Journal of Life Care Planning, Vol. 14, No. 3, (23-53)

The Editor's Introduction

In treatment as well as in life care plans, vestibular disorders are often overlooked and can add quality of life as well as enhance a life care plan. This article is one doctor's lifelong journey of defining and treating vestibular disorders.

A Clinical Approach to the Diagnosis and Management of Vestibular Disorders Utilizing a New System of Classification and Introducing a New Syndrome Into the Classification

Joel F. Lehrer, M.D., FACS

Introduction

This article is for clinicians and life care planners which will describe experiences and methods in the diagnosis and treatment of patients with vestibular disorders. A definition of a classification presented in 1994[1] was found to be successful in classifying vestibular disorders in over 90% of 327 consecutive patients seen in 1990. There have been advances in diagnosis, and new disorders have been identified since that time, such as dehiscence of the semicircular canals. The intention of this article is not to provide an encyclopedic list of the various pathologies that result in vestibular disorders and vestibular symptoms but to provide a pragmatic classification that will allow the clinician to evaluate patients with a diagnostic schema in mind and a method to arrive at a specific diagnosis and focused plan of management that can be useful in a life care plan. The goal of this article is to allow vestibular disorders to be evaluated and opined in a life care plan in the same fashion as disorders and diseases of other bodily systems.

A major problem in classifying and treating vestibular disorders has been the lack of the ability to image the actual pathology. Unfortunately, the testing is often non-definitive, with conclusions limited to whether a disorder is central or peripheral without defining the site of the pathophysiology or pathology. Examinations of the balance are not reported in detail and have not been viewed as important in the diagnostic and management process. Physical examinations have emphasized the examination of the eyes and the visual-ocular reflex (VOR), as well as a full cranial nerve examination, but have not focused on the evaluation of the balance. It is important that the life care planner understand the evaluation of balance and vestibular disorders so that the life care plan can accurately reflect which the individual needs.

The aim of this article is to provide clinicians and life care planners with definable diagnostic targets, the classification, a map to arrive at a proper diagnosis, and then a plan for effective treatments, which if successful, will serve to validate the diagnosis. Another aim is also to assist clinicians and life care planners in their mission of projecting appropriate recommendations through understanding what to ask the neurotolologist. In order to understand the classification, the process and treatment, a historical prospective will be reviewed of how Dr. Lehrer developed the classification, will describe the classification, and discuss the diagnostic methods and various treatments that Dr. Lehrer utilizes.

History of the Development of the Classification System

Fifty years ago, the differential diagnosis of patients with vestibular complaints included Meniere's disease, positional vertigo, labyrinthitis, vestibular neuronitis, loss of vestibular function and central vertigo. Vertigo was utilized as the descriptive term for these disorders. The use of the term "vestibular" had not yet been utilized to describe this category of disorders.

It should be noted that, aside from loss of vestibular function, the spinning symptom was the presenting one in all of these disorders. Vertigo, which derives from the Latin "to turn," is thus an apt term to generally describe these disorders. Dr. Francis Miskolczy Fodor was instrumental as a scientific practitioner who brought his European background and experiences to the United States and influenced the research and development of otologic disorders.

Dr. Fodor taught about the "Harmonic Symptom Complex," knowledge of which enabled professionals to differentiate between acute central and peripheral vertigo. The key is to analyze the direction of the fast component of the nystagmus and the direction of the body sway which is present. If they are opposite in direction, the disorder is peripheral which, in the absence of mastoid infection or trauma or hearing loss, would indicate a diagnosis of vestibular neuritis. This entity will be discussed later in this article. If the direction of the nystagmus and body sway is the same, the disorder is central, which today would indicate a disorder such as multiple sclerosis. Dr. Fodor explained that this technique was utilized in the early part of the 20th century, long before CT scans and MRIs were available, in order to differentiate between true labyrinthitis caused by erosion of the horizontal semicircular canal by mastoid infection, which would yield the peripheral finding, or

erosion of the infection into the posterior fossa causing brain abscess affecting the cerebellum, which would yield the central finding. It has been recognized this simple, straightforward examination is invaluable at the bedside or in the office when a patient presents with acute vertigo. Dr. Fodor taught to examine balance as well as eye movements in order to better evaluate the dizzy patient. It has since been recognized that the careful examination of the balance is crucial to diagnosis and management of vestibular disorders, be they of the spinning or non-spinning type.

During the 1960s, the general teaching at the time was that all vertigo would resolve in six months. This was based upon a paper by Silvoniemi.[2] However, community-based neurotologists started noticing patients, despite the passage of more than six months after the initial examination and prognosis, returned to practices with a nose or throat problem, and, when questioned about the "vertigo" symptom, informed the neurotologist that it was still present. It was realized that these cases needed more attention, a more careful examination, and a more precise diagnosis than simply "vertigo."

It was then that the focus on the balance examination in all patients with the complaint of dizziness was initiated and not limit the examination to patients with acute vertigo. The acute vertigo patients that Dr. Fodor treated could hardly stand, so the early examinations were usually performed sitting in the hospital bed. Ultimately, patients with vertigo were examined in community-based offices if they were not so sick as to require hospitalization. The examination involved having the sitting patient extend arms and hands and point the index fingers while the examiner placed his or her index fingers opposite and looked for a sway. If the patient could stand, the Romberg test was performed to look for a body sway or deviation. The Romberg test is preferred to be performed with the feet closely together and then observe the patient for at least 15 seconds while standing behind them and reassure them that they are protected so they do not fear falling. Currently, the Romberg test is performed in all patients if they are able to stand without provoking nausea or vomiting. A professional needs to look for shakiness or a sway. Sometimes the patient will start to fall, but they are protected against actually falling by the examiner. The body deviations on all of the tests that are performed are usually consistent, being in the same direction when repeated at the time of the initial examination and in follow-ups if the patient is not improved.

Dix and Hallpike, in their classic paper[3] described two groups of dizzy patients: one group with symptoms that could be interpreted as spinning and another group that was definitely not spinning. They did not suggest a particular treatment for either group. In 1965, a paper by Dr. Gordon Gilbert, a neurologist, was published in the *Journal of the American Medical Association* in which Gilbert opined that there was a relationship between cluster headache and vertigo.[4] He reported three cases in which he had discovered a link between these disorders. He suggested anti-migraine therapy for patients with vertigo as such therapy was successful in cases of cluster headache. Medications were prescribed, and it was found that they were effective in treating some of the "dizzy" patients who presented with findings of imbalance but did not suffer from spinning symptoms.

By the late 1960s, tests had been refined and balance examinations added standing tests, the Romberg, described above, and the Quix test.[5,6] The Quix test is performed by having the patient stand with feet together, with the arms and hands outstretched and the index fingers pointed outwards. The examiner stands behind the patient to protect against falling, and the assistant stands opposite the patient with index fingers pointed at the patient's index fingers. One looks for shakiness or a body sway. When performed in the sitting position as described above, it is called the sitting Quix test examination. These three examinations should be performed for at least 15 seconds and repeated without resting the patient in between in order to increase their sensitivity when necessary in a patient with the vestibular complaints. The addition of the standing tests increased the sensitivity of the balance examination. In cases where these tests were negative, where no sway or shakiness was seen, tests became even more sensitive by requiring the patient to perform a tandem Romberg test and then redo the Quix test, or by having the patient do the Quix test after getting up quickly after positional testing. The additional effort to do the Quix test resulted in increased sensitivity, which identified imbalance, and which was especially helpful in young or athletic patients who did not initially display imbalance on the Quix or Romberg tests. In the great majority of cases, when there was imbalance, there was imbalance only on the Quix test which is the most sensitive of the three. These examinations for the presence of imbalance were crucial for the demonstration of an objective physical finding in patients presenting with complaints of dizziness. The ability to demonstrate such a finding literally opened the way to providing a path to diagnosis and management in patients who presented with non-spinning dizziness, who were at that time, and even now, often classified as psychological or even psychiatric cases.

During the late 1960s, the past pointing test was beginning to be utilized. This test has the patient and examiner begin by each pointing their index fingers at each other. The patient, with eyes closed, then raises the hands and fingers and then lowers them to meet the examiner's fingers. A deviation is termed "past pointing" and is a sign of body deviation. The findings are similar to those found with the Quix test. The Unterberger or Fukuda marching tests, in which the patient marches in place with eyes closed, were not

as reliable and repeatable as the Romberg and Quix tests.

It was found that evaluation of the eye movements and eye responses to stimulation are a necessary part of the evaluation of vestibular disorders but are not sufficient to evaluate the non-spinning disorders. Examining the balance is also critical to differentiate peripheral from central vertigo as described above.

Dr. Lehrer began to utilize the anti-migraine drugs of the day to treat patients with dizziness and imbalance. These drugs were anti-serotonin drugs and included Methysergide and Cyproheptadine (CH). Dr. Lehrer found that patients responded symptomatically to these drugs and that the abnormal findings on balance examination responded as well. These drugs were anti-5HT2 drugs, as was Azatadine, a derivative of CH, which is no longer available, and Pizotifen. Dr. Lehrer presented these findings to the American Neurotology Society in 1976[7] and classified these cases as vestibular neuronitis.

In 1976, the International Audiology Society had a conference, and there was a presentation by Professor A. Bosatra on the utilization of the stapedius muscle reflex in the diagnosis of the nucleo-reticular vestibular syndrome (NRVS). This vestibular disorder had been described by Arslan and Sala[8] in 1956 as a vestibular disorder with spinning and non-spinning symptoms. Congresso Soc Ital Lar Otol demonstrated that the disorder's pathologic site was in the brain stem by analyzing three parameters of the stapedius muscle reflex, the latency, the slope and the amplitude.[9,10] Medical literature illustrates that serotonin and serotonergic neurons were well represented in the brain stem.[11,12,13] These authors described a group of dizzy patients with sudden and transient seizures accompanied by feelings of blackout, which was interpreted as a group with spinning symptoms. They also described a second group of patients with no severe paroxysms in which the disequilibrium might take the form of "feeling top heavy" or "off balance" especially on standing or walking, which appears to be a group with non-spinning symptoms.

The vestibular neuron's dendrites extend from the inner ear, synapse in Scarpa's ganglion, and then exit the ganglion as the vestibular nerve, and then synapse again in the brain stem. If the dizziness originates in the inner ear, it could be termed as a labyrinthitis; if in the ganglion, as a ganglionitis; if in the nerve, as a neuritis; and if in the brain stem, as a brain stem affectation, which is where Dix and Hallpike postulated the pathology to lie based upon their results with galvanic stimulation. The use of vestibular neuronitis as a diagnostic category[14] was analyzed, and it was concluded that it was a broad and non-specific category, defined in that fashion by Dix and Hallpike, and could be dissected into vestibular neuritis (the spinning group) and NRVS (the non-spinning group). Therefore, the term vestibular neuronitis was discarded from classification and replaced with vestibular neuritis (VN) when the presentation is one of spinning, to describe the first group of patients described by Dix and Hallpike, and NRVS, in which the presentation is one of disequilibrium associated with non-spinning vestibular symptoms of the second group.

Howard House wrote an impressive paper on post stapedectomy perilymphatic fistulas in 1956.[15] He described these patients as suffering from an "artificial" endolymphatic hydrops (ELH). This made sense in that a leakage of perilymph could result in a secondary swelling of the endolymphatic space. The knowledge that such fistulas could occur in other situations came when Fee published his classic paper in 1968[16] on post-traumatic perilymphatic fistulas in three patients. Interest was aroused, and confirmation of Fee's report occurred in 1974 when Healy, Friedman, and Strong published a series of 40 patients with post-traumatic perilymphatic fistulas.[17] They stated their criteria for exploration but cautioned that they did not want to initiate a rash of tympanotomies in dizzy patients. There is only 135.21 cubic millimeters of perilymph in the inner ear, or 0.135cc.[18] Dr. Lehrer found leakages of fluid at either the oval or round windows, or both, and grafted these areas with ear lobe fat, and found that patients had resolution of the dizziness and imbalance which had afflicted them. A professional needs to look for this entity especially among those patients who had not improved on medical therapy with Methysergide or CH. The next revelation was the description of spontaneous perilymphatic fistulas by Stroud and Calcaterra.[19] This entity could now be said to occur with or without trauma.

The fistula test with a pneumatic otoscope was utilized at first. It became recognized that a more calibrated pressure was easily obtainable utilizing the impedance bridge. Utilizing the bridge, with both positive and negative pressure, became popular after Kohut, Waldorf, Haenel & Thompson[20] reported the increased sensitivity of negative pressure in this test. We now use 400 daPa, as Causse, Causse, & Bel[21] reported, both positive and negative.

There was initial skepticism about the fistula diagnosis, which has increased over the years. The fistula test was not consistent in many patients. There were tests for ELH, particularly the glycerin/glycerol dehydration test.[22,23] Practitioners started to broaden the scope of this test in order to test not only for hearing as was done for Meniere's disease, but for balance, examining the Quix and Romberg tests as well as the fistula test.[24] The professional continues to use a dehydration test, preferring Furosemide at this time based upon Weider and Johnson's[25] report, and utilizes glycerin only when there is sulfa allergy which could raise issues with the use of Furosemide. The experience has been that a positive dehydration test, in which the findings improve over the two hours of the Furosemide test or the three hours of the glycerin test, provides excellent clinical confirmation of the diagnosis of PLFS.

A paper published in 1982 by Brookes, Morrison, &

Booth[26] on the use of Acetazolamide in the diagnosis of MD found that, in patients with a history of symptomatic endolymphatic hydrops, 500mg of Acetazolamide given intravenously caused a temporary elevation in the pure tone and speech audiometric thresholds or an increase in the negative summating potential (SP) during transtympanic electrocochleography (ECochG), or so affect both in 66% of patients so tested. The authors opined that this effect could be secondary to an increase in plasma osmolality. However, the effect is more likely secondary to lowering perilymph pressure through reducing cerebrospinal fluid (CSF) pressure. The physiological explanation for this effect is that Acetazolamide, a carbonic anhydrase inhibitor, as is Topiramide, is known for its effect of decreasing CSF pressure and also decreases perilymphatic fluid pressure through the connection of the perilymphatic space with the CSF through the cochlear aqueduct. Patients whose findings worsened during the glycerin and Furosemide testing, and improved on the same testing utilizing Acetazolamide, could have perilymphatic hypertension (PLHT).[27,28] The distortion in the inner ear fluid compartments would be opposite to that of ELH, a mirror image of ELH. The symptoms presented by these patients were similar to the PLFS, but due to a distortion in which the perilymphatic compartment was distended, rather than the endolymphatic one. Successful treatment of these patients with Acetazolamide and Topiramate has clinically confirmed this entity.

The Classification System of Vestibular Disorders

Patients with vestibular disorders are among the most difficult to diagnose and treat in otolaryngology and head and neck surgery. Clinicians are well served by modern imaging studies such as CT and MRI in most areas of our specialty, but the utility of these diagnostic techniques has been limited although advances continue. An effective classification of these disorders would allow the clinician to correlate the information such that it could lead to a rational diagnosis and a rational, definitive treatment which would result in a stabilization of vestibular dysfunction by resolving a disturbance in function or by assisting in vestibular compensation. Dr. Lehrer first published such a schema in 1987.[29] Dr. Lehrer subsequently abandoned the term "vertigo" as the all-inclusive term for vestibular disorders as he discovered that many patients with vestibular disorders present with non-spinning symptoms. It thus becomes essential to describe the symptoms of patients with non-spinning symptoms and to define which patients truly have a vestibular disorder, and not a cardiac, autonomic or psychogenic cause of their symptoms. NcNally and Stuart[30] addressed this issue in their classic paper entitled "Vertigo from the Standpoint of the Otolaryngologist." These authors broadened the term "vertigo" to dizziness and stated that labyrinthine dizziness is not necessarily rotatory. They described patient reports of symptoms while undergoing caloric tests as faintness, unsteadiness, a rocking sensation, a staggering sensation, a sensation of weakness, a sensation of backward swaying, and a wavy sensation. There are many systems of classification that ascribe these types of symptoms as non-vestibular and often as non-organic or psychogenic. It is crucial for the physical and mental health of patients who have a true vestibular disorder that they be correctly identified and correctly treated. Dr. Lehrer has seen patients whose lives and self-images have been destroyed by ascribing their vestibular complaints to psychogenic causes. This is not to say that psychogenic dizziness does not occur, but to say that, in the absence of identification of a physical cause, it is often over-diagnosed. Dr. Lehrer examined the 11-year-old son of a radiologist whose complaint was non-spinning dizziness. He was referred to a psychiatrist who told the father and son that "it was all in your body and not in your mind." The child had NRVS and was treated with success. This patient did have identifiable imbalance which is the key to identifying a physical cause for vestibular symptoms.

What are the non-spinning vestibular symptoms? McNally and Stuart described some of them in their paper. Table 1 is a list of the various vestibular symptoms that Dr. Lehrer has usually encountered. These complaints are the starting point that suggests to the clinician that a careful and complete examination of the vestibular system is warranted. That evaluation starts with a history and physician examination.

Table 1

Vestibular Complaints

- Spinning
- Dizziness
- Blurred vision
- Lightheadedness
- Motion sickness
- Ear or head fullness
- Feeling drunk
- Dizzy turning in bed or on getting out of bed
- Feeling spacey or disoriented imbalance or disequilibrium
- Staggering
- Dizziness or imbalance in an elevator, escalator or treadmill
- Feeling tilted
- Feeling floor moving
- Swaying
- Disoriented in stores or malls
- Dizziness on standing up, bending or turning.

Mallinson and Longridge,[31] in another classic paper entitled "Specific Vocalized Complaints in Whiplash and Minor Head Injury Patients," state that their report delineates complaints voiced by their patients and legitimates them as being organic. These complaints are usually non-spinning.

They reported the complaints of patients and their symptoms, such as lightheadedness, feels like the car is moving when stopped, unsteady with eyes closed, like sea legs, unsteady, clumsy, blurry vision, out of focus, like being on a boat pitching in the water, and queasy after driving for an hour. Regarding psychiatric disorders in patients with these complaints, they support Furman and Jacobs'[32] "new narrow definition of psychiatric dizziness" which states that this diagnosis should only be used when dizziness occurs exclusively in combination with other symptoms as part of a recognized psychiatric disorder. Furman and Jacobs are quoted as urging extreme caution in making the diagnosis of psychiatric dizziness. Table 2 describes the classification that Dr. Lehrer presently utilizes and is based upon the system presented to the American Neurotology Society in 1992. Dr. Lehrer reported that 96% of 327 consecutive new patients seen in 1990 were classified successfully and that 97% of known outcomes accorded with predictions made on the basis of the assigned diagnosis. Dr. Lehrer continued to use this classification with success and has found few patients that he could not classify. Of course, new disorders such as semicircular canal dehiscence have been identified and are included in this article. The major gap between the classification herein presented and other classifications has been in patients with non-spinning symptoms, which are generally classified as unknown or psychogenic. Table 2 describes the present classification.

Table 2

Classification of Vestibular Disorders

Disorders in which vertigo is the most prominent symptom:

1. Vestibular Neuritis (VN)
2. Benign Paroxysmal Positional Vertigo (BPPV)
3. Meniere's disease (MD)
4. Wallenberg's Syndrome

Disorders in which non-spinning symptoms are most prominent:

1. Nucleo-Reticular Vestibular Syndrome (NRVS)
2. Perilymphatic Fistula Syndrome (PLFS)
3. Perilymphatic Hypertension (PLHT)
4. Central Vestibular
5. Vestibular Loss
6. Motion Sickness
7. Psychogenic
8. Vestibular Migraine

Miscellaneous:

1. Drug Induced
2. Cervical Vertigo
3. Neurovascular Compression Syndrome
4. Sudden Hearing Loss with Vertigo (SHL)
5. Recurrent Vestibulopathy
6. Semicircular Canal Dehiscence

Unclassified

The most common disorders seen in Dr. Lehrer's practice are NRVS, BPPV, PLFS, VN, and PLHT. They made up 78% of the cases that Dr. Lehrer reviewed and still make up the dominant portion of his practice. Nucleo-reticular vestibular syndrome is essentially an unknown entity outside of the Italian neuro-otologic community in which it was first described. It is common in Dr. Lehrer's practice and was the diagnosis established in the 11-year-old boy described above. It is often mistakenly ascribed to psychogenic causes. Perilymphatic fistula syndrome is called "a controversial syndrome" because there is no definitive test to define the syndrome, such as imaging or eye findings. The dehydration test has been definitive in Dr. Lehrer's experience and will be described. Vestibular neuritis is well described, but has, in his opinion, been mislabeled as neuronitis. Benign paroxysmal positional vertigo is well known and actively sought in all dizzy patients, if not by the physician then by the patients themselves, because it is widely reported in the lay press because of its dramatic treatment and high and prompt cure rate. Perilymphatic hypertension is less well known but is manageable with medications.

The Examination of the Patient

The history. When a patient presents with any one or several of the vestibular complaints described earlier in this article, a history of the onset, circumstances, and other factors that the patient feels would be relevant is obtained. The initial questioning of the nature of the complaint should focus on whether the symptoms were of a spinning or a non-spinning nature. Several of the disorders described in the classification will have both spinning and non-spinning symptoms. A history of imbalance should be sought if that complaint is not volunteered by the patient. Patients may present with the personally most bothersome symptom of a disorder, such as ear pain, ear fullness, pressure in the ears or head, hearing loss, lightheadedness, feeling drunk or dizzy, blurry vision, neck pain, feeling weak or tired, or imbalance. One should at least ask if they get dizzy getting into or out of bed, whether they have to hold on when washing their hair in the shower when their eyes are closed, whether they have difficulty on stairs, or on escalators or getting off an elevator, whether they have difficulty climbing out of a bathtub, whether they have dizziness on turning, bending or sudden twisting movements. Symptoms such as lightheadedness, wooziness or feeling spacey are often volunteered by the patient or may be elicited on further questioning. Dr. Lehrer initially strongly encourages the patient to explain the symptoms, which are quite often vague, before offering any ideas which can help the patient clarify their symptoms. It is not unusual for these ideas to be helpful to patients as they often have difficulty with explicitly describing what they are feeling. This is understandable, considering the variety and non-specificity of many of the complaints related to vestibular dysfunction.

Regarding the circumstances of the complaint, one should question the patient about the onset of the dizziness or other complaint, whether it followed head or neck trauma, whether it occurred after a fall, whether it started at a specific time or was gradual in onset, whether it followed an ear or sinus infection. One should also question the patient specifically about symptoms of hearing loss and tinnitus and ear or head fullness and relate those symptoms to the onset and occurrences of the dizziness.

There can be symptoms of visual system dysfunction associated with PLFS, NRVS and PLHT, so the patient should be questioned about dizziness in stores or malls, dizziness in scrolling on their computer or reading, dizziness or discomfort in crowds or watching movies.

There may be symptoms of cognitive dysfunction in PLFS, NRVS and PLHT. The patient should be asked about any difficulties with memory and concentration. They can be asked if they have difficulty understanding what they have read, whether they need to keep lists to remind them of what they have planned for the day, or whether they forget why they went into a room. Those patients with cognitive dysfunction are especially prone to anxiety although all patients with chronic vestibular symptoms will be anxious to some extent.

A history of other diseases or disorders should be sought, especially those that affect the vestibular system or brain, such as Lyme disease and multiple sclerosis. The medication list should be examined. Loop diuretics affect the inner ear more than the thiazides. Dr. Lehrer had a case in which a simple switch from Furosemide to Hydrochlothiazide (HCTZ) relieved the imbalance in a patient with PLHT who was being treated for hypertension. Dr. Lehrer has seen many patients with the reverse, patients in whom a change from HCTZ or Dyazide to a loop diuretic resolved the dizziness symptoms and the associated imbalance. Dr. Lehrer will discuss the greater efficacy of loop diuretics versus HCTZ in ELH. Topiramate and Acetazolamide are effective in treating PLHT as well as benign intracranial hypertension, but can have adverse effects on ELH, whether primary as in Meniere's disease or secondary as in PLFS. A patient with dizziness and imbalance might respond to a change of these various medications if they are found to be associated with vestibular symptoms. Minocycline, sometimes used in acne, can cause vertigo. Of course, patients who have been treated with an Aminoglycoside should be evaluated for ototoxicity.

The physical examination. The physical examination provides the key to the diagnoses of VN, BPPV, NRVS, PLFS and PLHT. Vestibular neuritis and BPPV are well described in the literature and texts, but the other three are not so well described. Perilymphatic fistula syndrome is mentioned but diminished by being described as "controversial," PLHT is hardly mentioned and therefore obscure, and NRVS is never mentioned in modern literature or texts. This situation has resulted in a vacuum which has been filled by often attributing psychogenic causation to many patients because of their well-understood anxieties about their diagnosis, their inability to function as they did in the past, and their mental health in a situation in which they have been advised that there is no physical cause and no specific treatment for their complaints. Patients suffering from vestibular complaints after head injury are regularly told by neurologists that, since the brain imaging was normal, the symptoms will resolve over time, usually two years, although there is literature[33] that describes such symptoms as persisting for up to 10 years, which connotes a chronic, incurable illness. It is important to emphasize that a repeatable finding of an abnormality in balance on the physical examination identifies the physical reality of the vestibular complaint to the examiner and the patient and is a giant step in arriving at a diagnosis of a condition which is treatable. One should know the layout and size of a house, but one cannot enter that house smoothly without a key. As described above, that key is the physician examination.

The patient with any of the vestibular complaints that have been described should have a complete examination of the ears, nose, throat and neck as well as a complete evaluation of the cranial nerves. Middle ear or mastoid infection or tumor can be associated with symptoms of inner ear extension, which must be taken very seriously as it can be life threatening at worst, and injurious to the hearing and balance systems at the least.

Regarding the neurotologic evaluation, as part of the cranial nerve evaluation, Dr. Lehrer addresses eye movements and eye signs; looks for spontaneous nystagmus, by having the patient, with open eyes, follow his index finger to their left side, then have their gaze move to the center and then to the right side; aware of end position nystagmus and, if nystagmus is present in the extreme lateral positions, will guide the eyes slightly centrally away from the end position and re-evaluate the finding. Dr. Lehrer examines upward and downward gaze for nystagmus as well. If an acute vestibular vertigo syndrome is suspected to be present, Dr. Lehrer will look for vestibular nystagmus with eyes closed if the eyes open examination is negative and will utilize Frenzel's spectacles, which eliminate the suppressive effect of eyes open testing. A videonystagmography (VNG) examination is available should there be the need to go further. Dr. Lehrer also performs the head impulse test (HIT) to evaluate the vestibular-ocular reflex. If nystagmus is present, it will be classified, as per the literature, as first degree if it is present only in the direction of gaze, as second degree if it is also present when the eyes are centered, and third degree if it is present in all directions of gaze. Vestibular nystagmus presents with a rhythmic slow and fast component. Its direction is defined by the direction of the fast component. Ocular and central types of nystagmus have other characteristics and have been well described in the various texts that will be referenced later in this article.

Dr. Lehrer then examines the balance, performing the

various tests always with the chin up slightly which increases the sensitivity of the tests. The first test is the sitting Quix test in which the patient is seated squarely in the examining chair, arms and index fingers extended, first with eyes open and then with eyes closed. The balance examinations always begin with eyes open and then with eyes closed. The eyes closed examinations should reveal more imbalance than with the eyes open. If they do not, Dr. Lehrer would conclude that the results of the exam are non-physiologic and could possibly represent psychiatric entities or malingering. Posturography examinations for non-physiologic imbalance rely on a similar disparity between simpler and more difficult portions of the testing.

As for the results of the sitting Quix test, Dr. Lehrer looks for shakiness or a sway of the body and/or fingers and arms. Sometimes the patient's arms will exhibit the discus thrower motion in which one arm raises while the other lowers. Sometimes the fingers and arms will drop, or the patient's body will tilt backwards and their arms will tilt upwards. If there is a sway on sitting, one should take more than the usual care on examining the patient standing as there is more likelihood of a fall standing when there is a sway, sitting. A tilt of the entire body is often seen in patients with severe imbalance, such as is seen in acute VN.

Dr. Lehrer then has the patient stand for the Romberg test, with feet totally together, arms at the sides or crossed over the chest (which makes the test more sensitive), and with eyes open. The patient can be examined wearing shoes or sneakers, but not with footwear that extends above the ankles as this could provide more stability and decrease the sensitivity of the test. The patient is examined first with eyes open and then with eyes closed. One looks for shakiness, even slight shakiness, or a sway or a tilt, or even falling. The exam is repeated several times with eyes open and then closed. The sequences should last about 15 seconds, which is long enough to reveal imbalance. Exams that last one to three seconds are too short to reveal imbalance in most cases and will result in missing the imbalance, which is the most important finding in the non-spinning disorders. The Quix test is performed next, with the patient standing with feet together, as in the Romberg, and directed to do so by being told to stand in the smallest space possible so as to increase the sensitivity of these tests. One could see imbalance with the feet together and then re-examine the patient with the feet only an inch or so apart and discover that the finding of imbalance is no longer present. Increasing the sensitivity of the balance evaluation is crucial to demonstrating the abnormality in balance, especially in young or athletic patients. The abnormal Quix test is one in which the patient becomes shaky, which can be observed by looking at the outstretched hands and fingers or at the body, especially the midline of the back and buttocks, and the legs, as one can often see the movements of those areas more easily than the upper extremity movements. The examiner is standing behind the patient, with an assistant standing in front, extending his or her arms and fingers opposite to the patient's fingers. One must stand behind the patient in order to observe the response to the balance testing body but also to protect and reassure the patient that they will not fall and to support them early on should they start to fall. Patients will reach out to support themselves or spread their feet protectively or in fear and must be firmly, but kindly, instructed and reassured that the examination is important and that they will not fall nor suffer injury. Establishing confidence in a patient that you will protect them is crucial in the performance of an accurate examination of the balance. Dr. Lehrer has seen what he calls a "switch effect" on the Quix test when the patient closes the eyes in that there is a slight but observable motion or shaking of the fingers which starts immediately after the eyes are closed. This is a positive finding.

The Quix test can be made more sensitive by repeating it after having the patient perform a tandem Romberg test in which they stand heel to toe and try to keep balanced. Dr. Lehrer has found the tandem Romberg to be too difficult a test for patients without pathologic imbalance and, therefore, does not depend upon it to reveal pathology. But it is valuable in increasing the sensitivity of the Quix test. Another technique to increase the sensitivity of the Quix is to have patients perform it after getting up quickly from positional testing. This introduces a dynamic element to the balance examination and, again, is quite useful in evaluating the young or athletic patient for imbalance. It should be noted that patients with only mild imbalance on examination may still have significant symptoms of a vestibular disorder. These techniques will increase existing imbalance as it introduces a dynamic element into the examination. The increased imbalance that is usually present when the patient has dynamically stood up after positional testing often serves to convince both them and the examiner that a real balance problem is present. This is helpful to the clinician in gaining the patient's confidence that something objective has been found after the fruitless search many of them have pursued before the present consultation, and it confirms the need for further evaluations that would be required to establish a definitive diagnosis.

The modified Romberg test, performed on a foam pad as described by Agarawal, Carey, Hoffman, Sklare, & Schubert[34] can be performed as part of the balance examination. The time to a fall or significant loss of balance has been calculated for normal, based upon age and sex, and can provide documentation of a loss of balance. The patient should not have rigid footwear on for this test; Dr. Lehrer provides operating room booties for footwear.

The visual system, the vestibular system and the proprioceptive system are the three sensory systems that have been classically described as providing the sensory input to the brain, especially the cerebellum, for orientation and balance. The sense of touch in the feet also contributes to balance and orientation. Cerebellar functions include

participating in balance and spatial orientation, mainly through its connections to the vestibular nuclei, to fine tune body and limb movements, and in motor planning and evaluating sensory information for action as well as cognitive functions.

The neurotologic examination should evaluate proprioceptive function in the foot and ankle by testing position sense and vibration sense. Formal neurophysiologic testing, such as electromyography of the lower extremities, should be performed in patients whose imbalance is not explained by vestibular findings alone.

If there are abnormalities in balance on the initial examination, Dr. Lehrer has the patient tested by an audiologist. A complete audiologic evaluation is performed first. Pure tone testing, tympanometry and speech reception thresholds and speech discrimination are performed in the standard manner. Stapedius muscle reflex (SMR) testing is performed by the methods described by Bosatra et al. Lehrer and Poole described experiences with this test in patients and labeled the test the Brian Stem Arc Study (BSAS).[35] Fistula testing completes the initial evaluations performed by the audiologist.

In the Lehrer and Poole paper, they described the stapedius muscle reflex pathway as outlined by Borg.[36] The afferent pathway is transmitted through the cochlear nerve into the brain stem, with the intermediate pathways in the brain stem as described by Borg synapsing in the facial nuclei, which provide the efferent pathway through the facial nerve to the stapedius muscle, resulting in its contraction, which can be measured by the impedance bridge. Bosatra, Rossolo, and Poli evaluated the latency, amplitude and slope of the reflex in patients with normal hearing and normal facial nerve function, as Lehrer and Poole did in their study, to determine if there was an abnormality in the brain stem pathway between the afferent and efferent arms of the reflex. Dr. Lehrer has considered asymmetry of the responses as an additional abnormal finding and described abnormalities in this reflex in patients with NRVS. An abnormality in the BSAS provides objective evidence of abnormal brain stem transmission and is the objective pathophysiologic finding in NRVS. Of course, an acoustic neuroma (AN) could result in an abnormal SMR. The clinician should be aware that this is another possible diagnosis to consider. Interestingly, however, Dr. Lehrer has seen patients with known ANs, with disequilibrium, respond to medication for NRVS. The test is generally performed with contralateral stimulation (ipsilateral stimulation can also be utilized) at 0.5K, 1K and 2K at a level of stimulus of 110dB. Patients with abnormal middle ear pressure, perforated ear drums or poor compliance on tympanometry or conductive hearing loss cannot be tested, and patients with sound sensitivity and significant tinnitus should not be tested. Dr. Lehrer has performed this study on patients with hearing loss and has been able to detect asymmetries in response which have proved diagnostic although one cannot expect to obtain a classically normal reflex response when there is a hearing loss.

Clinical fistula testing, utilizing a Politzer bag to deliver positive pressure to the external auditory canal, dates back at least to the early 20th century. The test was utilized to evaluate patients with mastoid infections for a fistula of the horizontal semicircular canal. The pneumatic otoscope is presently being utilized for this purpose. When perilymphatic microfistulas were described by Fee and thereafter by others, the pneumatic otoscope was utilized for diagnosis, with a positive finding of elicited nystagmus, either by direct visualization, or on ENG or VNG, being the objective evidence of a positive fistula test. In a classic paper, Kohut, Waldorf, Haenel, and Thompson[37] reported that they had found perilymphatic fistulas in patients without hearing loss and also described their positive findings in these cases, utilizing Hennebert's sign and symptom. He described patients with surgically proven perilymphatic fistulas who had symptoms of dizziness and imbalance on fistula testing, but not the sign of nystagmus. This makes sense as the pathologic opening into the inner ear was not in the area of the horizontal semicircular canal, but through microfissures described by Harada, Sando, and Myers[38] and Kohut, Himojosa, and Budetti[39] in the wall between the middle and inner ear in the areas of the vestibule and the saccular and utricular organs. Kohut et al.found that negative pressure was more sensitive than positive pressure in the evaluation of these patients. We have utilized the impedance bridge to perform the fistula test. The patient's ear is sealed, as for tympanometry, and three pressure sweeps of positive to negative pressure, utilizing 400 daPa, are made with the patient sitting, with eyes open and then eyes closed, and then standing, with feet together with eyes open and then eyes closed. Symptoms of dizziness, disorientation, nausea and even vomiting have occurred as positive findings. The audiologist stands behind the patient for the standing tests and notes responses of swaying, shakiness or falling. Positive symptoms or positive signs are described and reported as mildly, moderately or severely positive, using a system of plus signs. It should be emphasized that Dr. Lehrer considers a positive fistula test as a positive screening test, with the diagnosis to be confirmed on dehydration testing utilizing a dehydration test, first for ELH (Furosemide or glycerin) and then for PLHT (Acetazolamide) if there is worsening or an inconclusive result on the first test. If Dr. Lehrer suspects PLFS or PLHT despite a negative screening fistula test, he will proceed with a dehydration test. The need for a dehydration test arises when there is a failure to respond to treatment for NRVS or in a patient who falls on the Quix or Romberg, which is sometimes seen in BPPV but almost never seen in NRVS.

The testing for positional nystagmus and vertigo completes the initial physical examination. The patient is seated on the examining table in a position in which the head would extend over the edge of the table when the patient is placed in the full supine position. The patient is instructed to

keep the eyes open at all times, told not to panic if dizziness occurs, told that the dizziness usually will last for less than 30 seconds, and told to relax the neck such that full extension and full turning of the head and neck can be performed. The standard Dix-Hallpike examination is performed. If there is positional nystagmus of the peripheral vestibular type, Dr. Lehrer primarily utilizes the Epley Maneuver for treatment.[40]

One should be alert as to central lesions that can cause positional vertigo. In these cases, in Dr. Lehrer's experience, there is a disparity between the symptoms and the nystagmus seen. The patient may have nausea and vomiting with only slight positional nystagmus, or may have active nystagmus with minimal or no vertigo. The nystagmus may not be transient, but prolonged for much longer than the 30 seconds expected in BPPV or persistent, often lasting as long as the position is maintained. These patients should have Imaging studies to evaluate them for a central lesion.

Videonystagmography (VNG) or electronystagmography (ENG) is performed as a standard test. If there is a loss of function on caloric testing, this can provide diagnostic information as to vestibular loss which is seen especially after administration of ototoxic drugs such as Gentamycin. Dr. Lehrer has not observed specific diagnostic information other than vestibular loss from this test although abnormalities can be present and should be integrated into the evaluation of the patient. There may be a reduced caloric response in patients with VN, but, in his experience, the clinical evaluation is paramount. Dr. Lehrer does not diagnose BPPV on the basis of positional nystagmus seen on VNG or ENG; he relies on the positional nystagmus observed on the Dix-Hallpike physical exam in concert with the patient's description of symptoms of vertigo at the time of testing. It is the symptoms and the clinical findings that are, in Dr. Lehrer's experience, paramount. Dr. Lehrer has seen patients treated for BPPV on the basis of positional nystagmus found on these studies without improvement in their vestibular complaints, which, in these cases, are usually associated with imbalance which was not detected.

The Vestibular Disorders – The Spinning Disorders

Vestibular neuritis. These patients present with acute spinning vertigo, associated with nausea and vomiting. There are no auditory symptoms. Most of these patients have normal hearing. The vertigo is continuous, is only relieved by remaining motionless, and can last for hours, days or weeks. Patients seen early in Dr. Lehrer's practice had shorter courses than those seen recently. These patients display vestibular nystagmus, with the fast component directed away from the involved ear. The body sway is towards the involved ear (The Harmonic Symptom Complex). Schuknecht and Kitamura identified the pathology in the vestibular nerve.[41]

Dr. Lehrer utilizes Steroids for treatment and has found them more effective the shorter the delay between the onset of the symptoms and the initiation of treatment. He utilizes 60mg of Prednisone per day, taken in the morning with food as the initial treatment in ambulatory patients, and intravenous Decadron (8mg every 6 hours) initially in hospitalized patients. The positive effects of such treatments are resolution of the nausea and vomiting and spinning, which occurs more quickly in those treated more promptly. Anti-emetics such as Ondansetron are used as necessary. The general use of Meclizine as a treatment for VN and other vestibular disorders is based upon its anti-emetic properties and its vestibular suppressant effects. There have been concerns that vestibular suppressants will retard central compensation in VN. Dr. Lehrer treats ambulatory patients with the full 60mg dose of Prednisone for at least a week. If there is resolution of the nystagmus, the imbalance and the nausea and vomiting, Dr. Lehrer will gradually wean them off the drug. However, Dr. Lehrer advises the patient that, should symptoms reappear while they are decreasing the dosage, they should increase the dose to the previous level. This process can take several weeks, with the final dosage being 10mg every other day for 4 to 6 days. There have been cases in which the course is quite prolonged. In these cases, Dr. Lehrer has utilized intratympanic Decadron treatments twice a week until the vertigo has subsided. Hospitalized patients are weaned off the IV Decadron as their condition improves. Oral Prednisone is substituted if symptoms persist after the IV therapy is no longer necessary.

Some patients with VN continue to have prolonged imbalance after the nystagmus and vertigo and nausea have subsided. These patients do not respond to continued treatment with steroids. They often respond to CH. Dr. Lehrer's explanation is that the inflammation, which primarily affected the vestibular nerve, has extended into the brain stem. The vertigo, of course, is the dominant early symptom in these cases. But after the vertigo has subsided, the chronic disequilibrium symptoms and the finding of continued mild imbalance appear and persist. Thus, one should be aware of this non-spinning complication of VN in order to recognize it and afford appropriate treatment. Acute vertigo has been described by Grimm, Hemenway, Lebray, and Black[42] at the onset of the PLFS, and this syndrome should be also be kept in mind in cases of prolonged dizziness and disequilibrium.

Silvionemi utilized the term "vestibular neuronitis" but described VN. Subsequent studies have correctly identified this acute vertigo syndrome as VN. Dix and Hallpike described two groups of patients whom they labeled as vestibular neuronitis, the first group having symptoms that were consistent with spinning, and which Dr. Lehrer now classifies as VN, and a non-spinning group, which Dr. Lehrer now classifies as NRVS. As noted above, patients may suffer an inflammatory process which affects the vestibular nerve and extends into the brain stem, with VN being dominant at the onset with acute symptoms of spinning and nausea and vomiting, treatable with steroids, followed by symptoms of

persistent dizziness and mild disequilibrium secondary to NRVS, which is treatable with Pizotifen or CH. When VN symptoms change from acute vertigo to a chronic mild imbalance with various vestibular complaints, the patients should be suspected of suffering from NRVS rather than being labeled as having had a psychogenic reaction to the acute vertigo.

Benign paroxysmal positional vertigo. There has been voluminous literature on this subject since Epley's classic paper in 1992. Dr. Lehrer utilizes the Epley Maneuver for most patients with BPPV. It is an amazing treatment, and, not unexpectedly, widely publicized. Dr. Lehrer examines the patient from the head, as Dr. Epley has described, rather than from the side. It is more comfortable, and there is better control and visualization. Dr. Lehrer moves the patient gradually into the various positions, rather than abruptly. If a vestibular type of positional nystagmus is induced, the Epley Maneuver is performed, allowing for 20 to 30 seconds at least; after the nystagmus has ended, the maneuver is done by gradually shifting the head from the involved side to the opposite side as the first step and keeping the patient in that position for the duration of any rebound nystagmus, and then at least for another 30 seconds. The patient is then rotated on to their side, again the side opposite the pathology, and positioned with the side of the nose on the examining table. After at least 5 minutes in this position, the patient then sits up rapidly, with assistance and encouragement, bending far forward so as to move their face between their legs. Dr. Lehrer has them straddle the table with their legs so as to facilitate this quick move up and through a sitting position, followed quickly by bending the head forwards between the legs, with chin tucked into the into the chest. Dr. Lehrer describes this to a patient as going into the "crash position" as described in the air safety card instructions. Dr. Lehrer encourages deep breaths after this move and then has the patient sit up. The best result is no dizziness or vertigo, but slight dizziness is acceptable. If there is severe dizziness, especially if there is nystagmus, the patient is rested and then the maneuver is repeated. There is often no dizziness or nystagmus on repeating the maneuver. If there is imbalance noted on the physical examination prior to the maneuver, the balance is rechecked, and often the imbalance has resolved. Dr. Lehrer has found imbalance, even falling, in patients without symptoms of positional vertigo who have BPPV on examination, and they are treated with improvement, or even resolution of the imbalance on the post maneuver examination. As a result of these observations, Dr. Lehrer always performs positional examinations even if the patients deny positional symptoms. If the imbalance does not resolve on follow-up examination and the positional testing has become negative, the clinician should consider the concurrence of NRVS or PLFS with the BPPV and perform the appropriate evaluations.

The initial Epley Maneuver has been said to be successful in 90% of patients. The remaining 10% require repeated maneuvers, and almost all will be successfully treated. Dr. Lehrer utilizes a vibrator, or oscillator, behind the affected ear in patients with recurrence. Dr. Lehrer has used the barbecue roll technique for horizontal canal BPPV, but this entity often responds to the usual Epley Maneuver. Dr. Epley developed a chair which can be quite helpful in patients with limitations of neck and body motion such that they cannot fully turn into the various positions for examination and treatment. The patient stays relatively upright for 48 hours after the maneuver, sleeping with the head elevated at 45 degrees although some practitioners do not so instruct their patients.

The Brandt-Daroff[43] exercises are most helpful in those patients with persistent mild to moderate symptoms. Dr. Lehrer has utilized the Semont Maneuver[44] regularly in the past but prefers the Epley Maneuver for classic BPPV and recognizes the application of the Semont Maneuver in cases of apparent cupulolithiasis.

Meniere's disease. There are certain concepts and findings related to Meniere's disease (MD). Firstly, as described in the 1981 International Symposium on Meniere's disease,[45] it was initially described by Prospiere Meniere in 1861 as characterized by repeated attacks of vertigo, associated with tinnitus and fluctuating hearing loss. Secondly, it is a disorder in which the pathologic conditions of endolymphatic hydrops, distortions and ruptures of the membranous labyrinth, atrophy of sensory and neural tissues and vestibular fibrosis have been thoroughly documented.[46] Vestibular fibrosis can result in fibrous strands connecting the stapes footplate to the membranous labyrinth, occasionally forming a thick fibrous layer on the vestibular surface of the stapes footplate. This may result in a positive Hennebert's sign in about a third of ears with MD.[47] The symptom complex described by Meniere had not changed since his original description. Pfaltz and Matefi[48] stated that MD is described as a triad of symptoms, tinnitus, fluctuating hearing loss and vertigo, which can present as a complete clinical picture after one year but can be incomplete during the first year. It is usually unilateral, has typical audiometric patterns, and is typically distinguished by the aforementioned triad of symptoms. Dr. Lehrer describes the approach to this disorder labeled by Schuknecht[49] as a pathophysiologic entity whose symptomatology is based upon progressive ELH, whose symptoms are of an episodic nature, with the episodes most logically explained by breaks in the membrane walls. Schuknecht believed that otologists in general have been severely deluded in their assessment of therapies by the variable nature of the symptomatology. Jongkees[50] stated that such terms such as pseudo MD or para MD indicate a lack of knowledge regarding the diagnosis in a particular patient. Having served on the Hearing and Equilibrium Committee of the American Academy of Otolaryngology and Head and Neck Surgery that reported criteria for diagnosis of MD in 1995,[51] Dr. Lehrer was therefore exposed to the difficulties of strictly labeling the criteria for diagnosis of this

disorder and can report that the criteria were therefore graded as to reliability in diagnosis. Possible, probable, certain and definite were the categories chosen. Medicine is best viewed as an Art, as Hippocrates has stated. He described it as "The Most Noble of All the Arts," so we as physicians can take some consolation that our uncertainties are not ignoble, and we must do our best for our patients in an uncertain environment.

Dr. Harold Schuknecht stated that the history is crucial in establishing the diagnosis. There are four symptoms of inner ear disorders: hearing loss, vestibular symptoms, tinnitus, and ear fullness. These symptoms can present in various ways, together or separately. In MD, the symptoms have been described as occurring in tandem, especially the hearing loss, the vestibular symptom (vertigo in this disease) and the tinnitus, with ear fullness occurring as well in some attacks. These symptoms, occurring acutely as an attack, describe the classical presentation of an attack of MD. In classical MD, there must be multiple attacks. The attacks last for hours, not days, and not minutes. They recur at varied intervals. There can be varied periods in which there are no attacks, described as remissions of the disease. Dr. Schuknecht emphasized the importance of the history by stating that a clinician could essentially make this diagnosis over the telephone. The Academy report described above emphasized the occurrence of multiple attacks as necessary to consider the diagnosis. In patients with symptoms of a vestibular disorder, the four symptoms described above may present together, or separately, or in combination, but the pattern of the symptom complex and the pattern of the attacks in MD is unique. It is most important not to categorize all dizziness as MD or as a variant of MD. The diagnosis of MD should be established by history and the finding of a sensory hearing loss, in the low frequencies early in the disease, often progressing to a flat loss later in the disease. High tone hearing losses are unusual in Dr. Lehrer's experience. The criteria described above are the most recently approved by the Academy. Others are being proposed but have not been codified.

Confirmation of the pathophysiologic disturbance (ELH) is important in confirming the diagnosis. The auditory glycerin test[52,53] improves hearing by dehydrating the endolymph and consequently improves the pure tone and/or discrimination levels. ECoghG[54] can identify ELH. Dr. Lehrer utilizes a canal electrode for this study. Others have used tympanic and transtympanic electrodes. The Acetazolamide test has been described as being utilized for diagnosis of ELH by temporarily worsening the ELH by dehydrating the perilymph, with positive tests being elevation of the pure and speech thresholds and increasing the negative SP on ECochG. The use of these tests has been questioned by some clinicians since they are not accurate in all cases. Dr. Lehrer's approach to the diagnosis of vestibular disorders is to utilize the history and focused testing and examinations, including imaging when appropriate, to arrive at a diagnosis that is classifiable according to the schema that Dr. Lehrer has herein presented. This will provide the opportunity for definitive treatment of a defined disorder or disease, and, in Dr. Lehrer's experience, has led to better outcomes in the management of vestibular disorders. There have been recent advances in the detection of ELH by Nakashima et al.[55] who have been able to identify ELH, utilizing 3 Tesla MRI studies in concert with intratympanic or IV Gadolinium. Kato et al.[56] have also correlated abnormal results on multifrequency tympanometry (MFT)[57] with the aforementioned MRI studies, allowing the clinician to utilize MFT in the evaluation of ELH. The identification of ELH does not necessarily confirm the presence of MD as the history and the physical and audiometric test results are still a necessary part of the diagnostic endeavor. PLFS is also associated with ELH. Dr. Lehrer has identified ELH in patients with PLFS on these MRI studies and on MFT, as well as by the dehydration tests described earlier in this article. Dr. Lehrer's hope is that advances will continue to be developed to assist in the definitive diagnosis of all vestibular disorders, especially those that his classification has added to the usual list of these disorders.

The medical management of MD should begin with an evaluation of metabolic factors that can predispose or aggravate the disease. Lehrer and Poole reported[58] that hyperinsulinism, impaired glucose tolerance and hypertriglyceridemia could result in vestibular symptoms in patients with and without MD and be successfully treated with diet. This pattern of abnormalities had been described by Olefsky, Farquhar, and Reaven[59] as a risk factor for atherosclerotic heart disease and had been labeled as Syndrome X in the 1980s and is now known as metabolic syndrome. Spencer[60] described hyperlipoproteinemia as a risk factor for inner ear disease. Hypothyroidism has been identified as an aggravating factor in Meniere's disease. Proper identification and treatment of such metabolic disorders provides the foundation for management of MD and other chronic vestibular disorders and can result in relief and/or resolution of symptoms. A low-salt diet should be instituted if the patient is not already limiting salt intake. Dr. Lehrer has not found it necessary to place patients on a low-sodium diet or to restrict caffeine, alcohol or smoking. If foods are found to be a trigger, they should be restricted. Inhalant allergies can aggravate MD and should be controlled.

The use of diuretics has long been a mainstay of treatment. Dr. Lehrer has found that the loop diuretics, which include Furesomide, Torsemide, Bumetanide, and Ethacrynic Acid (the only non-sulfa diuretic of this class of drugs), are more effective than the Thiazides, which are frequently prescribed, especially Dyazide because of the potassium-sparing effect of Triamterene. Dr. Lehrer's review of research studies on ELH supports his use of loop diuretics as these studies have utilized Furesomide rather than HCTZ.[61,62] Dr. Lehrer has successfully prescribed a loop diuretic as initial therapy without performing more than a

history and audiogram for patients who are in the midst of a crisis of MD, suffering attacks several days a week for months. These patients have usually been taking Dyazide without success and have responded to the loop diuretic in a short time. Dr. Lehrer has successfully utilized loop diuretics in MD patients who have failed chronic management with Dyazide. It is mentioned again that Acetazolamide actually increases the relative volume of endolymph because of its action on decreasing the cerebrospinal fluid volume. Dr. Lehrer would conclude that those patients who improve on this drug might be suffering from perilymphatic hypertension rather than ELH and MD.

Dr. Lehrer has found dilute sublingual histamine therapy, as described by Shambaugh,[63] a very effective treatment in about half the patients that he treats for MD. Those who do respond are subsequently treated indefinitely with a benign drug which can improve the tinnitus and vertigo, and, sometimes improve the hearing. The concurrence regarding the efficacy of this treatment is based upon experiences with patients who discontinued the treatment for one reason or another, had a recurrence of symptoms, especially an increase in their tinnitus, and then improved shortly after restarting the therapy. The key signal of change is the tinnitus symptom. If that symptom improves, it indicates that the therapy is appropriate and successful. Vertigo attacks have not occurred in patients if the tinnitus improves. The hearing may or may not improve. In about half the patients, there will be no change in the symptoms despite increasing the dose, even to stock levels. Histamine therapy is unhelpful in these cases. If symptoms are worsened as described above, Dr. Lehrer concludes histamine is active in the disease process, and adjusts the dose accordingly. The initial maintenance dose is 3 drops daily. The dose can be increased, based upon the clinical response (or lack of response). Histamine is seldom utilized, but it is worth the effort to employ as it is very effective if it is the appropriate drug. The theory would be that exogenous histamine would decrease endogenous histamine from being released in the inner ear and provoking the ELH. An exogenous overdose would itself provoke the MD symptoms. If histamine were not a provoking agent, there would be no effect on the inner ear with any dosage utilized. Betahistine (Serc and other trade names) tablets or solutions are also utilized in MD, especially in Europe, but are only available in the United States through compounding. Brandt[64] has described Betahistine's positive effect in patients with MD as being due to its action on histamine receptors. A vestibular suppressant such as Meclizine can be helpful in a minority of patients. The patients that Dr. Lehrer sees with histories of persistent vestibular disorders have generally been on Meclizine at some point, but it has not been helpful other than its sedating and anti-nausea properties. Many other anti-nausea drugs are used in MD and other vestibular disorders, but their effects are limited to their anti-nausea properties.

In patients with useful hearing in the involved ear, the most common intervention utilized for recalcitrant MD, with frequent attacks, is intratympanic Gentamycin (ITG), which has been quite efficacious. Dr. Lehrer performs one injection and then follows the patient. Others have performed several injections in one week and then follow the patient. There is a risk of inducing vertigo as a result of acutely reducing vestibular function in the injected ear. Dr. Lehrer's first case suffered from moderate vertigo for six weeks after one injection and then improved. His approach is to provide follow-up injections on a weekly basis if persistent vertigo attacks recur. ITG has largely replaced vestibular nerve section, which was often quite effective, but was often followed by imbalance which replaced acute vertigo attacks as the persistent symptom. Endolymphatic sac surgery (ELSS) is still widely utilized for recalcitrant MD[65] despite the "sham study" of Thomsen.[66] Dr. Lehrer utilizes ELSS in the management of the secondary ELH that occurs in selected cases of the PLFS.

In patients without useful hearing in the affected ear, destructive surgery of the entire labyrinth can be performed. These procedures would include either a formal labyrinthectomy by a transmastoid approach or a destructive labyrinthectomy by a middle ear approach. The transmastoid labyrinthectomy can, on occasion, fail to completely destroy the vestibular dysfunction in the operated ear. The procedure entails stapedectomy, followed by removal of the utricular and saccular organs as well as the ampullae of the semicircular canals with a right angle hook. This is well described in the literature and various texts. At the end of the procedure, Dr. Lehrer places ear lobe fat grafts soaked in Gentamycin into the vestibule, filling it completely. It is Dr. Lehrer's opinion that, by inducing a fibrous blockade in the vestibule, the movements of the inner ear fluids which stimulate the labyrinthine cells will be blocked such as to prevent stimulation. The Gentamycin will add to the destructive procedure by its toxic effects.

Wallenberg's syndrome and other central vestibular disorders. Wallenberg's syndrome results from an infarction of the brain stem, and in some cases, of the labyrinth as well. The patient presents with acute vertigo and a pulling of the body to the affected side. The patient also presents with nausea and vomiting, intractable hiccupping, ipsilateral facial pain, and diplopia, dysphagia, and dysphonia. Nystagmus is usually present towards the side of the lesion with the eyes closed but can be opposite in direction with the eyes open and fixated. There are multiple neurologic abnormalities on examination which may include ipsilateral loss of pain and temperature sensation on the face and an ipsilateral Horner's syndrome, ipsilateral paralysis of the palate, pharynx and larynx, ipsilateral facial and external rectus weakness, ipsilateral cerebellar signs and contralateral loss of pain and temperature sensation on the body. The vertigo is similar to that seen in vestibular neuritis, but the gravity of the patient's condition, and the abnormal neurologic symptoms and physical findings in Wallenberg's syndrome, are such that the

neurotologist should be able to differentiate this central nervous system disaster from the peripheral illness of vestibular neuritis.

As Dr. Lehrer stated earlier in this article, he does not intend to be encyclopedic. He is describing disorders and diseases which are principally peripheral and with which he has had the most experience and interest in sharing with the reader. There are excellent texts which describe the central vestibular disorders. Dr. Lehrer refers the interested reader to such outstanding texts written by professors such as Thomas Brandt,[67] Robert Baloh,[68] and Peter Rudge[69] and Robert Baloh and Vincente Honrubia[70] in order to review the panoply of central vestibular disorders.

This article was written to help describe some disorders that are omitted from the usual classifications and to emphasize the value of identifying and treating these disorders which are present in a large proportion of patients who present to Dr. Lehrer with the non-spinning vestibular symptoms, well described by McNally and Stuart, and Mallinson and Longridge. The non-spinning symptoms described by these authors are symptoms of a vestibular disorder. The clinician, when presented with a patient with these symptoms, should be aware that these patients can be suffering from a vestibular disorder, and, on the basis of what will be described below, perform the evaluations that can lead to a treatable definitive diagnosis in most patients. It has been Dr. Lehrer's experience that the non-spinning disorders have been the most difficult to diagnose and are the least recognized category of vestibular disorders.

The aim of the next section is to provide the clinician with an evidence-based approach to the diagnosis of these disorders as well as an experience-based approach to treatment and management. A clinician who is armed with the information that will be provided can believe that patients presenting with non-spinning symptoms can have a vestibular disorder. He or she can then proceed to an evaluation which takes abnormal eye movements into account but which also includes a careful and complete examination of the balance. The clinician can then utilize the test protocol which will be described to define the vestibular abnormalities and then be in a position to offer effective treatments.

The Vestibular Disorders – The Non-spinning Disorders

The nucleo-reticular vestibular syndrome. The first and most common of these disorders is the NRVS. As stated earlier, Dix and Hallpike described vestibular neuronitis as a "vertigo" syndrome, with the conspicuous absence of cochlear signs and symptoms. They described it as an organic disease confined to the vestibular apparatus localized to its peripheral nervous pathways up to, and including, the vestibular nuclei in the brain stem. They utilized galvanic testing to determine that the lesion was present in Scarpa's ganglion or the vestibular neurons. Their hypothesis was that there was a central affection of the vestibular neurones. In their introduction to the topic, they stated that the condition was confined to the vestibular apparatus and localized to its peripheral nervous pathways up to and including the vestibular nuclei in the brain stem. They described two groups of patients: one group with sudden and transient seizures accompanied by sensations of blackout and the other without severe paroxysms but disequilibrium with symptoms of feeling "top heavy" or "off balance" particularly when walking or standing. The disequilibrium was aggravated by head movements of all kinds. Spinning symptoms were not described in the second group, or in the first group. However, authors such as Silvoniemi interpreted the sudden seizures as true vertigo and labeled what has been come to be known as vestibular neuritis as vestibular neuronitis, which has led to confusion and clouding of the true nature of the disorder that Dix and Hallpike labeled as vestibular neuronitis, which they describe as follows: "When, therefore, it came to naming the condition, we required a term comprehensive enough to encompass this uncertainty. We chose the name 'vestibular neuronitis' and have since continued to use it." The uncertainty that Dix and Hallpike confessed to in 1952 can now be clarified by dissecting the term "neuronitis" into vestibular neuritis (an affection of the nerve) and the nucleo-reticular vestibular syndrome (an affection of the brain stem), as described in a paper so entitled.[71] It is those patients that Dix and Hallpike described as suffering from disequilibrium with vague, but vestibular, symptoms of feeling top heavy and off balance or some of the vague symptoms, described by McNally and Stuart and Mallinson and Longridge, that are suffering from NRVS.

The term NRVS was originated by Arslan and Sala in 1956. Bosatra et al. localized the pathophysiology of NRVS to the brain stem by utilizing the suprathreshold stapedius muscle reflex test (SSMR) in the 1970s by analyzing the parameters of latency, slope and amplitude of the reflex in patients with normal hearing. They found abnormalities in one or more of these parameters in patients presenting with the non-spinning symptoms described in this article. With the assistance of Professor Bosatra, Dr. Lehrer installed the necessary equipment for the SSMR and published the results in 1981.[72] Dr. Lehrer now had an objective test to localize pathology in the brain stem in patients who presented with symptoms of NRVS and who had abnormal findings on the Quix and Romberg tests. As described above, these examinations must be performed with care as the objective physical finding is crucial in establishing the imbalance that must be demonstrated to link the symptoms with an abnormality in the balance system. The Quix and Romberg tests have been described earlier in this article, and the methods of increasing the sensitivity of the tests have been described as well. A difference of 15 seconds in the examination of the balance can make the difference between leading the clinician on the correct path to diagnosis or missing the objective finding and having to ascribe the symptoms to a non-organic condition.

The approach to the patient with any non-spinning

vestibular symptoms begins with listening to the patient's complaints, allowing them to describe the onset, course, treatment, and any other factors which they feel are relevant to their complaints. These symptoms are notoriously difficult to describe, and, consequently, the patient should be encouraged to strive to define them as accurately as they can before the clinician injects common descriptions such as lightheadedness, wooziness, or spaciness into the conversation. The patients can be specifically questioned as to whether they lose balance in the shower with their eyes closed, whether they have to stabilize themselves on getting out of bed, or feel dizzy on turning in bed such as to look at their alarm clock, whether they lose balance on turning quickly when standing, or whether they have blurry vision, hearing loss, fullness or pain in the ears, or tinnitus. The patient may describe any of the various non-spinning vestibular symptoms previously described. Dizziness questionnaires can be utilized as well but should include both spinning and non-spinning symptoms. Non-spinning syndromes such as NRVS, PLFS and PLHT can occur spontaneously or follow head or neck trauma, with the onset of symptoms often being delayed, even for up to 6 months.[73] Nucleo-reticular vestibular syndrome can occur concurrently with VN or BPPV, with the NRVS symptoms and findings being present after the spinning symptoms of the other disorder resolve.

The patient can be specifically questioned as to whether they lose balance in the shower with their eyes closed, whether they have to stabilize themselves on getting out of bed, whether they lose balance on turning quickly, or whether they have blurry vison, hearing loss, fullness or pain in the ears, or tinnitus. The patient may describe any of the various non-spinning vestibular symptoms previously described.

The physical examination of the vestibular system, which has been described above, should then be performed. The clinician can initially identify findings on either the eye examinations, the examination of the balance or on positional testing in order to objectively verify the presence of a dysfunction of the vestibular system.

Dr. Lehrer's testing includes a complete audiometric evaluation, tympanometry (abnormal tympanometry precludes an accurate SSMR test), SSMR and a fistula test (which will be discussed in the next section), and is described in a request form as a vertigo evaluation. Normal hearing, normal tympanometry, negative fistula testing and an abnormal SSMR in a patient with the non-spinning symptoms described above who can be demonstrated to have imbalance on the Romberg and/or Quix tests can be diagnosed with NRVS. At this time that the imbalance in NRVS is mild, the patient does not fall on the Romberg or Quix tests. It must be emphasized that the clinician or an assistant must stand behind the patient to protect them from falling during these tests and also to instill confidence in the patient such that they will perform the test without spreading their feet or stepping or holding on to a countertop to prevent themselves from swaying or falling. The balance examinations demand patience from the clinician and confidence from the patient.

Dr. Lehrer utilizes otoacoustic emissions testing, with analysis of suppression in those patients who are intolerant of loud sounds or those patients with significant tinnitus who should not be exposed to the high-intensity tones utilized in the BSAS. Normally, emissions will be suppressed by contralateral masking, the pathway to suppression thought to be via the medial olivocochlear bundle and the inferior vestibular nerve in the brain stem. A lack of suppression is interpreted as a sign of a brain stem abnormality consistent with NRVS. The findings have not been published but find support from a poster by Koo, Chang, Song, and Kim[74] that described reduced responses on DPOAE testing with suppression in patients with VN.

The treatment of NRVS is medical.[75] Anti-serotonin drugs such as CH or Pizotifen (which are anti-migraine drugs) are utilized. Positive effects on the symptoms may not occur for 4 to 6 weeks but may occur sooner in many and much sooner in a few. The patient is re-examined three to four weeks after instituting therapy. If there is some improvement in the imbalance but a persistence of symptoms, the dosage can be gradually increased. Each patient requires individual management. The plan of treatment is to resolve the symptoms and abnormal physical findings completely, which takes one to two months on average; then plateau the dosage of the drug for the length of time that the symptoms had been present before therapy was instituted; and then phase off gradually over three to four weeks, restoring the dosage to the previous level if symptoms reoccur. Patients may stay on the drugs indefinitely, especially if their symptoms recur on decreasing or discontinuing the drugs as the drugs are safe. Patients who have successfully discontinued the drugs may use them as a "rescue therapy" should symptoms reoccur. In these cases, the use of the drugs is generally short term.

Dr. Lehrer began utilizing anti-serotonin drugs because of Dr. Gilbert's paper in which he suggested utilizing anti-migraine drugs for the treatment of vertigo. Migrainous vertigo was described by Liveing[76] in his classic publication. The attacks he described were truly vertiginous. Migrainous dizziness has been described as an entity and is included today in most classifications of vestibular disorders and will be addressed later in this article. The symptoms are variable and the connections to migrainous headaches are inconsistent. The migraine generator has been described as possibly being in the cerebral vasculature or in the brain stem. Diener demonstrated that the generator was in the brain stem, localizing it in the dorsal raphe nuclei.[77] This would indicate that the link between migraine and dizziness may well be in serotonergic mechanisms, as reflected by the density of serotonergic neurons in the brainstem, especially in the raphe nuclei, which have connections to the vestibular system, as will be described in the section on vestibular

migraine. At this time, it is important to remind the reader that, as described above, the pathologic site of NRVS has been localized in the brain stem. CH and Pizotifen have been demonstrated to act centrally on serotonin[78] as well as other central neurotransmitters, are known anti-migraine drugs, and have been described above as effective in NRVS.

The fistula test is utilized as a screening test; the more definitive dehydration tests will be discussed in the next section. Patients with NRVS present with the same symptoms as those with the PLFS and PLHT. The non-spinning symptoms described in this article apply to many disorders. There are patients with PLFS or PLHT with negative fistula tests who are consequently diagnosed as NRVS and unsuccessfully treated. Abnormal SSMRs can occur in PLFS, as described in Lehrer and Poole's 1981 paper on the SSMR, and in PLHT as well as in NRVS. The clinician should not exclude PLFS and PLHT on the basis of a negative fistula test. If a patient does not respond to therapy for apparent NRVS, despite increasing the dosage of medication, the presence of a disorder of the inner ear fluid system should be explored. Fistula tests should be repeated and further tests of the inner ear fluid system performed before the clinician decides that the patient's disorder is untreatable. These evaluations will be discussed in the next section.

Perilymphatic fistula syndrome. Fistulas of the horizontal semicircular canal have been recognized for over a century as arising from erosion of its bony covering by cholesteatoma. Fistula testing for this complication of mastoid infection utilized positive pressure induced by tragal compression or a Politzer Bag or more recently by a pneumatic otoscope. A positive test results from the active mobilization of perilymph through the defect and induces vertigo and/or nystagmus resulting from the applied pressure.

Fistulas did occur through oval window tissue or gelfoam seals in the 1950s after total stapedectomies for otosclerosis. Dr. Howard House described the resultant pathophysiolology of these microfistulas as an "artificial endolymphatic hydrops." Multiple clinical and research studies have confirmed the presence of ELH in microfistulas.[79,80,81,82] Fistula testing with a pneumatic otoscope is limited to positive pressure. As described, Dr. Lehrer utilizes the impedance bridge to standardize the pressures used and applies both positive and negative pressures in this test to increase its sensitivity, as Kohut has described.

The contemporary issue of fistulas of the inner ear focuses on the so-called microfistulas from the inner ear to the middle ear through preformed pathways so well described by Harada and Kohut. The mechanism of a positive fistula test has been posited by Nadol[83] as being secondary to vestibular fibrosis. The occurrence of adhesions noted in the areas of the oval and round windows in patients with PLFS could be viewed as the analogue of the inflammatory response in the vestibule of the inner ear.

The history is taken in the same manner as that performed for NRVS. There may be a history of initial spinning in PLFS, especially in post-traumatic cases as described by Grimm et al. Cognitive dysfunction has been described by Grimm et al. in cases of PLFS. Cognitive dysfunction has been described as occurring when there is vestibular dysfunction.[84,85] Dr. Lehrer has seen such complications in PLHT and NRVS as well as in PLFS. Questions about memory and concentration are relevant to the history in these disorders. The post trauma vision syndrome[86] in which the vision is intact but visual disorientations occur, especially in stores, malls or crowded moving environments, is not uncommonly associated with post traumatic PLFS and PLHT and can be present even in non-traumatic cases. Questions about intolerance to bright or fluorescent lights, dizziness in stores or malls, difficulties on computers, especially when scrolling, difficulty reading, or dizziness on escalators are appropriate, focusing on visual system dysfunction in addition to eliciting the already described vestibular complaints. The complication of visual system dysfunction can prevent vestibular compensation and prolong vestibular system dysfunction, even in a patient whose vestibular injury has been successfully addressed. It is crucial for the neurotologist to identify patients with visual system dysfunction when it presents concurrently with vestibular dysfunction so that appropriate treatment is afforded.

The vestibular physical examination has already been fully discussed. It is important to mention that patients with PLFS and PLHT are more likely to fall on Romberg and Quix testing as well as on the modified Romberg test. It is most important, as described previously, to protect and reassure the patient during the balance examination by standing close to the patient and even holding one's arms out to thus protect the patient posteriorly and laterally while the assistant protects the patient from the front and projects his or her arms and index fingers opposite to the patient's to better demonstrate a sway on the Quix test. Once again, it is important to objectively demonstrate that imbalance is present and that it is almost always repeatable on repeated sequences during the initial examination and on follow-up examinations. It is recommended to repeat the balance tests, with only seconds between each sequence, in order to increase sensitivity and to confirm the presence and direction of the observed imbalance.

The balance examination should be viewed critically to confirm that it is physiological, especially in post-traumatic or medicolegal cases. The imbalance that is noted should be worse when the examination places more stress on maintenance of balance. This would mean that the patient exhibits more imbalance with the eyes closed than with the eyes open and worse balance on the foam than on a smooth surface, such as the floor of the examining room. Dynamic posturography evaluation can be helpful if the examiner feels that the imbalance is not physiological.[87] This is a more

formal and quantitative examination but would utilize the same principle of noting whether the balance is better on easier conditions and worse on more difficult conditions. Aphysiologic imbalance can be secondary to psychological disorders, secondary gain situations, or malingering. The physician must use his or her judgment in evaluating the results as to the cause of the aphysiologic results should they occur. Regarding the office physical examination of the balance, imbalance is determined to be physiologic when the patient makes an effort to maintain balance at the beginning of the examination and then slowly exhibits a sway or tilt and tries to regain their centric position during the examination and exhibits a shakiness that is obviously not orchestrated. If they fall only with eyes closed but not with eyes open, or have increased imbalance or lose their balance more quickly with eyes closed than eyes open, I would judge that to be physiologic. The effort expended in keeping the balance is important in determining a physiologic response, as is the observation that there is better balance on easier conditions with less effort expended, and more effort and worse balance on more difficult tests.

After the history and physical examination are performed, the patient undergoes a complete audiologic evaluation, tympanometry, the BSAS and fistula testing. Hearing loss may or may not be present; but when there is a unilateral hearing loss, that ear can be identified as the pathologic ear with more confidence. Identifying the more involved ear also poses less of a dilemma when fistula testing is positive in only one ear, and is definite, or when a patient clearly identifies one ear as more involved than the other because of symptoms of pressure and fullness, or pain and tinnitus, or even a wet feeling. Other methods of identifying the more involved ear include comparing bilaterally positive fistula testing and identifying the ear that is consistently more positive, performing ECochG to identify an abnormal SP/AP ratio; or if both are abnormal, to identify the side which is more abnormal. The MFT has been found to be frequently abnormal in PLFS patients. The specialized 3 Tesla MRI already described can be performed to evaluate and compare the inner the ears for ELH. Caloric responses can also be compared, but Dr. Lehrer has not found these as helpful as the other evaluations and symptoms. The patient's subjective symptoms relating to one ear or the other are the most important factor in identifying the more involved ear.

Tympanometry and suprathreshold stapedial reflex testing (BSAS) are performed by the audiologist as well as fistula testing. Audiologists grade a positive fistula test as one, two or three plus and describe findings of imbalance that result from the pressures applied. If the fistula testing is positive to any extent, a dehydration test is performed.

As described, the glycerin dehydration test for Meniere's disease is adapted to evaluate patients for the ELH found in the PLFS by adding the balance examination and the fistula testing to the audiometric evaluation performed for the evaluation of the ELH in MD. Dr. Lehrer learned from Dr. Dudley Weider's report that Furosemide could also be utilized for this test and has since preferentially utilized Furosemide in patients who are not allergic to anti-bacterial sulfa drugs, with the knowledge that a history of allergy to sulfa antibacterial drugs may not preclude the use of loop diuretics.[88] The Furosemide test has minimal side effects as compared to the glycerin test which can result in nausea and vomiting. Glycerin ingestion also can result in headache, dryness of the throat and malaise, and a tremendous thirst even though no fluids are lost during this test. Patients are most grateful for the large drink of water given at the end of the test. If improvements occur, they will usually be reversed after rehydration. The dosage of glycerin is 1.2ml/kg given orally, as per Klockhoff and Snyder. The time frame for the glycerin test is 3 hours, with the patient being examined every 45 minutes. The time frame for the Furosemide test is 2 hours with the patient being examined every 30 minutes.

The format for the dehydration test is outlined below in Table 3. This format is utilized for the Furosemide, Acetazolamide and glycerin tests. The baseline examinations include a complete audiologic examination, pure tones, speech reception thresholds and discrimination scores, as well as fistula testing, which are performed after the initial balance examination and include testing of the balance on sitting Quix, standing Quix, Romberg and on the foam. The patient wears soft operating room booties throughout the test to standardize footwear, and stands, as for the initial balance examinations, in the smallest space possible, with feet tightly together at the toes and heels except for the examination on the Foam in which the feet are always comfortably apart.

FUROSEMIDE ACETAZOLAMIDE GLYCERINE Time administered: _____

RIGHT EAR

TIME	250	500	1K	2K	3K	4K	6K	8K

LEFT EAR

TIME	250	500	1K	2K	3K	4K	6K	8K

Time	SRT AD	% AD	RE:	SRT AS	% AS	RE:

Time	QUIX	QUIX SIT	STAND	FISTULA AD	FISTULA AS	FOAM

Table 3

Format for Dehydration Test

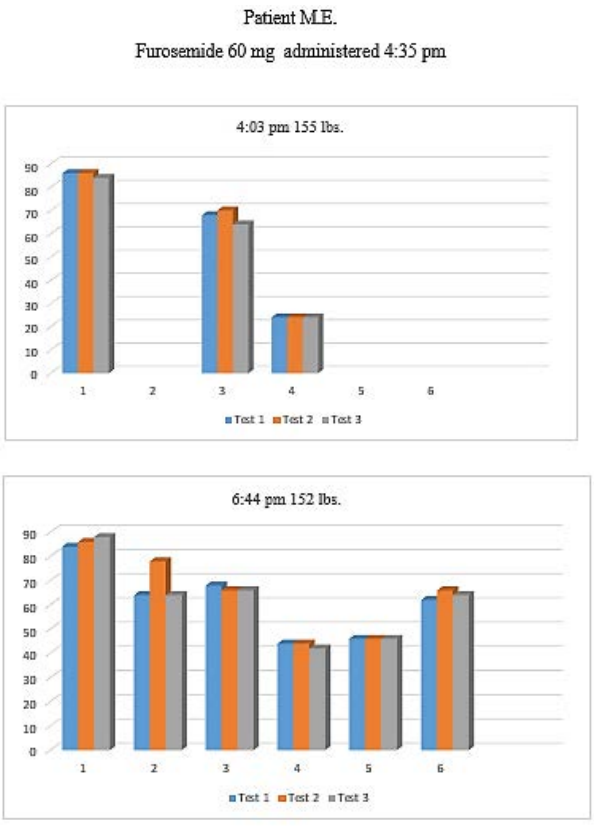
The dehydration test is generally performed initially utilizing Furosemide or glycerin in order to evaluate the patient for the ELH associated with the PLFS as this syndrome is much more common than PLHT. The dehydrating substance is administered after the baseline testing is complete. The patient is examined at 30-minute intervals during the Furosemide test and the Acetazolamide test, with a target time of 2 hours, sometimes longer, sometimes shorter depending upon the response, and at 45-minute intervals during the glycerin test, which has a target time of 3 hours. The patient's balance is examined by the clinician while the hearing and fistula testing is performed by the audiologist. One looks for improvement in one or more of the parameters to conclude that the test is positive for the ELH found in the PLFS. Often, it is the positive fistula testing and balance that improve as the initial hearing levels may be normal at baseline. If there is an improvement in the parameters, the patient rehydrates with water after the last sequence and to retest the balance 20 to 30 minutes after rehydration. Dr. Lehrer expects a return of the imbalance on retesting, which is explained as the wearing off of the dehydrating substance and the rehydration of the endolymphatic space by the water. This confirms to the examiner and the patient that the improvement in the imbalance was not the result of practice by the repetition of the balance sequences, but a physiological response to the test substance. Dr. Lehrer has seen patients who fall on the initial Quix and Romberg who are able to stand without falling or even swaying when the drug is active and exhibiting a return of imbalance after rehydration on both the glycerin and Furosemide tests. If there is worsening of the parameters, especially the balance, or a failure to improve, an Acetazolamide test is performed to evaluate the patient for PLHT.

Dr. Lehrer has utilized posturography with both glycerin and Furosemide, performing a baseline examination, administering the test substance, and then repeating the examination three hours later when glycerin is used and two hours later when Furosemide is used. The patient rehydrates with water after the last sequence. Posturography, performed with glycerin, has the advantage of decreasing the involvement of the physician as the audiologist administers the glycerin and performs the hearing testing and the fistula testing, and a technician can perform the posturography. The physician's involvement could be necessary if Furosemide were to be injected. The test results are displayed graphically, with the improvements clearly shown as well as the return of imbalance after rehydration.

Selected slides of posturographic examinations that were presented to the Neurocom Users Conference[89] are displayed in the figures below and will be discussed.

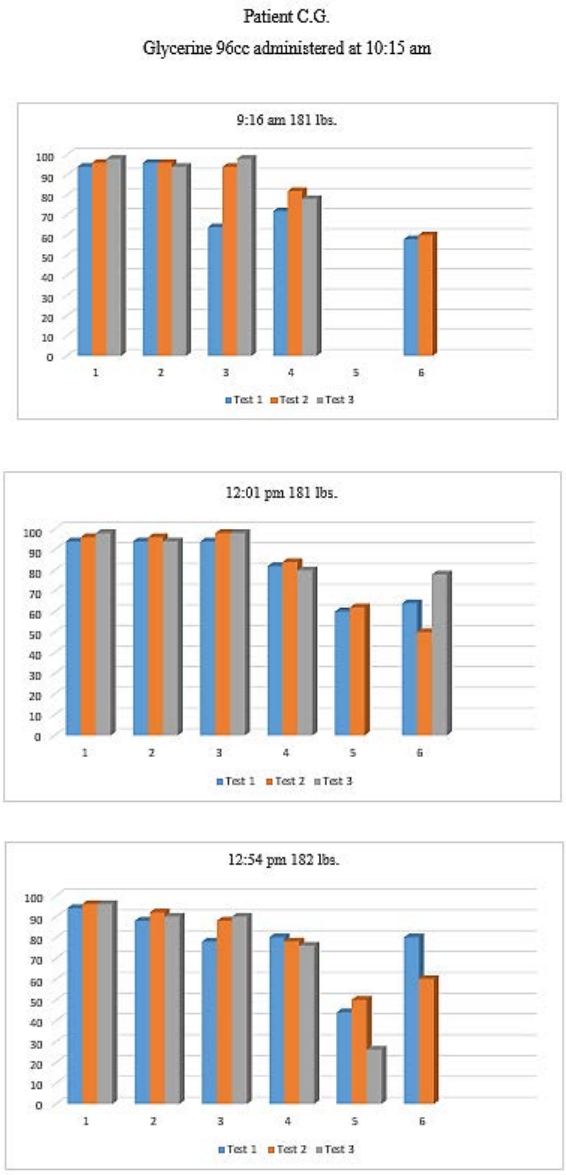
1. Patient M.E. had a baseline balance examination performed at 4:03 p.m., was administered 60mg of Furosemide by injection at 4:35 p.m. and re-examined at 6:44 p.m. One can see the abnormalities on the initial exam at 4:03 p.m., with falling on conditions 2 through 6, on all sequences in 2, 5, and 6, followed by the exam of 6:44 p.m. (2 hours, 9 minutes after Furosemide injection) which shows improved balance in all test conditions and dramatic improvement in conditions 2 and 6, in which there were no falls, and marked improvement in conditions 4 and 5, in which significantly improved test scores are displayed. The patient lost 3 pounds of fluids during the test (2 pounds are average). Foods and fluid intake are forbidden. The posturography setting was a 1.0 Gain for all cases.

Figure 1



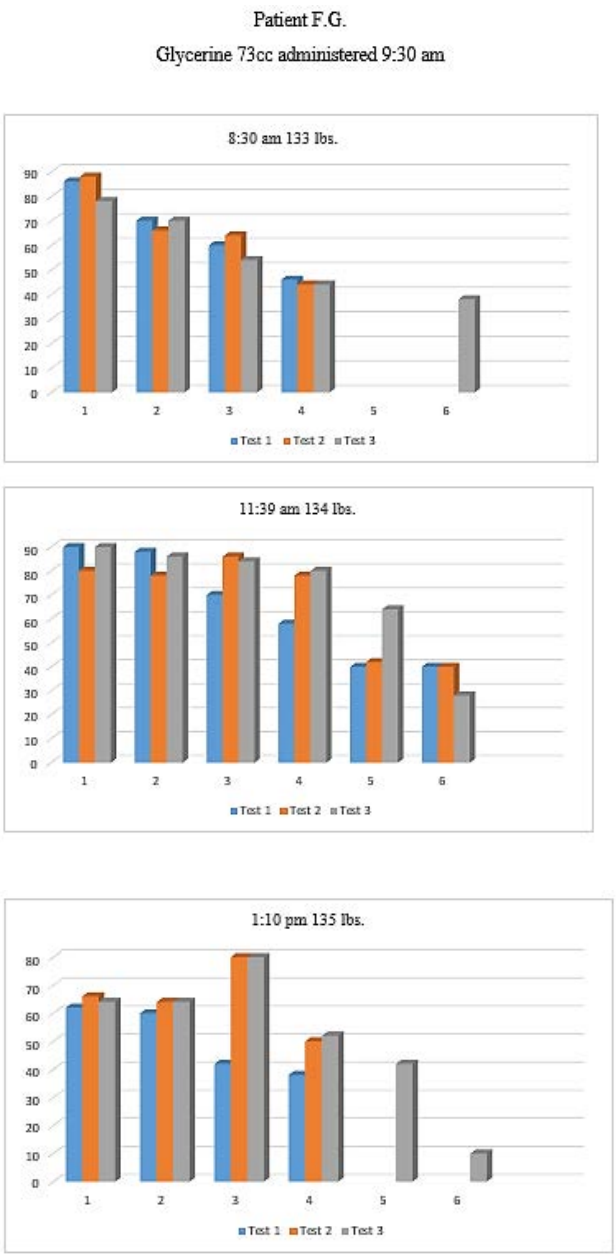
2. Patient C.G. had a baseline balance examination performed at 9:16 a.m. and was administered 96cc of glycerin (1.2ml/kg as per Klockhoff) at 10:15 a.m. One can see falls on conditions 5 and 6 at baseline. Testing at 12:01 p.m. shows improved scores (improved balance) in all conditions, with no falls on conditions 5 and 6. The weight of 181 pounds at baseline was unchanged; but after water was given at 12:35 p.m., there was a one-pound gain in weight at the time off the last test at 12:54 p.m. The test scores have now decreased in conditions 2 to 6, with a fall noted on condition 6 and a significant decrease in balance noted on condition 5. These tests demonstrate that both glycerin and Furosemide can demonstrably improve balance on posturography testing. It further shows that rehydration at the end of the test can significantly reverse the improvement in balance resulting from the glycerin. Dr. Lehrer has noted the same reversals on the Furosemide test. It is explained to the patient that the reversal demonstrates that their balance did not improve with practice, but was the result of a physiologic change in the inner ear fluid system from the administration of the test substance.

Figure 2



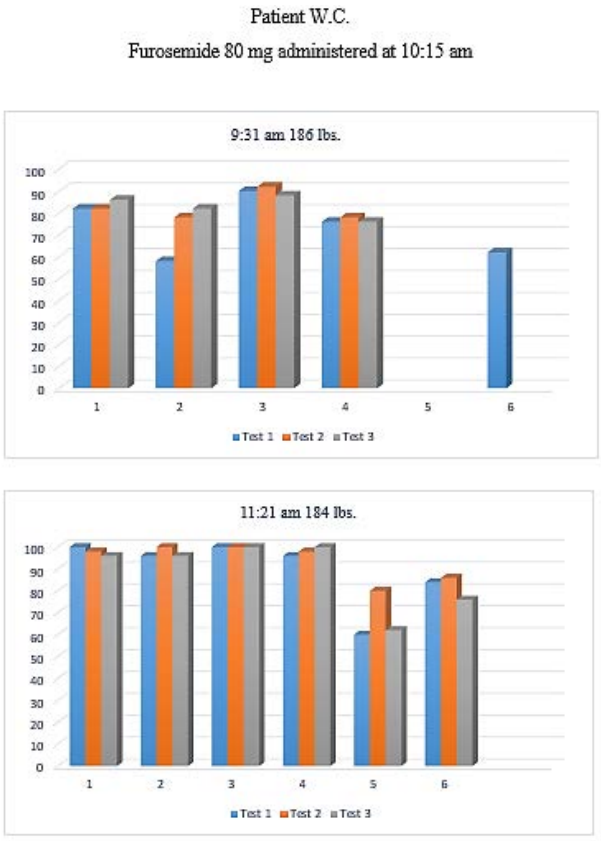
3. The glycerin test performed on patient F.G. shows essentially the same findings as the one performed on C.G.

Figure 3



4. The Furosemide test performed on W.C. shows essentially the same findings as those of M.E., including the weight loss.

Figure 4



5. The findings on a Furosemide test performed on another patient are presented in the next two tables. Table 4 shows that the Quix sway to the right resolved during the period of Furosemide activity and that the positive fistula test resolved as well. Table 5 shows the improvements in pure tones and discrimination scores that occurred during the Furosemide test. The criteria that are utilized are that pure tones have to increase on the tested ear by at least 10dB in 2 frequencies, or by 15dB at one frequency, or the discrimination score increase by at least 12%. These are the audiologic criteria used in evaluations. As described above, it is usually the balance improvement and the fistula testing that is more notable.

Table 4

Balance Test

Furosemide given at 09:30

Time	Quix	Standing	Fist AD	Fist AS
09:00	R	OK	+	neg
09:30	R	OK	+	neg
10:00	neg	OK	neg	neg
10:30	neg	OK	neg	neg
11:00	neg	OK	neg	neg

Table 5

Right Ear

Furosemide given at 09:30

Time	250	500	1000	2000	3000	4000	6000	8000	Dscrm
09:00	60	65	60	55	40	50	55	55	
10:00	55	55	50	40	40	40	50	55	76%
10:30	40	40	45	40	35	50	55	55	84%
11:00	35	35	30	40	35	45	50	50	96%
11:30	40	35	35	45	35	45	55	55	92%

The Furosemide, glycerin and Acetazolamide dehydration tests are time consuming and labor intensive. The main advantage is that the examinations reveal, in real time, to the examiner and the patient, changes in the balance, fistula testing and hearing that result from an induced change in the inner ear fluid system secondary to the dehydration of the endolymphatic space in the glycerin and Furosemide tests and the perilymphatic space in the Acetazolamide test.

In the cases of PLFS and PLHT, the dehydration tests reveal evidence of endolymphatic hydrops or perilymphatic hypertension. Both of these disorders have a clinical history and findings that will provide the roadmap for the clinician to establish the diagnosis. Both are disorders in which the spinning symptom is non-dominant although it may occur in acute PLFS and could occur in both intermittently. The principal vestibular disturbance is in the balance and not in the eye movements. The full gamut of vestibular symptoms can occur. There may be complications of visual and cognitive dysfunctions in both. However, the finding of imbalance on Quix or Romberg testing in a patient with chronic and persistent vestibular symptoms leads the clinician to have an evaluation of the hearing performed as well as tympanometry and stapedial reflex testing and fistula testing. If fistula testing is positive, the next step is a dehydration test. If the dehydration test reveals ELH in this patient who does not have the clinical picture of MD, e.g., spinning attacks with fluctuating hearing levels accompanied by tinnitus and ear fullness or pressure, then one can conclude that the ELH is a secondary ELH, as described by Howard House, and that the patient has PLFS. If the findings in this type of patient become worse or do not change on the Furosemide or glycerin testing and then improve on the Acetazolamide test, one can conclude that the diagnosis is PLHT. Both of these diagnoses can be established without the benefit of imaging. The clinician has been guided by the knowledge that these entities exist and are part of the

differential diagnostic algorithm and that they should be sought after if the road map leads to them on the basis of the history of vestibular complaints, the non-spinning nature of those complaints, the imbalance that has been found on examination, the positive fistula testing and the positive dehydration test.

Dr. Lehrer began evaluating patients for PLFS in 1977 after Fee had described post-traumatic cases in 1968 and Healy et al. had published their series in 1974. Lehrer, Poole, and Sigal published the paper on the glycerin dehydration test in 1980, describing only patients seen after trauma. Most clinicians will consider PLFS only in patients who have suffered head or neck trauma. However, Stroud and Calcaterra described Spontaneous PLFS in 1970, and Cole[90] described his experiences with spontaneous PLFS in 1995. A 1984 paper by Lehrer et al.[91] on post-traumatic PLFS noted that roughly half of 65 patients did not suffer trauma. Some clinicians have felt that there could be unrecognized traumatic events in these patients with spontaneous PLFS. That is a matter for future research in adults. However, there are multiple reports of PLFS occurring in young children with and without a history of trauma in their short and well observed lives.[92,93,94,95] This provides more evidence that spontaneous PLFS does occur.

PLFS has also been described as following neck injury from acceleration-deceleration forces.[96] Goodhill[97] described internal explosive forces such as straining as a cause of PLFS and implosive forces as well in the causation of PLFS. Such forces as barotrauma from external pressures and decompression incidents in diving have been well described. Dr. Lehrer has treated patients who suffered PLFS after blast trauma from explosions, especially gas explosions and air bag deployment. Both air bag deployments and blast trauma have been cited as a cause of inner ear injury.[98,99] The victims of the Boston Marathon bombers[100] were noted to have tympanic membrane perforations and dizziness which would indicate that the trauma from the explosions affected both the middle and inner ears.

The management of PLFS can be either medical or surgical. It was learned that if a patient improved during the Furosemide test they could be offered Furosemide or another loop diuretic orally as management. The four loop diuretics in the medical management of PLFS are Furosemide, Torsemide, Bumetanide, and in patients with a history of sulfa allergy whose dehydration test required glycerin, Ethacrynic Acid. All patients require potassium supplementation to compensate for the potassium loss associated with loop diuretic therapy. In those patients who have undergone a Furosemide test, the diuretic therapy is implemented two days later to avoid excessive dehydration. The patients are instructed to hydrate themselves and to replace the lost potassium by diet, utilizing a diet which provides them with 750mg/day of potassium or ingesting a 20meq Potassium tablet daily. The patient is initially placed on a moderate dose of the loop diuretic. The dosage can be adjusted on follow up, based upon an evaluation of their progress by history and physical examination of the balance. The diuretic therapy in those patients who are presently on a thiazide diuretic is changed, after consultation with their medical doctor, to a loop diuretic. If the patient is being treated for cardiovascular disease, the patient's medical doctor can be asked to integrate the diuretic therapy into the medical management. Medical consultation is always mandatory to assure that the proposed diuretic therapy will not result in hazardous volume depletion.

The patients are informed that diuretic therapy provides medical management of the inner ear disorder and is not intended as a cure. Many patients will cope with some imbalance rather than take medications. Patients are informed that rejecting therapy for the imbalance, which many feel they can cope with, does put their hearing at risk. Although the risk of hearing loss in untreated patients is quite small, it does exist.

One advantage of diuretic therapy is that both ears are treated concurrently, which would not be the case with surgery. Patients with an obvious case of bilateral PLFS can elect to receive medical treatment which would, if successful, obviate the need to operate on both ears. Patients should have the option to choose between medical and surgical management. Some will initially choose surgical management in order to achieve resolution of the disorder, which is the case in over 90% of cases in the first procedure. Those that choose diuretic therapy may ultimately elect to have surgery if the diuretic therapy disrupts their daily routine too much because of the frequency of urination. At this point, the choices are about evenly divided, and both groups have generally experienced sufficient relief to validate their choice.

The "cure" rate from surgery was higher in the past when the referrals were for simpler cases, as noted in the Lehrer et al. 1984 paper on post-traumatic PLFS. But, as time has passed, more cases have associated brain injuries as the number of cases with severe trauma has increased. There has been a recurrence rate of about 10% in successful cases. Secondary repair is performed, using the same technique as the primary repair. The secondary graft is post auricular fat and subcutaneous tissue whereas the primary graft is ear lobe fat. There is a greater risk to the chorda tympani after the first operation such that more care is required to dissect it from the flap. There is a slight risk of stapes fixation with the cement which must be carefully applied in the area of the fissula ante fenestram.

Surgery is recommended only in women in the stage of their life when a pregnancy is desired or is a consideration in the future. This is because they would have to abandon diuretic therapy should they become pregnant. If they are planning on a pregnancy, surgery is suggested and deferral of the onset of pregnancy for at least one year. Even after that year has passed, it is routinely recommended that the obstetrician accelerate the second stage of labor or consider

Caesarian section if there is a problem that prolongs and/or complicates the labor process.

If the patient elects to consider primary fistula surgery, it is discussed whether they need only a fistula repair or if an endolymphatic sac procedure is required as well. The technique of perilymphatic fistula (PLF) repair is discussed in detail at this point and endolymphatic sac surgery afterwards is briefly discussed.

Repair of PLFs is performed through a middle ear exploration approach. The first issue early on, and today, is whether a fistula is truly present. The "gold standard" of clear fluid appearing anterior to the oval window or in the round window niche that reappears after gentle suctioning is appropriate. Occasionally, one may see the anterior portion of the stapes footplate subluxed with fluid seeping from the anterior portion of the oval window itself. But, in most cases, the fluid appears from the area of the fissula ante fenestram, just anterior to the oval window.

The concepts of hydration and positioning are basic in the approach to detecting fistulas. Most surgeons operate with the patient in a horizontal position, and, if a fistulous leakage is not seen, have the anesthesiologist, if the anesthesia were general, increase intrathoracic pressure to increase cerebrospinal fluid pressure and encourage a leakage of perilymph, or compress the internal jugular vein to cause this effect. Dr. Lehrer has found that placing the patient in a mild Trendelenburg position accomplishes this increase in perilymph pressure secondary to the resultant increase in venous pressure in the head which causes an increase in cerebrospinal fluid pressure which is transmitted to the perilymph. This position results in more bleeding on raising the tympanomeatal flap. There will still be more bleeding with the head down so that the raising of the flap takes longer. There have been concerns that local anesthetic which penetrates into the middle ear can be mistaken for perilymph. Therefore, a diagnosis of a fistulous leakage occurs only when the fluid reoccurs on gentle suctioning in the areas of interest and careful examination of the area anterior to the stapes on the inner tympanic wall in the area of the fissula ante fenestram and the round window niche in the area inferior to the round window membrane. If this area is not readily visible, a microdrill is used to remove bony overhang such that the area is visible. Once again, fluid has to be noted to reoccur on gentle suctioning to confirm that it is fistulous fluid. In order to adequately visualize the round window niche, it is often necessary to turn the table towards the surgeon such that the area is visible or curette or drill away some posterior canal wall bone to expose the area. One will often see adhesions in the areas of interest, which may represent the middle ear analogue of the vestibular fibrosis discussed previously. Again, if the adhesions are lysed, either with a surgical instrument or a Laser, one has to confirm the presence of fluid recurrence from the areas of interest. The fistulous areas are found in bony clefts in the round window niche and in the fissula ante fenestram, or from a subluxed stapes, so these areas should be carefully inspected. The fissula is visualized anterior to the oval window between the incus and the malleus, so it is frequently necessary to extend the flap incision superiorly to provide for visualization of the anterior portion of the stapes footplate. If the fistulous fluid continues to leak and thus impair the placement of the grafts, the patient's head may be raised to a horizontal position or even higher into reverse Trendelenburg. Such repositioning can also be performed if there is excessive bleeding at any point in the procedure.

It has been Dr. Lehrer's experience that a positive dehydration test is highly sensitive in identifying patients with PLFS. The key to successful surgical identification is hydration and positioning as described above. Dr. Lehrer routinely grafts both windows whether or not he detects a fistulous leak. The above technique of positioning and hydration has resulted in detection of fistulas which were not apparent initially because of their tiny flow, especially at the second window when an obvious fistula was seen at the other.

Neurotologists primarily utilize ear lobe fat for tissue grafting as it can be cut into tiny pieces and conforms to the space into which it is inserted. Perichondrium or vein are difficult to work with in this regard, but post-auricular subcutaneous fat and areolar tissue are, like fat, readily available and conform well when grafted into the previously described microfissures. There are surgeons who describe "packing" the oval and round windows to repair a fistula. This technique suggests that the mucous membrane of the area is not denuded, which will consequently not provide the vascular bed for the graft to embed itself into the fistulous area. The second problem would be the non-conformity of a "pack" of tissue into the fistulous area, which would result in a failure to seal the fistulous track. The initial graft is covered with successively larger grafts to maintain pressure on the tiny graft that is the actual seal. The area is then covered with gelfoam to provide additional pressure. There are surgeons who use a tissue glue or a fibrin glue in order to seal a fistula.

Dr. Lehrer has performed endolymphatic sac surgery (ELSS)[101] when the physical evaluation results in falls on the Romberg and/or Quix tests or when there is a significant sensorineural hearing loss. Conner and Potter[102] described patients who developed Meniere's syndrome after PLF repair. Dr. Lehrer has seen this occur in two patients, one with a significant sensorineural hearing loss in the affected ear and one with imbalance such that she was falling on the Quix and Romberg tests on pre-surgical examinations. Dr. Lehrer's experience is that combining ELSS with fistula repair is more efficacious than fistula repair alone in restoring balance in patients who are falling on the balance examination and apparently avoids the complication of Meniere's syndrome after fistula repair. A valve or shunt can be inserted into the ELS; but if it is not possible because of the inaccessibility of the sac, a decompression has been

successful in Dr. Lehrer's experience. If there is a recurrence of the symptoms of the PLFS after ELSS, it is suggested to only repair the fistulas.

Perilymphatic hypertension. Dr. Lehrer's management of PLHT is only medical, utilizing Acetazolamide or Topiramate, both of which decrease cerebrospinal fluid pressure, and consequently, perilymph pressure, assuming that the cochlear aqueduct (CA) is open. Dr. Lehrer opines that the CA is open in these cases as it appears that the mechanism is similar to Papilledema in that the inner ear is affected by the increased CSF pressure rather than the eye. Marchbanks[103] has described symptoms of inner ear disease in patients with increased CSF pressure. Increased CSF pressure can occur in obese patients so that obesity and even recent weight gain may be factors in the development of PLHT. Topiramate can decrease CSF pressure as well as decreasing weight. Thus, it may play a dual role in treating PLHT. Conversely, the clinician should be aware that patients already on Acetazolamide or Topiramate who present with symptoms of an inner ear disorder may be suffering from ELH, primary or secondary. A change of medication has relieved the inner ear symptoms in such patients. As noted above, the reverse is also true, that patients on a loop diuretic who develop symptoms of an inner ear disorder may be suffering from PLHT and a change of diuretic to a thiazide has resolved those symptoms.

The symptoms of PLHT are similar to those of PLFS and NRVS. Positive fistula testing is present in most cases. However, the imbalance is generally worse in that these patients usually fall on Romberg and/or Quix testing. There can be a history of trauma, or the condition may be spontaneous. Obesity and recent weight gain can be factors. A dehydration test is performed, as usual, utilizing Furosemide or glycerin as the first step. If the findings worsen or do not improve during the Furosemide test, the Acetazolamide test should be performed. If there is an improvement on Acetazolamide testing, it is recommended to classify those cases as PLHT and place them on medication. If patients have a glycerin test performed because of sulfa allergy and become worse, Dr. Lehrer will classify them as PLHT and place them on a trial of Topiramate, as Acetazolamide is a sulfa drug. If the trial of Topiramate is unsuccessful, middle ear exploration is recommended even if the glycerin test resulted in worsening of the findings. More research into these entities are recommended.

Miscellaneous Causes of Vestibular Symptoms

The section of the classification describes disorders that Dr. Lehrer sees in less than 5% of his patients. Placing a disorder in this section is not intended to diminish the importance of the particular disorder or the clinicians who have uncovered or described these disorders. Some of these disorders, especially those related to a psychogenic cause, are diagnosed more frequently by other clinicians. Dr. Lehrer addresses the issues which appear to be responsible for the differences in the incidence of some of these disorders in the spirit of scientific discourse, with high respect for the clinicians who have described such disorders.

Delayed endolymphatic hydrops. Schuknecht[104] described this syndrome as occurring in patients who have sustained a profound hearing loss in one ear, usually from infection or trauma, and then after a prolonged period of time develop either episodic vertigo from the same ear or fluctuating hearing loss, sometimes with episodic vertigo, in the opposite ear. He stated that the ipsilateral form of the disease could be treated with labyrinthectomy, but that no satisfactory therapy was available for the contralateral form of the disease.

Recurrent vestibulopathy. This syndrome of recurrent vertigo was first described by Slater[105] as benign recurrent vertigo and was described by Leliever and Barber[106] as recurrent vestibulopathy. Recurrent attacks of vertigo without hearing loss, tinnitus or fullness occur at erratic intervals. Vestibular testing has not been definitive in localization.

Vestibular loss and drug-induced vestibular dysfunction. The most common cause of loss of vestibular function that Dr. Lehrer sees is drug induced from Gentamycin, an aminoglycoside antibiotic. Aminoglycosides can cause ototoxicity and cause hearing loss and vestibular loss. Macrolide antibiotics, such as erythromycin, can cause reversible hearing loss. Minocycline, used for acne, can cause vertigo. Use of loop diuretics, platinum-containing chemotherapeutic agents, heavy metals, quinine, and aspirin can result in hearing loss but vestibular symptoms are unusual.

Motion sickness. Patients with chronic motion sickness are treated with a variety of anti-emetics and vestibular suppressants. Dramamine and Bonine or Meclizine were the most frequently used drugs. Scopalamine has been utilized as well for motion sickness as it appears to affect neural pathways to the inner ear. Regarding space sickness, there is literature indicating that stimulants of the Dexedrine family are the most helpful drugs.[107]

Dehiscence of the semicircular canals (SSCD). Minor et al.[108] described SSCD in eight patients in an original article in 1998. They reported finding this abnormality in patients with vertigo, oscillopsia and/or disequilibrium related to sound. The dehiscence was identified by computed tomography (CT) of the temporal bones. Surgical plugging of the canal between the dehiscence and the ampulla relieved the symptoms but resulted in sensorineural hearing loss in one patient of two successfully operated. One patient's symptoms began after head trauma and another's after straining and exertion. The authors described sound-induced torsional nystagmus in the plane of the superior semicircular canal. They likened this to a positive Hennebert's sign and the Tullio phenomenon. One of the eight patients had undergone a negative exploration for a perilymphatic fistula. These patients had disequilibrium and a positive Hennebert's

sign, which has been alluded to previously as constituting a positive fistula test.

Minor reported on 17 patients in 2000.[109] He reported that 5 of 1,000 temporal bones in the Johns Hopkins Registry showed a complete dehiscence and that there was a thinning in the pathologic area to less than 0.1mm in 14 other bones. He suggested that minor head trauma or barotrauma could result in disruption in the area of the dehiscence and lead to the symptoms of the syndrome. He suggested that 0.5 to 1mm coronal cuts be utilized in the CT examinations. In 2003, he was a co-author on a paper[110] which concluded that the positive predictive value of CT improved with 0.5mm collimated helical CT with reformation in the plane of the superior semicircular canal (SSC). He described examinations that could reveal the eye signs of dehiscence, including stimulation with tones between 500 and 2000Hz with intensities of 100 to 110dB, pressure on the tragus and external auditory canal (essentially the fistula test), and Valsalva maneuvers performed by nose blowing against pinched nostrils and by raising intrathoracic pressure. Evoked eye movements were evaluated with Frenzel's glasses. Resurfacing and canal plugging surgical techniques were utilized.

More currently, Minor and Carey[111] reviewed their experiences with this syndrome for the Vestibular Disorders Association. The symptom of seeing things move can be elicited by loud noise, coughing, sneezing, pressing on the tragus, or straining. There may be a constant feeling of disequilibrium and imbalance. The sign of nystagmus evoked by sound and pressure results from inward or outward movements of the stapes footplate, much as in the fistula testing devised by Kohut, although in the PLFS it is rare to see eye movements.

A low-frequency conductive hearing loss may be present, suggesting otosclerosis. However, the acoustic reflex is present in dehiscence and should be absent in otosclerosis, especially if surgery is contemplated. Stapedectomy will not benefit patients with SSCD. Patients also complained of hearing their eye movements, having a distorted sensation of sound in the affected ear during activities such as running or hearing their own voice too loudly in the affected ear (autophony). Dr. Lehrer encountered a patient with SSCD who was a professor who could not lecture because of the feedback into his affected ear. He was beside himself with frustration and called Dr. Lehrer to ask whether he had PLFS. The patient informed Dr. Lehrer that a CT examination for SSCD had been negative; but, ultimately, when he had another consultation and a new CT, the dehiscence was identified. The CT examination must follow the criteria that Minor reported. However, Minor and Carey cautioned that a thin layer of intact bone could be missed on CT and that, consequently, the diagnosis also depends upon characteristic clinical findings and other physiologic tests. The addition of vestibular evoked myogenic potential (VEMP) testing to the evaluations plays an important role in diagnosis. Patients with SSCD typically have pathologically lowered thresholds and greater amplitudes on VEMP testing. Browaeys, Larson, Wong, and Patel[112] reported that MRI studies of the superior and posterior semicircular canals had a 100% negative predictive value in their series of 112 patients. They suggested CT evaluations only if there was a positive finding on MR imaging.

Surgical plugging of the affected canal has proven to be more effective than resurfacing although there is a small risk of hearing loss, more so when stapedectomy or prior surgery on the canal has been performed.

Posterior semicircular canal dehiscence was reported in 12 patients, which included children and adults, by Gopen, Zhou, Poe, Kenna, and Jones[113] in 2010. There was hearing loss in all patients as well as malformations which included a high riding jugular bulb, atresia/microtia, enlarged vestibular aqueduct and Alpert syndrome. Chronic disequilibrium was more common in adults. Vestibular evoked myogenic potentials were abnormal. A lateral (horizontal) case of dehiscence was reported on imaging by Bassim, Patel, and Buchman[114] in 2007.

Vascular compression syndrome. The vascular compression syndrome of the VIIIth cranial nerve was first described by Jannetta, Moller, and Moller in 1984.[115] The vestibular symptoms have been described as motion-induced dizziness that is constant when the patient is in motion. There are feelings that are described as rocking, swaying, disequilibrium, or imbalance that is relieved by standing still, sitting or lying down. Physical therapy, exercises, or antihistamine or benzodiazepine therapy do not provide relief. Any motion causes severe symptoms, and patients modify and may ultimately stop their daily routines. Patients may have the other three symptoms of inner ear dysfunction: tinnitus, ear fullness, or hearing loss. Abnormalities in waves II and III on auditory brain stem response testing were utilized to diagnose this condition in concordance with the history. The surgery was reported as successful in the nine patients relieved of the cross compression of the VIIIth nerve. They described this syndrome as disabling positional vertigo. The condition was disabling, but positional vertigo was not reported on examinations of these cases.

Brackmann, Kesser, and Day[116] reported in 2001 on their series of microvascular decompressions (MVD). They utilized air contrast CT studies and MRI imaging to identify cross compression of the VIIIth nerve and selected only those patients for surgery who had an identifiable compression on imaging. They reported generally successful results and concluded that the procedure was appropriate in carefully selected patients who had failed medical therapy and whose imaging studies suggested cross compression of the VIIIth nerve.

Cervical vertigo. Jongkees[117] presented a scholarly review of this issue which was published in the *Laryngoscope*. He described vestibular nystagmus that

followed cervical rotation, even with dead labyrinths. He reported that neck immobilization could relieve vertigo that occurred in certain positions of the neck. He noted that most authors explained the cervical influence on vertigo as most likely from either vertebro-basilar compression, a disorder of the sympathetic vertebral plexus or pathology in the sensory afferent fibers from the deep neck tissues, which he felt was the most likely source. He described personal experiences in which gentle physical therapy improved the vertigo that was present in the minority of his patients who also presented with pain in the neck and at the base of the skull. He concluded that palpation of the neck musculature was an integral part of the vestibular evaluation.

Ferrari[118] reported that neck pain may contribute to balance disturbances whether or not neck trauma had occurred. He referred to the Karlberg, Magnusson, Malstrom, Melander, and Moritz[119] study in which physiotherapy reduced neck pain and dizziness and improved postural performance. Inner ear injury can result from whiplash-type trauma, as reported by Chester and Grimm et al. A vestibular evaluation should include an examination of the neck as well as the vestibular system.

Vestibular migraine. Thomas Lempert[120] described vestibular migraine (VM) as presenting with attacks of spontaneous or positional vertigo, head motion-induced vertigo, and visual vertigo lasting 5 minutes to 3 days. He stated that the Barany Society and the International Headache Society jointly proposed that VM and probable VM be identified based on explicit criteria. Liveing[121] described giddiness and vertigo as occurring as part of a migraine attack or in migraineurs separately from the attacks of headache, which is a pattern that is still recognized. Epidemiological observations have revealed more than a chance association between migraine and dizziness, and migraineurs report more vertigo than patients with tension headaches. Benign paroxysmal vertigo of childhood has been recognized as a migrainous disorder. However, in adults, many of the presenting symptoms described as occurring in VM occur in the variety of vestibular disorders already discussed in this article and can be viewed as non-specific symptoms. Lempert stated that the diagnosis of VM is based upon the symptom type, severity and duration, a history of migraine, temporal association of migraine symptoms with vertigo attacks, and exclusion of other causes. He states that headache is often absent, that central spontaneous or positional nystagmus can be found as well as unilateral vestibular dysfunction. The only specific features appear to be a concurrent migraine attack or such migraine features as photophobia or auras. Despite the absence of definitive symptoms and definitive signs and tests in most cases of VM, there appears to be a relationship between migraine and vestibular disorders.

Lehrer and Poole[122] suggested that the link between a vestibular disorder such as NRVS and migraine lay in the serotonergic neurotransmission that appears to be common to both. Halberstadt and Balaban[123] described 5-HT transporter fibers as being most dense in the superior and medial vestibular nuclei but also being present in the lateral and caudal regions of the vestibular nuclear complex. Retrogradely labeled neurons were found in the raphe nuclei, principally the dorsal and raphe obscurus nuclei. They found that the vestibular nuclei received both 5-HT and non-5-HT projections from the raphe nuclei. They found projections from the raphe nuclei to terminal fields of cerebellar fibers and stated that these connections could influence the activity of cerebellovestibular circuits. They also stated that the majority of 5-HT neurons in the CNS are found clustered along the midline of the brainstem in raphe nuclei.

Such anti-migraine drugs as Methysergide and CH were found to be potent antagonists of transynaptically elicited effects of stimulation of the pathway from the dorsal raphe nucleus to the cingulate cortex.[124] These anti-migraine drugs have been described earlier in this article as being effective in NRVS but have not been described as being utilized in VM. Young, Khorana, Bondareva, and Glennon[125] reported that CH, and Pizotylline (Pizotifen), another anti-migraine drug, displayed high affinity for multiple subpopulations of serotonergic, dopaminergic, adrenergic, histaminergic and cholinergic receptors. Tajti, Uddman and Edvinsson[126] reported that evidence from animals and humans suggested that such brainstem nuclei as the raphe nuclei, the locus coeruleus and the periaqueductal grey matter are involved in the pathophysiology of migraine, in a paper devoted to the localization of the "migraine generator." Dahlem[127] postulated a migraine generator network involving the trigeminal nerve and an associated descending modulatory network of brainstem nuclei and parasympathetic vasomotor efferents.

Dr. Lehrer opines that VM is an umbrella diagnosis that includes more than one vestibular disorder and that the migrainous mechanism has been cited to be possibly involved in multiple vestibular disorders. Dr. Lehrer would suggest, however, that NRVS is a disorder that is closely linked to VM by its localization in the brainstem, the common involvement of serotonergic transmission and the efficacy of anti-5HT drugs in its management. Dr. Lehrer suggests that the common links of NRVS and VM that have been described would be fertile ground for further investigations into the mechanisms and disorders of neurotransmission and the testing and the treatment of these disorders. Regarding other vestibular disorders which have been associated with migraine, Dr. Lehrer would suggest further investigations that, recognizing the symptom complex of NRVS as one with migrainous connections by virtue of their common 5-HT neurotransmission, concentrate on symptom complexes other than that manifested by NRVS to attempt to determine whether there are migrainous factors involved.

Psychogenic dizziness. Experienced clinicians such as Staab, Ruckenstein and Brandt have described syndromes of psychogenic dizziness and vertigo. Staab and Ruckenstein

described the syndrome of chronic subjective dizziness[128] (CSD), and Brandt has described the syndrome of phobic postural vertigo[129] (PPV). Clinicians should be aware that psychogenic dizziness can occur but should also be wary of Furman's "new narrow definition of psychiatric dizziness" which states that this diagnosis should only be used when dizziness occurs exclusively in combination with other symptoms as part of a recognized psychiatric disorder. Furman also urged extreme caution in making the diagnosis of psychiatric dizziness because behaviors such as fears and phobias can be situations in which specific avoidance behaviors have developed as a consequence of a neurotologic disorder. Furman, Balaban, and Jacob stated[130] that anxiety may be the cause or effect of the complaint of dizziness. They stated that recognizing a vestibular disorder in a dizzy patient who presents with anxiety, or the converse, has important treatment implications and conclude that a combined treatment of both conditions could be beneficial. Balaban,[131,132] in two papers describing his findings of a link between balance control and anxiety, described findings that indicated that the parabrachial nucleus appeared to be an important node in a primary network that processes convergent vestibular, somatic, and visceral information processing to mediate avoidance conditioning, anxiety, and conditioned fear responses. He described the role of the parabrachial nucleus and the dorsal raphe nucleus pathway in processing vestibular and affective information as well as the role of serotonergic and non-serotonergic pathways to the vestibular nuclei and the serotonergic receptors expressed in the amygdaloid and cortical targets of the parabrachial nucleus. He hypothesized that the parabrachial and Kolliker-Fuse nuclei appear to constitute a primary source of information about whole body rotational velocity and body position relative to gravity to circuits that mediate anxiety responses. He concluded that "dorsal raphe nucleus projections to vestibular-parabrachial network pathways may contribute to both the implementation of a tradeoff between motor and sensory information gathering aspects of responses to self-motion, and calibrating the sensitivity of affective responses to aversive aspects of motion."

Staab and Ruckenstein described the use of selective serotonin reuptake inhibitors (SSRIs) in treatment of dizziness and described the serotonergic influences on the vestibular nuclei and cerebellum and commented on the possibility of physiologic effects of these drugs on the dizziness symptom.

Most of the dizzy patients that Dr. Lehrer treats describe anxiety, which may be part of the distress of the feelings of disorientation to their surroundings or part of the frustrations that they are experiencing on suffering an illness which previous doctors have been unable to diagnose, and the consequent fear that something terrible is afflicting them. Almost all of the patients that Dr. Lehrer has seen who have presented to an emergency room with dizziness have had CT scans of their brain performed to rule out a central cause, and, if they are admitted to hospital, have had MRI of their brain and ECGs performed to rule out heart disease. Brandt[133] describes "fear of death" as a clinical feature of the symptom complex in phobic postural vertigo. Dr. Lehrer's experience with his patients is that all patients with symptoms coming from their head, be it dizziness, headache or tinnitus, have a fear of something that could threaten their life, be it a tumor or a stroke or some other serious disease.

Dr. Lehrer does not doubt that the symptoms of anxiety and dizziness are intermingled. But his experience is that the symptoms of vestibular dysfunction he has observed and that have been described by McNally and Stuart and Mallinson and Longridge correlate with the organic vestibular entities and are treatable as organic physical entities. Many of the symptoms described as psychogenic by Staab, Ruckelstein and Brandt are descriptive of visually induced dizziness which Brandt[134] has described as a visual-vestibular interaction that would produce symptoms of spatial disorientation, apparent motion, postural imbalance and nausea. The clinician should analyze both the mind and the body, and those authors who describe psychogenic dizziness are correct to do so. Dr. Lehrer has found that the resolution, or even the identification of a physical entity, will relieve the anxieties that are generally present in his patients with vestibular disorders.

Dr. Lehrer closes this most important portion of this article by stating that more organic causes of the "dizziness" symptom can be found if the clinician utilizes a plan with a differential diagnosis in mind. The number of patients classified as unknown, non-organic or psychogenic has decreased in his practice by his use of the described classification, and by keeping up to date with the medical literature as new tests, new imaging techniques, and new disorders are identified and utilized.

Vestibular Rehabilitation and Posturography

Vestibular Rehabilitation was revolutionized in the United States by Anne Shumway Cook and Fay B. Horak in the 1980s and is, as they state with their co-authors in their chapter[135] on this subject, an important therapeutic option. As the authors state, "It is much more difficult for the CNS to compensate for a fluctuating vestibular deficit (e.g., Meniere's disease) or for multiple deficits than for a steady loss of any kind," Dr. Lehrer agrees and would add such disorders with fluctuating deficits such as NRVS, PLFS and PLHT which can be stabilized with successful treatment. Ideally, therapy follows the management of one of the specific disorders identified in this article rather than being the initial or the definitive therapy. Dr. Lehrer's patients with visual disorders have also benefited from the specialized therapy afforded by Occupational Therapists, which is usually provided in concert with neuro-optometric evaluations and management. The authors state that specific exercises for BPPV can result in successful treatment, and Dr. Lehrer wholeheartedly agrees. The utilization of the

clinical sensory interaction in balance (CTSIB) by Shumway Cook and Horak[136] is described, and they credit Nashner's[137] dynamic posturography protocol for the sensory organization test as the model for the CTSIB.

The aim of vestibular rehabilitation is to decrease dizziness and improve balance. The imbalance is continuously evaluated by the therapist. Reports to the referring physician describe the general condition and capabilities of the patient as well as a description of subjective symptoms of dizziness that are present and an objective evaluation of the balance.

Summary

Dr. Lehrer wishes to emphasize that the evaluation of outcomes in patients suffering from the vestibular disorders described above as having imbalance must include an evaluation of the balance as well as a review of the symptom complex. If the balance has not been evaluated initially, the clinician can rely only on the symptoms that the patient describes. These symptoms might be ameliorated by the medications prescribed for symptom relief and provide the clinician with a false picture of the results of the therapy that was provided. If the balance evaluation is performed initially and is abnormal, the clinician would then be in possession of an objective measure which, when evaluated after therapy, would provide evidence of the effects of therapy. Dr. Lehrer would therefore strongly recommend that all clinicians, especially those that publish studies, evaluate the balance of all patients with vestibular symptoms in order to provide the patient with a full evaluation and provide the clinician with an invaluable tool in the diagnosis and management of these patients as well as providing an objective measure when outcomes are reviewed.

Implications for Life Care Plans/Planners

The aim and hope on writing this article is to share experiences in managing vestibular disorders with life care planners. Dr. Lehrer hopes that the information and experiences provided herein will be helpful to his fellow physicians, surgeons, therapists in their ceaseless quest to improve the care of their patients, and to life care planners to project appropriate recommendations in a life care plan.

Bibliography

1. Lehrer, J. (1994), *Evaluation of a new classification* of vestibular disorders. Presented to the American Neuro-otology Society, Palm Beach, FL.
2. Silvoniemi, P. (1988). Vestibular neuronitis. An otoneurological evaluation. *Acta Oto-Laryngologica Suppl,* 453, 1-72.
3. Dix, M. R. & Hallpike, C. S. (1952). The Pathology, symptomatology and diagnosis of certain common disorders of the vestibular system. *Proceedings of the Royal Society of Medicine, 45*(6), 341-354.
4. Gilbert, G. (1965). Meniere's syndrome and cluster headaches: Recurrent paroxysmal focal vasodilatation. *Journal of the American Medical Association, 191*(9), 691-694.
5. Quix, F. H. (1924). The Past Pointing Test in Otology. *Ned Tijdschr Geneeskd* (2), 276.
6. Hart, C. W. (1983). The Quix Test. *Laryngoscope 93*(9), 1160-1161.
7. Lehrer, J. (1976). Periactin in the treatment of vertigo. Presented to the American Neuro-otology Society, Palm Beach, FL.
8. Arslan, M., & Sala, O. (1956). *Fistiopatalogia e clinica delle vie vestibolaari centrali.* Atti del 44th Congresso Soc Ital Lar Otol, Bologna, Italy.
9. Bosatra, A., Rossolo, M., & Poli, P. (1975). Modifications of the stapedius reflex under spontaneous and experimental brain stem impairment. *Acta Oto-Laryngologica, 80*, 61-66.
10. Bosatra, A., Rossolo, M., & Poli, P. (1976). Oscilloscopic analysis of the stapedius muscle reflex in brain stem lesion. *Arch Otolaryngol, 102*(5), 284-285.
11. Palkovits, M., Brownstein, M., & Saavedra, J. M. (1974). Serotonin content of the brain stem nuclei in the rat. *Brain Research, 80*(2), 237-249.
12. Saavedra, J. M. (1977). Distribution of serotonin and synthesizing enzymes in discrete areas of the brain. *Federation Proceedings, 36*(8), 2134-2141.
13. Hillarp, N. A., Fuxe, K., & Dahlstrom, A. (1966). Demonstration and mapping of central neurons containing dopamine, nor-adrenaline and 5-hydroxytrptamine and their reactions to psychopharmacia. *Pharmacological Reviews, 18*(1), 727-741.
14. Lehrer, J. F., & Poole, D. C. (1991). Vestibular neuronitis dissected into vestibular neuritis and the nucleo-recticular vestibular syndrome. Proceedings of the Third International Symposium and Workshops on Surgery of the Inner Ear, Aspen, CO. In I. K. Arenberg (Ed.), *Surgery of the Inner Ear* (pp. 431-436). Amsterdam, New York: Kugler Publications.
15. House, H. P. (1967). The fistula problem in otosclerosis surgery. *Laryngoscope, 77*(8), 1410-1426.
16. Fee, G. A. (1968). Traumatic perilymphatic fistulas. *Archives of Otolaryngology, 88*(5), 477-480.
17. Healy, G. B., Friedman, J. M., & Strong, M. S. (1976). Vestibular and auditory findings of perilymphatic fistula: a review of 40 cases. *Transactions - American Academy Opthalmology and Otolaryngology, 82*(1), ORL44-49.
18. Takagi, A., & Sando, I. (October, 1988). Computer-aided three-dimensional reconstruction and measurements of semicircular canals and their cristae in man. Presented at the Collegium Oto-Rhino-Laryngologicum Amicitiae Sacrum Meeting, Hakone, Japan. *Acta Oto-Laryngologica, 107*(5-6), 362-365.
19. Stroud, M. H., & Calcaterra, T. C. (1970). Spontaneous perilymph fistulas. *Laryngoscope, 80(*3), 479-487.
20. Kohut, R. I., Waldorf, R. A., Haenel, J. L., Thompson, J.

N. (1979). Minute perilymph fistulas: vertigo and Hennebert's sign without hearing loss. *Annals of Otology, Rhinology & Laryngology, 88*(2), 153-159.
21. Causse, J., Causse, B., & Bel, J. (1983). Tympanometry and fistula test. *Audiology, 22*(5), 451-462.
22. Klockhoff, I., & Lindbloom, V. (1966). Endolymphatic hydrops revealed by glycerol test: Preliminary report. *Acta Oto-Laryngologica, 61*(5), 459-462.
23. Snyder, J. M. (1974). Extensive use of diagnostic test for Meniere's disease. *Archives of Otolaryngology, 100*(5), 360-365.
24. Lehrer, J. F., Poole, D. C., & Sigal, B. (1980). Use of the glycerin test in the diagnosis of post traumatic perilymphatic fistulas. *American Journal of Otolaryngology, 1*(30), 207-210.
25. Weider, D. J., & Johnson, G. D. (1988). Perilymphatic fistula: a New Hampshire experience. *American Journal of Otolaryngology, 9*, 184-196.
26. Brookes, G., Morrison, A., & Booth, J. (1982). Acetazolamide in Meniere's disease: Evaluation of a new diagnostic test for reversible endolymphatic hydrops. *Otolaryngology−Head and Neck Surgery, 90*(3), 358-366.
27. Goode, R. (1981). Perilymph hypertension and the indirect measurement of cochlear pressure. *Laryngoscope, 9*1(10), 1706-1713.
28. Paparella, M., Schachern, P., & Goycoolea, M. (1988). Perilymphatic hypertension. Otolaryngology–Head and Neck Surgery, 99(4), 408-413.
29. Lehrer, J. F., & Poole, D. C. (1987). Diagnosis and management of vertigo. *Comprehensive Therapy, 13*(9), 31-40.
30. McNally, N. J., & Stuart, E. A. (1941). Vertigo from the standpoint of the otolaryngologist. *Transactions - American Academy of Ophthalmology and Otolaryngology, 46*, 33-37.
31. Mallinson, A., & Longridge, N. (1998). Specific vocalized complaints in whiplash and minor head injury patients. *The American Journal of Otology, 19*(6), 809-813.
32. Furman, J. F., & Jacob, R. G. (1997). Psychiatric dizziness. *Neurology, 48*(5), 1161-1166.
33. Roberts, H. A. (1979). *Severe accidental head injury: an assessment of long-term prognosis.* (pp 96-98). London: Macmillan Press Ltd; .
34. Agrawal, Y., Carey, J., Hoffman, H., Sklare, D., & Schubert, M. (2011). The modified Romberg balance test: Normative data in U.S. adults. *Otology & Neurology, 32*(8), 1309-1311.
35. Lehrer, J. F., & Poole, D. C. (1981). Abnormalities of the stapedius reflex in patients with vertigo. *The American Journal of Otology, 3*(2), 96-103.
36. Borg, E. (1973). On the neuronal organization of the acoustic middle ear reflex. A physiological and anatomical study. *Brain Research, 49*(1), 101-123.
37. Kohut, R. I., Waldorf, R. A., Haenel, J. L., & Thompson, J. N. (1979). Minute perilymph fistulas: Vertigo and Hennebert's sign without hearing loss. *Annals of Otology, Rhinology & Laryngology, 88*(2), 153-159.
38. Harada, T., Sando, I., & Myers, E. N. (1981). Microfissures in the oval window area. *Annals of Otology, Rhinology, and Laryngology, 90*(2), 174-180.
39. Kohut, R. I., Himojosa, R., & Budetti, J. A. (1986). Perilymphatic fistula: a histopathological study. *Annals of Otology, Rhinology, and Laryngology, 95*(5), 466-471.
40. Epley, J. M. (1992). The canalith repositioning procedure: for treatment of benign positional vertigo. *Otolaryngology−Head and Neck Surgery, 107*(3), 399-404.
41. Schuknecht, H. F., & Kitamura, K. (1981). Vestibular neuritis. *Annals of Otology, Rhinology, and Laryngology, 90*(1, Pt 2), 1-19.
42. Grimm, R. J., Hemenway, W. G., Lebray, P. R., & Black, F. O. (1989). The perilymph fistula syndrome defined in mild head trauma. *Acta Oto-Laryngologica Suppl, 464*, 1-40.
43. Brandt, R., & Daroff, R. B. (1980). Physical therapy for benign paroxysmal positional vertigo. *Archives of Otolaryngology, 106*(8), 484-485.
44. Semont, A., Freyss, G., & Vitte, E. (1988). Curing the BPPV with a liberatory maneuvre. *Advances in Otorhinolaryngology, 42*, 290-293.
45. International Symposium. (1981). *Meniere's disease: Pathogenesis, diagnosis and treatment.* New York: Thieme Stratton Inc.
46. *Ibid*., 13.
47. *Ibid*., 13.
48. *Ibid*., pp 1-9.
49. *Ibid*., p. 242.
50. Jongkees, L. B. W. (1972). Meniere's–a syndrome or disease? *Acta Oto-Laryngologica, 74*(Stockholm Suppl 305), 9.
51. Committee on Hearing and Equilibrium. (1995). Guidelines for the diagnosis and evaluation therapy in Meniere's disease, American Academy of Otolaryngology, Head Neck Surgery Foundation, Inc. *Otolaryngology−Head and Neck Surgery, 113*(3), 181-185.
52. Klockhoff, I., & Lindbloom, V. (1966). Endolymphatic hydrops revealed by glycerol test: Preliminary Report. *Acta Oto-Laryngologica, 61*(5), 459-462.
53. Snyder, J. M. (1974). Extensive use of diagnostic test for Meniere's disease. *Archives of Otolaryngology, 100*(5), 360-365.
54. Gibson, W. P. R., Moffat, D. A., & Ramsden, R. T. (1977). Clinical electrocochleography in diagnosis and management of Meniere's disease. *Audiology, 16*(5), 389-401.
55. Nakashima, T., Naganawa, S., Teransihi, M., Tagaya, M., Nakata, S., Sone, M., Otake, H., …Nishio, N. (2010). Endolymphatic hydrops revealed by intravenous gadolinium injection with Meniere's disease. *Acta Oto-*

Laryngologica, 130(3), 338-343.
56. Kato, K., Yoshida, T., Teranishi, M., Sano, R., Oake, H., Sone, M., …Nakashima, T. (2012). Peak width in multifrequency tympanometry and endolymphatic hydrops revealed by magnetic resonance imaging. *Otology & Neurotology, 33*(6), 912-915.
57. Iacovou, E., Vlastarakos, P., Ferekidis, E., & Nikolopoulos, T. (2013). Multi-Frequency tympanometry: Clinical applications for the assessment of the middle ear status. *Indian Journal of Otolaryngology and Head & Neck Surgery, 65*(3), 283-287.
58. Lehrer, J. F., Poole, D. C., Seaman, M., Restivo, D., & Hartman, K. (1986). Identification and treatment of metabolic abnormalities in patients with vertigo. *Archives of Internal Medicine, 146*, 1497-1500.
59. Olefsky, J. M., Farquhar, J. W., & Reaven, G. M. (1974). Reappraisal of the role of insulin in hypertriglycemia. *The American Journal of Medicine, 57*(4), 551-560.
60. Spencer, J. T. (1975). Hyperlipoproteinemia and inner ear disease. *Otolaryngologic Clinics of North America, 8*(2), 438-492.
61. Tsundo, R., Fukaya, T., & Komatsuzaki, A. (1998). The furosemide test and vestibular status in Meniere's disease. *Acta Oto-Laryngologica, 118*(2), 157-160.
62. Kimura, H., Aso, S., & Watanabe, Y. (2003). Prediction of progression from atypical to definite Meniere's disease using electrocochleography and glycerol and furosemide tests. *Acta Oto-Laryngologica, 123*(3), 388-395.
63. Shambaugh, G. (1967). Surgery of the ear. (p. 620). Philadelphia & London: W.B. Saunders Company.
64. Brandt, T. (1991). Vertigo: Its multisensory syndromes. London, Great Britain: Springer- Verlag.
65. Kitahara, T., Horii, A., Imai, T., Ohta, Y., Morihana, T., Inohara, H., & Sakagami, M. (2014). Effects of endolymphatic sac decompression surgery on vertigo and hearing patients with bilateral Meniere's disease. *Otology & Neurotology, 35*(10), 1852-1857.
66. Thomsen, J., Bretlau, P., Tos, M. & Johnsen, N.. J. (1981). Placebo effect in surgery for Meniere's disease. *Archives of Otolaryngology, 107*(5), 271-277.
67. Brandt, T. (1991). V*ertigo: Its multisensory syndromes.* London, Great Britain: Springer- Verlag.
68. Baloh, R. (1984). *Dizziness, hearing loss, and tinnitus: The essentials of neurotology.* Philadelphia: F. A. Davis Company.
69. Rudge, P. (1983). *Clinical neurology and neurosurgery monographs, Volume 4.* Edinburgh; New York: Churchill Livingstone.
70. Baloh, R., & Honrubia, V. (1979). *Contemporary neurology series: clinical neurophysiology of the vestibular system.* Philadelphia: F. A. Davis Company.
71. Lehrer, J. F. , & Poole, D. C. (1991). Vestibular neuronitis dissected into vestibular neuritis and the nucleo-recticular vestibular syndrome. Proceedings of the Third International Symposium and Workshops on Surgery of the Inner Ear, Aspen, CO. In I. K. Arenberg (Ed.), *Surgery of the Inner Ear* (pp. 431-436). Amsterdam, New York: Kugler Publications.
72. Lehrer, J. F., & Poole, D. C. (1981). Abnormalities of the stapedius reflex in patients with vertigo. *The American Journal of Otology, 3*(2), 96-103.
73. Mallinson, A., & Longridge, N. (1999). Authors' Reply in response to R. Ferrari, Letters to Editor. *The American Journal of Otology, 20*(5), 700-701.
74. Koo, J., Chang, M., Song, J., & Kim, J. (2014). Contralateral suppression of distortion product otoacoustic emissions: a potential diagnostic tool to evaluate the vestibular nerve. *Journal of Vestibular Research, 24*(51), 83.
75. Lehrer, J. F. (2004). The use of anti-serotonin drugs in the nucleoreticular vestibular syndrome: Preliminary observations. *International Tinnitus Journal, 10*(1), 84-86.
76. Liveing, E. (1997 and 1873). *On megrim, sick-headache, and some allied disorders: a contribution to the pathology of nerve-storms.* (pp. 120-130). London: J and A Churchill. (1873); Nijmegen, Netherlands: Arts & Boeve (1997).
77. Tepper, S. J. (2004). *Understanding migraine and other headaches.* (pp. 25-27). Jackson, MS: University of Mississippi Press.
78. Young, R., Khorana, N., Bondareva, T., & Glennon, R. A. (2005). Pizotyline effectively attenuates the stimulus effect of N-methyl-3, 4-methylenedioxyamphetamine (MDMA). *Pharmacology Biochemistry and Behavior, 82*(2), 404-410.
79. Arenberg, I. K., Ackeley, R. S., Ferraro, J., & Muchnik, C. (1988). ECOG results in perilymphatic fistulas: Clinical and experimental studies. *Otolaryngology−Head and Neck Surgery, 99*(5), 435-443.
80. Nomura, Y., & Hara, M. (1986). Experimental perilymphatic fistula. *American Journal of Otolayrngology, 7*(4), 267-275.
81. Nomura, Y., Hara, M., Funai, H., & Okuna, T. (1987). Endolymphatic hydrops in perilymphatic fistula. *Acta Oto-Laryngologica, 103*(5-6), 469-476.
82. Meyerhoff, W. L., & Yellin, M. W. (1990). Summating potential/action potential ratio in perilymph fistula. *Otolaryngology−Head and Neck Surgery, 102*(6), 678-682.
83. Nadol, J. B. Positive Hennebert's sign in Meniere's disease. (1977). *Archives of Otolaryngology, 103*(9), 524-530.
84. Smith, P., Zheng, Y., Horii, A., & Darlington, C. L. (2005). Does vestibular damage cause cognitive dysfunction in humans. *Journal of Vestibular Research, 15*(1), 1-9.
85. Hanes, D., & McCollum, G. (2006). Cognitive-vestibular interactions: A review of patient difficulties and possible mechanisms. *Journal of Vestibular Research, 16*(3), 75-91.
86. Padula, W. V., & Argyris, S. (1996). Post trauma vision syndrome and visual midline shift syndrome. *NeuroRehabilitation, 6*(3), 165-171.
87. Cevette, M. J., Puetz, B., Marion, M. S., Wertz, M. L., &

Muenter, M. D. (1995). Aphysiologic performance on dynamic posturography. *Otolaryngology−Head and Neck Surgery, 112*(6), 676-688.
88. Healy, R., & Jankowski, T. A. (2007). Which diuretics are safe and effective for patients with a sulfa allergy? *The Journal of Family Practice, 56*(6), 488-490.
89. Lehrer, J. F. (1988). *Dehydration testing for perilymphatic fistula utilizing posturography.* Neurocom Users Conference, Dallas, TX.
90. Cole, G. (1995). Validity of spontaneous perilymph fistula. *The American Journal of Otology, 16*(6), 815-819.
91. Lehrer, J. F., Rubin, R. C., Poole, D. C., Hubbard, J. H., Wille, R., & Jacobs, G. B. (1984). Perilymphatic fistula: A definitive and curable cause of vertigo following head trauma. *The Western Journal of Medicine, 141*(1), 57-60.
92. Bluestone, C. D. (1988). Otitis media and congenital perilymphatic fistula as a cause of sensorineural hearing loss in children. *The Pediatric Infectious Disease Journal, 7*(11), S141-S145.
93. Myer, C. M., 3rd, Farrer, S. M., Drake, A. F., & Cotton, R. T. (1989). Perilymphatic fistulas in children: Rationale for therapy. *Ear and Hearing, 10*(2), 112-116.
94. Kielmovitch, I. H., & Friedman, W. H. (1988). Unilateral sensorineural deafness in children. *Otolaryngology−Head and Neck Surgery, 99*(6), 548-551.
95. Parnes, L. S., & McCabe, B. F. (1987). Perilymph fistula: An important cause of Deafness and dizziness in children. *Pediatrics, 80*(4), 524-528.
96. Chester, J. (1991). Whiplash, postural control and the inner ear. *Spine, 16*(7), 716-720.
97. Goodhill, V. (1979). *Ear diseases, deafness and dizziness.* Hagerstown, MD: Harper & Row Publishers.
98. Yaremchuk, K., & Dobie, R. A. (2001). Otologic injuries from airbag deployment. *Otolaryngology−Head and Neck Surgery, 125*(3), 130-134.
99. Serrador, J., Blatt, M., Haber, Y., Ghobreal, B., & Acosta, A. (2014). Blast exposure is associated with unilateral vestibular damage in US veterans. *Journal of Vestibular Research, 24*(51), 83-84.
100. Remenschneider, A., Lookabaugh, S., Aliphas, A., Brodsky, J., Devaiah, A., Daher, W., …Quesnel, A. (2014). Otologic outcomes after blast injury: the Boston Marathon experience. *Otology & Neurology, 35*(10), 1825-1834.
101. Lehrer, J. F., Quraishi, A. U., & Poole, D. C. (1987). The role of endolymphatic sac surgery in the management of secondary endolymphatic hydrops associated with perilymphatic fistulas: Preliminary observations. *The American Journal of Otology, 8*(2), 93-95.
102. Conner, G. H., & Potter, C. R. (1983). Hydrops following perilymph fistula repair. *Laryngoscope, 93*(6), 810-812.
103. Marchbanks, R. Why monitor perilymphatic pressure in Meniere's disease. (1997). *Acta Oto-Laryngologica, 117*(Stockholm Suppl 526), 27-29.
104. Schuknecht, H. F. (1978). Delayed endolymphatic hydrops. *The Annals of Otology, Rhinology & Laryngology, 87*(6 Pt 1), 743-748.
105. Slater, R. Benign recurrent vertigo. (1979). *Journal of Neurology, Neurosurgery, and Psychiatry, 42*(4), 363-367.
106. Leliever, W., & Barber, H. (1981). Recurrent vestibulopathy. *Laryngoscope, 91*(1), 1-6.
107. Kohl, R., Calkins, D., & Mandell, A. (1986). Arousal and stability: the effects of five new sympathomimetic drugs suggest a new principle for the prevention of space motion sickness. *Aviation, Space and Environmental Medicine, 57*(2), 137-143.
108. Minor, L., Solomon, D., Zinreich, J., & Zee, D. (1998). Sound- and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal. *Archives of Otolaryngology−Head & Neck Surgery, 124*(3), 249-258.
109. Minor, L. (2000). Superior canal dehiscence syndrome. *The American Journal of Otology, 21*(1), 9-19.
110. Belden, C., Weg, N., Minor, L., & Zinreich, J. (2003). CT evaluation of bone dehiscence of the semicircular canal as a cause of sound- and/or pressure-induced vertigo. *RSNA Radiology, 226*(2) 337-343.
111. Minor, L. & Carey, J. P., & the Vestibular Disorders Association. (n.d.). *Superior Semicircular Canal Dehiscence* (SSCD). Retrieved from http://vestibular.org/superior-canal-dehiscence-scd
112. Browaeys, P., Larson, T. L., Wong, M. L., & Patel, U. (2013). Can MRI replace CT in evaluating semicircular canal dehiscence. *AJNR American Journal of Neuroradiology, 34*(7), 1421-1427. doi: 10.3174/ajnr.A3459
113. Gopen, Q., Zhou, G., Poe, D., Kenna, M., & Jones, D. (2010). Posterior semicircular canal dehiscence: First reported case series. *Otology & Neurotology, 31*(2), 339-344.
114. Bassim, M., Patel, K., & Buchman, C. (2007). Lateral semicircular canal dehiscence. *Otology & Neurotology, 28*(8), 1155-1156.
115. Jannetta, P., Moller, M., & Moller, A. (1984). Disabling positional vertigo. *The New England Journal of Medicine, 310*(26), 1700-1705.
116. Brackmann, D., Kesser, B., & Day, J. D. (2001). Microvascular decompression of the vestibulocochlear nerve for disabling positional vertigo: The House Ear Clinic experience. *Otology & Neurology, 22*(6), 882-887.
117. Jongkees, L. B. W. (1969). *Cervical vertigo. Laryngoscope, 79*(8), 1473-1484.
118. Ferrari, R. (1999). Letter to the Editor. *The American Journal of Otology, 20*(5), 700.
119. Karlberg, M., Magnusson, M., Malstrom, E. M., Melander, A., & Moritz, U. (1996). Postural and symptomatic improvement after physiotherapy in patients with dizziness of suspected cervical origin. *Archives of Physical Medicine and Rehabilitation, 77*(9), 874-882.
120. Lempert, T. (2013). Vestibular migraine. *Seminars in Neurolology, 33*(3), 212-218.

121. Liveing, E. (1997 and 1873). *On megrim, sick-headache, and some allied disorders: a contribution to the pathology of nerve-storms.* (pp. 120-130). London: J and A Churchill. (1873); Nijmegen, Netherlands: Arts & Boeve (1997).
122. Lehrer, J. F., Poole, D. C. (1993). Letter to the Editor: The link between migraine and dizziness. *Headache: The Journal of Head and Face Pain, 33*(3), 162-163.
123. Halberstadt, A. L., & Balaban, C. D. (2003). Organization of projections from the raphe nuclei to the vestibular nuclei in rats. *Neuroscience, 120*(2), 573-594.
124. Olpe, H. (1981). The cortical projection of the dorsal raphe nucleus: Some electrophysiological and pharmacological properties. *Brain Research, 216*(1), 61-71.
125. Young, R., Khorana, N., Bondareva, T., & Glennon, R. A. (2005). Pizotyline effectively attenuates the stimulus effect of N-methyl-3, 4-methylenedioxyamphetamine (MDMA). *Pharmacology Biochemistry and Behavior, 82*(2), 404-410.
126. Tajti, J., Uddman, R., & Edvinsson, L. (2001). Neuropeptide localization in the "migraine generator" region of the human brainstem. *Cephalalgia, 21*(2), 96-101.
127. Dahlem, M. (2013). Migraine generator network and spreading depression dynamics as neuromodulation targets in episodic migraine. *Chaos, 23*(4), 46-101.
128. Staab, J., & Ruckenstein, M. (2005). Chronic dizziness and anxiety, effect of course of illness on treatment outcome. *Archives of Otolaryngology−Head & Neck Surgery, 131*(8), 675-679.
129. Brandt, T. (1996). Phobic postural vertigo. *Neurology, 46*(6), 1515-1519.
130. Furman, J. M., Balaban, C. D., & Jacob, R. G. (2001). Letter to the Editors. Interface Between Vestibular Dysfunction and Anxiety: More than Just Psychogenicity. *Otology & Neurology, 22*(3), 426-427.
131. Balaban, C. (2002). Neural substrates linking balance control and anxiety. *Physiology & Behavior, 77*(4-5), 469-475.
132. Balaban, C. (2004). Projections from the parabrachial nucleus to the vestibular nuclei: Potential substrates for autonomic and limbic influences on vestibular responses. *Brain Research, 996*(1), 126-137.
133. Brandt, T. (1991). Vertigo: Its multisensory syndromes. (p. 299). London, Great Britain: Springer-Verlag..
134. Brandt, T. (1991). Man in motion. Historical and clinical aspects of vestibular function: a review. *Brain, 114*(Pt 5), 2159-2174.
135. Bronstein, A., Brandt, T., & Woollacott, M. (1996). *Clinical disorders of balance, posture and gait.* New York: Oxford University Press.
136. Horak, F. (1986). Clinical measurement of postural control in adults. *Physical Therapy, 67*(12), 1881-1885.
137. Nashner, L. M. (1982). Adaptation of human movement to altered environments. *Trends in Neurosciences, 5*(5) 358-361.

Acknowledgement

I wish to express my gratitude to numerous colleagues and teachers who have shared their thoughts and insights and have provided suggestions that have encouraged me to pursue the paths that have led to my present understanding of vestibular disorders. Some are referenced in this article and others that educated and enlightened me did so through personal contacts.

I wish to thank Ms. Jennifer Seymour for her invaluable assistance, without which this article could not have been completed.

I wish to acknowledge my longtime collaborator and co-author, Ms. Donrue Poole, who is no longer at my side to provide ideas, information and her expertise in the endeavors described above.

About the Author

Dr. Joel F. Lehrer is one of the foremost ear and dizziness (balance) specialists in New Jersey. He has lectured and published extensively with particular emphasis in the areas of dizziness and imbalance and disorders of the inner ear. One of the founding partners of Northern Jersey Ear, Nose and Throat, PA, Dr. Lehrer was trained in Otolaryngology and completed a Fellowship in Otology and Neurotology at the Mount Sinai Hospital in New York City. He is board certified in Otolaryngology and Head and Neck Surgery and is a member of the American College of Surgeons. Dr. Lehrer was a member of the teaching staff at the Mount Sinai Hospital in Otolaryngology and Neurotology and at the Flower Fifth Avenue Hospital in Otolaryngology and has appointments at the Mount Sinai Medical School and the New York Medical College. From 1985 to 2014, Dr. Lehrer was Clinical Associate Professor of Otolaryngology at the University of Medicine and Dentistry of New Jersey. He is a Fellow of the American Neurotology Society and a member of the Barany Society and of the Neurotological and Equilobriometric Society, both of which are societies devoted to neurotology. Dr. Lehrer's practice provides information on dizziness, imbalance and disorders of the inner ear, diagnosis and treatments for ear disorders as well as rehabilitative services for hearing loss.