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Case report

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# Vestibular assessment of a patient with myasthenia gravis: case report

## Abstract

Vestibular Assessment in patients with Myasthenia Gravis (MG) is challenging, as diagnostic evaluation requires good recording of eye movements. Reports on Vestibular Function Testing (VFT) in MG patients have been scant and it is arguable that VFT will have little clinical value in the MG population. A 75-year-old man, with late onset acquired autoimmune MG presented with dizziness for evaluation. He completed VFT with no significant abnormalities in all tests and was elated to have vestibular ruled out as a contributing factor to his dizziness and imbalance. However, his functional impairments were still addressed and managed regardless of the test results. MG is a heterogenous condition that may be well-controlled with treatment. Patients with dizziness can still be diagnostically evaluated to rule in or out a vestibular involvement and should not be precluded from VFT. Patients should also be assessed for their functional impairments and not based on symptom checklist and objective test results alone. Hence, patients with normal VFT results can still benefit from a hybrid of vestibular rehabilitation therapy (VRT) with focus on habituation.

**Keywords:** myasthenia gravis, dizziness, therapeutics, exercise, education

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Kenneth Chua,[1,2] Sakumura J[1]

[1]The American Institute of Balance (AIB), United States
[2]Depart of Otolaryngology, Changi General Hospital (CGH), Singapore

**Correspondence:** Kenneth Chua, Department of Otolaryngology, Allied Health (Audiology), Changi General Hospital EHN 2B, 2 Simei St 3, Singapore, Tel +6569365261, Email Kenneth_chua@cgh.com.sg

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**Abbreviations:** MG, myasthenia gravis; AChR, acetylcholine receptors; LRP4, lipo-protein receptor related protein; MuSK, muscle-specific kinase; VRT, vestibular rehabilitation therapy; VFT, vestibular function testing; CDP, computerized dynamic posturography; SOT, sensory organization test; VNG, videonystagmography; KRC, kinetic rotatory chair; vHIT, video head impulse test; VAST, vertebral artery screen test; c-VEMP, cervical-vestibular evoked myogenic potential; ABR, auditory brainstem response; BRT, balance retraining therapy

## Introduction

Myasthenia Gravis (MG) is a rare neuromuscular junction disease that causes fluctuating skeletal muscle weakness of the eye, face, oropharyngeal, axial and limb muscles.[1] It was first described by Fredrich Jolly in 1895 as a form of severe (gravis) muscle and weakness forming (myasthenia).[2] This rare disease has a reported prevalence of 7.77 cases per 100,000 person-years and can be congenital or acquired.[2] The age of onset defines the subgroups of MG patients with 50 years as the upper-limit cut-off for early onset.[3] Regardless of MG subgroups, the main mechanism of MG are auto-antibodies, which target the post-synaptic muscle receptors at the neuromuscular junction. 80% of MG patients have antibodies that target Acetylcholine Receptors (AChR), 5% targeting Muscle-Specific Kinase (MuSK), <5% targeting low density lipo-protein receptor related protein 4 (LRP4) and the remaining with low or undetectable serum levels of antibodies via conventional assays, or other antibodies not known.[2,3] MG patients have reported variable degree of ocular weakness, including ptosis, diplopia and ocular misalignment that makes oculomotor assessment difficult during vestibular testing. In this case, however, patient was well controlled with a combination of immunotherapeutic and pharmacologic treatments[4] for MG and did not have significant ocular weakness or fatigue. Patients who present with vertigo/dizziness and with a background of MG may present as a challenge, as diagnostic vestibular testing often requires a good recording of eye movements and posturography. Thus far, there has been no reports on the assessment of vestibular function in MG patients with dizziness. We report a case of assessing an MG patient where VFT was of clinical value.

## Case report

The patient presented at the American Institute of Balance, Largo facility with the primary complaint of dizziness and imbalance. Balance problems have been ongoing for years, worsening gradually with a reported recent increase in falls. He reported a relatively complex neurologic history including a diagnosis of myasthenia gravis, which is currently managed by neurology. Orthopedic history is also remarkable for significant knee pain and he has bilateral total knee replacement procedures scheduled for January of 2020. He reportedly uses a cane on and off when he feels necessary but has never worked with physical therapy. Current lifestyle is reportedly relatively sedentary with very limited activity due to knee pain and fatigability. The patient denies any recent changes to his hearing and has no complaints of aural fullness, otalgia, otorrhea or tinnitus. The patient also denies blurring of vision in response to head movements or a true sensation of vertigo. Self-reported medical history aside from myasthenia gravis includes, cardiac stents, ventral hernia, arthritis, cataracts, anxiety, depression and rectal cancer. The patient remarked on extreme disappointment for being without a definitive diagnosis until recently, when his symptoms along with history were identified as MG. He was anxious for a quick explanation and solution to his dizziness and imbalance.

The patient was first evaluated with the Computerized Dynamic Posturography (CDP), using the Sensory Organisation Test (SOT) component (Figure 1). The SOT results were abnormal for conditions 5 and 6 with vision denied or conflicted on dynamic surface. Even though this suggested a vestibular pattern of abnormality, it was noted however that the patient was hip-dominant, left weight-bearing and remarked on significant knee discomfort throughout the test due to bilateral osteoarthritis. As the patient had weak orthopaedic strength, he became slightly intolerant to the test battery, there were hence limited number of trials on some conditions of the SOT. Motor control and adaptation tests were also deferred as a result of his significant orthopaedic history.

Otoscopy of his ears were normal, with bilateral intact tympanic membranes with normal ear canals. The patient had no

spontaneous nystagmus or gaze-evoked nystagmus Figure 2 on videonystagmography (VNG). Oculo-motor assessment of smooth pursuit Figure 3 and saccades Figure 4 were negative for central signs, with no cog-wheeling, stepwise saccadic eye movements on smooth pursuit and no dysmetria (hypometria/hypermetria) on saccades. Optokinetic response Figure 5 and post-head shaking nystagmus Figure 6 were negative for any pathological nystagmus. Despite the slightly degraded ocular motility observed that affected some of the eye recordings, this is likely influenced by his neurologic history and is of limited clinical significance.

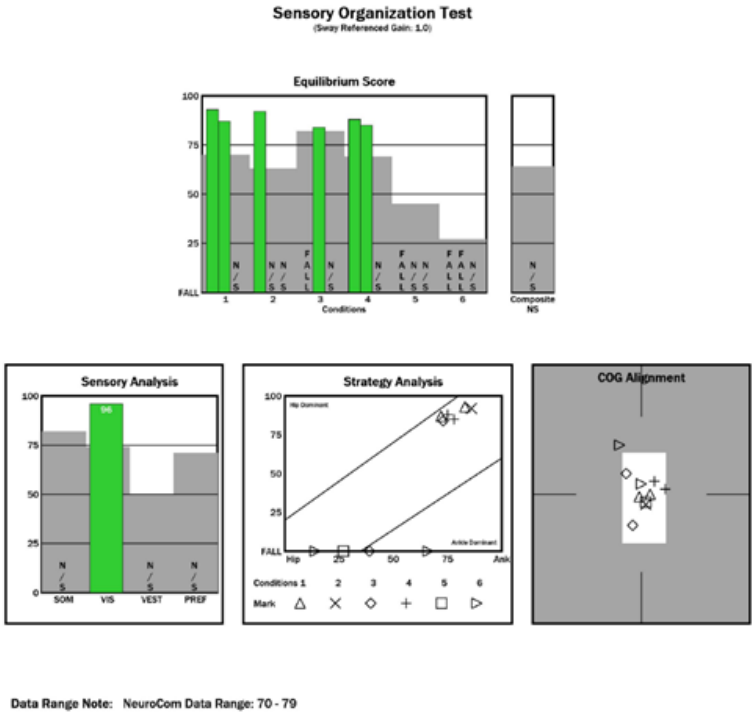


**Figure 1** Computerized Dynamic Posturography (CDP): Abnormal for conditions 5 and 6 with vision denied and vision conflicted on dynamic surface.

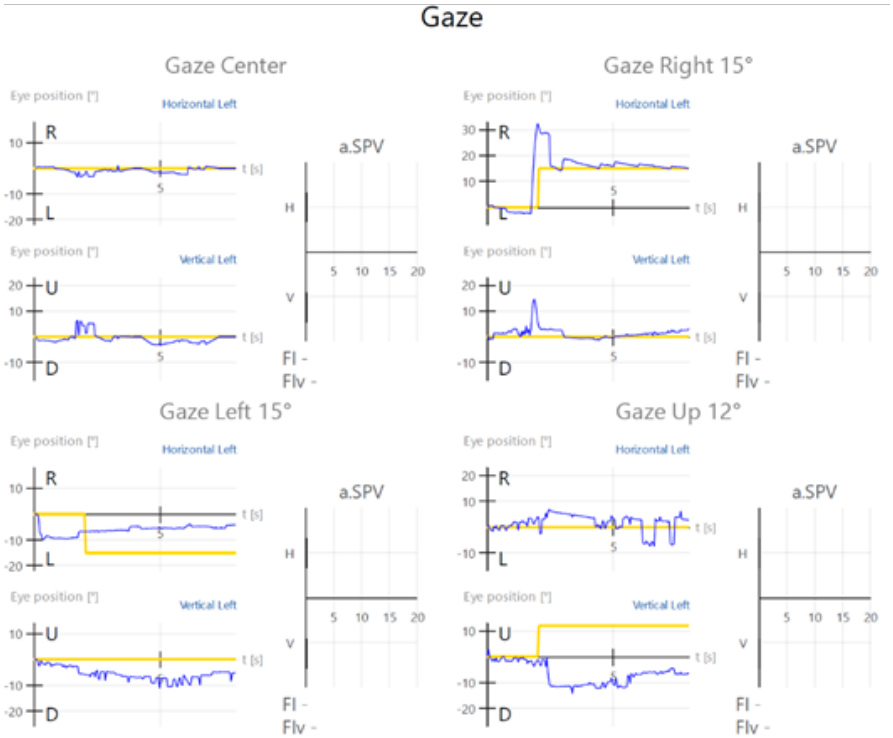


**Figure 2** Gaze evoked test for nystagmus.

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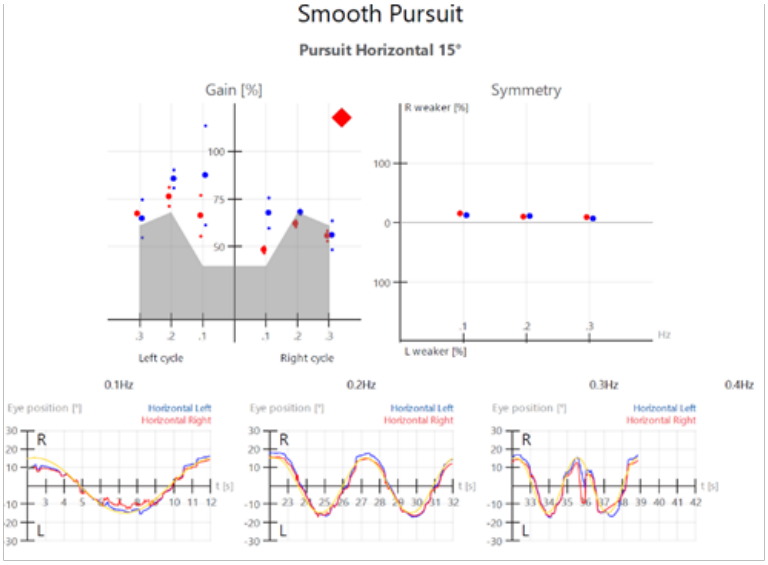


**Figure 3** Smooth pursuit oculo-motor test.

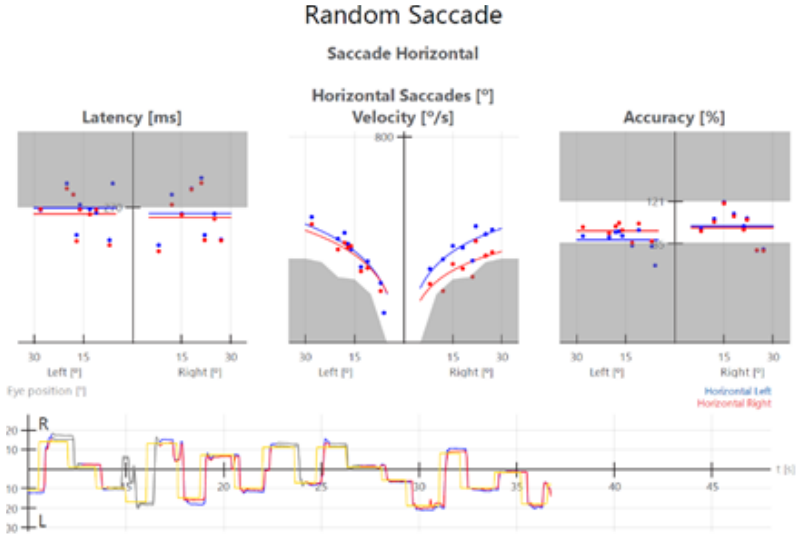


**Figure 4** Saccadic oculo-motor test.

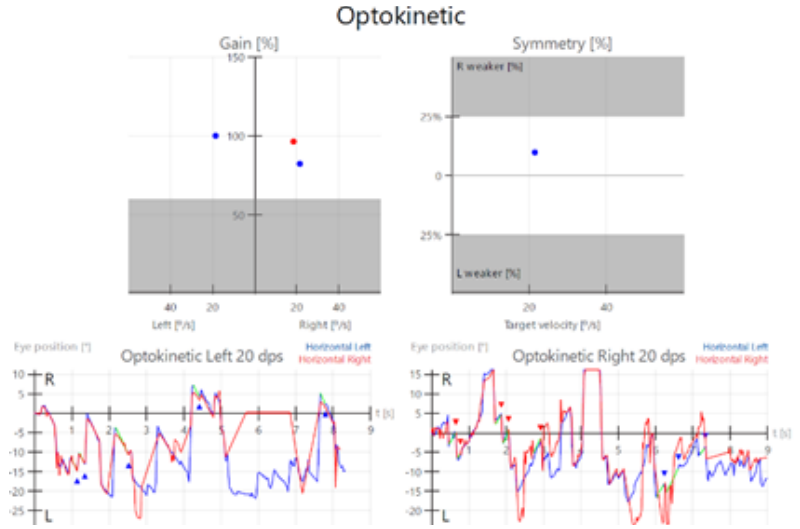


**Figure 5** Optokinetic test.

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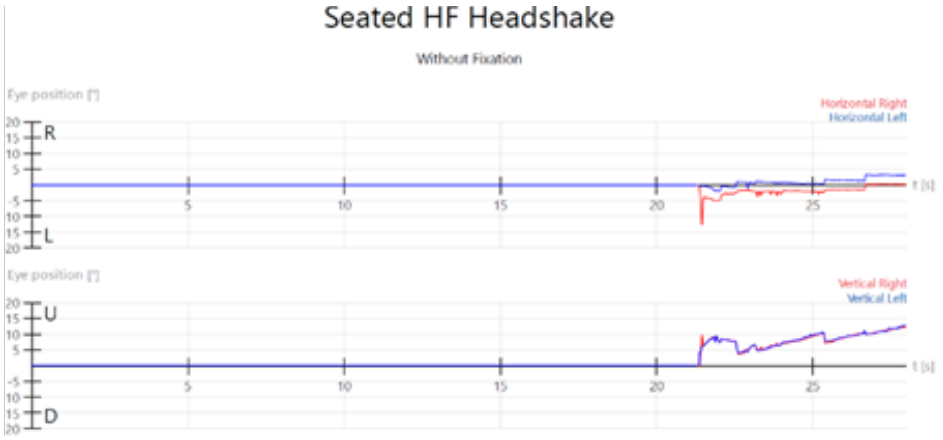


**Figure 6** High-frequency head shake test for post-head shake nystagmus.

Furthermore, the patient completed both the kinetic rotatory chair (KRC) Figure 7 and video-head impulse test (vHIT) Figure 8, which assesses vestibular function in physiological ranges. Despite the lower KRC left per rotatory gain (outlier response) and some saccades on the vHIT, these findings are of little clinical relevance in the absence of the functional impairments of oscillopsia and/or vertigo. There were symmetrical time constants on step velocity rotation to both sides, and eye movement gain within normal limits on the vHIT. The saccades observed in the vHIT could be volitional or influenced by disturbances in the neural firing of the vestibular nucleus centrally, not atypical in patients with a neurologic history.[5] Hence these saccades taken together with outlier are of little clinical significance.

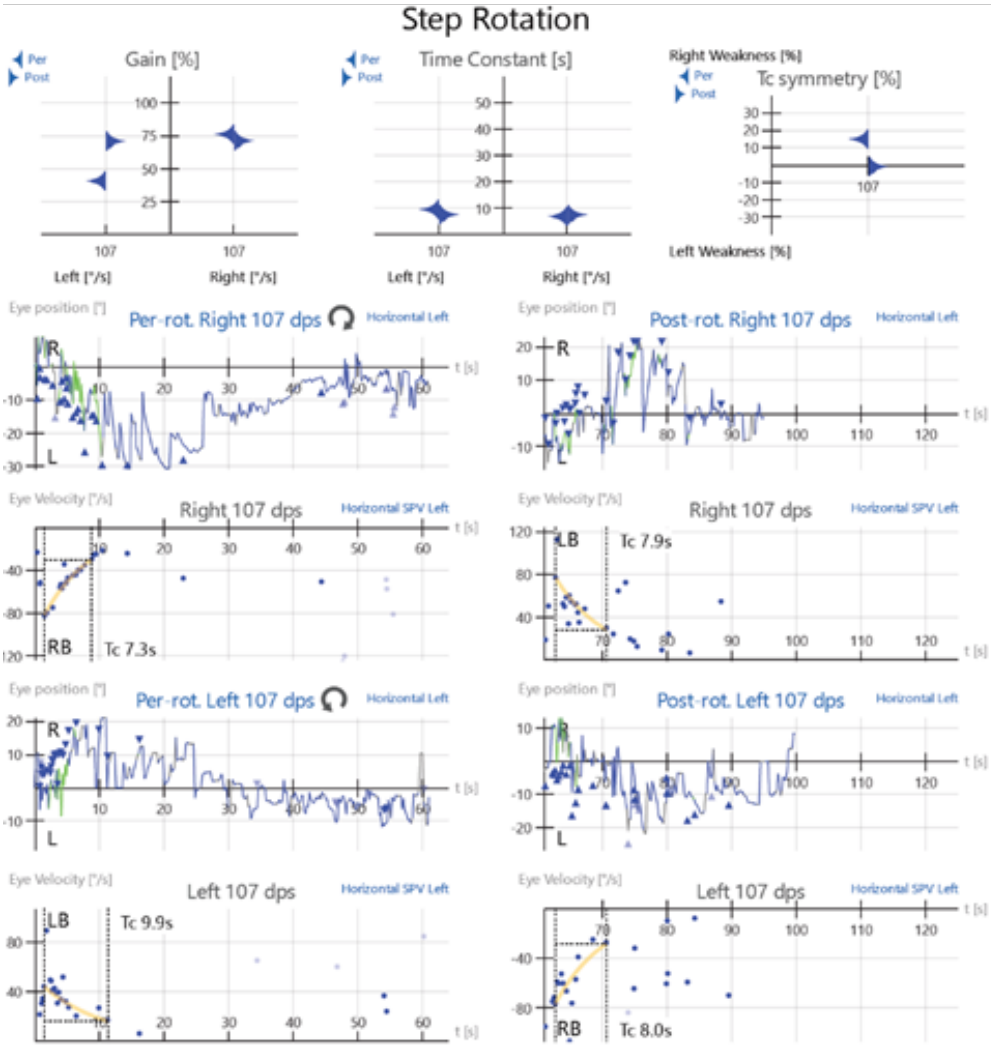


**Figure 7** Kinetic rotatory chair: step-velocity test.

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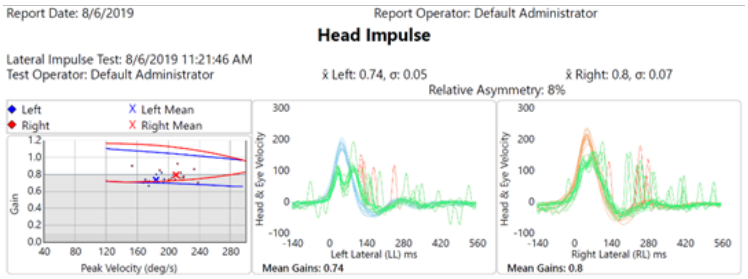


**Figure 8** Video-head impulse test (vhit)-lateral.

The patient was also screened for any compromise in the vertebral artery, with the Vertebral Artery Screening Test (VAST) before subjecting him to dix-hallpike manoeuvres on both sides. The patient was negative for BPPV of any variant in both ears. Assessment of the lateral canals were also negative for any further otolith involvement. Cervical Vestibular Evoked Myogenic Potentials (cVEMP) Figures 9a&b was however, abnormal on the right with degraded waveform morphology. This may be again influenced by the patient's neurologic history as repetitive nerve stimulation to different muscle groups, including the Sternocleidomastoid have shown degraded morphology in patients with MG.[6] There was also an isolated finding of low intensity, positional and direction fixed nystagmus on supine and body left. The slow-phase eye velocity was, however, all less than 3 beats in a ten-second window and were hence of little clinical relevance.[7] The findings were overall negative for any peripheral vestibular involvement as best can be determined by the scope and limitations of the tests. Bilateral neural conduction timings up to the level of the inferior colliculi are also normal, as evident by wave V seen on the Auditory Brainstem Response (ABR) despite a poorer morphology on the left (Figures 9c&d).

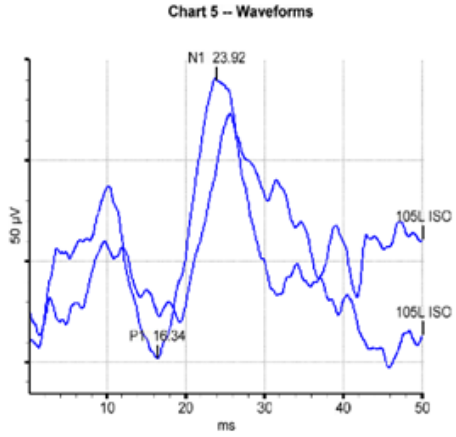


**Figure 9A** Left cervical vestibular evoked myogenic potential.


**Figure 9B** Right cervical vestibular evoked myogenic potential.


**Figure 9C** Right auditory brainstem response.

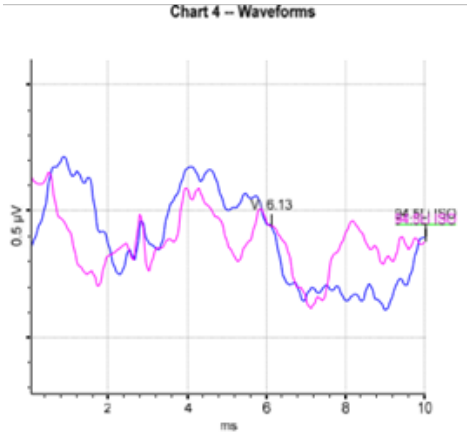


**Figure 9D** Left auditory brainstem response.

## Discussion

The test results for the patient's VFT were all negative for any active/acute or uncompensated peripheral vestibular involvement. This is supported by the absence of pathological nystagmus in any of the sub-tests, symmetrical and robust labyrinthine reactivity in the kinetic rotary chair and video- head impulse test. Despite some abnormalities with eye movements and electrophysiological responses, the patient still had relatively good VFT results. Although the patient tested negative for vestibulopathy and we can rule out any problem with the vestibular function, the patient is still experiencing dizziness, which is the functional impairment that limits his activities of daily living. Hence, while we are unable to determine that the dizziness is of labyrinthine origin, the patient still has significant functional impairment and may benefit from components of VRT. While the patient denies oscillopsia or provocation of symptoms with active head rotation, he may benefit from a hybrid balance retraining habituation type exercise regime.

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As the goal of VRT is to recalibrate the brain to "erroneous signals", which may be lacking/insignificant in patients without functional impairments, patients with functional impairments may not have vestibulopathy (a diagnosis) but may benefit from counselling, education and therapeutic exercises that will condition the brain to adapt and habituate to overcome the impairment.

On the contrary, a common clinical mistake is to send patients tested positive for vestibulopathy but with no functional impairment (no evidence of oscillopsia, vestibular recruitment, visual-vestibular dysfunction or subjective complains of dizziness and disequilibrium) for VRT. These patients do not benefit from it as there is nothing for VRT to correct. Dizziness is but a symptom that may or may not be accompanied with a diagnosis. A diagnosis alone with a symptom checklist and objective test findings are insufficient to determine a functional impairment. Functional impairment is a consequence that ensues as a result of the symptoms, such as a major limitation in activities of daily living, relative to most people. Hence, patients with vestibulopathy determined by objective VFT may not be functionally impaired and will not benefit from VRT. This supports the need for both diagnostics and rehabilitation professionals to have in-depth conceptual and procedural knowledge of vestibular and equilibrium mechanisms. Furthermore, there are extensive cross-linking neural networks between the vestibular nuclei and higher centres of the brain believed to link the vestibular and limbic systems, via neuroanatomical and neurochemical connections.[8] The modulation of mood and emotions may hence depend on vestibular stimulation. As mood disorders are co-morbidities commonly reported in MG patients,[9] guided vestibular exercises to stimulate the labyrinthine and vestibular nucleus centrally may modulate mood and influence emotions,[10,11] regardless of vestibular function.

Hence, there is value even if the test results are negative as it is of benefit to have objective testing to determine the relationship between functional impairment and physiological status of the vestibular system. The normal VFT results was thus of clinical value, as it was used as a powerful counselling and education tool to empower the patient and to help the patient readily accept the most efficacious diagnosis-based plan and rehabilitation strategies, which focused on the functional and not organic impairment. The patient was thus given a series of prophylactic self-directed vestibular rehabilitation exercises, guided with instructions and demonstrations. It was also suggested to the patient to involve cognitive tasking with these exercises at home as cognitive engagement is important for optimal outcomes.[12] Lastly, the patient was asked to return for Balance Retraining Therapy (BRT) with a physical therapist for strengthening, conditioning and balance rehabilitation after his bilateral knee replacements and, if there are no contraindications for it after consulting his neurologist. All the educational information, counselling and self-directed exercises were given to the patient in both verbal and written form and the patient exhibited good understanding. In MG patients, VFT is commonly overlooked as it is believed that dizziness is not of a labyrinthine aetiology. While that may be true, MG patients should not be precluded from VFT as it not only serves to rule in/out the vestibular system's involvement with dizziness but has other clinical applications. MG patients with vertigo can be sent for vestibular function testing if the MG is well-controlled. Muscle fatigue and weakness may not necessarily preclude a good recording of eye movements. Diagnostic results from vestibular testing can be useful to inform patient and other healthcare providers to exclude or include, the contribution of peripheral vestibular function as a cause of dizziness and to correlate with any functional impairments.

Regardless of the VFT results, clinicians should focus on treating the patients' functional impairments through education, counselling and rehabilitation exercises as part of a cognitive-behavioral intervention. Objective test results should support and supplement clinical expertise when making management decisions. Patients without a vestibulopathy (diagnosis) but with functional impairments should not be readily turned away. In a dizzy patient, the further they are from the diagnosis of a neurotological syndrome, the greater the overlap of psychiatric disorder[13,14] such as anxiety and panic disorders that may exacerbate their functional impairments. Hence the empowerment of patients through education and counselling, not only reduces the effects of psychiatric co-morbidities, but also improves mood and mediate balance when combined with cognitive engagement and habituation exercises. Extensive neural networks between the cerebral cortices, thalamic relay stations and vestibular nuclei are believed to be providing the platform for such an integration. On the other hand, clinicians should exercise caution when making management decisions based solely on objective test results, as these test results should be correlated with the patients' functional impairments.

## Acknowledgments

None.

## Conflicts of interest

The author declares that there is no conflict of interest to disclose.

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