Vestibular migraine: Diagnostic criteria (Update)[1]

Literature update 2021

Consensus document of the Bárány Society and the International Headache Society

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Abstract. This paper presents diagnostic criteria for vestibular migraine, jointly formulated by the Committee for Classification of Vestibular Disorders of the Bárány Society and the Migraine Classification Subcommittee of the International Headache Society (IHS). It contains a literature update while the original criteria from 2012 were left unchanged. The classification defines vestibular migraine and probable vestibular migraine. Vestibular migraine was included in the appendix of the third edition of the International Classification of Headache Disorders (ICHD-3, 2013 and 2018) as a first step for new entities, in accordance with the usual IHS procedures. Probable vestibular migraine may be included in a later version of the ICHD, when further evidence has accumulated. The diagnosis of vestibular migraine is based on recurrent vestibular symptoms, a history of migraine, a temporal association between vestibular symptoms and migraine symptoms and exclusion of other causes of vestibular symptoms. Symptoms that qualify for a diagnosis of vestibular migraine include various types of vertigo as well as head motion-induced dizziness with nausea. Symptoms must be of moderate or severe intensity. Duration of acute episodes is limited to a window of between 5 minutes and 72 hours.

Keywords: Migraine, vertigo, dizziness, vestibular, diagnostic criteria, Bárány Society, International Headache Society

1. Preface to the update

The current consensus criteria for the diagnosis of vestibular migraine were first published in 2012 by the International Bárány Society [1]. In 2013 they were introduced in the International Classification of Headache Disorders (ICHD-3 beta version 2013 [2] and ICHD-3 2018 [3]) where they appear in the appendix for new syndromes. This literature update leaves the diagnostic criteria unchanged and adds recent developments and findings, particularly in the *Comments* section. Probably the most relevant progress is the separate publication of diagnostic criteria for childhood vertigo syndromes related to migraine [4].

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[1]This is a revision of the 2012 paper.

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2. Introduction

The link between migraine and vertigo was recognized by some of the early neurologists in the 19th century [5], but systematic studies of vertigo caused by migraine started only a hundred years later [6, 7]. In the past decades vestibular migraine has taken shape as a diagnostic entity that may affect up to 2.7% of the general population [8]. Despite this recognition, the lack of a universally accepted definition of vestibular migraine has hampered patient identification in clinical and research settings. In response, the Bárány Society, which represents the international community of basic scientists, otolaryngologists and neurologists committed to vestibular research, mandated a classification group to develop diagnostic criteria for vestibular migraine. The definition of vestibular migraine is part of a larger endeavor for classification of neuro-otological disorders steered by the Committee for Classification of Vestibular Disorders of the Bárány Society. Individual disorders are defined by classification groups which include otolaryngologists and neurologists from at least three continents [9]. The format of the classification is modeled on the International Classification of Headache Disorders. As a first step and prerequisite for the classification of vestibular disorders, the Classification Committee of the Bárány Society published a consensus on the definitions of vestibular symptoms [10].

A principal aim of the definition of vestibular migraine is its broad acceptance within both the vestibular and headache community. Therefore, drafts of this classification were discussed with the Migraine Classification Subcommittee of the Classification Committee of the International Headache Society (IHS). Controversies centered on the balancing of the sensitivity and specificity of the criteria. This conflict is inherent to all medical classification efforts, because highly specific criteria will inevitably exclude patients affected by the condition (false negatives), while highly sensitive criteria will include patients who do not have the condition (false positives). As a result of these discussions, a proposed category, *possible vestibular migraine,* was eliminated while the categories *vestibular migraine* and *probable vestibular migraine* were retained. Other changes that resulted from discussions between the Bárány Society and the IHS included a narrower definition of the duration of acute episodes and a sharpened focus on the careful exclusion of differential diagnoses. The final approved diagnostic criteria were the product of an accord between the IHS Classification Committee and the Committee for Classification of Vestibular Disorders of the Bárány Society. The ICDH-3 includes only *vestibular migraine* in the appendix for research purposes, while the Bárány classification also contains the category *probable vestibular migraine*.

3. Diagnostic criteria for vestibular migraine

Previously used terms: migraine-associated vertigo/dizziness, migraine-related vestibulopathy, migrainous vertigo.

1. Vestibular migraine

A. At least 5 episodes with vestibular symptoms[1] of moderate or severe intensity[2], lasting 5 min to 72 hours[3]

B. Current or previous history of migraine with or without aura according to the International Classification of Headache Disorders (ICHD-3)[4]

C. One or more migraine features with at least 50% of the vestibular episodes[5]:
   - headache with at least two of the following characteristics: one sided location, pulsating quality, moderate or severe pain intensity, aggravation by routine physical activity
   - photophobia and phonophobia[6],
   - visual aura[7]

D. Not better accounted for by another vestibular or ICHD diagnosis[8]

2. Probable vestibular migraine

A. At least 5 episodes with vestibular symptoms[1] of moderate or severe intensity[2], lasting 5 min to 72 hours[3]

B. Only one of the criteria B and C for vestibular migraine is fulfilled (migraine history *or* migraine features during the episode)

C. Not better accounted for by another vestibular or ICHD diagnosis[8]

Notes

1. Vestibular symptoms, as defined by the Bárány Society's Classification of Vestibular Symptoms [10] and qualifying for a diagnosis of vestibular migraine, include:

- spontaneous vertigo including
    - internal vertigo, a false sensation of self-motion, and
    - external vertigo, a false sensation that the visual surround is spinning or flowing,
- positional vertigo, occurring after a change of head position,
- visually-induced vertigo, triggered by a complex or large moving visual stimulus
- head motion-induced vertigo, occurring during head motion,
- head motion-induced dizziness with nausea. Dizziness is characterized by a sensation of disturbed spatial orientation. Other forms of dizziness are currently not included in the classification of vestibular migraine.

2. Vestibular symptoms are rated "moderate" when they interfere with but do not prohibit daily activities and "severe" if daily activities cannot be continued.
3. Duration of episodes is highly variable: About 30% of patients have episodes lasting minutes, 30% have attacks for hours and another 30% have attacks over several days. The remaining 10% have attacks lasting seconds only, which tend to occur repeatedly during head motion, visual stimulation, or after changes of head position. In these patients, episode duration is defined as the total period during which short attacks recur. At the other end of the spectrum, there are patients who may take four weeks to fully recover from an episode. However, the core episode rarely exceeds 72 hours [11–13].
4. Migraine categories 1.1 and 1.2 of the ICDH-3 [3].
5. One symptom is sufficient during a single episode. Different symptoms may occur during different episodes. Associated symptoms may occur before, during or after the vestibular symptoms.
6. Phonophobia is defined as sound-induced discomfort. It is a transient and bilateral phenomenon that must be differentiated from recruitment, which is often unilateral and persistent. Recruitment leads to an enhanced perception and often distortion of loud sounds in an ear with decreased hearing.
7. Visual auras are characterized by bright scintillating lights or zigzag lines, often with a scotoma that interferes with reading. Visual auras typically expand over 5–20 minutes and last for less than 60 minutes. They are often, but not always restricted to one hemifield. Other types of migraine aura, e.g. somatosensory or dysphasic aura, are not included as diagnostic criteria because their phenomenology is less specific and most patients also have visual auras.
8. History and physical examinations do not suggest another vestibular disorder *or* such a disorder is considered but ruled out by appropriate investigations *or* such disorder is present as a comorbid or independent condition, but episodes can be clearly differentiated. Migraine attacks may be induced by vestibular stimulation [14]. Therefore, the differential diagnosis should include other vestibular disorders complicated by superimposed migraine attacks. See *Comments* for discussion of specific differential diagnoses.

4. Comments

4.1. Epidemiology

Vestibular migraine is one of the most common vestibular disorders affecting between 1% and 2.7% of the general population [8, 15], 11% of patients in specialized dizziness clinics [11] and 13% of patients in headache clinics [16]. About 65% to 85% of the patients are female [8, 12]. Familial occurrence of vestibular migraine with autosomal dominant inheritance has been documented in several families [17].

4.2. Relation to migraine aura and migraine with brainstem aura (formerly termed basilar-type migraine)

Both migraine aura and migraine with brainstem aura are terms defined by the ICHD-3 [3]. Only a minority of patients with vestibular migraine experience their vertigo in the time frame of 5–60 minutes as defined for an aura. Even fewer have their vertigo immediately before headache starts, as would be required for the ICHD-3 category *typical aura with headache* [3]. Therefore, episodes of vestibular migraine cannot be regarded as migraine auras.

Although more than 60% of *patients suffering from migraine with brainstem aura (formerly termed basilar-type migraine)* have vertigo, the ICHD-3 requires at least two reversible brainstem symptoms each lasting between 5 and 60 minutes for a diagnosis of *migraine with brainstem aura* [3]. Less than 10% of patients with vestibular migraine fulfill these crite-

ria [11]. Thus, *vestibular migraine* and *migraine with brainstem aura* are not synonymous, although individual patients may meet the diagnostic criteria for both conditions. Further studies are needed to define overlap and delineation of the two conditions.

4.3. Relation to benign paroxysmal vertigo of childhood/vestibular migraine of childhood

The ICHD-3 defines *benign paroxysmal vertigo (of childhood)* as an episodic syndrome that may be associated with migraine. The diagnosis requires five episodes of severe vertigo, occurring without warning and resolving spontaneously after minutes to hours. In between episodes, neurological examination, audiometry, vestibular functions and EEG must be normal [3]. Since the introduction of the ICHD-3 in 2018 the recurrent childhood vertigo syndromes have been jointly redefined by the Bárány Society and the International Headache Society. The proposed classification distinguishes among three presentations based on their relation to migraine: vestibular migraine of childhood, probable vestibular migraine of childhood and recurrent vertigo of childhood [4]. The delineation of these syndromes aligns terminology for children and adults and will promote both research and clinical diagnosis of pediatric vertigo syndromes.

4.4. Other symptoms

The current classification restricts the diagnosis of vestibular migraine to patients with vertigo and accepts patients with dizziness only when they have head-motion induced dizziness with nausea. This delineation aims at the specificity of the criteria by exclusion of non-vestibular complaints such as orthostatic dizziness or panic symptoms. On the other hand, there are numerous patients who fulfill all criteria of vestibular migraine but complain of dizziness rather than vertigo. Particularly for research purposes the application of the strict criteria is encouraged to ensure a clear-cut case definition. A future version of the classification may redefine the required symptoms for vestibular migraine.

Transient auditory symptoms [7], nausea, vomiting, prostration, unsteadiness and susceptibility to motion sickness may be associated with vestibular migraine. However, as they also occur with various other vestibular disorders they are not included as diagnostic criteria.

4.5. Results of vestibular testing

Vestibular migraine is classified entirely on the basis of clinical features as reported by the patient. Just as in migraine itself, there are no biological markers for vestibular migraine. Vestibular findings and testing results can be pathological, particularly during or shortly after an episode [18, 19]. However, acute findings are not readily available in most patients and interictal testing results are not sufficiently specific to serve as diagnostic criteria. Profound abnormalities in the symptom-free interval such as severe hearing loss and complete unilateral or bilateral vestibular loss are usually indicative of another cause.

4.6. Overlap with Menière's disease

Migraine is more common in patients with Menière's disease than in healthy controls [20]. Patients with features of both Menière's disease and vestibular migraine have been repeatedly reported [20, 21]. In fact, migraine and Menière's disease can be inherited as a symptom cluster [22]. Fluctuating hearing loss, tinnitus and aural pressure may occur in vestibular migraine, but hearing loss does not progress to profound levels [23]. Moreover, chronic hearing loss in VM is usually bilateral and downsloping as compared to unilateral or asymmetric and flat in chronic Menière's disease [23]. Similarly, migraine headaches, photophobia and even migraine auras are common during Menière attacks [20, 24]. Occasional patients with vestibular migraine have endolymphatic hydrops on MRI [25]. However, the pathophysiological relationship between vestibular migraine and Menière's disease remains uncertain [26]. In the first year after onset of symptoms, differentiation of vestibular migraine from Menière's disease may be challenging, as Menière's disease can be monosymptomatic with vestibular symptoms only in the early stages of the disease.

When the criteria for Menière's disease [27] are met, particularly unilateral hearing loss as documented by audiometry, Menière's disease should be diagnosed, even if migraine symptoms occur during the vestibular attacks. Only patients who have two different types of attacks, one fulfilling the criteria for vestibular migraine and the other for Menière's disease, should be diagnosed with the two disorders. A future revision of this classification may include a vestibular migraine/Menière's disease overlap syndrome [21].

4.7. Other differential diagnoses

Benign paroxysmal positional vertigo (BPPV). Vestibular migraine may present with purely positional vertigo, thus mimicking BPPV. Direct nystagmus observation during the acute phase may be required for differentiation. In vestibular migraine, positional nystagmus is usually persistent and of constant, mostly low to moderate velocity rarely exceeding 30°/s [19]. Symptomatic episodes tend to be shorter with vestibular migraine (minutes to days rather than weeks) and more frequent (several times per year with vestibular migraine rather than once every few years with BPPV). Typical BPPV is more common in patients with migraine for reasons that are unclear.

Transient ischemic attacks (TIAs). A differential diagnosis of vertebrobasilar TIAs must be considered particularly in elderly patients. Suggestive features include vascular risk factors, sudden onset of symptoms, total history of attacks of less than one year, and angiographic or Doppler ultrasound evidence for vascular pathology in the vertebral or proximal basilar artery. Sudden neck pain may point to vertebral artery dissection.

Vestibular paroxysmia. Vestibular paroxysmia presents with brief attacks of vertigo, lasting from one to several seconds, which recur many times per day. Successful prevention of attacks with carbamazepine supports the diagnosis [28].

Psychiatric dizziness. Anxiety and depression may cause dizziness and likewise complicate a vestibular disorder. Anxiety-related dizziness is characterized by situational provocation, intense autonomic activation, catastrophic thinking and avoidance behaviour. More than 50% of patients with vestibular migraine have comorbid psychiatric disorders [29].

4.8. Migraine induced by vestibular activation

Caloric stimulation often triggers migraine attacks within 24 hours in patients with migraine [14], which shows that migraine attacks can be a secondary effect of vestibular activation rather than its cause in susceptible individuals. Possibly, the high rate of headaches and other migraine symptoms during Menière attacks can be explained by this mechanism. Thus, migraine symptoms during vertigo attacks do not prove a diagnosis of vestibular migraine and consideration of other potential causes remains mandatory.

4.9. Provoking factors

Provocation of an episode can be a diagnostic clue. Menstruation, stress, lack of sleep, dehydration, and certain foods may all trigger migraine attacks, but are not included as diagnostic criteria for vestibular migraine, because their sensitivity and specificity have not been adequately studied.

4.10. Response to antimigraine medication

A favorable response to anti-migraine drugs may support the suspicion of an underlying migraine mechanism. However, the apparent efficacy of a drug may be influenced by confounding factors including spontaneous improvement, placebo response, and multiple drug effects (e.g. anxiolytic or antidepressant). Also, drug responses are useful for diagnosis only when the sensitivity and specificity of the criterion is high. So far, the evidence for treating vestibular migraine with anti-migraine drugs is insufficient as it is mostly based on observational studies rather than randomized controlled trials [30]. Consequently, a positive drug response is not regarded as a reliable criterion for the diagnosis of vestibular migraine.

4.11. Chronic vestibular migraine

In this classification, vestibular migraine is conceptualized as an episodic disorder. However, a chronic variant of vestibular migraine has been reported [31]. Between attacks, many patients experience some degree of visually-induced, head motion-induced or persistent dizziness [29]. A distinction between chronic vestibular migraine, motion sickness and comorbid persistent postural-perceptual dizziness seems particularly challenging in these patients [32–34]. In the future, following additional research, chronic vestibular migraine may become a formally recognized category of a revised classification.

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