

Vestibular paroxysmia: diagnostic criteria

Michael Strupp¹, Jose A. Lopez-Escamez², Ji-Soo Kim³, Dominik Straumann⁴, Joanna Jen⁵, John Carey⁶, Alexandre Bisdorff⁷, Thomas Brandt¹

¹Dept. of Neurology and German Center for Vertigo and Balance Disorders, Munich, Germany

²Otology and Neurotology Group CTS495, Dept. of Genomic Medicine - Centre for Genomics and Oncology Research - Pfizer/Universidad de Granada/Junta de Andalucía (GENyO), PTS, Granada and Dept. of Otolaryngology, Granada University Hospital Granada, Spain,

³Dept. of Neurology, Seoul National University College of Medicine, Seoul National University Bundang Hospital, South Korea

⁴Dept. of Neurology, University Hospital Zurich, University of Zurich, Switzerland

⁵Dept. of Neurology, University of California, Los Angeles, USA

⁶Dept. of Otorhinolaryngology, Johns Hopkins University School of Medicine, Baltimore, USA,

⁷Dept. of Neurology, Centre Hospitalier Emile Mayrisch, Esch, Luxemburg

Corresponding author:

Michael Strupp, MD, FANA, FEAN Dept. of Neurology and German Center for Vertigo and Balance Disorders, Munich, Germany, University Hospital Munich, Campus Grosshadern, Marchioninistrasse 15, 81377 Munich, Germany

Phone: ++49 89 44007 3678

Fax: ++49 89 44007 6673

E-mail: michael.strupp@med.uni-muenchen.de

Abstract

This paper describes the diagnostic criteria for vestibular paroxysmia as defined by the Classification Committee of the Bárány Society. The diagnosis of *definite* vestibular paroxysmia is mainly based on the patient history and requires: A) at least ten attacks of spinning or non-spinning vertigo; B) duration less than 1 minute; C) spontaneous occurrence; D) stereotyped phenomenology in a particular patient; E) response to a treatment with carbamazepine/oxcarbazepine; and F) not better accounted for by another diagnosis. Probable vestibular paroxysmia is defined as follows: A) at least five attacks of spinning or non-spinning vertigo; B) duration less than 5 minutes; C) spontaneous occurrence or provoked by certain head-movements; D) stereotyped phenomenology in a particular patient; E) not better accounted for by another diagnosis.

Neurovascular cross-compression in the proximal part of the 8th cranial nerve, covered by oligodendrocytes, is the assumed mechanism. Important differential diagnoses are Menière's disease, vestibular migraine, benign paroxysmal positional vertigo, epileptic vestibular aura, paroxysmal brainstem attacks (in multiple sclerosis or after brainstem stroke), superior canal dehiscence syndrome, perilymph fistula, transient ischemic attacks and panic attacks. Current areas of uncertainty in the diagnosis of vestibular paroxysmia are: a) MRI findings of vascular compression which are not diagnostic of the disease or predictive for the affected side because they are also observed in about 30% of healthy asymptomatic subjects; and b) response to treatment with carbamazepine/oxcarbazepine may support the diagnosis but there are so far no randomized controlled trials for treatment of vestibular paroxysmia.

Introduction

The Bárány Society representing the international community of basic scientists, otolaryngologists and neurologists committed to vestibular research mandated a Classification Committee for an International Classification of Vestibular Disorders (ICVD). Individual disorders are defined by subcommittees, which include otolaryngologists and neurologists from at least three continents. Since the beginning of the process, consensus papers on the classification of vestibular symptoms [3], vestibular migraine [18], Menière's disease [20] and benign paroxysmal positional vertigo (BPPV) (von Brevern et al., in press) have been published.

The term “vestibular paroxysmia” was introduced by Brandt and Dieterich in 1994 [4]. Prior to their description, a heterogeneous syndrome of vertigo with symptoms of various durations (from seconds to days), various characteristic features (spinning vertigo, light-headedness or gait instability without vertigo), and varying accompanying symptoms was called “disabling positional vertigo” [13]. The initial criteria of vestibular paroxysmia were based on an observational study on 11 patients, were as follows [4]: 1) short attacks of spinning or non-spinning vertigo lasting from seconds to minutes; 2) attacks frequently dependent on a particular head position; 3) hyperacusis or tinnitus (7 patients) permanently or during the attack; and 4) auditory or vestibular deficits measurable by neurophysiological methods. To make the diagnosis three out of four criteria had to be fulfilled and the patient had to respond to treatment with carbamazepine.

In 2008, based on a study on 32 patients, the suggested diagnostic criteria were modified as follows to include two categories [11]: “*Definite vestibular paroxysmia*”: at least five attacks and the patient also fulfils criteria A-E: A) vertigo spells lasting seconds to minutes. The individual attack is self-limiting and subsides without specific therapeutic intervention; B) one or several of the following provoking factors induce the attacks: 1. occurring at rest; 2. certain head/body positions (not BPPV-specific positioning maneuvers); 3. changes in head/body position (not BPPV-specific positioning maneuvers); C) none or one or several of the following characteristics during the attacks: 1. disturbance of stance; 2. disturbance of gait; 3. unilateral tinnitus; 4. unilateral pressure/numbness in or around the ear; 5. unilaterally reduced hearing; D) one or several of the following additional diagnostic criteria: 1. neurovascular cross-compression demonstrated on MRI (CISS sequence, TOF MR angiography); 2. hyperventilation-induced nystagmus as measured by oculography; 3. increase of vestibular deficit at follow-up investigations as measured by oculography; 4. treatment response to antiepileptics (not applicable at first consultation); E) the symptoms cannot be explained by another disease. The diagnostic criteria for “*Probable vestibular paroxysmia*” were as follows: at least five attacks and the patient fulfils criterion A, and at least three of criteria B-E.

In 2009, an analogous clinical syndrome in two patients with brief, spontaneous, recurrent attacks of tinnitus and vertigo which responded to low-dose gabapentin was described as “audiovestibular paroxysmia” [24].

Methods

This work forms part of an ongoing multi-year project to develop an International Classification of Vestibular Disorders (ICVD) which uses a structured process to develop international consensus definitions for vestibular symptoms, syndromes, disorders, and diseases. This process, overseen by the Classification Committee of the Bárány Society (CCBS), is based on expert, multi-disciplinary committees with international representation developing diagnostic criteria for subsequent comment and refinement prior to publication. These criteria are based on a critical appraisal of current best scientific evidence. All definitions are supported by notes, comments, and written discussion according to a template established by the CCBS for ICVD. The criteria for vestibular paroxysmia were developed iteratively over a three-year period (2013-15) through discussion, presentation, and refinement. Special care was taken that the criteria are practical and can be applied in every country all over the world; this is applies particularly to the use of laboratory examinations not available everywhere.

Diagnostic criteria for vestibular paroxysmia

Definite vestibular paroxysmia (each point needs to be fulfilled)

- A) At least ten attacks¹ of spinning or non-spinning vertigo
- B) Duration less than 1 minute³
- C) Spontaneous occurrence⁴
- D) Stereotyped phenomenology in a particular patient^{5, 6}
- E) Response to a treatment with carbamazepine/oxcarbazepine⁷
- F) Not better accounted for by another diagnosis.

Probable vestibular paroxysmia (each point needs to be fulfilled)

- A) At least five attacks¹ of spinning or non-spinning vertigo²
- B) Duration less than 5 minutes³
- C) Spontaneous occurrence⁴
- D) Stereotyped phenomenology in a particular patient^{5, 6}
- E) Not better accounted for by another diagnosis.

Notes

1. The number of attacks was chosen because vestibular paroxysmia is an episodic vestibular disorder which usually presents with a high frequency of attacks. The frequency of attacks varies greatly between patients: from 30 attacks per day to a few attacks per month. The course of the disease is usually chronic (i.e., longer than three months) with some patients suffering many hundreds attacks per year.

2. The type of vertigo (spinning or non-spinning) or directional pulsion is intraindividually quite uniform. If the attacks occur while standing or walking patients usually experience unsteadiness.

3. Duration: in most patients the attacks last only a fraction of a second to up to one minute; in other patients the duration of the attacks may be longer or increase during the course of the disease. Differential diagnoses with short attacks to consider are Tumarkin's otolith crisis, paroxysmal brainstem attacks, perilymph fistula and, rarely, epilepsy with vestibular aura (for "differential diagnosis" see below). During the course of the disease the duration of attacks may increase, lasting up to many minutes. In patients with longer-lasting attacks, other disorders, in particular vestibular migraine and Menière's disease should be considered. 4. The majority of attacks occur spontaneously ("out of the blue"). In some patients the attacks may be induced by turning the head to the right or left in the upright position. This seems to be similar to the induction of attacks in trigeminal neuralgia due to a sensory input. The triggering head or body movements typically do not have the same pattern as those in BPPV. In some of the patients attacks of vertigo and nystagmus may be provoked by hyperventilation (5). If the attacks are

reproducibly induced by a sustained lateral turn of the head one should consider rotational vertebral artery occlusion syndrome as a differential diagnosis.

5. Some patients may report auditory symptoms, such, unilateral tinnitus or hyperacusis during the attack. Conclusions about the affected ear and the affected nerves can be drawn from the type of complaints - vestibular (originating from the canals or otolith organs) or cochlear symptoms [24]). If there is a combination of symptoms of the 9th and other cranial nerves, the side of the lesion can possibly be deduced. Thus, for example, simultaneously occurring symptoms of the 7th and 8th cranial nerves (with vertigo, tinnitus and hemifacial spasm) [25] indicate an irritation of both cranial nerves in the internal acoustic meatus, where both lie in close proximity to each other.

6. If a patient is examined during an attack, a nystagmus time-locked to the attack and beating toward the affected ear may be observed (personal observation and recording).

7. The majority of patients responds to a treatment with carbamazepine (200 to 800 mg per day) or oxcarbazepine (300 to 900 mg per day) as also already stated in previous studies on the classification of vestibular paroxysmia [4, 11] and others [2, 8, 27]. Although yet not proven in a state-of-the-art randomized controlled clinical trial it strongly supports the diagnosis. This criterion is similar to the treatment responds to indomethacin in paroxysmal hemicrania (International Headache Society Classification ICHD-II, 3.2). If a patient had not already treated it cannot be applied at the first consultation.

Comments

Epidemiology

Since only case series and single cases have been published best 2013 [2, 4, 8, 10, 11, 17, 27] there are so far no data on the life-time prevalence of vestibular paroxysmia. The relative frequency of vestibular paroxysmia in a group of more than 17,000 patients with vertigo and dizziness is around 4% [5]. The mean age of the patients in the three case series with more than 10 patients was 51 yrs (range 25-67 yrs) [4], 48.0 ± 15.3 yrs (mean \pm SD) [11] and 48.4 ± 14.5 (range 25-77 yrs). Vestibular paroxysmia was also described in children with features similar to those in adults [17] and appears to have often a good long term prognosis with spontaneous remission with age [6]. From the three studies mentioned above [5, 11, 17] of a total number of patients affected $n = 63$, 32 were female.

Pathophysiology and etiology

As in trigeminal neuralgia, hemifacial spasm, glossopharyngeal neuralgia or myokymia of the superior oblique muscle [12], it is assumed that a neurovascular cross-compression is the cause of the short attacks of vertigo in vestibular paroxysmia [2, 4, 21, 27]. Arteries in the cerebellopontine angle are thought to be the pathophysiological cause of a segmental pressure-induced lesion with demyelination of the central (oligodendroglia) myelin, proximal to the “transition zone”. This corresponds to the first 15 mm after the nerve exit [15]. Therefore, in a strict anatomical sense, vestibular paroxysmia can be classified as a central vestibular disorder, although physiologically it is peripheral. A loop of the AICA seems to be involved most often, less often the PICA, the vertebral artery, or a vein [2, 11]. It is assumed that the symptoms are triggered by direct pulsatile compression and/or ephaptic discharges, i.e. pathological paroxysmal interaxonal transmission between neighboring and in part demyelinated axons. Finally, in addition to elongation and increased looping, a vascular malformation or arterial ectasia in the posterior fossa can also cause the nerve compression.

Laboratory examinations

Approximately 50% of patients undergoing testing of vestibular and audiological function exhibit signs of a mild to moderate unilateral hypofunction during the attack-free intervals [11]. Hearing loss is usually less prominent than in patients with Menière’s disease. Identification of the affected side is often not possible. If the attacks are accompanied by strictly unilateral audiological symptoms and there are vestibular and audiological deficits on the same side, one can determine the affected ear in exceptional cases [25]. The role of MRI (see below) in identifying the affected side also has to be evaluated further [8, 11].

MR Imaging

In a study on 32 patients with vestibular paroxysmia, neurovascular compression of the 8th cranial nerve was detected in 95% of the patients; bilateral neurovascular compression was found in 42% of the patients [11]. In another study on 20 patients with vestibular paroxysmia, neurovascular compression of the 8th cranial nerve was found in all patients, but also in 7 out of 20 control subjects (sensitivity: 100%, specificity: 65% for the diagnosis of vestibular paroxysmia by MRI) [2]. The distance between the brainstem and compressing vessels varied between 0.0 and 10.2 mm. This part of the nerve is proximally to the transition zone and covered by oligodendrocytes. In 15 cases, the compressing vessel was the anterior inferior cerebellar artery (75%), in one the posterior inferior cerebellar artery (5%), in two a vein (10%), and in another two the vertebral artery. Thus, a high-resolution MRI with CISS/FIESTA sequences of the brainstem can support the diagnosis.

Seven Tesla MRI was performed in six patients with vestibular paroxysmia [23]. It confirmed neuro-vascular cross-compression also seen on 1.5 and 3.0 MRI. No structural abnormalities were detected in any of the patients in 7 Tesla MRI. These findings imply that the symptoms of vestibular paroxysmia are not caused by structural nerve lesions supporting the view that the attacks are caused by excitation of the 8th cranial nerve and being not related to vestibular hypofunction.

In trigeminal neuralgia, high-resolution diffusion tensor imaging revealed significant lower anisotropy and a higher apparent diffusion coefficient in the affected trigeminal root, which correlates with structural atrophic nerve changes [16]. Comparable findings are not yet available for the 8th cranial nerve due to methodological limitations by the short course of the 8th cranial nerve from the brainstem to the internal acoustic meatus and the adjacent temporal bone. A cranial MRI should also be performed to exclude the presence of a tumor in the area of the cerebellar pontine angle, arachnoid cysts [1], megalodolichobasilaris, brainstem plaques in MS (paroxysmal brainstem attacks with or without ataxia) or other brainstem lesions.

Differential diagnosis

The leading symptom of vestibular paroxysmia is *recurrent spontaneous attacks* of vertigo. The diagnosis is generally straightforward because of the characteristic brief duration (seconds up to one minute), the frequently recurring attacks of vertigo and the response to a treatment with carbamazepine or oxcarbazepine. There are only a few other disorders which may present with this leading symptom:

- Menière's disease: duration of the attacks 20 min to 12 hours, low-to medium-frequency sensorineural hearing loss (> 30 dB, < 2000 Hz) [20].
- Tumarkin's otolithic crisis ("vestibular drop attacks"). These sudden falls are usually not accompanied by vertigo and occur most often in patients with known Menière's

disease, typically while standing, whereas in vestibular paroxysmia the attacks occur in any body positions.

- Paroxysmal brainstem attacks with vertigo, dysarthria or ataxia, (after stroke or in multiple sclerosis) may be difficult to distinguish, as they also respond to low doses of carbamazepine. It was shown that they may be caused by a brainstem lesion due to MS plaques or lacunar infarctions [19], which also leads to ephaptic discharges of neighboring fibers of the brainstem paths. In such cases the use of MRI with thin brainstem slices is useful for establishing the diagnosis.
- Vestibular migraine [18]: officially the duration of the attacks is 5 min to 72 hours, current or previous history of migraine, most attacks being accompanied by other migraineous symptoms. In vestibular migraine, short spells of vertigo may be induced by changes of head or body position when patients are motion sensitive during an episode of vestibular migraine.
- Vertebrobasilar transient ischemic attacks: if the leading symptom of these attacks is vertigo they are most often monosymptomatic [22].
- Panic attacks: according to DSM-5, the diagnostic criteria for a panic attack include a discrete period of intense fear or discomfort, in which four (or more) of the following symptoms develop abruptly and reach a peak within minutes: feeling dizzy, unsteady, lightheaded, or faint; nausea or abdominal distress; palpitations, and/or accelerated heart rate; sweating; trembling or shaking; sensations of shortness of breath or being smothered; feeling of choking; chest pain or discomfort; de-realization or depersonalization; fear of losing control or going insane; sense of impending death; paresthesias; chills or hot flashes. Panic attacks are often longer than typical attacks of vestibular paroxysmia. It may be helpful to ask the patient which of the symptoms come first to differentiate between the two.
- Perilymph fistula: The cardinal symptoms of perilymph fistula (and superior canal dehiscence syndrome) are attacks of vertigo caused by changes in pressure, for example, by coughing, pressing, sneezing, lifting, or loud noises and accompanied by illusory movements of the environment (oscillopsia) and instability of posture and gait with or without hearing disorders. The attacks, which can last seconds to days, may also occur during changes in the position of the head (e.g., when bending over) and when experiencing significant changes in altitude (e.g., mountain tours, flights) [5].
- Episodic ataxia type 2: the duration of the attacks varies from several minutes to hours and more than 90% of the patients have cerebellar signs, in particular gaze-evoked nystagmus and downbeat nystagmus [14, 28]. Manifestation after the age of 20 is exceptional.
- Epilepsy with vestibular aura: Vestibular auras can manifest with short attacks of vertigo and nystagmus. Vestibular aura with additional symptoms, so-called non-isolated vestibular aura, is much more prevalent than isolated vestibular aura, which

is rare. Vestibular aura is primarily associated with temporal lobe seizures. Isolated vestibular aura spells often last only a few seconds, but longer spells are also reported [29].

Other differential diagnoses are characterized by recurrent attacks of vertigo that are induced by certain maneuvers. These differential diagnoses include BPPV, central positional vertigo/nystagmus, “rotational vertebral artery occlusion syndrome” (RVAOS), and orthostatic hypotension. In BPPV the attacks are induced by changes of head or body position relative to gravity and the diagnosis can be proven by the diagnostic positional maneuvers. However, if they are negative, vestibular paroxysmia remains an important differential diagnosis. In central positional/positioning nystagmus the positioning maneuvers induce a similar nystagmus in different head positions [7]. In RVAOS the attacks are induced by rotation of the head either to the right or left and diagnosis is proven by angiography. Similar to vestibular paroxysmia the symptoms are also caused by an excitation of the peripheral vestibular system [26]. In orthostatic hypotension the symptoms occur when the patients stands up and may be associated with vertigo and downbeat nystagmus; the key to this diagnosis is measurement of supine and orthostatic blood pressure [9].

Therapy

Medical Treatment. A therapeutic trial of low dose of carbamazepine (200–600 mg/day) or oxcarbazepine (300–900 mg/day) is often effective. Moreover, a positive response supports the definite diagnosis. The exact specificity of the drug response for the establishment of the diagnosis still needs to be shown by ongoing research. A study on the course of the disease in 32 patients over a 3-year period revealed a significant and continuing decrease in the attack frequency down to 10% of the initial value as well as a reduction in the intensity and duration of the attacks [11]. In case of intolerance, phenytoin, and valproic acid are possible alternatives; however, there are no study data available yet.

Surgical Treatment. Despite the report of partial successes [21] and a clinically well-documented single case [27], operative microvascular decompression should be avoided because, on the one hand, there is the risk of a brainstem infarction due to intra- or post-operative vasospasm and it is frequently difficult to determine the affected side with certainty.

Areas of uncertainties

- Epidemiological and clinical features of vestibular paroxysmia, including the nystagmus during the attacks, are not well known. Since there are no tests to prove the diagnosis of vestibular paroxysmia, further multicenter studies are needed to improve phenotyping.

- The role of imaging in the diagnosis and identification of the affected side is not clear because of the high rate of vascular compression of the 8th cranial in healthy subjects' nerves. Further, MRI with the necessary sequences (CISS, FIESTA) to identify a neuro-vascular compression is not easily available in all countries.
- The role of therapy, i.e. the response to carbamazepine/oxcarbazepine, as a diagnostic criterion has to be further established, in particular, since there are no published randomized controlled trials on treatment.

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