



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

Jeremy Hornibrook<sup>1,2,3\*</sup>

<sup>1</sup>Department of Otolaryngology-Head and Neck Surgery, Christchurch Hospital, New Zealand

<sup>2</sup>University of Canterbury, Christchurch, New Zealand

<sup>3</sup>University 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 otolith 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.

### OPEN ACCESS

#### \*Correspondence:

Jeremy Hornibrook, Department of Otolaryngology-Head and Neck Surgery, Christchurch Hospital, New Zealand,

E-mail: [jeremy@jhornibrook.com](mailto:jeremy@jhornibrook.com)

Received Date: 24 Apr 2018

Accepted Date: 24 May 2018

Published Date: 31 May 2018

#### Citation:

Hornibrook J. The Postural and Cognitive Disabilities of Chronic Perilymph Fistula (PLF) after Mild Head Trauma. *Ann Clin Case Rep.* 2018; 3: 1514.

ISSN: 2474-1655

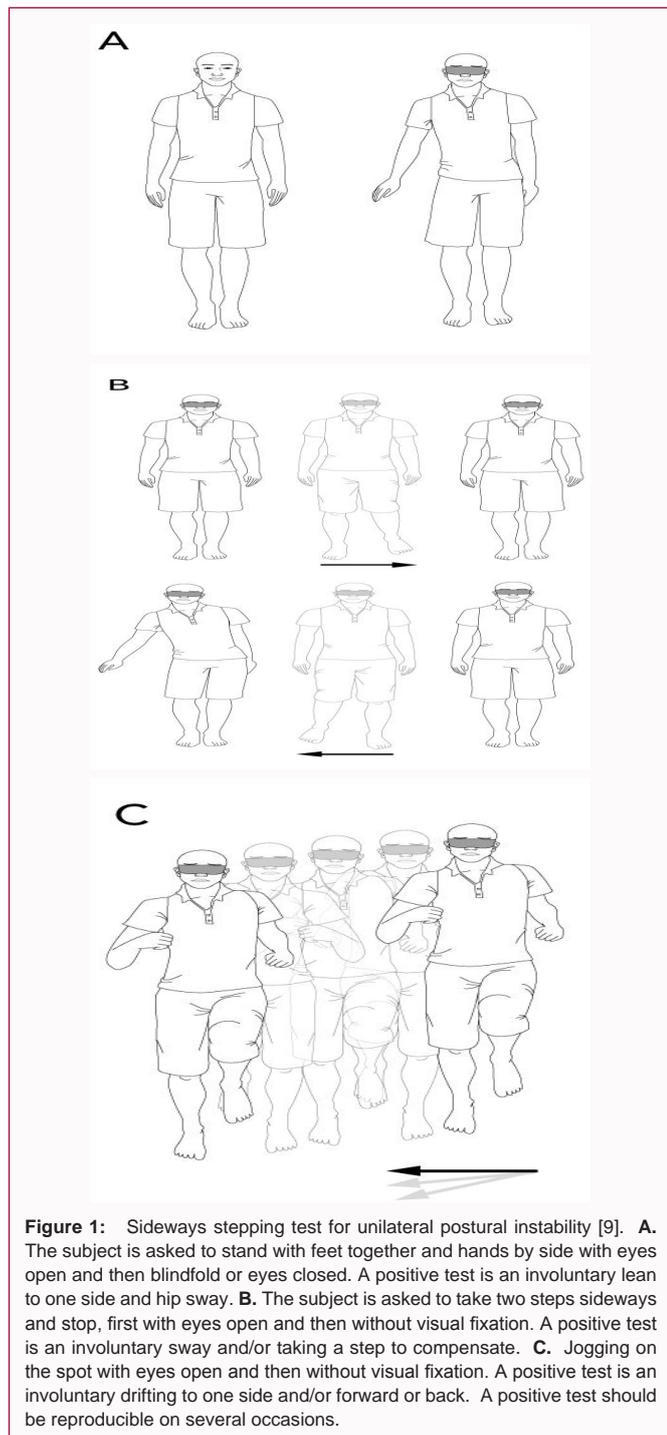
Copyright © 2018 Jeremy Hornibrook. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

**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 dehiscence 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 five. 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].



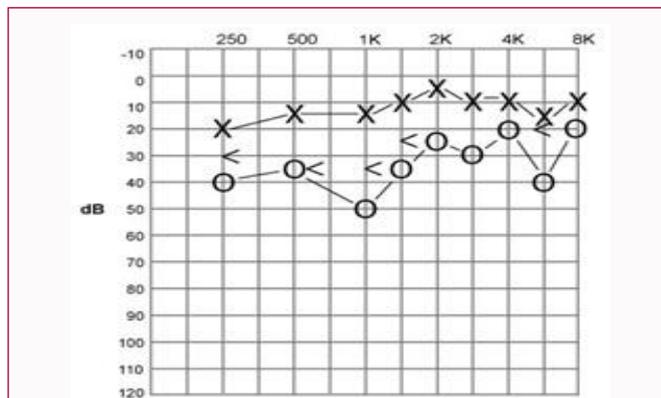
**Figure 1:** Sideways stepping test for unilateral postural instability [9]. **A.** The subject is asked to stand with feet together and hands by side with eyes open and then blindfold or eyes closed. A positive test is an involuntary lean to one side and hip sway. **B.** The subject is asked to take two steps sideways and stop, first with eyes open and then without visual fixation. A positive test is an involuntary sway and/or taking a step to compensate. **C.** Jogging on the spot with eyes open and then without visual fixation. A positive test is an involuntary drifting to one side and/or forward or back. A positive test should be reproducible on several occasions.

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].

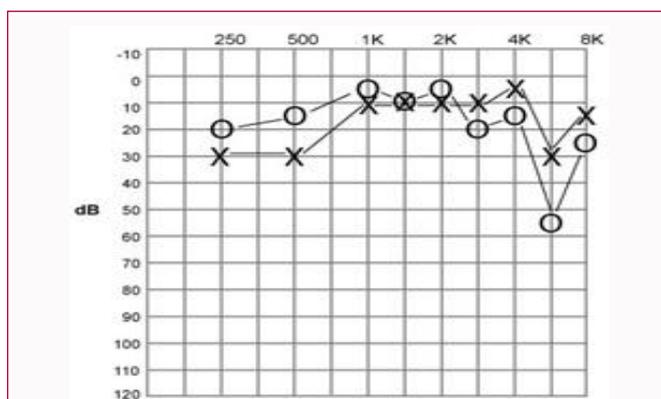
**Protocol**

**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



**Figure 2:** Preoperative audiogram showing a sensorineural loss in the right ear.



**Figure 3:** Postoperative audiogram at 3 months showing hearing recovery in the right ear.

[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 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 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].

Patient	Sex	Age	Symptoms For	Ear	Site	Repair	Preceding event	Symptoms	Follow up
1	F	51	4 yr	Left	OW (FA)	CT	Nil	Disequilibrium to left; nausea; subtle memory difficulty; normal hearing	23 yr
2	F	51	5 mo	Right	RW	CT	Fainted→concussion	Disequilibrium to right; motion intolerance, tinnitus right ear; normal hearing	22 yr
3	F	35	6 mo	Left	OW(FA)	CT	2 whiplash injuries	Acute otitis media→vertigo + vomiting→disequilibrium to left; nausea; motion intolerance; subtle memory difficulty; normal hearing	21 yr
4	F	59	8 mo	Right	OW(FA)	CT	Nil	Disequilibrium to right; nausea; motion intolerance; subtle memory difficulty; normal hearing	20 yr
5	F	40	1 yr	Left	OW(FA)	CT	Face hit by cricket ball; knocked down by a sheep	Vertigo after acute otitis media; disequilibrium to left; nausea; motion intolerance; subtle memory difficulty; normal hearing	20 yr
6	F	37	3 mo	Left	OW (central footplate)	CT	Nil	Acute otitis media→disequilibrium to left; motion sickness; subtle memory difficulty; normal hearing	20 yr
7	F	63	15 yr	Left	OW(FA)	CT	MVA→whiplash	Disequilibrium to left, nausea, motion intolerance, tinnitus and sensorineural hearing loss left ear. Positive Hennerbert's test.	19 yr; see Table 2
8	F	19	6 mo	Left	RW	CT + glue	Struck over left ear by milking cups	Disequilibrium to left; nausea; motion sickness; subtle memory difficulty; normal hearing	17 yr
	F	28	1 yr	Right	RW	CT + glue	Concussion and whiplash	Same symptoms	9 yr
9	F	53	2 yr	Right	RW	CT + glue	Nil	Disequilibrium to right; nausea; motion intolerance; tinnitus right ear; normal hearing	16 yr
10	F	45	1 yr	Left	RW(air bubble)	CT + glue	Nil	Disequilibrium to left; nausea; subtle memory difficulty;	16 yr
11	F	34	18 mo	Right	RW	CT + glue	Nil	Disequilibrium to right; nausea; motion sickness; popping tinnitus right ear; normal hearing	15 yr
12	F	40	6 mo	Left	OW(FA)	CT + glue	Punched on left ear	Disequilibrium to left; nausea; motion sickness; tinnitus left ear; normal hearing	13 yr
13	F	40	6 mo	Right	OW(FA)	CT + glue	Nurse. Hit head on bed frame	Disequilibrium to right; nausea; motion sickness; subtle memory difficulty; normal hearing	12 yr
14	F	26	7 yr; worse 9 mo	Left	OW(FA)	CT + glue	Recent head injury; previous whiplash and prior fall from horse→ head injury	Disequilibrium to left; nausea; motion intolerance; tinnitus left ear; normal hearing	11 yr
15	F	42	10 yr	Left	OW(FA)	CT + glue	Whiplash in train crash	Disequilibrium to left; falls; nausea; motion intolerance; normal hearing	9 yr
16	F	78	3 yr	Right	OW(FA)	CT + glue	Fall from a horse; mastoidectomy at age 2 yr	Disequilibrium to right; motion intolerance; mixed hearing loss right ear	7 yr
17	F	63	3 yr	Left	OW(FA) (+crack in footplate)	CT + glue	Nil	Disequilibrium to left; nausea; motion sickness; normal hearing	6 yr
18	F	61	6 mo	Left	RW	CT + glue	Mastoidectomy presenting as meningitis 13 yr prior	Disequilibrium to left; nausea; motion intolerance; tinnitus; left mastoid cavity; no hearing left ear	6 yr
19	F	47	17 mo	Left	OW(FA)	CT + glue	MVA whiplash injury	Disequilibrium to left; nausea; motion intolerance; Nausea from loud sounds; normal hearing	6 yr
20	F	32	8 mo	Left	OW (crack in footplate)	CT + glue	Head injury in fall from horse	Disequilibrium to left; nausea; nausea from vestibular therapy; normal hearing	4 yr later repair small drum perforation.

**Table 2:** Eventual cause found in 3 of 7 patients with a forgotten or denied trauma history [9].  
OW= Oval Window, RW= Round Window.

Sex	Age	Symptoms for	Ear and Site	Eventual Confirmation of Traumatic Event
F	51	4 Years	Left OW	Struck on head by swinging ladder; hit head on a plank
F	59	8 Months	Right OW	Not found
F	37	3 Months	Left OW	4 years prior hit head on gallery roof in plunging air carft
F	53	2 Years	Right RW	Not found
F	45	1 Year	Left RW	Admitted her husband had 'smacked' her ear
F	34	18 Months	Right RW	Not found
F	63	3 years	Left OW	Not found

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 xylocaine and adrenaline containing fluorescein to delineate local anesthetic from perilymph [11]. Via a standard endaural incision a tympanometal 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 window 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 from straining, 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 imply 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 fissula 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

- Hornibrook J. Perilymph fistula: Fifty years of controversy. *ISRN Otolaryngol*. 2012;31:281248.
- Shea JJ. Stapedectomy-a long-term report. *Ann Otol Rhinol Laryngol*. 1982;91(5):516-520.
- Fee GA. Traumatic perilymph fistulas. *Arch Otolaryngol*. 1968;88(5):43-6.
- Stroud MH, Calceterra TC. Spontaneous perilymph fistulas. *Laryngoscope*. 1970;80(3): 479-87.
- Shea JJ. The myth of spontaneous perilymph fistula. *Otolaryngol Head Neck Surg*. 1992;107(5): 613-6.
- Hughes GB, Sismanis A, House JW. Is there consensus in perilymph fistula management? *Otolaryngol Head Neck Surg*. 1990;102(2), 102-111.
- Selzer S, McCabe BF. Perilymph fistula: the Iowa experience. *Laryngoscope*. 1986;96:37-49.
- Grimm RJ, Hemenway, Lebray PR, Black FO. The perilymph fistula syndrome in defined mild head trauma. *Acta Otolaryngologica Suppl*. 1989;464:1-40.
- Hornibrook J. A balance test for chronic perilymph fistula. *Int J Otolaryngol*. 2012;2012:163691.
- Singleton GT. Diagnosis and treatment of perilymph fistulas without hearing loss. *Otolaryngol Head Neck Surg*. 1986;94(4), 426-9.
- Arenberg IK, Wu CM. Fluorescein as an easy, low-cost, indirect, or reverse intraoperative marker to rule out perilymph versus local injection. *Am J Otol*. 1996;17:259-62.
- Goodhill V. Sudden deafness and round window rupture. *Laryngoscope*. 1971;81:1462-74.
- Okano E, Myers EN, Dickson DR. Microfissure between the round window niche and posterior canal ampulla. *Ann Otol Rhinol Laryngol*. 1977;86:49-57.
- Kohut RI, Hinjosa R, Budetti JA. Perilymphatic fistula: a histopathologic study. *Ann Otol Rhinol Laryngol* 1986;95:466-71.
- Harrison WH, Shambough GE, Derlacki EL, Clemis JD. Perilymph fistula in stapes surgery. *Laryngoscope* 1967;77(5), 836-49.
- Kohut RI, Hinjosa R, Thompson JN, Riu JH. Idiopathic perilymphatic fistulae: Atemporal bone histopathologic study with clinical, surgical, and histopathologic correlations. *Arch Otolaryngol Head Neck Surg*. 1995;121:412-20 .
- Bisdorff A, von Brevern M, Lempert T, Newman-Toker DE. Classification of vestibular symptoms: towards an international classification of vestibular disorders. *J Vestib Res*. 2009;19(1-2):1-13.
- Brandt T. Basel Otolithic vertigo. *Otolith Function and Disorders*eds. Karger, editors. Tran Ba Huy, P Toupet M. 2001;58:34-47.
- Grimm RJ. Personal communication. International Symposium on Perilymph Fistula, Portland, Oregon, USA; 1990.
- Smith PF, Horii A, Russell N, Bilkey DK, Zheng Y, Liu P, et al. The effects of vestibular lesions on hippocampal function in rats. *Prog Neurobiol*. 2005;75(6), 391-405.
- Baek JH, Zheng YZ, Darlington C, Smith PF. Evidence that spatial memory deficits following bilateral vestibular deafferentation in rats are probably permanent. *Neurobiol Learn Mem*. 2010;94(3):402-13.
- Smith PF, Haslett S, Zheng Y. A multivariate statistical and data mining analysis of special memory-related behaviour following bilateral vestibular loss in rat. *Behav Brain Res*. 2013 Jun 1;246:15-23.
- Brandt T, Schautzer F, Hamilton DA, Brüning R, Markowitsch HJ, Kalla R, et al. Vestibular loss causes hippocampal atrophy and impaired spacial memory in humans. *Brain*. 2005;128(11):2732-41.
- Smith PF, Darlington CL. Personality changes in patients with vestibular dysfunction. *Front Hum Neurosci*. 2013;7:678.
- Lotta W, Tillmann HCK, Jean L, Harald E, Manfred S, Dominik S, et al. Vestibular stimulation on a motion-stimulator impacts on mood states. *Front Psychol*. 2012;3:499.
- Wackym PA, Balaban CD, Mackay HT, Wood SJ, Lundell CJ, Carter DM, et al. Longitudinal cognitive and neurobehavioural functional outcomes before and after repairing otic capsule dehiscence. *Otol Neurotol*. 2016;37(1):70-82.