

# Vestibular migraine and recurrent vertigo of childhood: Diagnostic criteria consensus document of the Classification Committee of Vestibular Disorders of the Bárány Society and the International Headache Society

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Received 3 June 2020

Accepted 25 November 2020

**Abstract.** This paper describes the diagnostic criteria for “Vestibular Migraine of Childhood”, “probable Vestibular Migraine of Childhood” and “Recurrent Vertigo of Childhood” as put forth by the Committee for the Classification of Vestibular Disorders of the Bárány Society (ICVD) and the Migraine Classification subgroup of the International Headache Society. Migraine plays an important role in some subgroups of children with recurrent vertigo. In this classification paper a spectrum of three disorders is described in which the migraine component varies from definite to possibly absent. These three disorders

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are: Vestibular Migraine of Childhood, probable Vestibular Migraine of Childhood and Recurrent Vertigo of Childhood. The criteria for *Vestibular Migraine of Childhood* (VMC) include (A) at least five episodes with vestibular symptoms of moderate or severe intensity, lasting between five minutes and 72 hours, (B) a current or past history of migraine with or without aura, and (C) at least half of episodes are associated with at least one migraine feature. *Probable Vestibular Migraine of Childhood* (probable VMC) is considered when at least three episodes with vestibular symptoms of moderate or severe intensity, lasting between five minutes and 72 hours, are accompanied by at least criterion B or C from the VMC criteria. *Recurrent Vertigo of Childhood* (RVC) is diagnosed in case of at least three episodes with vestibular symptoms of moderate or severe intensity, lasting between 1 minute and 72 hours, and none of the criteria B and C for VMC are applicable. For all disorders, the age of the individual needs to be below 18 years old. It is recommended that future research should particularly focus on RVC, in order to investigate and identify possible subtypes and its links or its absence thereof with migraine.

Keywords: Vestibular, children, childhood, vertigo, Benign Paroxysmal Vertigo of Childhood, dizziness, imbalance, vertigo attacks, migraine, Bárány Society

## 1. Introduction

The most frequent conditions believed to cause vertigo and dizziness during childhood are currently classified as “Benign Paroxysmal Vertigo of Childhood” and Vestibular Migraine (VM). “Benign Paroxysmal Vertigo of Childhood” is assumed to have a prevalence of around 3% in children up to 18 years of age [2, 4, 18]. It was described as follows: recurrent spontaneous attacks of vertigo which may be associated with vomiting, pallor, fearfulness, postural imbalance, ataxia, and/or nystagmus in otherwise healthy children. In the original description by Basser, the condition begins before the age of 4 and resolves spontaneously by age 8 to 10 [5]. Its onset may be at younger ages but is often not recognized until children are old enough to properly describe their symptoms. The current criteria for Benign Paroxysmal Vertigo as presented in the International Classification of Headache Disorders does not include strict age limits and states that the condition is not limited to childhood [19].

The established diagnostic criteria of VM require at least five episodes with vestibular symptoms of moderate or severe intensity, lasting between five minutes and 72 hours, a life time diagnosis of migraine headaches and migraine symptoms during most of the attacks [19]. These criteria can be applied to children as well, but have not yet been validated for them. The exact prevalence of VM in the pediatric population is so far unknown [13].

It is likely that a substantial proportion of pediatric patients with episodic vertigo fit both “Benign Paroxysmal Vertigo of Childhood” criteria as well as VM criteria, and past research has shown different likelihoods of children with “Benign Paroxysmal Vertigo of Childhood” developing migraine later in

life [6, 11, 31]. Consequently, it is debatable whether “Benign Paroxysmal Vertigo of Childhood” and VM in children are distinct entities or part of the same spectrum.

Guidelines for diagnostic testing and treatment have been neither established for “Benign Paroxysmal Vertigo of Childhood” nor so far for VM in children. Furthermore, the battery of diagnostic tests that children with suspected “Benign Paroxysmal Vertigo of Childhood” and/or VM undergo, varies widely between centres. It typically includes neurological/neuro-ophthalmological examinations, vestibular examinations, ENT/audiological evaluation, and/or Magnetic Resonance Imaging studies. These tests mainly serve to ensure that the symptoms are “not attributable to another disorder”. Treatment recommendations for children are sparse as placebo controlled studies are lacking [1, 20, 22]. As the boundaries and overlaps of Benign Paroxysmal Vertigo of Childhood and VM in children has become clearer, future studies may clarify which children would benefit from targeted treatments.

This consensus document aims to classify “Vestibular Migraine of Childhood” (VMC) and “probable Vestibular Migraine of Childhood” (probable VMC) as well as to introduce a new term and classification of recurrent vertigo in children, named “Recurrent Vertigo of Childhood” (RVC), which should replace the term “Benign Paroxysmal Vertigo of Childhood”.

Additional goals are to facilitate research on 1) the relationship between VMC, probable VMC and RVC, and 2) RVC itself. This could help in defining possible subgroups (including their etiologies) and pave the way for new recommendations on diagnostic criteria, guidelines for diagnostic testing and treatment protocols in this population. It could also improve

personalized counseling and care to both parents and children.

## 2. Methods

The work presented here is part of an ongoing project to develop an International Classification of Vestibular Disorders (ICVD). The ICVD uses a structured process to develop consensus diagnostic criteria for vestibular symptoms and disorders. The process of establishing criteria is overseen by the Classification Committee of the Bárány Society. For each diagnostic category, an international team of content experts from multiple disciplines is established to propose initial criteria based on the best available scientific evidence. For the classification of “Vestibular Migraine of Childhood”, “probable Vestibular Migraine of Childhood” and “Recurrent Vertigo of Childhood”, one member of the Headache Classification Committee of the International Headache Society has been delegated to this team in order to add the expertise from the view of headache experts.

The initial criteria were proposed and circulated to the subcommittee members in 2019 and a first draft was presented to the Committee for the Classification of Vestibular Disorders of the Bárány Society. Comments of (sub)committee members were gathered and synthesized. Modified criteria were presented in 2020 to the Committee for the Classification of Vestibular Disorders of the Bárány Society for tentative approval. The definitions presented here are supported by a process of discussion and refinement as established by the classification committee for the ICVD. The criteria presented below have been carefully considered to account for broad applicability to the international community of pediatricians, otolaryngologists, neurologists, neurosurgeons, neuro-otologists, physiotherapists, neurophysiologists, and audiologists who may be seeing patients with these syndromes.

## 3. Diagnostic criteria

### 3.1. Vestibular Migraine of Childhood (VMC)

- A. At least five episodes with vestibular symptoms of moderate or severe intensity<sup>1,2</sup>, lasting between five minutes and 72 hours
- B. A current or past history of migraine with or without aura

- C. At least half of episodes are associated with at least one of the following three migraine features:
  1. Headache with at least two of the following four characteristics:
    - a) One sided location
    - b) Pulsating quality
    - c) Moderate or severe pain intensity
    - d) Aggravation by routine physical activity
  2. Photophobia and phonophobia
  3. Visual aura
- D. Age < 18 years
- E. Not better accounted for by another headache disorder, vestibular disorder, or other condition

### 3.2. Probable Vestibular Migraine of Childhood (probable VMC)

- A. At least three episodes with vestibular symptoms of moderate or severe intensity, lasting between five minutes and 72 hours
- B. Only one of the criteria B and C for Vestibular Migraine of Childhood
- C. Age < 18 years
- D. Not better accounted for by another headache disorder, vestibular disorder, or other condition

### 3.3. Recurrent Vertigo of Childhood (RVC)

- A. At least three episodes with vestibular symptoms of moderate or severe intensity, lasting between 1 minute and 72 hours
- B. None of the criteria B and C for Vestibular Migraine of Childhood
- C. Age < 18 years
- D. Not better accounted for by another headache disorder, vestibular disorder, or other condition

## 4. Notes

1. Children are, even more so than adults, not always able to precisely describe their vestibular symptoms. Reported vertigo, dizziness, or parental observation of recurrent attacks presenting with unsteadiness may all be present in children [19]. The vestibular symptoms occur spontaneously and without loss of consciousness. They may include nausea, vomiting, and/or pallor.
2. Vestibular symptoms qualifying for VM were previously defined by the ICVD together with the International Classification of Headache Disorders. *Vestibular symptoms* include spontaneous

vertigo (internal vertigo (false sensation of self-motion) and external vertigo (false sensation of spinning or flowing of the visual surround)), positional vertigo (after change of head position), visually induced vertigo (triggered by visual stimuli), head motion-induced vertigo, and head motion-induced dizziness with nausea (dizziness defined as a sensation of disturbed spatial orientation). Vestibular symptoms of *moderate intensity* interfere with, but do not prevent daily activities. Vestibular symptoms of *severe intensity* prevent continuation of daily activities [9, 19, 24].

## 5. Comments

### 5.1. Vestibular migraine in the pediatric population

Vestibular migraine is nowadays a well-known entity that can occur at any age. Although the diagnostic criteria for vestibular migraine do not include any age limit [24], the subcommittee decided that separate criteria for Vestibular Migraine of Childhood would be beneficial for two main reasons. First, it facilitates structured description of three disorders in which the migraine component varies from definite to probable to possibly absent: Vestibular Migraine of Childhood, probable Vestibular Migraine of Childhood and Recurrent Vertigo of Childhood. This captures most likely the spectrum of recurrent vertigo in the pediatric population, which is not caused by any other recognized disorder (see “Differential diagnoses of recurrent vertigo in the pediatric population”). Secondly, it allows to redefine criteria, tailored to the pediatric population. The minimum number of episodes for probable VMC was reduced to three as compared to probable vestibular migraine in adults which requires five episodes, as consensus in the subcommittee agreed that children and parents usually seek medical help after a more limited number of attacks.

Regarding migraine headache characteristics, children and adolescents (aged under 18 years) more often present with bilateral migraine headache than adults. Usually, in late adolescence or early adult life, unilateral pain emerges [19]. Although unilateral pain only occurs in a subset of children with migraine [14], the subcommittee decided to keep this characteristic as part of the criteria for (probable) Vestibular Migraine of Childhood. After all, the sensitivity and specificity of this characteristic is still undetermined for these entities and taking it out of the criteria,

would probably only lead to loss of sensitivity, without increasing specificity of the criteria.

### 5.2. Terminology: Vestibular Migraine of Childhood and Recurrent Vertigo of Childhood instead of Benign Paroxysmal Vertigo of Childhood

It was decided to change terminology from “Benign Paroxysmal Vertigo of Childhood (BPVC)” to Vestibular Migraine of Childhood (VMC), probable Vestibular Migraine of Childhood (probable VMC) and Recurrent Vertigo of Childhood (RVC), for multiple reasons:

- In the International Classification of Vestibular Disorders the term “paroxysmal” is reserved for vestibular spells of short duration of less than one minute, which is at odds with the use in BPVC.
- In the definition of BPVC no accompanying migraine features are required, but it is inferred to be a migraine precursor. Follow-up studies have shown that a variable proportion, but by far not all children, were developing migraine later in life [6, 20, 25]. Since many children with episodic vertigo also present with migraine features, the currently presented criteria make a distinction between populations with and without prominent migraine features, using a spectrum varying from VMC, to probable VMC and RVC. This allows to conduct research with better defined groups (VMC, probable VMC and RVC) and should promote the identification of possible subgroups and their evolution in time (e.g. probable VMC might evolve into VMC).
- Several different clinical features for “Benign Paroxysmal Vertigo of Childhood” have been proposed in the past [5, 11, 20, 43]. This has led to heterogeneity and might therefore render meta-analyses more difficult, when attempting to pool study results.

### 5.3. Does Recurrent Vertigo of Childhood represent one or several entities? Aims for future research

The subcommittee members are aware that RVC is not a homogeneous new entity, but a mixture of different / emerging entities, of which the available data do not yet allow more precise definitions. Some children presenting with pure episodic vertigo (and possibly non-specific features such as nausea, vomiting,

imbalance, pallor) and with normal examination, might actually suffer from a migraine precursor. However, this will not be the case for all of them. The clinical observations of members of the subcommittee suggest several lines of thought for subtypes within RVC, that could be more precisely defined in the future: one type that presents with some migraine features but not (yet) enough features to fulfill the criteria for VMC or probable VMC, and one type without any migraine features. The latter seems to predominantly occur in children below the age of ten and presents with brief attacks of vertigo (less than five minutes) and disappears spontaneously after weeks or months. Another subgroup could be linked to weakness of ocular vergence [44]. Since the subcommittee decided that there is currently insufficient evidence to support this clinical experience to propose explicit definitions for these subtypes, it is therefore recommended that future research focusing on RVC explicitly collects data on age of onset and age of end of symptoms, the duration and type of symptoms, all migraine features present during childhood (and possibly later in life), as well as the family history and subtle abnormalities on clinical examination (such as on binocular vision). Only by structured data collection, it will be possible to investigate and identify subtypes within RVC. This could provide the basis for future data driven revisions of these criteria (e.g. including subtypes of RVC).

#### 5.4. Duration of symptoms

In the criteria for RVC the minimum duration of symptoms was reduced from 5 minutes to 1 minute. This shortening of symptom duration is based on clinical observations of members of the subcommittee. The maximum duration was kept at 72 hours, although clinical experience suggests that the duration of symptoms is usually much shorter. At the moment, there are insufficient data in the literature about the duration of symptoms (see above). When future studies provide new insights about symptom duration, this will be integrated into future revisions of the criteria.

#### 5.5. Physical and laboratory examinations

Previous criteria of “Benign Paroxysmal Vertigo of Childhood” included normal neurological examination, audiometry, vestibular function and even EEG. This was not included in the current criteria, since it is known that nystagmus, fluctuating hearing loss and

vestibular hypofunction may also occur in VM [14] as it may obviously in Menière’s disease, yet the type of nystagmus and the hearing loss are important criteria for the differential diagnosis [26]. All children should have a thorough clinical examination of their balance and eye movements, including the head impulse test and vergence testing. If the neurological examination suggests other disorders than migraine in the asymptomatic interval [30], other diagnoses should be considered (see “differential diagnoses”). In case the eye movement testing suggests abnormalities, further neuro-ophthalmological assessment should be carried out to rule out refractive errors and ocular motor pathologies such as insufficient convergence [44].

#### 5.6. Differential diagnoses of recurrent vertigo in the pediatric population

##### 5.6.1. Sensorineural hearing loss due to cochleo-vestibular anomalies

While many children (20–85%) with sensorineural hearing loss have associated vestibular impairment [3, 12, 16], the typical clinical features in these children are disequilibrium and motor developmental delay, not attacks of vertigo. The reason for this is that most children present with congenital profound vestibular impairment (areflexia) and not with sudden, stepwise progressive loss in vestibular function that would lead to a recurrent transitory perception of vertigo. The exceptions to this would include etiologies of hearing loss that are known to affect the vestibular organ and are progressive in nature. Most commonly these would include children with cochleo-vestibular anomalies (i.e. enlarged vestibular aqueduct or incomplete partition type 1 to 3) or children with congenital cytomegalovirus infection [8, 28, 39]. Sudden changes of vestibular function can be associated with episodic attacks of vertigo and disequilibrium. In children who present with concomitant hearing loss, it is necessary to look for anatomic anomalies with imaging, preferably MRI [32], and to test for congenital cytomegalovirus on a specimen obtained within the first 3 weeks of life, if available, in an older child.

##### 5.6.2. Functional and psychiatric causes of dizziness

A major part of the differential diagnosis of VMC, probable VMC, and RVC includes functional and psychiatric causes of vestibular symptoms. Differentiation may be difficult because of overlapping

symptoms and comorbidity. Episodic psychiatric disorders (e.g., panic attacks) and exacerbations of chronic functional and psychiatric disorders (e.g., exacerbations of persistent postural-perceptual dizziness (PPPD) [34] or generalized anxiety disorder, respectively) may cause both vestibular and autonomic symptoms. In patients with migraine features during the attacks, a diagnosis of VMC or probable VMC can be made, even if additional functional or psychiatric features are apparent. These may warrant a separate functional or psychiatric diagnosis [34]. In cases of RVC, it can remain difficult to determine whether the vertigo/dizziness has a vestibular, functional, or psychiatric origin. Careful history taking as well as repeated history taking over time may reveal typical triggers for vertigo of functional or psychiatric origin (e.g. vertigo triggered by undesired events at school, at home, or with other children).

#### 5.6.3. Benign paroxysmal positional vertigo (BPPV)

This can be differentiated from RVC by the trigger (specific head movements) and a positive diagnostic maneuver for BPPV (Dix-Hallpike/diagnostic Semont maneuver or lateral roll) [30]. Triggers can be challenging to decipher as children find themselves in head hanging positions much more often than adults. Therefore, obtaining the details of all episodes, not just the most distinct one, can help to differentiate these two entities. During provocative positional maneuvers, the typical nystagmus in the plane of the tested canal should be present: a vertically upbeat torsional nystagmus with the upper pole of the eye beating towards the lower ear in patients with posterior canal BPPV or a horizontal apogeotropic or geotropic nystagmus in patients with lateral canal BPPV. It should be noted that VM may also present with positional vertigo [24, 40]. The associated nystagmus is more often horizontal than vertical or torsional, and usually persists as long as the offending head position is maintained [45]. The prevalence of BPPV in children is significantly lower than in adults with the biggest risk factors being head trauma, inner ear anomaly or cranial surgery that involves the use of high speed drills [15].

#### 5.6.4. Vestibular paroxysmia

This diagnosis should be considered in children presenting with frequent episodes of vertigo lasting less than one minute [23, 37]. Diagnostic criteria were previously published and include, among others, spontaneous attacks of spinning or non-spinning

vertigo which last less than one minute and response to treatment with a sodium channel blocker [7, 37, 38]. Vestibular paroxysmia can be differentiated from VMC and probable VMC by the absence of migraine features (as mentioned in criterion C of the VMC criteria). Furthermore, duration of attacks can help differentiate vestibular paroxysmia from VMC, probable VMC and RVC: typically less than one minute versus at least five minutes (VMC and probable VMC) and at least one minute (RVC). However, in some patients with vestibular paroxysmia the attacks or some of the attacks may last up to a few minutes [37]. In these cases, response to treatment might help to further differentiate between vestibular paroxysmia and RVC. Response to treatment can be expected in days to weeks, depending on the frequency of attacks [23].

#### 5.6.5. Menière's disease

This also occurs in children [10, 42]. Fluctuating hearing loss, tinnitus and aural pressure can also be symptoms of VM (and possibly RVC). However, the sensorineural hearing loss typically does not progress to severe hearing loss in VM. In VM, it is often bilateral and more pronounced in the high rather than the low frequencies. In patients with two different types of attacks (e.g. Menière's disease and VMC), both diagnoses can be made. Some patients with Menière's disease have migraine features during their attacks, which might imply an overlap syndrome between Meniere's disease and VMC. In these cases, patients may fulfill the diagnostic criteria for both [1, 29]. This needs further investigation, before it will be considered in a future revision of these criteria.

#### 5.6.6. Vestibular epilepsy/epileptic vertigo and nystagmus

This is a very rare cause of vertigo. It is usually caused by lesions around the temporo-parietal junction/insular cortex. It can be accompanied by spontaneous nystagmus and is usually associated with other epileptic phenomena, typical of partial or complex partial seizures such as aura, smacking or impaired consciousness.

#### 5.6.7. Episodic ataxias

So far eight subtypes have been described. The most frequent subtype is episodic ataxia type 2 (EA2), an autosomal dominant disorder, in which attacks of ataxia (stance, gait, limb), dizziness, and vertigo occur that last from many minutes up to days.

It often manifests in early childhood. EA2 is an important differential diagnosis of VMC. Possible accompanying symptoms are headache, oscillopsia, diplopia, dysarthria or muscle weakness. Nystagmus, most often downbeat and gaze-evoked, and other central cerebellar ocular motor signs, such as saccadic smooth pursuit and impaired fixation suppression, are present in more than 90% of patients between the attacks and are more pronounced during the attacks [36]. Therefore, if patients have these signs and symptoms, the diagnosis of EA2 is more likely than VMC or RVC and genetic testing is justified. However, in the early course of EA2, interictal nystagmus and ocular motor abnormalities may not be prominent. Over years, it is a slowly progressive disorder.

#### 5.6.8. Tumors and other lesions in the posterior fossa

They may rarely lead to attacks of vertigo and/or unsteadiness in the pediatric population. An expanding tumor typically presents with persistent and progressive symptoms such as ataxia, dysarthria, double vision, hearing loss and other cranial nerve dysfunction. MRI is indicated in such cases.

#### 5.6.9. Hemodynamic orthostatic dizziness/vertigo

This should be considered when the following criteria are met: 1) Five or more episodes of dizziness, unsteadiness or vertigo triggered by arising (i.e. a change of body posture from lying to sitting/standing or sitting to standing), or present during upright position, which subsides by sitting or lying down; 2) Orthostatic hypotension, Postural tachycardia syndrome or syncope documented on standing or during head-up tilt test; 3) Not better accounted for by another disease or disorder [21]. For “probable” hemodynamic orthostatic dizziness/vertigo, the criteria are less strict. *Orthostatic hypotension* is defined as a sustained reduction of systolic blood pressure of at least 20 mmHg or diastolic blood pressure of 10 mmHg within 3 minutes of standing or during head-up tilt test. *Initial orthostatic hypotension* is characterized by a transient blood pressure decrease (systolic >40 mmHg or diastolic >20 mmHg) within 15 seconds of standing. When a person stands, blood pools in the abdomen and legs. Normally, the autonomic nervous system will compensate by constricting blood vessels and pushing the blood to the brain. When autonomic pathways are affected, these reflexes do not function adequately. As a result,

the person becomes symptomatic. *Postural tachycardia syndrome* can be considered in case of a sustained heart rate increase of at least 30 beats per minute, or a heart rate of 120 beats per minute or more, within 10 minutes of standing or during head-up tilt test in the absence of orthostatic hypotension. For younger individuals (aged 12–19 years), a minimum increase of 40 beats per minute is required for the diagnosis. Approximately one third of teenagers will experience symptoms of lightheadedness with standing [33]. It is a common problem in otherwise healthy teenagers, particularly after a growth spurt, but generally improves with time. It occurs more frequently in patients with a history of anxiety, or of chronic pain [35]. Hemodynamic orthostatic dizziness/vertigo can be differentiated from VMC, probable VMC and RVC by their trigger: standing up.

#### 5.6.10. Recurrent episodes of serous otitis media

They may also lead to recurrent periods of unsteadiness in children by involvement of the inner ear, although the evidence for the association between serous otitis media and vestibular impairment remains debated [27]. Vestibular symptoms due to serous otitis media can be differentiated from VMC and RVC. The periods of unsteadiness are related to the presence of serous otitis media, and therefore often last longer than 72 hours. The following signs can be found with serous otitis media on the affected ear(s): a history of hearing loss, fluid behind the tympanic membrane with otoscopy, negative Rinne tuning fork test, type B or C<sub>2</sub> curve (curves indicating otitis media or negative pressure in the middle ear respectively) with tympanometry, and a conductive hearing loss on the audiogram. From clinical experience, the unsteadiness can disappear when placing ventilation tubes or when the serous otitis media has resolved due to its natural course or appropriate treatment (although evidence from controlled studies is lacking). It is therefore advised to perform otoscopy in children complaining of recurrent vertigo.

#### Conflict of interest

The working meetings for the International Classification of Vestibular Disorders (ICVD) are financially supported by the International Bárány Society and by Neuro+e.V., Berlin, Germany, a non-profit association for neurological research.

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