

# Perilymphatic Fistula: an approach to diagnosis and management that provides surer diagnosis and provides medical and surgical management options: report of six illustrative recent cases

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## Abstract

Perilymphatic Fistula (PLF) remains a controversial topic and its management is challenging. Various etiologic events have been proposed and spontaneous PLF has been described [1]. Diagnostic methods and strategies have been reported in the literature but no standard algorithm exists. The most accepted diagnostic method remains intra-operative confirmation of perilymph leak at oval window or round window. However, concerns have been raised regarding excessive surgical intervention given the lack of pre-operative confirmatory tests. We have developed a diagnostic strategy that has been successful in our hands. The aim of this study is to present six patients with PLF who underwent surgical repair. We describe our approach in the evaluation and management of these patients. We discuss the diagnostic tests and the operative technique used in management of these patients and we provide a review of the literature in support of our approach.

**Keywords:** dizziness, hearing loss, sensorineural, vertigo, vestibular diseases, vestibular function tests.

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## INTRODUCTION

Perilymphatic fistula (PLF) has become an integral part of modern Neurotology. PLF is an abnormal communication between the perilymphatic space and middle ear space. PLF most commonly occurs in the round window niche and/or the oval window at the fissula ante fenestram. Various causes of PLF have been described including stapedectomy, barotrauma, physical exertion, head and neck trauma, congenital and idiopathic<sup>2-4</sup>. However, the diagnosis of PLF remains a subject of controversy due to the variability in patient presentation and difficulty in pre-operative evaluation. Multiple tests have been described but no definitive diagnostic tool is available at present to confirm presence of PLF preoperatively. Thus there are no firm guidelines for medical and surgical intervention. Because PLF is a controversial diagnosis, it is crucial to discuss it in our literature and perhaps come to a consensus. In this paper, we describe 5 patients with post-traumatic PLF, and one patient with spontaneous PLF. We describe our algorithm in diagnosing PLF and review the literature supporting our approach. Additionally, we describe our surgical technique in treatment of PLF.

## MATERIALS AND METHOD

Evaluation of a patient with suspected PLF begins with a thorough history including a detailed history regarding the mechanism of trauma. A standard ear, nose, throat and neck examination is performed. A focused neuro-otological examination is performed including cranial nerve examination, examination of spontaneous and positional nystagmus, and balance examination that includes Romberg and Quix testing (Table 1).

**Table 1.** Evaluation of PLF.

History
Physical Examination
Complete Head and Neck Examination
Cranial Nerve Examination
Spontaneous and Positional Nystagmus
Balance Examination - Romberg and Quix test
Audiological Evaluation
Pure tone
Tympanogram
Suprathreshold Stimulus Stapedial Testing
Fistula Testing
ENG
Lasix Dehydration Test (or Glycerin)

A complete audiometric evaluation is then performed, as is the suprathreshold stimulus stapedial testing and fistula testing. Suprathreshold stimulus stapedial testing was performed with stimulation being presented to the contralateral ear at 0.5k, 1k, and 2k Hz at 110 dB. Evaluation of symmetry of response, and abnormalities in latency, amplitude and slope of the response are performed<sup>20</sup>. Fistula testing was performed with an impedance bridge using positive and negative pressure, sweeping the pressure from +400 dapa to -400 dapa three times in each of four test situations, the patient sitting eyes open, the patient standing eyes open, the patient sitting eyes closed, and the patient standing eyes closed<sup>13</sup>. The audiologist performs this test, looking for nystagmus or a sway or complaints of dizziness, lightheadedness, disorientation or nausea.

ENG has not shown to be helpful in diagnosing PLF but it was performed in all patients with suspected PLF to evaluate for the presence of vestibular function. Vestibular function was intact in all of our cases.

The Lasix dehydration test, as previously described in the literature, was administered to patients with suspected PLF<sup>5</sup>. Patients with sulfa allergy underwent the Glycerin test<sup>6</sup>. Intramuscular injection of 60mg of Lasix was administered in the left buttock. Pure tone thresholds, speech reception thresholds, fistula testing, and Quix<sup>21</sup> and Romberg testing were performed before the injection and every 30 minutes subsequently for 2 hours and changes in pure tone responses, speech discrimination and speech reception thresholds, fistula testing, and Quix and Romberg balance testing were observed. If the findings improved, it indicated presence of Endolymphatic Hydrops which confirmed presence of PLF<sup>21</sup>. Endolymphatic Hydrops also occurs in Ménière's disease. However, patient's history and the definitive episodes of spinning vertigo differentiate the Endolymphatic Hydrops due to Ménière's disease from PLF. According to the 1995 American Academy of Otolaryngology - Head and Neck Surgery guidelines describing probable, possible, definite, and certain Ménière's disease, one of the required criteria is to rule out other possible causes of patient's symptoms, one of which can be PLF<sup>23</sup>.

During the Lasix dehydration test, patients may show improvement, worsening, or have no change in their audiometric and physical examination findings. (Figure 1) If the symptoms worsen, perilymphatic hypertension may be suspected<sup>24</sup>. If there is no change, it indicates no evidence of inner ear fluid disorder. If the patient's symptoms and physical exam findings improve during the Lasix test, management options include medical therapy with oral Lasix or surgical treatment with middle ear exploration.



**Table 2. Patients Testing and Results.**

Patient	Injury	Time to presentation	Time to surgery	Chief Complaint	Romberg	Quix	Positional Nystagmus	Spontaneous Nystagmus	Audiogram	Fistula Test	Lasix Test	Intervention	Post-operative	Comments
1	MVA	12 months	33 months	Persistent Dizziness	slight shaky	Shaky	Absent, buy dizzy on sitting up	Absent	Normal	Left > Right	Improvement	OW, RW Repair	Normal Quix and Romberg	Abdominal Pain with oral Lasix
2	MVA	3 months	8 months	Persistent Dizziness	slight shaky	right shaky sway	Absent, buy dizzy on sitting up	Absent	Normal	Left = Right	Improvement	OW, RW Repair	Normal Quix and Romberg	Wants to get pregnant; Lasix contraindicated
3	MVA	5 months	18 months	Persistent Dizziness	shaky	Shaky	Absent	Absent	Moderate SNHL	Left > Right	Improvement	OW, RW Repair	Normal Quix and Romberg	Severe weakness with oral Lasix
4	MVA	3 months	26 months	Persistent Dizziness	shaky	Shaky	Absent	Absent	Normal	Right > Left	Improvement	OW, RW Repair	Partial Improvement	Excessive urination with oral Lasix
5	spontaneous	4 months	9 months	Persistent Dizziness	falls back	falls back	Absent	Absent	Normal	Left > Right	Improvement	OW, RW Repair	Normal Quix and Romberg	Excessive fatigue with oral Lasix
6	Fall	60 months	78 months	Persistent Dizziness	shaky	Shaky	Absent, buy dizzy on sitting up	Absent	Normal	Left Only	Improvement	OW, RW Repair	Normal Quix and Romberg	Could not tolerate oral Lasix after 1 year

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need not be severe. Even whiplash injury to the neck can cause PLF<sup>10,11</sup>. Contrary to popular belief, only half of patients with traumatic PLF have hearing loss.

Chronic or persistent combinations of disequilibrium, headache, adult-onset motion sickness, neurocognitive disruption exacerbated by exertion or physical activity, and hearing loss have all been described as symptoms of PLF. In patients with these symptoms, findings of positive Romberg, ataxia and positional nystagmus should raise the suspicion of PLF. Healy et al. described episodic positional vertigo and ataxia between the episodes as chief complaints in their series of patients with PLF<sup>12</sup>. Their criteria for middle ear exploration included abnormal gait, positional nystagmus, positive Romberg, and positive fistula test. Hearing loss was a less common complaint in their cohort, whereas others have reported hearing loss to be the primary complaint in patients with PLF<sup>7,10</sup>.

Although multiple tests are available to assist a physician in diagnosing PLF, controversy exists in the utility of these tests. The use of fistula test in addition to the classic physical exam findings in identifying patients with PLF has been well documented<sup>12</sup>. During fistula testing, dizziness may occur in patients with PLF by applying positive and negative pressure in EAC, which results in pressure changes in inner ear, with positive fistula testing more likely occur in the symptomatic ear. The usual fistula test is performed using a pneumatic otoscope which only provides positive pressure. However, Kohut et al. have found that negative pressure is a more sensitive test, and therefore important to perform as part of the fistula testing<sup>13</sup>.

Multiple diagnostic modalities have been described to help diagnose PLF. Moving platform fistula test has not been widely used<sup>14</sup>. Transtympanic endoscopy and the use of fluorescein have been described as methods of documenting a fistula without operation<sup>15,16</sup>. However, injection of this dye has been found to be inaccurate in the diagnosis of PLF as dye has been found in serum as well as perilymph. This procedure has not been reported in recent literature because of its lack of efficacy. Standard ECOG, transtympanic ECOG, and intraoperative ECOG have also been described in the diagnosis of Ménière's Disease and in the evaluation of PLF but these tests also have not been used widely<sup>17,18</sup>. In our practice, we have utilized tympanic electrodes to perform ECOG.

Most recently, magnetic resonance imaging of inner ear after intratympanic injection of gadolinium has been described for evaluation of Ménière's disease<sup>19</sup>. If a patient has a history more suggestive of PLF, an increased signal on MRI, indicating hydrops secondary to PLF should be considered because although Endolymphatic Hydrops is a pathologic correlate of Ménière's disease, it has also been observed in PLF. Thus, a diagnostic test

utilized in Ménière's disease can be helpful in diagnosing PLF, as long as the patient's history is appropriately considered.

Inner ear dehydration tests have been described for diagnosis of Ménière's disease. Since endolymphatic hydrops is also present in PLF as described in the literature, various inner ear dehydration tests have been used in PLF<sup>4-6</sup>. We use the Lasix dehydration test because it is the most convenient for patients and it provides the opportunity for medical therapy. Patients are themselves able to witness improvement in their imbalance during the test. Loop diuretics are effective in our hands whereas thiazide diuretics are not. If patients' symptoms get worse on the Lasix dehydration test, a presumptive diagnosis of perilymphatic hypertension can be made<sup>24</sup>.

If medical therapy is contraindicated or not tolerated by the patient, surgical intervention is offered. During the surgical procedure, certain steps are crucial to provide the best outcome. It is important to have a broad view with a large ear speculum. A superior endaural incision can provide a wider surgical field before entering the middle ear. Maintaining a bloodless field prior to raising the annulus and entering in the middle ear avoids pooling of blood around the oval window or the round window niche. In order to better visualize the leak, we place the patients in Trendelenberg which increases intracranial and perilymph pressure. Other techniques have been described including Valsalva and Internal Jugular compression to assist in visualizing the leakage of clear fluid from the oval and the round window. We confirm the leak by suctioning and observing the reaccumulation of fluid according to the method of Kohut<sup>13</sup>. Before placing the fat grafts, we denude the mucosa in the area of the fissula ante-fenestram in the oval window area and the floor of the round window niche inferior to the round window membrane in order to provide a raw surface for the graft to adhere. If a leak is seen at one of the two areas, we graft both because of the possibility of an inapparent fistula at the second window.

We have discussed our evaluation and management of patients with PLF. We encourage our colleagues to include PLF in their differential diagnosis and to consider our diagnostic approach and testing in their evaluation of these patients. Our experience is that imbalance can be reversed long after the insult but hearing loss cannot be reversed after several weeks, so that prompt attention (usually within a week) is required in cases of PLF with sudden hearing loss, such as those seen in SCUBA divers.

## CONCLUSION

PLF is a condition that has multiple causes and can present with varying symptoms. PLF should be considered as part of the differential diagnosis in patients

with complaints of dizziness, imbalance, or hearing loss. The diagnosis of PLF is challenging. The absence of hearing loss does not eliminate the possibility of PLF. Fistula testing followed by Lasix dehydration test is helpful in determining inner ear fluid imbalance as a cause of disequilibrium and hearing loss. Medical therapy with diuretics can provide benefit and avoid surgery. We are conservative in offering surgery although surgical treatment for PLF is safe and effective. The diagnostic method we utilized provided positive findings pre-operatively in all these cases as illustrated. The time and effort in evaluation and diagnosis is worthwhile in our experience. This is a retrospective study that provides an illustration of our methods. We plan to do a prospective study to further define the role of our diagnostic methodology.

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