



General Vestibulopathy

By Gerard J. Gianoli, MD

So, you've been diagnosed with "General Vestibulopathy." Is it a meaningful or a worthless diagnosis? What does it mean and what do you do from here?

VESTIBULOPATHY

A disorder characterized by dizziness, imbalance, nausea and vision problems whose cause is unknown.

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I recently saw WM, an established patient of mine who was doing quite well, for an annual follow up visit. Thinking back on her story reminded me of the importance of receiving a correct diagnosis when you have an inner ear problem. At 35 years of age, WM tripped while attending a wedding, hitting her head and causing a severe laceration. Blood was everywhere. She was brought to the emergency room (ER) where they sewed up her forehead and diagnosed her with a concussion. Her forehead healed, but she was left with horrible imbalance that required her to use a walker to get around. Life was not easy for this patient, who was the mother of two young children at the time. She had vertigo spells once a week with associated vomiting, but the constant imbalance was her main disability. As one can imagine, this hampered her independence. She went from specialist to specialist receiving multiple diagnoses, but no one identified a vestibular component to her condition. Many of the doctors told her that her balance problem had nothing to do with the fall and head injury. The ones that did think it was related told her that she injured the balance portion of her brain, and that she would have to learn to live with it. She went through an untold number of physical therapy treatments that did not help her. Unfortunately, it was over 35 years before she received any indication that she had an inner ear problem.

Good luck trying to find the definition of "General Vestibulopathy" on the internet. In a Google search I found only four matches and in none of these is the term defined. I've been practicing neurotology with a strong emphasis on vestibular disorders for 24 years, and I've never used the term until I was asked to write this column. Don't get me wrong. I know what "general" means and I know what "vestibulopathy" means, but I've never used them together or heard them used together until now.

There is an ICD 10 code (the codes that doctors and hospitals use for billing purposes) that describes General Vestibulopathy - H81.9 "unspecified disorder of vestibular function." This is defined as:

- A disorder characterized by dizziness, imbalance, nausea, and vision problems.
- Pathological processes of the vestibular labyrinth which contains part of the balancing apparatus. Patients with vestibular diseases show instability and are at risk of frequent falls.

Given that there are numerous disorders that can cause “dizziness, imbalance and nausea,” as well as numerous “pathologic processes of the vestibular labyrinth,” this diagnosis may not seem very helpful.

I disagree. I think the term “General Vestibulopathy” has significant utility.

As a Neurotologist, my practice is heavily weighted toward vertigo and dizziness. Approximately 90% of my patients have vertigo/dizziness as their main complaint. Many (most?) of these patients, during the onset of their vestibular disorder, encountered one of the following diagnostic dilemmas:

1. They could not adequately describe their symptoms (e.g. dizziness, vertigo) to their doctors.
2. Symptoms frequently associated with inner ear problems (hearing loss, tinnitus, aural fullness) were not recognized by the patient or the doctors to be part of the disease process. Consequently, these symptoms may not have been relayed to the doctors or were not elicited by the doctors.

If one of these scenarios occurred, the physicians seeing the patient would likely be perplexed as to the cause. The immediate reaction by a competent physician is to first rule out life-threatening causes – heart disease, cerebrovascular disease, seizure disorder, meningitis, brain tumor, etc. While it may be reassuring to the patient that there is no life-threatening cause, a serious life-altering problem still exists. Unfortunately, the causes for ill-defined “dizziness” are numerous.

When someone is diagnosed as having “General Vestibulopathy” it identifies the inner ear as the source of the problem. True, it is not specific to the cause, but it does essentially eliminate other organ systems as causative. The main benefit of this is the elimination of the fruitless endeavor of testing every possible organ system. No need for a thallium stress test. No need for an EEG. No need for a psychiatric evaluation. I think you get the picture. Of course, it is always possible to have more than one thing wrong. Certainly someone can have an inner ear problem AND heart disease (or a number of other conditions).

The point here is that, **once the diagnosis of “General Vestibulopathy” has been made**, at least for the symptoms of dizziness/vertigo, **diagnostic**

ONCE THE DIAGNOSIS OF “GENERAL VESTIBULOPATHY” HAS BEEN MADE, DIAGNOSTIC EFFORTS GOING FORWARD CAN FOCUS ON THE VESTIBULAR SYSTEM AND CAN EXCLUDE OTHER PHYSIOLOGIC SYSTEMS OUTSIDE OF THE INNER EAR.

efforts going forward can focus on the vestibular system and can exclude other physiologic systems outside of the inner ear.

The next step after the diagnosis of “General Vestibulopathy” is to identify the underlying inner ear problem. The process is one of narrowing down the possibilities.

It has been often quoted that “90% of the diagnosis is history.” While that may be true when making the diagnosis of “General Vestibulopathy” it is not so true when trying to get a more specific diagnosis – i.e. BPPV, bilateral vestibular loss, Meniere’s disease, Migraine Associated Vertigo, etc. For the diagnosis of “General Vestibulopathy” history remains important. But for more specifics, testing is mandatory.

Most testing for vestibular problems can be classified into two categories 1) anatomic testing and 2) physiologic testing.

Anatomic tests are imaging studies – e.g. MRI and CT scan. These are helpful in identifying lesions such as tumors, cysts, inflammation, ischemia and aberrant anatomy (congenital, developmental or acquired). While these processes are important to identify, they do not tell the whole story. For example, most patients are sent for an MRI scan if they have persistent dizziness. Some will inevitably be found to have a tumor on their MRI scan.



However, for about half of these patients, the tumor is an incidental finding and has nothing to do with the cause of the patient's dizziness.

Physiologic testing (Video ENG, Audiometry, Rotational Testing, etc.) of the vestibular system tells us the status of the vestibular system. Has there been loss of vestibular function on one or both sides? Has the brain adjusted for the loss of function - central compensation? Is there low frequency dysfunction, high frequency dysfunction, or some combination? Physiologic testing can also directly identify specific diagnoses, such as Benign Paroxysmal Positional Vertigo, during the Dix-Hallpike Test. In other cases it may not be diagnostic, but is highly suggestive of a specific diagnosis, such as Superior Semicircular Canal Dehiscence with VEMP testing.

My patient WM, who I discussed at the beginning of this article, saw many doctors at some of the best medical centers in the US. However, for 35 years no clinician seemed to put together that her progressive asymmetric hearing loss and her episodes of vertigo were related to her chronic imbalance. Imagine using a walker or a cane for 35 years due to a correctable condition that was overlooked! About one year prior to referral to see me, an astute physician finally identified her problem as possibly due to an inner ear disorder. She was eventually referred to our office where she

was diagnosed as having a perilymphatic fistula and associated BPPV. Within 6 weeks of starting medical therapy she no longer needed her walker, her imbalance resolved, vertigo ceased and her hearing stabilized. That was four years ago and she is still doing great.

The important turning point for WM was a doctor making the correct diagnosis of an inner ear problem (i.e. General Vestibulopathy). This finally directed her down the path to an appropriate treatment regimen. So, although General Vestibulopathy may not be a specific diagnosis, it is helpful in narrowing efforts in the appropriate direction and prevents unnecessary evaluations/treatments.

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