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### Mal De Debarquement Syndrome

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# Mal de débarquement syndrome

## Review and proposed diagnostic criteria

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### Summary

Mal de débarquement syndrome (MdDS) is a rare cause of imbalance encountered in a neurology practice. However, it consists of classic features that the practicing neurologist should be able to recognize when evaluating a patient with dizziness. It is characterized by a feeling of rocking and swaying, and typically follows prolonged exposure to motion, such as being on a boat or plane. In this review article, we provide the clinical neurologist with the history of this disorder followed by illustrative cases of patients diagnosed with MdDS. Next we present proposed criteria for aiding in diagnosis. Finally, we will discuss a differential diagnosis for the syndrome, insights into the possible pathophysiology, current treatments, and future directions in management.



**M**al de débarquement syndrome (MdDS) is not commonly encountered in clinical practice, but it is a unique cause of dizziness that can often be diagnosed clinically by the astute neurologist. It is characterized by classic features of rocking or swaying that begin following passive motion exposure, most commonly boat travel.<sup>1</sup> It results in a false perception of self-motion that the individual begins to experience only upon return to land. MdDS can be either self-limited or persistent; when it is persistent, it may have less likelihood of resolution. Due to the lack of awareness of this syndrome among neurologists, patients are often subjected to numerous tests and referrals prior to receiving a diagnosis. With increased understanding of this syndrome, patients may be more accurately and efficiently diagnosed by neurologists.

### History

MdDS has become increasingly recognized following Brown and Baloh's 1987 case series publication of 6 patients with a persistent motion-induced subjective balance disorder. However, the condition was first described long before then. It was perhaps initially recognized by Hippocrates, who first described nausea, taken from the root word "naus" or "ship" in the

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*The differential diagnosis for MdDS includes other entities such as vestibular migraine, chronic subjective dizziness, motion sickness, and various otologic causes of dizziness.*

Greek language. In his chapter on aphorisms, Hippocrates described his observation that “sailing on the sea shows that motion disorders the body.”<sup>2</sup> A more detailed description was provided in 1881 by J.A. Irwin, who remarked on “the unsteady gait sometimes observable in a non-drunken sailor during his first few hours on shore after a long and stormy voyage.”<sup>3</sup> Thus, throughout history, MdDS has been recognized time and again as a distinct cause of imbalance. The name of this syndrome, mal de débarquement, originates from a French term dating back to 1575 that translates to “sickness of disembarkment.” Disembarking refers to leaving an aircraft or going ashore after being on a ship. The word “mal” is Latin for “malam,” which translates to “evil.” And for those who have this syndrome, it can certainly seem like they are experiencing something evil!

### Illustrative cases

**Case 1** A 46-year-old woman presented to the neuro-otology clinic with a chief complaint of dizziness. When asked to describe her dizziness, she responded, “It’s like I’m drunk!” She had returned from a 7-day Caribbean cruise approximately 1 year prior and had felt off-balance from the moment she stepped off the ship. She also had felt a constant swaying sensation since departing from the cruise ship. While on board the ship, she had had no such symptoms. Upon disembarking, she had felt as though the ground was moving. This feeling had not subsided at all for the past year since the cruise ended. She did note that if she was driving in her car the self-motion perception improved a great deal. However, when she stopped driving and got out of the car, her disabling symptoms would immediately return. She was continuing to work as a manager of an electronics store, but she felt that she needed to be constantly walking around while at work. She preferred not to sit at her desk because her symptoms worsened when remaining still. She had seen several physicians and undergone extensive workup including various laboratory studies, brain MRI, videonystagmography (VNG), and audiogram. All studies were within normal limits. She also had been tried on medications, including verapamil, nortriptyline, alprazolam, and methylphenidate, none of which provided any relief. She did find partial relief with diazepam as needed.

**Case 2** A 40-year-old woman presented to the neuro-otology clinic for evaluation of “rocking motion.” She had a past history of migraine but no history of seasickness or car sickness. She related that she had begun feeling the rocking sensation the morning after disembarking from a 7-day cruise to Puerto Rico almost 6 months earlier. She denied nausea and vomiting. Hearing was preserved. The rocking sensation was constant but worsened if she remained still. She tried meclizine, escitalopram, and venlafaxine without any improvement in symptoms. She did have mild temporary improvement in symptoms when she took clonazepam. She tried vestibular therapy but found no relief. She was able to continue working as an engineer, but she felt as if she was moving all the time. She had a CT of the head, audiogram, and VNG performed; all of the studies were within normal limits.

### Clinical history

MdDS is defined by a persistent perception of self-motion that occurs following a period of exposure to passive motion.<sup>1</sup> This is often seen following an ocean cruise. The syndrome is more common in women, usually in their 40s.<sup>4</sup> The most common triggers are boat and ship travel, but MdDS can also occur after airplane trips. As was seen in the 2 illustrative cases, the

**Table 1** Diagnostic criteria of persistent mal de débarquement syndrome (MdDS)

- A. Onset of symptoms after exposure to passive motion,<sup>a</sup> such as following travel on a boat/cruise ship or airplane
- B. Main symptom is illusory rocking motion
- C. Symptoms improve with re-exposure to passive motion, such as boat or car travel
- D. Nausea is not a prominent symptom
- E. Minimum 3-month duration of symptoms<sup>b</sup>
- F. Not better accounted for by another International Classification of Headache Disorders-3 diagnosis or by another vestibular disorder<sup>c</sup>

<sup>a</sup>Exposure to passive motion must last at least several hours and more likely will have lasted days.

<sup>b</sup>A diagnosis of MdDS may still be made with symptoms lasting less than 3 months; however, for a diagnosis of persistent MdDS symptoms must have lasted at least 3 months.

<sup>c</sup>History and physical do not suggest another vestibular disorder, or another disorder was considered but ruled out with appropriate testing, or another disorder is present but as an independent condition with episodes that can be clearly differentiated.<sup>21</sup>

onset of symptoms is usually immediate after stepping foot on dry land. The symptoms are variable but all center on the concept of movement despite not actually physically moving. Common descriptive terms used by patients to describe their symptoms are rocking and swaying.<sup>4</sup> These patients usually do not experience nausea or vomiting and do not feel a sense of spinning. Hearing is preserved. Patients with MdDS classically report that their symptoms greatly improve or even dissipate if they are re-exposed to the movement that predated their symptoms.<sup>1,4,5</sup> For example, they feel markedly better if they return to a boat or cruise ship and get back on board. They also report feeling much better while driving a car. However, once they stop driving or return to land again, their symptoms not only return but may also temporarily worsen.<sup>4</sup> Typically, MdDS does not persist for an extended period of time. Most patients experience spontaneous resolution of symptoms within days to several months. As a result, these patients are not usually the ones presenting to specialty clinics for consultation. For an unfortunate subset of patients with MdDS, however, the symptoms do not remit within several months, and these patients may then be classified as having persistent MdDS. It is much more likely that patients with persistent MdDS will present to a clinic for formal evaluation. Studies have shown that the longer the duration of symptoms, the less the likelihood of resolution.<sup>5</sup> It is not uncommon for patients to develop anxiety and/or depression as a result of their persisting symptoms, especially if they have not been given a definitive diagnosis yet. Finally, most patients with MdDS report that they have been prescribed antivertiginous medications for their symptoms but note little relief from any of them.<sup>4,6</sup>

## Diagnosis

Simply put, MdDS remains a clinical diagnosis. The features detailed in the clinical history are what support the diagnosis of MdDS in a patient. Of these, the most important are a history of rocking illusory motion following a boat or airplane trip, a relative lack of nausea, and reduction of symptoms with re-exposure to motion (such as when driving or boating). However, the workup in a patient complaining of dizziness could potentially include testing such as VNG, MRI brain, CT temporal bone, audiogram, laboratory studies, or cardiologic evaluation. The tests vary depending on the clinical presentation of the patient. Testing is useful only in helping to exclude other disorders that might present with features similar to MdDS. However, in most classic cases of MdDS, no specific testing is necessary to make the diagnosis. The neurologic examination of a patient with MdDS is notably benign, although patients will convey that they feel constantly disabled by their symptoms or they might appear hesitant on balance tests.<sup>6</sup> VNG typically shows normal vestibular responses, hearing is normal, and no structural

**Table 2** Comparison of mal de débarquement syndrome (MdDS), vestibular migraine, and anxiety-related dizziness

	MdDS	Vestibular migraine	Anxiety-related dizziness
<b>Onset</b>	Abrupt, following boat or air travel	Spontaneous, or may follow stress or certain food triggers	More likely after stress, can also be spontaneous
<b>Description of vertigo</b>	Rocking, swaying	Spinning, floating, rocking, motion sensitivity, nausea <sup>7,9</sup>	Rocking, floating, internal spinning <sup>7</sup>
<b>Presence of migraine headaches</b>	Similar to population	Often present	Similar to population
<b>Clinical examination</b>	Normal	Normal, may have unsteadiness on Romberg testing	Normal
<b>Vestibular laboratory testing</b>	Normal	Normal or low-velocity static positional nystagmus <sup>9,15</sup>	Normal
<b>Response to medication</b>	No response usually	Often responds to same medications used to treat migraine <sup>8</sup>	May respond to anxiolytics <sup>16</sup>

abnormalities are found on brain imaging to identify MdDS.<sup>1</sup> Table 1 lists proposed diagnostic criteria of persistent MdDS.

### Differential diagnosis

The differential diagnosis for MdDS includes other entities such as vestibular migraine, chronic subjective dizziness, motion sickness, and various otologic causes of dizziness. A detailed clinical history will provide the key to differentiating among each of these (see table 2). Onset of symptoms in MdDS is typically abrupt and begins soon after disembarking from a ship or airplane. Persistent MdDS lasts months with spontaneous resolution but can persist for years. It is not preceded by an infection or other illness. It rarely responds to medications and there is no definitive diagnostic test.

**Vestibular migraine** In contrast to MdDS, vestibular migraine often consists of recurrent vertigo attacks that sometimes but not always include additional symptoms such as headache, photophobia, phonophobia, or aura. Vestibular migraine is the currently accepted terminology that replaces past descriptors such as migraine-associated vertigo. Patients may return to normal in between attacks of vestibular migraine. Onset is not classically triggered by specific events such as travel or an illness. Vestibular migraine might respond to some of the more common medications used to treat migraine conditions,<sup>7,8</sup> but MdDS usually will not. Nausea is much more prominent in vestibular migraine.<sup>9</sup> Testing is not particularly helpful in differentiating between the 2, as most often testing is normal in both conditions.

**Chronic subjective dizziness** This is a term applied to conditions such as panic disorder or anxiety-related dizziness. The dizziness is sometimes described as rocking, just as with MdDS. However, it also could be more of a floating or internal spinning sensation.<sup>7,10</sup> It is either constant or occurs in attacks, and dizziness is only one of a myriad of symptoms that might also include somatic complaints such as weakness, fatigue, gastric distress, and heart palpitations. The symptoms often begin after a period of stress such as a prolonged illness or a death in the family, and they may last months to years. In contrast, MdDS occurs after passive motion exposure. Anxiety is common in both patients with chronic subjective dizziness and those with MdDS, which could make it more likely to get the 2 mixed up.

**Motion sickness** This results from vestibular or visual stimulation occurring with motion, and symptoms of dizziness, nausea, vomiting, and sweating are common. Symptoms begin during the time of actual motion; this distinguishes it from MdDS, which begins after passive motion has ended. Medications such as central-acting anticholinergics and H1 antihistamines are most effective in treating motion sickness,<sup>11</sup> but they will not effectively treat MdDS.



*MdDS might result from visual-vestibular conflict as the person is in motion while the visual system sends signals indicating a stationary environment.*

**Otologic causes** These are easily differentiated from MdDS because they rarely cause the unending rocking sensation that is so typical of MdDS. Meniere disease is distinguished by the presence of unilateral muffled and fluctuating hearing with tinnitus, none of which are usually seen in MdDS since hearing is expected to be normal. Benign paroxysmal positional vertigo is also easily distinguishable because it is associated with characteristic nystagmus and responds to canalith repositioning, which has no effect on patients with MdDS. Vestibular neuritis is an inflammatory condition that presents clinically with a single spontaneous attack of vertigo that is usually spinning and accompanied by nausea and nystagmus, all features that are unlikely to be seen in MdDS. VNG testing in vestibular neuritis will show unilateral hypofunction but is likely to be normal in MdDS. Conditions such as superior canal dehiscence (SCD) and perilymph fistula can cause episodes of vertigo induced by cough or noise causing intracranial or middle ear pressure changes. They can result in a “third window phenomenon” due to labyrinth defects sometimes caused by trauma or cholesteatoma.<sup>12</sup> Hearing tests are usually abnormal in SCD and perilymph fistula, in contrast to MdDS. A history of airplane travel triggering symptoms might lead one to consider either perilymph fistula or MdDS, but classic perilymph fistula presents with a sudden “popping” noise in the ear and is soon followed by hearing loss, vertigo, and tinnitus.<sup>12</sup> Again, MdDS could begin following a history of air travel but it would not present with hearing changes. Finally, a condition called idiopathic bilateral vestibular loss can result in dizziness and gait unsteadiness that is worse in darkness. Patients with this condition can be differentiated from patients with MdDS because they also commonly report “bouncing vision” when walking (oscillopsia) and they have reduced vestibular responses with head impulse testing and quantitative vestibular testing.<sup>13,14</sup>

### Pathophysiology

The pathogenesis of MdDS is not well-understood. A number of theories have been proposed. One possibility is that MdDS is a disorder of neuroplasticity and more specifically a disorder of vestibular adaptation.<sup>1</sup> When on a boat, the movements consist of roll in combination with pitch and yaw plane movements.<sup>17</sup> The natural adaptation to this environment is mediated by the vestibulo-ocular reflex (VOR) adaptation processes, possibly including velocity storage mechanisms.<sup>18,19</sup> When returning to land, “sea legs” is a common phenomenon.<sup>20</sup> A small number of susceptible individuals may not be able to “turn off” or revert to the prior state of vestibular adaptation and have a persisting sensation of rocking that is the hallmark of persistent MdDS.<sup>6</sup> If vestibular adaptation and velocity storage are involved, it would imply that individuals with impaired vestibular function (and thus short VOR time constant values) would not be susceptible; indeed, most patients with MdDS have normal vestibular function.<sup>6</sup>

It is also possible that there is a genetic predisposition to MdDS as with motion sickness.<sup>11,21</sup> Patients with MdDS and motion sickness both utilize the somatosensory system more than the vestibular system to maintain balance, so, similar to the theory of motion sickness, MdDS might result from visual-vestibular conflict as the person is in motion while the visual system sends signals indicating a stationary environment.

### Treatment

There are no effective treatments for MdDS. The resolution of MdDS is spontaneous, so it often remits without any specific treatment. Most medications for dizziness or motion sickness

such as meclizine, dimenhydrinate, and scopolamine are not useful in MdDS. Anxiolytics may temporarily improve symptoms, with benzodiazepines likely to provide the most benefit.<sup>4</sup> Clonazepam is often preferred due to its longer half-life. Some physicians have found benefit with the use of selective serotonin reuptake inhibitors as baseline treatment and use benzodiazepines only as needed.<sup>1</sup> In a recent study, Cha et al. explored the use of repetitive transcranial magnetic stimulation for MdDS and found overall good tolerability and possible short-term improvement in rocking sensation reported by participants using a visual analog scale. Further trials with longer treatment course are needed to support this finding.<sup>22</sup> Vestibular rehabilitation has been suggested by some<sup>6</sup> as potentially helpful, yet other studies report no significant response to therapy or exercise.<sup>1,4</sup> To date, no randomized controlled clinical trials of potential treatments for MdDS have been performed.

## DISCUSSION

This disorder is largely unrecognized by most physicians treating patients who present with the symptoms of MdDS, mainly because most clinicians are unaware of the syndrome. Therefore, increased awareness of MdDS in the medical community is necessary. Macke et al. recently published a study examining the social, societal, and economic burden of MdDS and found that the average cost related to obtaining a diagnosis was about \$3,000 per patient. They noted that patients with MdDS report poor overall quality of life and visit no less than 19 physicians on average before receiving a diagnosis.<sup>23</sup> Currently, most patients with MdDS are diagnosed after consultation with one of a small number of specialists in our country that are familiar with the disorder. This often does not take place until months after the patient's onset of symptoms and numerous referrals have been made. With increased awareness of MdDS, patients could be diagnosed at an earlier stage and could bypass extensive medical testing and multiple physician referrals. Earlier diagnosis would also help reduce the anxiety these patients experience as a result of not knowing the cause of their unrelenting symptoms. In terms of treatment, there is no first-line treatment recommendation for MdDS. New potential treatments such as repetitive transcranial magnetic stimulation, certain vestibular therapy exercises, or pharmacotherapy may prove useful, but more trials are needed before any strong conclusions can be made.

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