surgeons of the American Otological Society and the American Neurotological Society in 1990 the number of PLF explorations a year varied from none to fifty with an average of 5. The primary indications were a history of barotrauma (61%), head trauma (52%), “dizziness” (19%) and sudden hearing loss (17%). Of concern is that 75% said that they would graft a window even if a fistula was not found [6].

Two notable institutional series detail symptoms. In 177 patients at the University of Iowa the most common symptom presentation was a combination of hearing loss, tinnitus and vestibular symptoms, predominantly disequilibrium and motion intolerance [7]. In the Portland experience 79 fistulas were found in 90 ears, nearly all of whom had a trauma history. The main symptom was “disequilibrium”, and many patients reported subtle cognitive problems [8].
For the possibility that PLF patients have an identifiable balance/postural control abnormality most studies are based on posturography. The author has sought to develop a simple and reproducible clinic test demonstrated here. The sideways stepping test (Figure 1) is a logical modification of Singleton’s eyes-closed turning test [9,10].

**Protocol**

**Background**

The author’s results on 21 chronic PLFs in 20 patients with unilateral postural instability on the sideways stepping test are documented in regard to age, symptoms, length of symptoms, ear and window of leak, mode of repair and outcomes have been published [9] as in Table 1. When the cause is blunt head trauma the leak is nearly always at the oval window. In 7 with no apparent or a denied trauma history the cause was later found in 3 as in Table 2. Two patients in the series are the subject of two YouTube videos: https://www.youtube.com/watch?v=CXWnsh0TSrM; https://www.youtube.com/watch?v=DHH5i7BuaOs. A subsequent patient is the subject of a YouTube video: https://youtu.be/2DXgQMnlgbw

The most recent patient is the subject of this presentation.

**New patient history (see YouTube video)**

A 52 year-old professional rugby coach had fainted and hit his head on a bedside cabinet one year prior. He developed headaches and had buzzing tinnitus in the right ear. He complained of being “dizzy” and nauseated, impaired mood and concentration, deteriorating balance and further falls. When travelling in an elevator he would feel as if falling when it stopped. There was a past history of 4 concussions playing rugby. Initial CT head scan, subsequent MRI and CT inner ear scans were normal.

On pure tone audiometry there was a sensorineural hearing loss in the right ear (Figure 2). Standard head impulse (VOR) test, VOR suppression and Dix-Hallpike provocative positional tests were normal.

Balance was significantly abnormal. On three separate occasions he was unable to stand steadily with eyes closed and tilted to the right. On stepping sideways blindfolded or with eyes closed he was stable to the left but unstable to the right. Without visual fixation he could not...
<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age</th>
<th>Symptoms For</th>
<th>Ear</th>
<th>Site</th>
<th>Repair</th>
<th>Preceding event</th>
<th>Symptoms</th>
<th>Follow up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>51</td>
<td>4 yr</td>
<td>Left</td>
<td>OW (FA)</td>
<td>CT</td>
<td>Nil</td>
<td>Disequilibrium to left; nausea; subtle memory difficulty; normal hearing</td>
<td>23 yr</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>51</td>
<td>5 mo</td>
<td>Right</td>
<td>RW</td>
<td>CT</td>
<td>Fainted—concussion</td>
<td>Disequilibrium to right; motion intolerance; tinnitus right ear; normal hearing</td>
<td>22 yr</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>35</td>
<td>6 mo</td>
<td>Left</td>
<td>OW(FA)</td>
<td>CT</td>
<td>2 whiplash injuries</td>
<td>Acute otitis media—vertigo + vomiting—disequilibrium to left; nausea; motion intolerance; subtle memory difficulty; normal hearing</td>
<td>21 yr</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>59</td>
<td>8 mo</td>
<td>Right</td>
<td>OW(FA)</td>
<td>CT</td>
<td>Nil</td>
<td>Disequilibrium to right; nausea; motion intolerance; subtle memory difficulty; normal hearing</td>
<td>20 yr</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>40</td>
<td>1 yr</td>
<td>Left</td>
<td>OW(FA)</td>
<td>CT</td>
<td>Face hit by cricket ball; knocked down by a sheep</td>
<td>Acute otitis media—disequilibrium to left; motion sickness; subtle memory difficulty; normal hearing</td>
<td>20 yr</td>
</tr>
<tr>
<td>6</td>
<td>F</td>
<td>37</td>
<td>3 mo</td>
<td>Left</td>
<td>OW (central footplate)</td>
<td>CT</td>
<td>Nil</td>
<td>Disequilibrium to left; nausea; motion intolerance; tinnitus and sensorineural hearing loss left ear. Positive Hennerbert’s test.</td>
<td>19 yr; see Table 2</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>63</td>
<td>15 yr</td>
<td>Left</td>
<td>OW(FA)</td>
<td>CT</td>
<td>MVA—whiplash</td>
<td>Disequilibrium to left, nausea, motion intolerance, tinnitus and sensorineural hearing loss left ear. Positive Hennerbert’s test.</td>
<td>19 yr; see Table 2</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>19</td>
<td>6 mo</td>
<td>Left</td>
<td>RW</td>
<td>CT + glue</td>
<td>Struck over left ear by milking cups</td>
<td>Disequilibrium to left; nausea; motion sickness; subtle memory difficulty; normal hearing</td>
<td>17 yr</td>
</tr>
<tr>
<td>9</td>
<td>F</td>
<td>53</td>
<td>2 yr</td>
<td>Right</td>
<td>RW</td>
<td>CT + glue</td>
<td>Concussion and whiplash</td>
<td>Same symptoms</td>
<td>9 yr</td>
</tr>
<tr>
<td>10</td>
<td>F</td>
<td>45</td>
<td>1 yr</td>
<td>Left</td>
<td>RW (air bubble)</td>
<td>CT + glue</td>
<td>Nil</td>
<td>Disequilibrium to right; nausea; motion intolerance; tinnitus right ear; normal hearing</td>
<td>16 yr</td>
</tr>
<tr>
<td>11</td>
<td>F</td>
<td>34</td>
<td>18 mo</td>
<td>Right</td>
<td>RW</td>
<td>CT + glue</td>
<td>Punched on left ear</td>
<td>Disequilibrium to right; nausea; motion sickness; popping tinnitus right ear; normal hearing</td>
<td>15 yr</td>
</tr>
<tr>
<td>12</td>
<td>F</td>
<td>40</td>
<td>6 mo</td>
<td>Left</td>
<td>OW(FA)</td>
<td>CT + glue</td>
<td>Head injury to left ear</td>
<td>Disequilibrium to left; nausea; motion sickness; tinnitus left ear; normal hearing</td>
<td>13 yr</td>
</tr>
<tr>
<td>13</td>
<td>F</td>
<td>40</td>
<td>6 mo</td>
<td>Right</td>
<td>OW(FA)</td>
<td>CT + glue</td>
<td>Nurse. Hit head on bed</td>
<td>Disequilibrium to right; nausea; motion sickness; subtle memory difficulty; normal hearing</td>
<td>12 yr</td>
</tr>
<tr>
<td>14</td>
<td>F</td>
<td>26</td>
<td>7 yr; worse 9 mo</td>
<td>Left</td>
<td>OW(FA)</td>
<td>CT + glue</td>
<td>Recent head injury; previous whiplash and prior fall from horse→ head injury</td>
<td>Disequilibrium to left; nausea; motion intolerance; tinnitus left ear; normal hearing</td>
<td>11 yr</td>
</tr>
<tr>
<td>15</td>
<td>F</td>
<td>42</td>
<td>10 yr</td>
<td>Left</td>
<td>OW(FA)</td>
<td>CT + glue</td>
<td>Whiplash in train crash</td>
<td>Disequilibrium to left; falls; nausea; motion intolerance; normal hearing</td>
<td>9 yr</td>
</tr>
<tr>
<td>16</td>
<td>F</td>
<td>78</td>
<td>3 yr</td>
<td>Right</td>
<td>OW(FA)</td>
<td>CT + glue</td>
<td>Fall from a horse; mastoidectomy at age 2 yr</td>
<td>Disequilibrium to right; motion intolerance; mixed hearing loss right ear</td>
<td>7 yr</td>
</tr>
<tr>
<td>17</td>
<td>F</td>
<td>63</td>
<td>3 yr</td>
<td>Left</td>
<td>OW(FA) (+crack in footplate)</td>
<td>CT + glue</td>
<td>Nil</td>
<td>Disequilibrium to left; nausea; motion sickness; normal hearing</td>
<td>6 yr</td>
</tr>
<tr>
<td>18</td>
<td>F</td>
<td>61</td>
<td>6 mo</td>
<td>Left</td>
<td>RW</td>
<td>CT + glue</td>
<td>Mastoidectomy presenting as meningitis 13 yr prior</td>
<td>Disequilibrium to left; nausea; motion intolerance; tinnitus; left mastoid cavity; no hearing left ear</td>
<td>6 yr</td>
</tr>
<tr>
<td>19</td>
<td>F</td>
<td>47</td>
<td>17 mo</td>
<td>Left</td>
<td>OW(FA)</td>
<td>CT + glue</td>
<td>MVA whiplash injury</td>
<td>Disequilibrium to left; nausea; motion intolerance; Nausea from loud sounds; normal hearing</td>
<td>6 yr</td>
</tr>
<tr>
<td>20</td>
<td>F</td>
<td>32</td>
<td>8 mo</td>
<td>Left</td>
<td>OW (crack in footplate)</td>
<td>CT + glue</td>
<td>Head injury in fall from horse</td>
<td>Disequilibrium to left; nausea; from vestibular therapy; normal hearing</td>
<td>4 yr later repair small drum perforation.</td>
</tr>
</tbody>
</table>

Table 1: Author’s published results on 20 patients with 21 surgically repaired PLFs who had a positive sideways stepping test. All were female. Follow up: 23 years - 4 years. OW= 14, RW= 7. Symptoms for 3 months to 10 years (median 10.5 months). When the leak site is OW the cause is nearly always blunt head trauma or whiplash. OW= Oval Window, RW= Round Window, FA= Fissula Antefenestram, CT= Connective Tissue [9].
run on the spot but drifted right (Figure 1) [9].

The history of a mild head injury, prior head injuries, hearing loss in the right ear, nausea and a demonstrable unilateral postural abnormality raised the possibility of a perilymph leak in the right ear.

**Operation**

The right ear canal was infiltrated with xylcaine and adrenaline containing fluorescein to delineate local anesthetic from perilymph [11]. Via a standard endaural incision a tympanomeatal flap was raised. For visual access to the stapes footplate posterior annulus bone was curetted with preservation of the corda tympani nerve. The round window membrane in the round barotrauma niche was clearly seen with no leak. The anterior footplate was partly obscured by the long process of the incus, but there was a leak of perilymph, likely from the fissula ante fenestram. Mucosa around the leak was elevated. Two tiny pieces of connective tissue from the wound were placed and covered with TISSEEL tissue glue, and the eardrum replaced.

**Results**

**New patient postoperative recovery**

On the first postoperative day and on all subsequent occasions he reported and demonstrated normal balance and eventual resolution of all cognitive problems (Video). At 3 months a pure tone audiogram showed recovery of hearing in the right ear (Figure 3). He was able to return to an increasingly successful international coaching career.

**Discussion**

A well accepted two-way mechanism of implosive and explosive routes was proposed by Good hill to illustrate likely causes of sudden hearing loss and vestibular damage from barotrauma, nose-blowing, diving and flying barotrauma, and explosive noise trauma [12]. For blunt head trauma, information from temporal bone histology has been more relevant.

Early temporal bone studies in the 1930s showed that a crack between the round window niche and the posterior canal ampulla was not uncommon, but assumed to be an artefact. Subsequently it has been shown to be developmental [13]. These findings were the impetus for Kohut's temporal bone studies on patients whose histories suggested that they might have had a PLF [14]. On the assumption that PLF ears would have endolymphatic hydrops (to explain their symptoms) [15] he examined the paired temporal bones of patients with histological hydrops and from patients with normal hearing and no vestibular symptoms in regard to oval and round window features. In all the normal temporal bones the fissula ante fenestram was closed by cartilage and the round window fissure was sealed by collagen or bone. In bones with hydrops one had a “patent” fissula ante fenestram and a history of vertigo attacks diagnosed as Meniere’s disease. One had a "patent" fissula ante fenestram containing only fibrous tissue and a history of “waxing and waning” disequilibrium that could have been a PLF. Kohut suggested that a patent or potentially patent fissula ante fenestram could be a pre-existing congenital feature predisposing to a PLF. Unfortunately there is only one published confirmatory case of a post-mortem on both ears with a premortem diagnosis of a unilateral oval window PLF cured surgically [16]. In both ears there was no evidence of hydrops.

In the PLF literature there is a wide and confusing range of terms in regard to its vestibular symptoms: dizziness, imbalance, unsteadiness, disequilibrium and vertigo, which often implicate the author’s belief that the symptoms are due to endolymphatic hydrops [15]. Vertigo has always had a well understood definition of a hallucination of motion, but in the PLF literature dizziness and vertigo have been used loosely and probably mean any vestibular symptom. The Barany Society has sought to refine the definition of common vestibular symptoms [17]. Dizziness is defined as "the sensation of disturbed special orientation without a false or distorted sense of motion". Chronic PLF patients do not describe either of these. The most predominant symptom is being "off balance”. Once called "disequilibrium” the new Barany name is “lateral pulsion”, a neurological term which may not necessarily be the most appropriate. PLF patients appear to have a unique postural control abnormality which Brandt proposes is due to chronic otolith organ dysfunction via vestibulospinal pathways [18]. There is as yet no recognized vestibular condition in which a balance test is the only or essential diagnostic feature. Clearly the sideways stepping test described here requires objective verification on PLF subjects, normal subjects and in patients with conventional unilateral vestibular (canal) hypo function [9].

Grimm and colleagues [8] performed detailed neurotological studies on adults with mild defined craniocervical trauma with a confirmed PLF; noting some complained of significant memory loss and poor coping, which had been blamed on traumatic brain injury. He has suggested that these subtle cognitive chronic PLF symptoms make it a neurological as well as an otological syndrome [19].

There is increasing evidence from experimental animal studies that permanent vestibular lesions can impair performance in learning and memory tasks requiring special information processing [20-22] and in humans [23,24]. Vestibular stimulation on a motion-simulator has been shown to alter mood states [25]. Patients with otic capsule dehiscences can exhibit deficits in verbal memory, visual memory, attention/concentration and working memory [26].

After an inner ear injury there is nearly always recovery or central adaptation. A chronic PLF is a rare example of an unstable peripheral organ. The vestibular system is a very primitive aspect
of brain function which is preoccupied with calculating gravity and orientation to earth vertical, so when it becomes perpetually confused higher brain function is subtly affected [19].

In summary, chronic PLF following head trauma is usually at the oval window fistula ante fenestram in individuals with a congenital predisposition. There is often a history of previous head injuries. There does not have to be a hearing loss (Table 1). The predominant symptoms are a chronic unilateral postural instability (suggesting otolith organ dysfunction), nausea, motion intolerance, and often subtle cognitive difficulties. Eventually advances in imaging may image a window fistula. Meanwhile, when a window PLF is strongly suspected a simple tympanotomy is justified [1].

Disclosures

The author has nothing to disclose.

Acknowledgements

The author is grateful to the patient for permission to present his case and to Robert Ashford at Department of Medical Illustrations at Christchurch Hospital for the recordings on this patient and for the presentations on three who are the subject of Youtube videos. Table 1 and Figure 1 are from International Journal of Otolaryngology, Volume 2011, Article ID 835672, doi: 10.1155/2011/835671 with copyright owned by the author. The illustrations are by Slipstream Creative (http://slipstreamcreative.co.nz).

References
