

Decompensated labyrinthine weakness presenting as *de novo* peripheral vertigo: a discrete clinical entity?

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Abstract

Objective: A distinct subgroup of patients, presenting with apparently spontaneous onset of vertigo, is described.

Results: Although vestibular evaluation revealed caloric weakness, the proximate cause of vertigo was not labyrinthine dysfunction, but rather the loss of vestibular compensation for an older and previously compensated labyrinthine injury.

Conclusion: Instead of addressing the vestibular weakness, effective management needs to focus on the condition that has caused the loss of compensation.

Key words: Vertigo; Dizziness

Introduction

Dizziness, especially in older patients, often has multiple causes, and accurate diagnosis may be difficult. While dizziness is common, peripheral labyrinthine vertigo is relatively unusual in this age group. We have identified a group of patients who appear to present with recent-onset peripheral vestibular dysfunction, but whose vertigo is in fact due to deterioration of a non-vestibular balance modality which unmasks a previously compensated labyrinthine weakness.

When unilateral labyrinthine injury leaves the patient with a fixed vestibular deficit, balance is usually restored through the process of vestibular compensation. In cases of unilateral loss of labyrinthine function, a predictable sequence of events initially occurs, which has been classically described by McCabe and others.^{1,2} This includes a rapid onset of central inhibition ('cerebellar clamp'), which dampens signals from the vestibular nuclei on the undamaged side in order to decrease the discrepancy in activity between the damaged and the normal side. This is followed by a series of compensatory steps, which eventually leaves the patient with increased reliance on the undamaged contralateral vestibular system, as well as on visual and proprioceptive input.

Balance is maintained by the triad of vestibular, visual and proprioceptive systems. When one system is defective, balance can be maintained as long as two of these three mechanisms are intact. If, however,

either of the remaining two mechanisms is weakened, the patient develops imbalance. For example, a patient with bilateral peripheral vestibular weakness will lose their balance in the dark (loss of visual input) or when swimming underwater (loss of meaningful proprioceptive information).

In young and otherwise healthy patients, vestibular compensation occurs rapidly and often imperceptibly: vertigo from complete loss of unilateral vestibular function can completely disappear within a few weeks. Lesser degrees of damage can be compensated for even more easily, and the patient may not even be aware that they have been left with a partial but permanent vestibular deficit. In older patients, compensation is increasingly slower, and the elderly may never fully compensate. Several factors may be responsible for this, including decreased brain plasticity, impaired circulation and general deterioration of peripheral sensory systems. In 1988, Katsarkis and Segal reviewed factors that may facilitate or impede compensation.³ Physical activity speeds compensation, whereas lack of physical activity delays it. Animal studies suggest that drugs also affect compensation, and can be either facilitatory (caffeine, methamphetamine) or inhibitory (phenobarbital, chlorpromazine).³

This paper describes decompensated labyrinthine weakness, a discrete clinical entity of dizziness. It presents as *de novo* labyrinthine vertigo, but is in fact an unmasking of long-existing and previously compensated

peripheral vestibular weakness. Six illustrative cases are presented below.

Case reports

Case one

A 78-year-old man, presented with a 2-month history of worsening balance. He described dizziness triggered by head movements, but no nausea, vomiting or other vegetative symptoms. He had no tinnitus, and was unaware of hearing loss. In addition to his presenting complaint of imbalance, he had also become aware of recent deterioration in his vision. Six years earlier, he had been hospitalised with deep vein thrombosis and septic pulmonary emboli. He was given a Greenfield™ vena cava filter, and was treated with intravenous gentamicin for four weeks. He had no vertigo at that time.

Head and neck examination findings were normal. Neurotological examination revealed no spontaneous or gaze-associated nystagmus. Head shaking elicited a brief burst of oscillopsia. He past-pointed to the right. Stance and gait testing revealed mild ataxia, but no lateralisation.

Audiometry demonstrated a bilaterally symmetrical, high-frequency, moderate sensorineural hearing loss (SNHL), with good discrimination and normal middle-ear function. Electrocochleographic findings were normal. Video-nystagmography revealed a 26 per cent right-sided vestibular weakness.

Decompensated labyrinthine weakness was diagnosed. The patient was treated with eyeglasses that corrected his vision, followed by vestibular therapy. He made a full recovery.

Case two

A 72-year-old man presented with a 1-year history of vertigo. The episodes occurred daily and could last for hours. They were brought on by walking. There was no complaint of hearing loss or tinnitus, with no nausea or vomiting. The onset of dizziness coincided with complaints of tingling in the extremities. Past history was significant for two episodes of neurocardiogenic syncope requiring hospitalisation. The condition was medically controlled. Neurological examination led to a diagnosis of early Parkinson's disease.

Head and neck examination findings were unremarkable. Neurotological examination revealed no spontaneous or gaze-associated nystagmus. He past-pointed to the right side, and stance and gait testing confirmed lateralisation to the right. Audiometry revealed bilateral, moderate, high-frequency SNHL, significantly worse on the right side. Video-nystagmography confirmed a right peripheral vestibular weakness. Electrocochleography showed bilateral, mildly elevated endolymphatic fluid pressure, more on the right side.

In view of his history of hypotension, diuretics could not be used to treat his hydrops. Sinemet® was prescribed for the Parkinson's disease, and this completely

eliminated his vertigo. This response suggested that the hydrops may have been an incidental finding, and that his dizziness was due to decompensated labyrinthine weakness. The dizziness was relieved by addressing his worsened gait.

Case three

A 65-year-old woman, presented with a loss of balance and 'fogginess' of 2-months duration. She consistently veered to the left side, and felt like falling to the left side on walking. There was no true vertigo, tinnitus, hearing loss or nausea. Thirty years earlier she had suffered attacks of vertigo, with nausea and vomiting, and was diagnosed with Ménière's disease (affecting the left side). However, she received no treatment, and has been completely symptom-free for over 10 years.

A few months ago, she became aware of impaired vision, and was diagnosed with significant bilateral cataracts. She had difficulty with night driving.

On examination, there was a mild, high-frequency SNHL in the left ear. Video-nystagmography revealed significant left-sided caloric weakness.

She was diagnosed with left-sided labyrinthine weakness, possibly burned-out Ménière's disease to the left side, and recent decompensated labyrinthine weakness due to impaired vision. Her balance improved significantly after cataract surgery.

Case four

A 46-year-old woman presented with a 2-month history of dizziness, described as a sensation of movement and 'walking into walls'. The first episode occurred after a stressful argument with a co-worker, and lasted two weeks. Subsequent episodes were attributed to hypertension. She had fallen twice to the left side, and noted right-sided tinnitus. There was no history of hearing loss, nausea or vomiting with the dizzy spells.

Medical and neurological evaluation suggested cardiac arrhythmia, and 'white spots on the brain', interpreted as possible mini strokes. She was also hypertensive and had diabetes mellitus type 2, managed with metformin. Back surgery had left her with a right leg weakness. This had mostly recovered, but she was left with patchy hypaesthesia in the right foot. She complained of visual problems, stating 'the right eye goes in and out of focus'.

Head and neck examination was non-contributory. On neurotological examination, there was no spontaneous or gaze-associated nystagmus. On stance and gait testing, she lateralised slightly to the right side. The audiogram was normal. Electrocochleography was bilaterally normal. Video-nystagmography showed a right-sided vestibular weakness.

This patient had a right-sided caloric weakness of unknown age and aetiology, but with a clear history of visual and proprioceptive weakness, which presented two months earlier as dizziness. The coincidence with a stressful argument would suggest that the decompensation was worsened by cortical

distraction. She was referred to an ophthalmologist for management of her eye problem.

Case five

A 74-year-old man was referred for recent onset of imbalance and veering to the right side. He also became dizzy on turning to the right in bed. There was no significant associated nausea. He has had right-sided hearing loss and tinnitus since childhood due to a firecracker explosion. He had suffered polio as a child, and noticed recent and progressive weakness in his legs due to post-polio syndrome.

Head and neck examination findings were essentially normal. There was no spontaneous or gaze-associated nystagmus. Stance and gait testing was deferred because of leg weakness. The audiogram revealed bilateral, mild-to-moderate SNHL, with good discrimination. Video-nystagmography revealed a 39 per cent caloric weakness in the right ear. Electrocochleography findings were normal.

The diagnosis of decompensated labyrinthine weakness was made, triggered by his post-polio syndrome. He was referred for balance therapy, but has thus far not experienced any significant improvement.

Case six

An 81-year-old priest presented with a 2-month history of slight vertigo. He has had no nausea. He stated that when he gets up, he might fall back down at times. He has had difficulty walking, especially in his home and when looking down. Outdoors, he is able to walk 15 blocks with no difficulties. He has diabetes mellitus type 2, well controlled on metformin. He had recently developed 'slight cataracts' in both eyes.

Otological evaluation revealed bilateral, moderate-to-severe SNHL, with excellent discrimination. There was no spontaneous or gaze-associated nystagmus. Video-nystagmography demonstrated a 26 per cent left-sided caloric weakness.

The clinical impression was of decompensated labyrinthine weakness, due to recent deterioration in vision. Of interest, his vision was still adequately compensating in the well-lit outdoors, but not in darker surroundings, or when he looked down. He was referred to an ophthalmologist to address his visual impairment.

Discussion

Differential diagnosis

While dizziness is commonly seen among older patients, it is relatively uncommon for these patients to present with new-onset peripheral vertigo. When a patient gives a clear history of dizziness with a sensation of movement, and clinical evaluation confirms a unilateral peripheral vestibular weakness, the clinician may reasonably attribute the vertigo to the recent onset of labyrinthine dysfunction. This diagnosis appears to be supported by a unilaterally decreased caloric response. However, as suggested by the above

case reports, the triggering event in decompensated labyrinthine weakness patients is not a recent loss of peripheral vestibular function, but the recent loss of compensation for a previously acquired labyrinthine weakness.

Over the past 15 years, we have identified a number of patients, exemplified by the cases presented, who fit the diagnostic criteria for decompensated labyrinthine weakness syndrome.

Several clinical features distinguish decompensated labyrinthine weakness from *de novo* labyrinthine disease. The vertigo is often not episodic; rather, the patient complains of fluctuating, prolonged or constant dizziness, with a tendency to veer or fall to one side. Unlike most types of labyrinthine disease, the vertigo is persistent over weeks and months, with no abatement. The imbalance may be momentarily worsened by visual distraction or deprivation. For example, these patients may state that their vertigo is even worse when walking down a supermarket aisle, scrolling on the computer or walking to the bathroom at night.

A history of severe peripheral vertigo, head injury or other peripheral labyrinthine damage is usually not elicited; nonetheless, the patient should be specifically questioned.

Ipsilateral auditory symptoms (hearing loss, tinnitus, hyperacusis) may be absent in patients with decompensated labyrinthine weakness syndrome; although, depending on the nature of the original labyrinthine injury, there may be some cochlear damage. When auditory symptoms do exist, however, they predate the recent onset of vertigo, and do not worsen with the dizziness.

The patient's perception of vertigo is linked to the onset of a seemingly unconnected condition, which causes weakening of one of the compensatory mechanisms. Typical examples are the onset of macular degeneration or cataracts, a Parkinsonian gait disturbance, or diabetic peripheral neuropathy. While it is this aspect of decompensated labyrinthine weakness syndrome that makes the onset of vertigo more common among older patients, the condition could also develop in younger patients (as in case four) if the patient develops impairment of one of the compensatory mechanisms.

A key distinguishing feature of decompensated labyrinthine weakness syndrome is the absence of vegetative symptoms which normally accompany acute or recent-onset peripheral vertigo. Thus, although the vertigo is labyrinthine in type (i.e. peripheral), patients do not complain of symptoms such as nausea, vomiting, tachycardia, tachypnoea, hypersalivation or diaphoresis. This historic feature alone, in the context of a calorically demonstrated unilateral labyrinthine weakness, should raise the suspicion of decompensated labyrinthine weakness syndrome.

On examination, decompensated labyrinthine weakness patients usually do not have nystagmus, but they point towards the weaker side, and on stance and gait tasking they will veer or fall towards the weaker

labyrinth. Caloric testing is further misleading, as it confirms a decreased response on the affected side, at times with some additional minor age-related central signs such as impaired smooth pursuit. Although the absence of nystagmus and of vegetative symptoms point towards a central problem, the sensation of vertigo and caloric weakness will lead the clinician to believe that the primary cause is the peripheral weakness.

Table I compares the features of decompensated labyrinthine weakness with two well-known causes of dizziness, Ménière's disease and central vertigo. Decompensated labyrinthine weakness differs significantly from both these conditions. Typical peripheral vertigo occurs either once or episodically, and is accompanied by vegetative symptoms and often auditory symptoms. The discrete and brief episodes in Ménière's disease are triggered by factors known to increase endolymphatic fluid pressure, and are separated by asymptomatic intervals.

Typical peripheral vertigo does not persist for weeks and months. Central vertigo is ill-defined, but persistent. It does not cause an illusion of rotation, or a sensation of falling or veering to one particular side.

A common form of central dizziness has been termed 'multiple sensory deficit syndrome' by Baloh.⁴ The condition, which is the cumulative result of an otherwise subclinical weakness in vestibular, visual and proprioceptive (sensorimotor) functions, causes instability, with no clear single aetiology. Each of the three systems may be only mildly impaired, but becomes unable to compensate for impairment in the other two. Decompensated labyrinthine weakness differs from multiple sensory deficit syndrome in that there is a demonstrable vestibular deficit on video-nystagmography, which clinically reappears as one or more of the other systems is damaged. Further, the damage to these other systems is also clinically apparent, and usually carries a diagnosis.

Decompensated labyrinthine weakness also differs from temporary impairment of vestibular compensation as described by Katsarkis and Segal.³ These authors described episodic decompensation as a complaint of imbalance when mental alertness decreases, particularly when exposed to a moving environment. Drugs, in particular alcohol, may also bring about temporary decompensation in such patients. By contrast, decompensated labyrinthine weakness patients have chronically persistent vertigo, rendered symptomatic by recent-onset clinical disease affecting one of the compensatory systems.

It is interesting that in most decompensated labyrinthine weakness patients there is no re-emergence of nystagmus, as might be expected. This may be because suppression of nystagmus during initial compensation is primarily visually driven, whereas the ability to overcome the subjective sensation of vertigo is cortical, and compensation for stance and gait problems is proprioceptive and spinovestibular.

Therapeutic implications

As the proximate cause of vertigo in decompensated labyrinthine weakness is not the pre-existing labyrinthine weakness but the recent impairment of compensation, treatment should not be directed at vestibular suppression. These patients do not improve with meclizine or diazepam, and may in fact become worse. Nor do they improve with salt restriction and diuretics.

Treatment instead requires the identification and aggressive management of the condition that led to decompensation. Thus, improving vision by removal of cataracts or the use of corrective lenses will allow visual compensation to reassert itself, and the symptom of vertigo will improve or disappear. Improving sensory input to the soles of the feet by walking on harder surfaces and wearing thin-soled shoes can maximise proprioception, even in some cases of peripheral neuropathy. Vestibular therapy,

TABLE I
CLINICAL CHARACTERISTICS DIFFERENTIATING DECOMPENSATED LABYRINTHINE WEAKNESS FROM CLASSIC PERIPHERAL VERTIGO AND CENTRAL VERTIGO

Clinical characteristic	Peripheral vertigo*	Central vertigo†	Decompensated labyrinthine weakness
Triggering cause	Labyrinthine	CNS disease	Visual or proprioceptive loss
Historic features			
– Characteristics of dizziness	Episodic vertigo	Constant imbalance	Constant or fluctuating vertigo
– Auditory symptoms	+/-	–	–
– Vegetative symptoms	+	–	–
Clinical features			
– Horizontal nystagmus	+/-	–	–
– Stance or gait lateralisation	+/-	–	+
Audiogram & VNG findings			
– Horizontal nystagmus	+/-	–	–
– Stance or gait lateralisation	+/-	–	+
– Hearing loss or tinnitus	+	–	–
– Unilateral caloric weakness	+/-	–	+
– Central VNG findings	–	+	+/-

*For example, Ménière's disease. †For example, multiple sensory deficit syndrome. CNS = central nervous system; VNG = video-nystagmography

aimed at strengthening the contralateral labyrinth and increasing reliance on remaining visual and proprioceptive function also has some value, but only if these other systems are first made maximally operative.

- **Compensation mechanisms following peripheral vestibular deficit are well known, and rely largely on visual and proprioceptive input**
- **The clinical presentation of delayed vestibular decompensation is less clear**
- **These patients present with a discrete clinical picture, distinct from both classical peripheral and central vertigo**
- **Appropriate treatment requires identification and treatment of the cause of decompensation**

Finally, as the onset of decompensated labyrinthine weakness may be the presenting symptom of diabetes, Parkinson's disease or neurological disorders, patients with recent decompensation require thorough medical evaluation to identify and treat these potentially serious non-vestibular diseases.⁵

Conclusion

A discrete clinical entity, decompensated labyrinthine weakness, is described. The condition usually develops in older patients, although it may also be seen in younger patients. It has several clinical features that distinguish it from both *de novo* peripheral vestibular

weakness and central forms of dizziness. As the proximate cause is not vestibular, but rather in the visual or sensorimotor areas, awareness of decompensated labyrinthine weakness syndrome has important therapeutic implications. Effective treatment should be directed at the proximate cause of the vertigo, rather than the old and previously compensated labyrinthine weakness.

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