Overview of the International Classification of Vestibular Disorders

Alexandre R. Bisdorff, MD, PhD[a,*], Jeffrey P. Staab, MD, MS[b], David E. Newman-Toker, MD, PhD[c]

KEYWORDS

• Vestibular • Vertigo • Symptoms • Classification • Bárány Society

KEY POINTS

- Classifications and definitions are essential to facilitate communication between clinicians and researchers and promote diagnostic criteria and research in mechanisms epidemiology and treatment.
- To build the International Classification of Vestibular Disorders (ICVD), the Bárány Society organized a systematic internal process and processes for encouraging consensus with other scientific societies.
- The ICVD is organized in 4 layers: (1) symptoms and signs, (2) syndromes, (3) disorders and diseases, and (4) mechanisms.
- Definitions for vestibular symptoms and vestibular migraine have been published. Those for benign paroxysmal positional vertigo, Menière disease, and behavioral aspects should follow in 2015.

INTRODUCTION

Symptom and disease definitions are a fundamental prerequisite for professional communication in clinical, research, and public health settings. The need for structured criteria for epidemiologic, diagnostic and therapeutic research is more obvious for disciplines that rely heavily on syndromic diagnosis, such as psychiatry and headache, where there are currently no histopathologic, radiographic, physiologic, or other confirmatory diagnostic tests available. However, diagnostic standards and

[a] Department of Neurology, Centre Hospitalier Emile Mayrisch, rue Emile Mayrisch, Esch-sur-Alzette 4005, Luxembourg; [b] Department of Psychiatry and Psychology, Mayo Clinic, 200 1st Street SW, Rochester, MN 55905, USA; [c] Departments of Neurology, Otolaryngology, and Epidemiology, The Johns Hopkins University School of Medicine & Bloomberg School of Public Health, The Johns Hopkins Hospital, CRB-II, Room 2M-03 North, 1550 Orleans Street, Baltimore, MD 21231, USA
* Corresponding author.
E-mail address: alexbis@pt.lu

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classifications are also crucial in areas of medicine, such as epilepsy and rheumatology, where, although confirmatory tests do exist, there is substantial overlap in clinical features or biomarkers across syndromes. Vestibular disorders are similar to the latter examples. Scientific and therapeutic progress, as well as public awareness of psychiatric and headache disorders, vastly increased after the introduction of the first modern version of the Diagnostic and Statistical Manual of Mental Disorders (DSM-III) by the American Psychiatric Association in 1980[1] and the first International Classification of Headache Disorders by the International Headache Society in 1988.[2]

Although numerous advances in basic and clinical vestibular research have been made, progress in the field likely has been hampered by the lack of explicit and uniform criteria for the description of symptoms, syndromes, and clinical disorders. Other than the definition of Menière disease by the Japanese Society for Equilibrium Research and the American Academy of Otolaryngology—Head and Neck Surgery[3] as well as the Classification of Peripheral Vestibular Disorders by the Spanish Society of Otorhinolaryngology,[4] there have not been systematic efforts to create widely accepted classification criteria before the initiative by the Bárány Society to build the International Classification of Vestibular Disorders (ICVD).[5]

There are probably several reasons why the classification and unification of definitions for vestibular disorders has lagged other illnesses. Classifications are usually created by scientific societies of subspecialists within a medical specialty or under the lead of 1 medical specialty. Vestibular disorders cross medical specialist boundaries and, despite being very prevalent,[6] are the province of small subspecialties within otolaryngology and neurology. Additionally, for neurologists and otolaryngologists to be able to cover competently the spectrum of differential diagnoses of vestibular disorders, they need to acquire elaborate knowledge about the inner ear, vestibular, postural, and oculomotor pathways in the brain, and related systems that control autonomic and threat responses, which goes beyond the standard curricula during residency training of any specialty. It was, therefore, necessary that an international scientific society with an interdisciplinary membership of clinicians and basic scientists with expertise in vestibular disorders, like the Bárány Society,[7] assume responsibility for developing the ICVD.

GOALS AND SCOPE FOR THE INTERNATIONAL CLASSIFICATION OF VESTIBULAR DISORDERS INITIATIVE

The goal of the ICVD initiative is to develop a comprehensive classification scheme and definitions of individual vestibular diseases disorders that is acceptable worldwide. To achieve the goal of wide acceptance, the Bárány Society is actively seeking the input of members from other associations dealing with vestibular disorders, such as the Société Internationale d'Otoneurologie and the Comisión de Otoneurología de la Sociedad Española de Otorrinolaringología in Europe, the American Academy of Otolaryngology – Head and Neck Surgery (AAO-HNS) in the United States, the Japanese Equilibrium Society, and the Korean Balance Society, as well as individual scientists and clinicians from the international vestibular community. Beyond cooperation with individuals and associations within the vestibular community, the Bárány Society is also seeking cooperation and consensus with scientific associations from related disciplines, if there are important aspects of diseases that touch more than 1 society. One example is vestibular migraine, where the Bárány Society cooperated with the International Headache Society to publish a consensus document on diagnostic criteria.[8]

The term "vestibular disorders" refers to disturbances arising from the vestibular system, but the definition of the vestibular system itself can be understood broadly

or narrowly. The vestibular system contributes to gait, stance, locomotion, balance, vision, spatial orientation, navigation, and spatial memory because of the widespread use of vestibular information in the brain. Furthermore, brain dysfunction of almost any cause, whether primary or secondary, may affect balance adversely. It is therefore necessary to limit the scope of problems classified as "vestibular disorders" within the ICVD, although some decisions will ultimately be arbitrary.

The ICVD will include diseases that affect the vestibular labyrinth of the inner ear, connections from the labyrinth to the brain at the brainstem, the cerebellum, subcortical structures that process spatial stimuli, and the vestibular cortex. The ICVD will also include illnesses that are primarily the province of other specialties, but produce symptoms that mimic vestibular disorders. The ICVD will focus on the vestibular presentations of these conditions, but will not seek to redefine or reclassify the primary nonvestibular disorder. Examples include syncope, seizures, stroke, headache disorders, cerebellar ataxia syndromes, extrapyramidal movement disorders, and behavioral disorders that are already defined by other groups. The classification will eventually include controversial and emerging entities, such as cervical vertigo,[9] in the hope that identifying limits of current scientific knowledge will prompt research to fill these gaps.

Before starting the task of elaborating definitions of disorders the problem of variable use of terminology for describing core vestibular symptoms, such as dizziness and vertigo, had to be addressed. Even when studied in a single, English-speaking country, the term "vertigo" has been shown to have diverse meanings for patients,[10] generalist physicians,[11] and even otologists.[12]

METHODOLOGY AND PROCESS FOR DEVELOPING THE INTERNATIONAL CLASSIFICATION OF VESTIBULAR DISORDERS

In 2006 the Classification Committee of the Bárány Society (CCBS) convened its first meeting to begin structuring the approach to developing the ICVD. The group needed to develop a conceptual framework, a list of initial topics, and a process for consensus building. To permit terminological consistency in defining vestibular disorders as part of the ICVD, it was decided to first define key vestibular symptoms and build consensus around these formalized definitions (**Boxes 1** and **2**); the product of this initial work has already been published.[5] Initial topics (eg, Menière disease, vestibular migraine, benign paroxysmal positional vertigo) were chosen for their frequency and importance to the vestibular disorders community. Specific issues are delegated to subcommittees working according to predetermined guidelines. Subcommittees

Box 1
Glossary of primary vestibular symptom definitions in the International Classification of Vestibular Disorders

Dizziness is the sensation of disturbed or impaired spatial orientation without a false or distorted sense of motion.

Vertigo is the sensation of self-motion (of head/body) when no self-motion is occurring or the sensation of distorted self-motion during an otherwise normal head movement.

Vestibulovisual symptoms are visual symptoms that usually result from vestibular pathology or the interplay between visual and vestibular systems. These include false sensations of motion or tilting of the visual surround and visual distortion (blur) linked to vestibular (rather than optical) failure.

Postural symptoms are balance symptoms related to maintenance of postural stability, occurring only while upright (seated, standing, or walking).

Box 2
Glossary of secondary vestibular symptom definitions in the International Classification of Vestibular Disorders

Spontaneous vertigo (*or dizziness*) is vertigo (or dizziness) that occurs without an obvious trigger.

Triggered vertigo (*or dizziness*) is vertigo (or dizziness) that occurs with an obvious trigger.

- *Positional vertigo* (*or dizziness*) is vertigo (or dizziness) triggered by and occurring *after* a change of head position in space relative to gravity.
- *Head motion vertigo* (*or dizziness*) is vertigo (or dizziness) occurring only *during* head motion (ie, that is time locked to the head movement).
- *Visually induced vertigo* (*or dizziness*) is vertigo (or dizziness) triggered by a complex, distorted, large field, or moving visual stimulus, including the relative motion of the visual surround associated with body movement.
- *Sound-induced vertigo* (*or dizziness*) is vertigo (or dizziness) triggered by an auditory stimulus.
- *Valsalva-induced vertigo* (*or dizziness*) is vertigo (or dizziness) triggered by any bodily maneuver that tends to increase intracranial or middle ear pressure.
- *Orthostatic vertigo* (*or dizziness*) is vertigo (or dizziness) triggered by and occurring on arising (ie, a change of body posture from lying to sitting or sitting to standing).

Vestibulovisual symptoms

- *External vertigo* is the false sensation that the visual surround is spinning or flowing.
- *Oscillopsia* is the false sensation that the visual surround is oscillating.
- *Visual lag* is the false sensation that the visual surround follows behind a head movement with a delay or makes a brief drift after the head movement is completed.
- *Visual tilt* is the false perception of the visual surround as oriented off the true vertical.
- *Movement-induced blur* is reduced visual acuity during or momentarily after a head movement.

Postural symptoms

- *Unsteadiness* is the feeling of being unstable while seated, standing, or walking without a particular directional preference.
- *Directional pulsion* is the feeling of being unstable with a tendency to veer or fall in a particular direction while seated, standing, or walking. The direction should be specified as latero-, retro-, or anteropulsion. If lateropulsion, the direction (right or left) should be specified.
- A *balance-related near fall* is a sensation of imminent fall (without a completed fall) related to strong unsteadiness, directional pulsion, or other vestibular symptom (eg, vertigo).
- A *balance-related fall* is a completed fall related to strong unsteadiness, directional pulsion, or other vestibular symptom (eg, vertigo).

have a designated chair, who is a member of the Bárány Society. He or she composes his or her subcommittee, which must include members from 3 continents, at least 1 clinician must be an otolaryngologist and 1 a neurologist. Clinicians and scientists from other fields are included, based on needs for additional expertise.

The work of a subcommittee is part of a consensus process based on the best available evidence at present. Each subcommittee's work is subject to review by the CCBS. Draft definitions are presented publically at the biennial Bárány Society congresses and posted online for a period of commentary by Bárány Society members. Input is also

solicited from other scientific societies based on the topic treated. The CCBS oversees the whole process to ensure that all the parts of the classification are coherent with each other. The final document is then published in the *Journal of Vestibular Research* without further peer review and is then available as open access publication.

INTERNATIONAL CLASSIFICATION OF VESTIBULAR DISORDERS STRUCTURE

The proposed structure of the ICVD currently includes 4 layers. Layer I, symptoms and signs; layer II, clinical syndromes; layer III-A, diseases and disorders; and layer III-B, pathophysiologic mechanisms (Fig. 1). Each layer contains elements (eg, specific symptoms or diseases) that are important in their own right, but are also important in their links with other elements. This structure will allow the ICVD to depict conceptual connections between elements within and across layers. Because knowledge of these connections is incomplete, it is recognized that some links may also "skip" layers.

A multilayer approach was considered essential to accommodate the breadth of clinical and research applications, now and in the future. Some clinicians and researchers must organize their approach beginning with signs and symptoms, whereas others must lead with a primary focus on specific diseases or pathophysiologic mechanisms.

INTERNATIONAL CLASSIFICATION OF VESTIBULAR DISORDERS LAYER I: SYMPTOMS AND SIGNS

Definitions for specific vestibular symptoms in layer I of the ICVD have been written and published.[5] Work is underway on specific signs, particularly pathologic eye

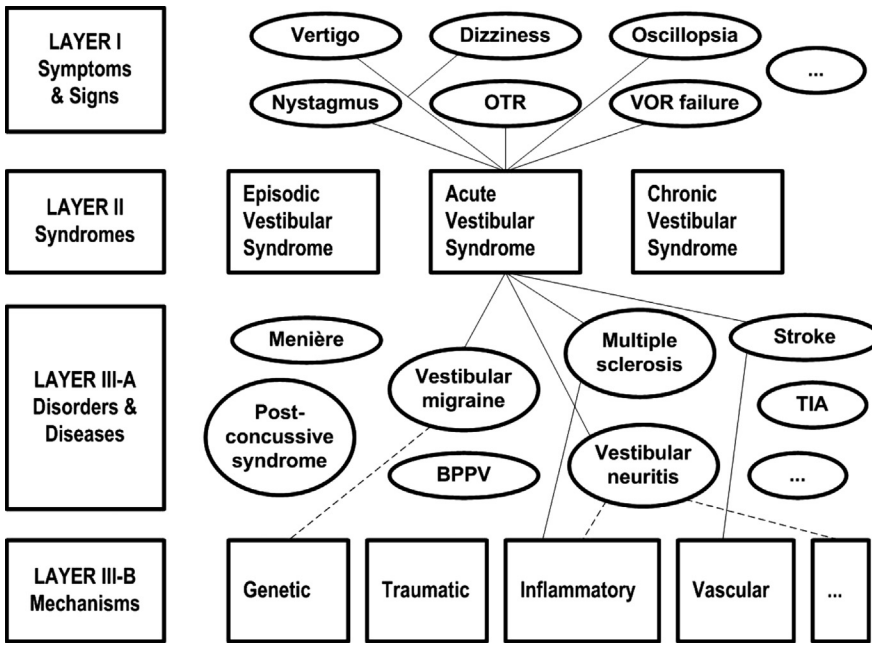

Fig. 1. Four-layer framework of the international classification of vestibular disorders. Links between layers are shown demonstrating, in this case, the conceptual relations between the "acute vestibular syndrome" (layer II), its component symptoms (layer I), its etiologic causes (layer III-A), and their mechanistic underpinnings (layer III-B). Solid lines represent definite links, and dashed lines represent uncertain links. OTR, ocular tilt reaction; TIA, transient ischemic attack.

movements. This layer was addressed first because it was considered foundational to the development of all subsequent definitions, the vast majority of which will be based on clinical phenomena. It was decided to limit the scope of this work to defining cardinal vestibular symptoms, representing the primary clinical symptoms typically resulting from vestibular disorders but excluding secondary symptoms such as nausea, fatigue, and anxiety.

The principles followed for developing symptom definitions were as follows:

- No "vestibular" symptom has a totally specific meaning in terms of topology or nosology and its pathogenesis is likely to be incompletely understood.
- Symptom definitions should be as purely phenomenological as possible, without reference to a specific theory on pathophysiologic causes of a particular disease.
- Definitions for symptoms are clearest if they are nonoverlapping and nonhierarchical, allowing for 1 or more symptoms to coexist in a particular patient.

The ICVD describes 4 categories of cardinal vestibular symptoms: (1) vertigo, (2) dizziness, (3) vestibulovisual symptoms, and (4) postural symptoms, including subtypes for each.[5] The new nomenclature distinguishes *vertigo* (a false sense of motion of spinning or nonspinning quality) and *dizziness* (disturbed spatial orientation without a false sense of motion), which represents an important departure from typical practice in the United States, where dizziness is an umbrella term encompassing vertigo, unsteadiness, imbalance, presyncope, and other 'nonspecific' sensations.[11] Although vertigo and dizziness are distinguished from one another in the ICVD, neither is considered pathognomonic in its links to underlying vestibular pathology. Both symptoms are frequently encountered in patients with vestibular or nonvestibular disorders, whether acute or chronic.[10,13] Vertigo and dizziness are each divided into 2 categories, spontaneous and triggered.

A separate category on vestibulovisual symptoms was developed, because vestibular dysfunction can result in a range of visual disturbances. Because "internal" and "external" vertigo are sometimes dissociated clinically (eg, in a patient who sees the world spinning or rotating from jerk nystagmus, but feels no spinning with eyes closed), the visual sense of motion could not simply be incorporated into the definition of vertigo. The term "external vertigo," referring to false sensations of motion in the visual surround, was listed among the vestibulovisual symptoms. Although some prior studies[6] have called this sense of visual flux "oscillopsia," group consensus was that oscillopsia should be restricted to describing a bidirectional, oscillating visual motion that incorporates complaints such as "jumping" or "bouncing" vision.

The ICVD definitions for postural balance symptoms use *unsteadiness* as the preferred descriptive term for postural instability when upright (ie, sensations of swaying, rocking, or wobbling when sitting, standing, or walking), rather than the more linguistically ambiguous terms "disequilibrium" or "imbalance." If the unsteadiness has a particular directional bias, the term *directional pulsion* should be used and the direction specified (eg, lateropulsion to the right). A subcommittee devoted to signs is currently establishing a classification and definitions of various forms of nystagmus.

INTERNATIONAL CLASSIFICATION OF VESTIBULAR DISORDERS LAYER II: SYNDROMES

Layer II, syndromes, offers an intermediate layer of syndromic classification that bridges between constellations of symptoms and signs and the diseases or disorders causing them. For example, sudden-onset vertigo, nausea, vomiting, head motion intolerance, gait unsteadiness, and nystagmus would constitutive an "acute vestibular syndrome" that has underlying causes, such as vestibular neuritis and acute

cerebellar infarction. Currently proposed are 3 specific syndromes comprising the bulk of all vestibular presentations: (1) acute vestibular syndrome comprising diseases and disorders that usually manifest with a single episode of sudden onset vestibular symptoms and signs (eg, vestibular neuritis or acute stroke), (2) episodic vestibular syndrome comprising diseases and disorders that are recurrent by nature (eg, Menière disease, vestibular migraine, or transient ischemic attack), and (3) chronic vestibular syndrome comprising diseases and disorders that produce persistent vestibular symptoms and signs over an extended period of time (bilateral vestibular failure or cerebellar degeneration). Layer II will facilitate the development of clinical care pathways and standardized inclusion criteria for research studies focused on diagnostic accuracy.

INTERNATIONAL CLASSIFICATION OF VESTIBULAR DISORDERS LAYER III: DISORDERS AND DISEASES

Layer III-A contains vestibular diseases and disorders and seeks to be relatively comprehensive. The ICVD will use existing terms for vestibular disorders and diseases wherever possible. New terms will be developed only for conditions not included in previous classifications or, rarely, for conditions having multiple names that are all incompatible with ICVD nomenclature. If several existing names are used to describe the same condition, the CCBS will endeavor to include the most suitable one in the ICVD classification system. Other terms will be designated as "terms not used in this nomenclature."

For most vestibular diseases and disorders, no single pathognomonic finding or definitive set of confirmatory tests are available. Therefore, the definitions use operational criteria for symptom dimensions (eg, type, timing, or triggers) or symptom clusters and ancillary test results, as appropriate. Both supporting and negating criteria will be considered. Criteria will be graded to define a range of certainty about a particular diagnosis from *definite* (clear and certain) to *probable* (less clear and less certain). The former will be more restrictive (ie, more specific), whereas the latter will be more inclusive (ie, more sensitive). Denoting a degree of diagnostic certainty is important for both clinical care and research. For example, clinicians will likely apply a high-risk therapy (eg, vestibular neurectomy) only to patients with "definite" disease, although they may be willing to apply a low-risk therapy (eg, dietary modification) to a patient with probable or even "possible" disease.

The CCBS established the first 4 disease-oriented subcommittees to address the definitions and diagnostic criteria for diseases where consensus is most urgently needed because of their epidemiologic importance or ongoing controversy. These subcommittees are focused on Menière disease, benign paroxysmal positional vertigo, vestibular migraine, and behavioral neuro-otologic conditions. The diagnostic criteria for vestibular migraine were the first to be published.[8] The definition of vestibular migraine identifies this condition as a subcategory of migraine, similar to retinal migraine, with vestibular symptoms as a predominant sensory manifestation. The criteria are based on definitions first proposed by Neuhauser and colleagues[14] in 2001 and later formalized by Furman and colleagues.[15,16] The Menière Subcommittee built on the AAO-HNS definition of Menière disease, which has been used worldwide since its publication in 1995.[3] The new definition will only include 2 categories of certainty (definite and probable) and will be more precise regarding hearing loss and the diagnostic overlap with other conditions presenting the episodic vestibular syndrome, mainly vestibular migraine. The Benign Paroxysmal Positional Vertigo Subcommittee prepared detailed definitions of symptoms and signs for all variants of canalolithiasis

and cupulolithiasis currently described in the medical literature, subdividing them into established and emerging entities according to available evidence. These definitions are expected to be published in 2015.

The Behavioral Subcommittee was the last of the current working groups to be formed and met for the first time in August 2010 in Reykjavik during the 26th Bárány Society meeting. That subcommittee was charged with 2 tasks: (1) to identify the primary and secondary psychiatric disorders that manifest vestibular symptoms and modify their standard definitions for ease of use by otologists and neuro-otologists, and (2) to evaluate the available data on phobic postural vertigo, chronic subjective dizziness, space–motion discomfort, and visually induced dizziness (previously visual vertigo) to determine whether these entities represent 1 or more disorders, and then prepare a suitable definition or definitions for the ICVD. This task was complicated by the fact that both sets of standardized psychiatric nomenclature were in the process of revision when the subcommittee was formed. The updated version of the DSM (DSM-5) was published in 2013 after a lengthy revision cycle [17]. Its definitions align better with those of the 11th edition of the International Classifications of Diseases (ICD-11),[18] which is still in beta draft version awaiting finalization in 2017. The subcommittee prepared modified definitions of anxiety and depressive disorders that manifest vestibular symptoms for use in neuro-otologic clinical and research settings. It also determined that phobic postural vertigo and chronic subjective dizziness were different descriptions of a single clinical entity with space–motion discomfort and visually induced dizziness as important symptoms. Therefore, the subcommittee defined 1 chronic vestibular syndrome that captures key features of this condition and termed it persistent postural–perceptual dizziness in keeping with ICVD nomenclature. The definition of persistent postural–perceptual dizziness will be posted to the Internet for commentary in spring 2015, with the remaining behavioral vestibular disorders to follow.

INTERNATIONAL CLASSIFICATION OF VESTIBULAR DISORDERS LAYER III-B: MECHANISMS

Layer III-B contains the pathoanatomic, pathophysiologic, and etiologic mechanisms underlying vestibular disorders. It is anticipated that this layer will be developed last and will be the most incomplete in the first iteration of the ICVD, but will expand and grow the most with future scientific discovery. This layer has been created with the knowledge that, eventually, clinical phenomena (ie, symptoms and signs) may be linked directly with mechanistic understanding (eg, genetic mutation) for the purposes of diagnosis and treatment, skipping intermediate steps in the diagnostic process that are unavoidable at present (eg, “diagnosis” of Menière disease).

INTERNATIONAL CLASSIFICATION OF VESTIBULAR DISORDERS AND FUNCTIONAL OUTCOMES

Finally, it is recognized that the functional impact of vestibular diseases and disorders is substantial and that a schema for standardized assessment of disability or handicap is needed. A diagnosis itself does not provide information about functional consequences for the affected individual. The World Health Organization has created the International Classification of Functioning, Disability and Health, which describes the adverse effect of disease on daily activities and makes the assessment of functional impairment and disability caused by diseases of all types systematic and comparable. A working group is developing an instrument to use International Classification of Functioning, Disability and Health measures to assess function in patients with vestibular disorders.

SUMMARY

An initiative under the aegis of the CCBS is currently underway to develop a comprehensive classification structure and formal definitions for vestibular symptoms, syndromes, and diseases. The success of the ICVD will depend on its ability to improve communication among scientists, clinicians, patients, policymakers, and the general public, to advance knowledge about vestibular disorders, and to reduce the morbidity of those afflicted by the conditions it defines.

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