Diagnostic Criteria for Persistent Postural-Perceptual Dizziness (PPPD):
Consensus document of the Committee for the Classification of Vestibular Disorders of the
Bárány Society

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Abbreviations:

BPPV – benign paroxysmal positional vertigo
CCBS – Committee for the Classification Vestibular Disorders of the Bárány Society
CSD – chronic subjective dizziness
fMRI – functional magnetic resonance imaging
ICD-11 – International Classification of Diseases, 11th edition (beta draft)
ICVD – International Classification of Vestibular Disorders
MdDS – mal de debarquement syndrome
PIVC – parieto-insular vestibular cortex
PPPD – persistent postural-perceptual dizziness
PPV – phobic postural vertigo
SMD – space-motion discomfort
VV – visual vertigo
Abstract

This paper presents diagnostic criteria for persistent postural-perceptual dizziness (PPPD) to be included in the International Classification of Vestibular Disorders (ICVD). The term PPPD is new, but the disorder is not. Its diagnostic criteria were derived by expert consensus from an exhaustive review of 30 years of research on phobic postural vertigo, space-motion discomfort, visual vertigo, and chronic subjective dizziness. PPPD manifests with one or more symptoms of dizziness, unsteadiness, or non-spinning vertigo that are present on most days for three months or more. Upright posture, active or passive movement, and exposure to moving or complex visual stimuli may exacerbate symptoms. Precipitating disorders include conditions capable of triggering vertigo, unsteadiness, or dizziness or disrupting balance, such as peripheral or central vestibular disorders, other medical illnesses, and psychological distress. PPPD may be present alone or co-exist with other conditions. Possible subtypes await identification and validation in future investigations. The pathophysiologic processes underlying PPPD are not fully known. Emerging research suggests that it may arise from functional changes in postural control mechanisms, multi-sensory information processing, or cortical structures linking spatial orientation and threat assessment. Thus, PPPD is classified as a chronic functional vestibular disorder. It is not a structural or psychiatric condition.
1. Introduction

This paper introduces the diagnostic criteria for persistent postural-perceptual dizziness (PPPD), a chronic functional vestibular syndrome. PPPD is a new term, but the core features of the disorder can be found in medical writings dating back to the 19th century [1-3]. After a brief review of this historical context, the contemporary background of PPPD is presented, followed by its diagnostic criteria with explanatory notes to guide their application. Then, the differential diagnosis is discussed in detail. Lastly, data suggesting possible pathophysiologic mechanisms is summarized.

1.1. Historical background

In the 1870s, three German physicians described syndromes of dizziness and discomfort in motion rich environments, accompanied by autonomic arousal, anxiety, and avoidance of provocative circumstances [1-3]. Benedikt [1] emphasized a neuro-ophthalmologic process in Platzschwindel (vertigo in a plaza or square), whereas Cordes [2] focused on a psychological genesis in Platzangst (fear in a plaza or square) [4]. Westphal [3] proposed that postural control, locomotion, conscious appraisal of spatial orientation, and threat assessment were “part of one process” in Die Agoraphobie (fear of the marketplace) [5]. Other European and American physicians added commentary [4,6,7], including observations that otologic diseases could trigger Westphal’s agoraphobia, especially in people with pre-existing anxiety [6], but differing views of these three syndromes and debates about whether they were predominantly neurologic or psychiatric in nature were never resolved. As otology, neurology, and psychiatry matured into separate specialties in the early 20th century, Platzschwindel and Platzangst faded from use and agoraphobia became a psychiatric disorder, losing its space and motion context [8,9].
A century later small case series were published describing various syndromes of spatial disorientation and aberrant motion sensations, including supermarket syndrome [10], space phobia, [11,12], motorist’s vestibular disorientation syndrome [13], visually induced motion symptoms [14], and physiologic height vertigo [15].

1.2. Contemporary context

Sustained investigations in larger numbers of patients began in the 1980s. From clinical observations in their tertiary otorhinolaryngologic practice, Brandt and Dieterich [16] defined *Phobischer Attacken-Schwankschwindel* (phobic postural vertigo, PPV) in 1986 as a syndrome of postural dizziness and fluctuating unsteadiness accompanied by mild anxiety and depression in patients with obsessive compulsive personality traits. Other features are listed in Table 1. Brandt, Dieterich, and their colleagues [17-19] showed that PPV was common, persistent, and distinct from other vestibular diseases and psychiatric disorders.

Starting in the mid-1980s, Jacob and colleagues conducted a series of investigations into potential links between anxiety symptoms, persistent dizziness, and vestibular dysfunction in patients from a tertiary anxiety disorders clinic [20-22]. In 1989, they described [23] and subsequently validated [24] *space-motion discomfort* (SMD) as a combination of uneasiness about spatial orientation and increased awareness of motion stimuli. Active or passive movement (e.g., walking down a supermarket aisle, riding in a vehicle) and exposure to moving or patterned objects in the environment (e.g., passing traffic, striped curtains, crowds) increased symptoms in affected individuals.

In 1995, Bronstein [25] identified *visual vertigo* (VV) in a portion of patients in his tertiary otorhinolaryngologic clinic. This syndrome, which followed acute peripheral or central vestibular losses, manifested with sensations of unsteadiness or dizziness on exposure to
complex or moving visual stimuli that persisted despite patients seeming to recover from their acute vestibular deficits. The visual cues that exacerbated VV overlapped with environmental stimuli that activated SMD [26,27].

Finally, in 2004, Staab and colleagues [28] described *chronic subjective dizziness* (CSD) based on observations of patients in their tertiary balance center, and defined it more explicitly in 2007 [29]. This syndrome was similar in many ways to PPV, but focused on physical symptoms of persistent nonvertiginous dizziness or unsteadiness with heightened sensitivity to motion of self or objects in the environment and difficulty with precision visual tasks. Their definition was highly sensitive (>85%) and specific (>90%) for identifying CSD versus Menière’s disease, vestibular migraine, or benign paroxysmal positional vertigo (BPPV), in patients with or without additional comorbidity [30].

1.3. Current considerations for classifying vestibular diseases and disorders

The International Classification of Vestibular Disorders (ICVD) divides diseases and disorders based on duration of symptoms into acute, episodic, or chronic syndromes [31]. PPPD may last for months to years making it a chronic vestibular syndrome. Vestibular diseases and disorders also are divided into structural, functional, and psychiatric conditions based on proven or presumed pathophysiologic mechanisms. Here functional conditions are considered as they were in the early 19<sup>th</sup> century as disorders “arising from a change in the mode of action of an organ” [32], unrelated to structural or cellular deficits. As revived in the modern era, most notably in gastroenterology [33], this concept of functional conditions also distinguishes them from psychiatric illnesses. Studies of PPV, VV, SMD, and CSD identified a number of functional alterations in vestibular and balance mechanisms [34-43] and separated these conditions from primary psychiatric disorders.
These findings, which are reviewed below, appear largely applicable to PPPD, indicating that it is a functional, not structural or psychiatric, vestibular disorder.

2. Methods

In 2006, members of the Bárány Society created a working group to standardize nomenclature for vestibular diseases and disorders worldwide. This led to formation of the Committee for Classification Vestibular Disorders of the Bárány Society (CCBS) to oversee development of the first International Classification of Vestibular Disorders (ICVD) [31]. To date, this process has generated consensus documents defining vestibular symptoms [45], vestibular migraine [46], BPPV [47], and Menière’s disease [48]. Additional definitions are in the offspring. In 2010, the CCBS chartered a Behavioral Subcommittee to identify primary and secondary psychiatric disorders that cause or amplify vestibular morbidity and review evidence about the nature of PPV, SMD, VV, and CSD. In keeping with established procedures for the classification process [49], the Behavioral Subcommittee included an otologist (A.H.), neurologist (M.S.), and members with special expertise in psychosomatic medicine (J.P.S., A.E.H.) and psychiatry (J.P.S., R.J.). Members hailed from three continents (Asia, Europe, and North America). Two senior neuro-otologists (T.B., A.B.) graciously agreed to advise subcommittee members on their deliberations.

The subcommittee met for the first time in August 2010 during the Bárány Society’s biennial congress in Reykjavík, Iceland. From 2010-2012, the chair (J.P.S.) consulted with subcommittee members individually. These deliberations produced a consensus that PPV, SMD, VV, and CSD included a core set of physical symptoms that represented a distinctly definable vestibular disorder. Subcommittee members prepared a draft definition of this
disorder that was updated iteratively after review by the general membership of the Bárány Society in June 2012 in Uppsala, Sweden, the CCBS in November 2013 in Mondorf-les-Bains, Luxembourg, and then again by the general membership in May 2014 in Buenos Aires, Argentina. The disorder was named persistent postural-perceptual dizziness to reflect its main diagnostic criteria of persistent non-vertiginous dizziness, unsteadiness, and non-spinning vertigo that are exacerbated by postural challenges and perceptual sensitivity to space-motion stimuli. A separate, 100-word narrative definition was prepared for the World Health Organization as part of the Bárány Society’s recommendations for updates to the vestibular disorders section of the forthcoming 11th edition of the International Classification of Diseases (ICD-11) [50].

3. Criteria for the diagnosis of Persistent Postural-Perceptual Dizziness (PPPD)

Criteria A-E must be satisfied to make a diagnosis of PPPD.

A. One or more symptoms of dizziness, unsteadiness, or non-spinning vertigo are present on most days for 3 months or more.1-3

1. Symptoms are persistent, but wax and wane.

2. Symptoms tend to increase as the day progresses, but may not be active throughout the entire day.

3. Momentary flares may occur spontaneously or with sudden movements.

B. Symptoms are present without specific provocation, but are exacerbated by:

1. Upright posture,

2. Active or passive motion without regard to direction or position, and

3. Exposure to moving visual stimuli or complex visual patterns,
although these three factors may not be equally provocative.4

C. The disorder usually begins shortly after an event that causes acute vestibular symptoms or problems with balance, though less commonly, it develops slowly.5

1. Precipitating events include acute, episodic, or chronic vestibular syndromes, other neurologic or medical illnesses, and psychological distress.
   a) When triggered by an acute or episodic precipitant, symptoms typically settle into the pattern of criterion A as the precipitant resolves, but may occur intermittently at first, and then consolidate into a persistent course.
   b) When triggered by a chronic precipitant, symptoms may develop slowly and worsen gradually.

D. Symptoms cause significant distress or functional impairment.

E. Symptoms are not better attributed to another disease or disorder.6

Notes

(1) The primary symptoms of PPPD include non-motion sensations of disturbed or impaired spatial orientation (dizziness), feelings of being unstable while standing or walking (unsteadiness), and false or distorted sensations of swaying, rocking, bobbing, or bouncing of oneself (internal non-spinning vertigo) or the surroundings (external non-spinning vertigo) [45].

(2) Symptoms must be present for more than 15 of every 30 days. Most affected individuals experience symptoms every day or nearly every day.

(3) Symptoms need not be continuous, but must be present for prolonged (hours-long)
periods throughout the day. Momentary symptoms alone do not fulfill this criterion.

(4) All three provocative factors of criterion B must be discernable in the clinical history, but do not have to be equally troublesome.

a. Upright posture means standing or walking.

b. Active motion refers to a person’s self-generated movements. Passive motion refers to a person being moved by conveyances or other individuals (e.g., riding in a vehicle or elevator/lift, being jostled in a crowd).

c. Visual stimuli may encompass large or small portions of the visual field. Full field stimuli (e.g., passing traffic, rooms filled with busy décor, graphics displayed on large screens) are the most provocative, but smaller stimuli (e.g., books, computers, mobile electronic devices) may be troublesome when they are the focus of sustained visual attention.

(5) The most common precipitating events are peripheral or central vestibular conditions (25-30% of cases), attacks of vestibular migraine (15-20%), panic attacks or generalized anxiety that manifest prominent dizziness (15% each), concussion or whiplash injuries (10-15%), and autonomic disorders (7%). Other events that are capable of producing vertigo, unsteadiness or dizziness, or altering balance function (e.g., cardiac dysrhythmias, adverse drug reactions) trigger the disorder less commonly (collectively ~3%) [29,30]. The majority of conditions that trigger PPPD are acute or episodic in nature. Patients report the onset of chronic symptoms of PPPD following their acute illnesses. However, precipitants such as generalized anxiety disorder, autonomic disorders, and peripheral or central degenerative
conditions may develop insidiously. In these cases, patients are less likely to report a distinct onset. When a specific precipitant cannot be identified, particularly when symptoms slowly worsen, re-evaluation of the diagnosis is indicated and a period of prospective monitoring may be needed to confirm it.

(6) PPPD may co-exist with other diseases or disorders. Evidence of another active illness does not necessarily exclude a diagnosis of PPPD. Rather, clinical judgment must be exercised to determine the best attribution of the patient’s vestibular symptoms to all identified illnesses [44,51].

4. Comments

4.1. Further descriptions of diagnostic criteria

The diagnostic criteria of PPPD are explained in greater detail below, based on information derived from numerous reports on PPV, SMD, VV, and CSD. Table 1 lists the diagnostic criteria of PPPD and the characteristics of PPD, SMD, VV, and CSD that informed the definition. Citations given in the subsections below relate to the four precursors.

4.1.1. Core vestibular symptoms

The primary symptoms of PPPD are dizziness, unsteadiness, and certain types of non-spinning vertigo [16,44]. The dizziness of PPPD is a non-motion symptom [45] that patients may describe variously as cloudiness, fuzziness, fullness, heaviness, or lightness in the head, or a feeling that their spatial orientation is not sharp or visual focus is not clear. Unsteadiness is a sensation of instability or wobbling when upright, or a feeling of veering from side to side when walking without a directional preponderance [45]. Non-spinning
vertigo encompasses feelings of swaying, rocking, bouncing, or bobbing that patients may describe as motion inside their heads, involving their entire heads or bodies, or occurring in the environment. Tilting and sliding sensations are included in the Bárány Society's definition of non-spinning vertigo [45], but these are not typical symptoms of PPPD [17,44,51]. Intermittent, momentary sensations of illusory movement that last no more than a few seconds (Criterion A.3.) may include both spinning and non-spinning vertigo [16,17].

4.1.2. Temporal pattern of symptoms

PPV was defined by fluctuating postural symptoms and momentary illusions of movement [16,17]. SMD was described as a situational phenomenon, occurring during exposures to provocative stimuli [22,23]. The earliest reports of VV [25] also focused on situational symptoms, though recent investigations identified a link between persistent VV and chronic dizziness [41]. Definitions of CSD [29,44] emphasized chronic symptoms lasting throughout the day and exacerbated by motion stimuli. One study found that symptoms of CSD were absent or mildest for about an hour after patients awoke in the morning, but then increased as the day progressed [52]. These observations identified two temporal patterns incorporated into PPPD (Table 1). Most patients experience a background of vestibular symptoms throughout the day, nearly every day [17,44]. Symptom-free intervals tend to be brief (minutes to hours), though a distinct minority of patients may experience symptom-free periods lasting for days to weeks. Symptoms wax and wane spontaneously, but are aggravated by the three provocative factors of Criterion B. When free of these circumstances, patients' symptoms may be innocuous, limited to non-motion dizziness with minimal unsteadiness or non-spinning vertigo. However, all
Symptoms are susceptible to provocation with upright posture, motion, and exposure to complex visual stimuli. With these provocative situations, unsteadiness and non-spinning vertigo tend to dominate the clinical picture.

4.1.3. Provocative factors

The provocative factors included in Criterion B were described in all or most of the precursors of PPPD (Table 1). Upright posture, active or passive movement, and exposure to visually complex or moving stimuli aggravate the core symptoms of PPPD. There are no data to identify one of these factors as being more provocative than the others or to determine the sensitivities and specificities of demanding the presence of one, two, or all three factors to make a diagnosis of PPPD. Therefore, Criterion B requires that affected individuals will report at least some difficulty with all three factors, while recognizing that patients will not be equally sensitive to all of them, and that the most provocative factor will vary among patients with PPPD.

Symptoms may not increase immediately on standing, moving, or entering visually stimulating environments, but build throughout continued exposure to these situations. Symptoms usually do not return to baseline immediately on cessation of provocations, but may last for hours thereafter. This pattern differs from that experienced by patients with structural deficits whose symptoms increase and decrease in close temporal relationship to motion exposures. Patients with PPPD may express concerns about having to endure an increase in their noxious symptoms in these provocative situations, but this differs from individuals with anxiety disorders who tend to focus more on fears of becoming incapacitated, injuring themselves or others, or attracting unwanted attention.
4.1.3.1. Upright posture

Most patients with PPPD report more severe symptoms when standing or walking than when sitting or lying down [16,17,44]. Individuals who are particularly sensitive to postural changes also may experience increased symptoms when sitting upright without back or arm support. Patients may not have a complete resolution of symptoms when lying down, but recumbent postures are the least troublesome. Patients may minimize the adverse effects of upright posture by touching fixed objects, using gait aids, or holding onto other people. Patients do not have to hold tightly to support their weight. Rather, a light touch is sufficient to obtain the stabilizing effect of somatosensory input.

4.1.3.2. Active or passive movement

Provocative movements may occur in any direction or position. Most patients find that active and passive motions are troublesome in direct proportion to their intensity. Speed, duration, and repetition of movement appear to be more important than whether motion is actively or passively induced. High velocity movements that are prolonged or repeated are universally provocative whether encountered actively or passively. Responses to less intense movements vary among affected individuals. Most patients feel best when still, but others report that moving at a modest pace (e.g., walking or riding a bicycle) is more tolerable than remaining stationary when upright [51].

4.1.3.3. Visual stimuli

Challenging visual stimuli may be moving or stationary. Environments that contain full field visual flow (e.g., passing traffic, milling crowds), large complex patterns (e.g., busy carpeting, large store displays), or wide-open spaces with distant or indistinct visuospatial
reference points (e.g., large fields, warehouses, atria) are most troublesome [22-27,44]. Exposure to full field visual stimuli may exacerbate symptoms for hours, even after brief exposures. Smaller visual targets are generally less troublesome unless sustained attention to them is required. Performing tasks that require continuous visual focus (e.g., reading a book, using a computer, or watching television) may exacerbate symptoms even when patients are sitting still [29,44]. The increasing need in the modern world to view information on electronic screens is a bane for many patients with PPPD.

4.1.4. Clinical course

In most cases, PPPD develops as the acute symptoms of precipitating events remit [51]. Patients do not experience symptom-free intervals. Instead, as their acute vertiginous symptoms fade, they develop the characteristic chronic symptoms of PPPD. In other cases, PPPD has a stuttering onset. Patients may experience PPPD-like symptoms lasting days to weeks, until recurrences settle into a persistent course. This stuttering course is more likely when precipitants are short-lived, recurrent events (e.g., attacks of BPPV, migraine, or panic). Least often, PPPD has a gradual onset. Precipitants such as generalized anxiety, autonomic disorders, and degenerative diseases of the peripheral vestibular apparatus or cerebellum are chronic conditions that may develop slowly. With these triggers, symptoms of PPPD may appear gradually and worsen slowly.

Specific precipitants of PPPD cannot be identified in all patients, especially those who have been symptomatic for many years and lack adequate documentation of their initial clinical presentations. Even in these situations, however, most patients describe an acute, subacute, or stuttering onset of illness. Patients with clinical histories of gradually worsening chronic vestibular symptoms or balance problems that have no identifiable
starting points, particularly those without generalized anxiety or autonomic problems, are less likely to have PPPD than those with more definitive onsets of illness. They warrant prospective observation over a period of several months to verify the diagnostic picture and properly screen for other diseases, especially slowly emerging degenerative disorders.

4.2. Possible variants or subtypes of PPPD

Commonalities among PPV, SMD, VV, CSD, and the syndromes that predated them formed the basis of the diagnostic criteria for PPPD. However, semiologic similarities, even those stretching over more than a century, do not necessarily mean that PPPD is a single entity. The current state of scientific knowledge left two important questions unanswered: (1) Is PPPD a single disorder with one principal pathophysiologic process or is it the common manifestation of multiple conditions that produce similar symptoms from different pathophysiologic mechanisms? (2) If PPPD is a single disorder, does it have clinically meaningful and validly distinguishable subtypes?

4.2.1. A single disorder or a group of related conditions?

The precipitating factors of PPV [16], SMD [23], VV [25], and CSD [29] span a variety of neuro-otologic, other medical, and psychological events. It is not known if these act through one pathophysiologic process to produce a single syndrome or via separate mechanisms that share enough of a final common pathway to generate symptoms and susceptibilities that closely resemble one another. The former situation would be akin to posttraumatic stress disorder in which a wide variety of traumatic experiences produce one syndrome with four clusters of symptoms that can be provoked by internal and external factors [8]. The latter circumstance would be like hypertension in which
chronically elevated blood pressure can arise from multiple diseases that produce sustained increases in intravascular pressure via different physiologic mechanisms.

4.2.2. Do subtypes exist?

Although PPV, SMD, VV, and CSD share a number of features, they also have different areas of emphasis (Table 1). Postural provocation is a distinguishing feature of PPV [16,17]. It was not part of the original description of CSD [29], but was added later [45]. Difficulty with self-motion is part of PPV [16,17], SMD [23], and CSD [29,44]. Trouble with visual motion stimuli is the primary feature of VV [25-27] and is emphasized in SMD [22-24] and CSD [29,44]. Thus, PPV, SMD, VV, and CSD may reflect different perspectives on a single, multifaceted clinical entity or they may offer insights into potentially distinguishable subtypes of PPPD (e.g., posturally predominant subtype, visually predominant subtype). Mild anxiety and depressive symptoms and phobic behaviors were included in the descriptions of PPV [16,17], but were considered comorbidities of CSD [44,51,53]. This raises the possibility that PPV may be either a distinct phobic subtype of PPPD or encompass PPPD plus a specific phobia of dizziness-related experiences.

In the absence of definitive scientific data on these possible subtypes, the Behavioral Subcommittee chose to define only PPPD for the ICVD, but allow for clinical variability as reflected in Criterion B. The World Health Organization permits related terms to be listed in the ICD-11. Therefore, PPV, SMD, VV, and CSD were retained as index terms for cross-referencing in the ICD-11 beta draft definition of PPPD [50].

4.2.3. Probable (subthreshold) PPPD

Members of the subcommittee concluded that there were not enough published data
to define a clinically meaningful probable or subthreshold version of PPPD. Clinical experience with PPV and CSD suggests caution in applying the diagnosis of PPPD to patients who do not fulfill all of its diagnostic criteria.

4.3. Making a diagnosis of PPPD

The diagnosis of PPPD is made by gathering clinical history relevant to Criteria A-D. Data from physical examinations and clinically indicated diagnostic testing help to determine if PPPD is the best diagnosis, either alone or in combination with other diseases or disorders (Criterion E). PPPD is not a diagnosis of exclusion [17,44,51]. It should not be given to patients who report only non-specific chronic vestibular symptoms or those who have enigmatic complaints that do not fulfill its definition. In such cases, prospective monitoring may provide the clinical evidence needed to verify or exclude the diagnosis.

4.4. Differential diagnosis

The differential diagnosis of PPPD includes chronic sequelae of acute precipitants, recurrent attacks of episodic precipitants, ongoing manifestations of chronic precipitants, other chronic vestibular syndromes, medical or psychiatric disorders that produce persistent unsteadiness or dizziness, and adverse effects of regularly consumed prescription or non-prescription medications [16,25,29,51].

4.4.1. Chronic sequelae of acute precipitants

Some precipitants of PPPD are acute disorders that have the potential for chronic symptomatic complications (e.g., vestibular neuritis or stroke leading to persistent uncompensated vestibulopathies). For these disorders, the diagnostic question is whether
patients’ presenting symptoms are due to PPPD alone, chronic manifestations of its precipitants, or both [51]. This diagnostic dilemma is resolved by careful attention to the clinical history and assessment of patients’ compensation status. A history of persistent non-vertiginous dizziness and unsteadiness provoked by upright posture, patients’ own movements, and exposure to visual motion stimuli plus physical exam and laboratory evidence of good compensation (e.g., no spontaneous nystagmus or abnormal responses to head thrust, headshake, or stepping tests) indicates that PPPD is the only active diagnosis. In contrast, the presence of ongoing episodes of head motion-provoked vertigo or unsteadiness and exam findings of incomplete compensation without persistent dizziness argues against PPPD. A third possibility is the combination of persistent dizziness and motion sensitivity plus head motion-provoked symptoms and exam findings of incomplete compensation, which would indicate coexisting PPPD and uncompensated vestibulopathy.

4.4.2. Recurrent attacks of episodic precipitants

PPPD may be triggered by episodic vestibular disorders such as vestibular migraine [46], BPPV [47], and Menière’s disease [48] that cause distinct bouts of vestibular symptoms in contrast to the persistent, waxing and waning dizziness, unsteadiness and non-spinning vertigo that are hallmarks of PPPD. When PPPD co-exists with these disorders, proper diagnosis rest on identifying the characteristic symptoms of each active disorder. Episodic disorders add distinctive vestibular symptoms to the background of PPPD [54], such acute attacks of vertigo plus cephalalgia, photophobia and phonophobia, with or without visual aura for vestibular migraine [46], short-lived positional vertigo for BPPV [47], or attacks of vertigo, tinnitus, and fluctuating hearing for Menière’s disease [48].
4.4.3. **Ongoing manifestations of chronic precipitants**

Some precipitants of PPPD are chronic conditions themselves (e.g., chronic anxiety and depressive disorders, post-concussive syndrome, autonomic disorders, and heart diseases). They may cause persistent unsteadiness or dizziness with or without triggering PPPD. When present alone, they are not as greatly affected by the motion provocations of Criterion B as is PPPD. The strategy for differential diagnosis in these cases is to determine if Criteria A-D for PPPD are present and evaluate key elements of clinical histories, physical examinations, and laboratory testing to determine which conditions explain the patients’ symptoms best (i.e., PPPD alone, precipitating events alone, or both) (Criterion E).

4.4.3.1. **Chronic anxiety and depressive disorders**

Chronic anxiety due to generalized anxiety disorder, agoraphobia, social phobia, obsessive compulsive disorders, and traumatic stress disorders may manifest with persistent dizziness [8,9]. Depressive disorders also may cause dizziness [55]. These disorders are diagnosed according to the latest versions of the International Classification of Diseases [9] or Diagnostic and Statistical Manual of Mental Disorders [8]. However, in neurologic, otologic, and primary care settings where patients with vestibular symptoms are most commonly encountered, simple self-report questionnaires offer a valid and efficient means of detecting psychiatric morbidity. The 7-item Generalized Anxiety Disorders Scale (GAD-7) may be used to screen for pathological anxiety [56]. The 9-item Patient Health Questionnaire (PHQ-9) may be used to screen for depression [57]. The 14-item Hospital Anxiety and Depression Scale (HADS) covers both anxiety and depressive symptoms [58]. Positive results indicate that an anxiety or depressive disorder is likely, either as the cause of vestibular symptoms or co-existing with PPPD [17,29,44,53].
A history of troubling life circumstances or recent stressful events cannot be used as evidence for or against the presence of functional or psychiatric diagnoses, including PPPD [51]. A recent study found that childhood and adulthood adversity were equally prevalent in patients with structural versus functional or psychiatric causes of vestibular symptoms [59]. Furthermore, panic attacks and generalized anxiety disorder frequently occur in the absence of identifiable stressors [8,9]. Therefore, a diagnosis of PPPD rests on fulfillment of its diagnostic criteria, regardless of patients’ histories of adversity.

4.4.3.2. Postconcussive syndrome

Patients with postconcussive syndrome following a traumatic brain injury or whiplash often experience chronic dizziness in addition to headache, insomnia, cognitive symptoms, and mood lability [60]. Patients who fulfill all of the diagnostic criteria for PPPD after a traumatic brain injury or whiplash should receive the diagnosis. The presence or absence of other sequelae of injury will determine if additional diagnoses are warranted. On the other hand, patients who complain of chronic dizziness after head injury should not be given a diagnosis of PPPD if they do not manifest Criteria A-D.

4.4.3.3. Autonomic disorders

Autonomic disorders frequently cause dizziness. The autonomic disorders most like to trigger PPPD, based on research from CSD [61], are type 1 neurocardiogenic (vasovagal) syncope and postural orthostatic tachycardia syndrome, which are most often encountered in adolescents and young adults. Orthostatic intolerance with or without hypotension from neurologic and cardiovascular illnesses (e.g., autonomic neuropathy) is more common in older adults, in whom it may be part of multi-factorial dizziness. Patients with autonomic
disorders tend to have more pronounced orthostatic and exertional dizziness than those with PPPD [61]. However, symptoms overlap considerably, so the differential diagnosis depends on examination of autonomic integrity. PPPD does not cause abnormal changes in heart rate or blood pressure. Autonomic disorders do not create difficulties with complex or moving visual stimuli in patients who are sitting still. Thus, vital signs during autonomic challenges and sensitivity to visual stimuli when seated at rest best distinguish PPPD from autonomic disorders, recognizing the potential for the two problems to co-exist.

### 4.4.4 Other chronic vestibular syndromes

Chronic vestibular syndromes in the differential diagnosis of PPPD include bilateral peripheral vestibulopathy [62], neurodegenerative disorders (e.g., downbeat nystagmus syndrome, cerebellar diseases) [63, 64], and mal de debarquement syndrome (MdDS) [65].

#### 4.4.4.1 Bilateral peripheral vestibulopathy

Bilateral peripheral vestibulopathy is best distinguished from PPPD by its characteristic findings on physical examination and laboratory testing [62], such as bilaterally positive head thrusts and diminished responses to caloric stimulation. Clinical history may offer additional clues, but these are not as definitive. For example, PPPD does not cause oscillopsia, but oscillopsia is present in only 30-40% of patients with bilateral peripheral vestibulopathy. Individuals with bilateral vestibulopathy typically have less trouble with complex visual stimuli when sitting still than patients with PPPD, but this symptom varies among patients with both disorders.

#### 4.4.4.2 Chronic neurologic disorders
Neurodegenerative disorders that affect posture and gait such as Parkinson’s disease, cerebellar degeneration [64], and downbeat nystagmus syndrome [63] may manifest with dizziness or unsteadiness when standing or walking before motor signs can be detected on physical examination. The gradual onset of these complaints in the relative absence of difficulties with complex or moving visual stimuli should arouse suspicion that PPPD is not the correct diagnosis. The best approach in this situation is a period of prospective monitoring (typically 6-12 months) with a symptom log kept by the patient and serial examinations performed by the clinician before making a definitive diagnosis.

Bilateral peripheral neuropathy of the feet and orthostatic tremor [66-68] may cause or contribute to dizziness and unsteadiness when patients are upright, though neither one causes trouble with complex or moving visual stimuli. Peripheral neuropathy is much more likely to manifest with sensory loss and pain, or present as one part of a multi-factorial picture, than to be the sole cause of dizziness. Orthostatic tremor is diagnosed by identifying its characteristic 13-18 Hz tremor in the lower legs on electromyographic or posturographic testing [66-68].

4.4.4.3. Mal de debarquement syndrome

Mal de debarquement syndrome (MdDS) [65] is a condition of persistent unsteadiness triggered by traveling on boats, aircraft, or automobiles, usually for at least a few hours. Symptoms characteristically decrease during passive motion (e.g., riding in a car) and then increase again when motion ceases. That is opposite the pattern found in most patients with PPPD, though a minority of individuals with PPPD experience temporary decreases in symptoms during modest motion, such as walking at a medium pace or riding a bicycle on a smooth path. A “spontaneous onset” version of MdDS has been
described, though most patients in those reports had migraine or anxiety disorders [65], which are known precipitants of PPPD. A major difference between MdDS and PPPD is the effect of treatment. MdDS improves very little with medications or vestibular habituation, whereas treatment studies of PPV [69], VV [70,71], CSD [28], and PPPD, itself [72], showed significant improvements with serotonergic antidepressants or vestibular habituation.

4.4.5. Adverse effects of medications

Prescription medications, over the counter preparations, and dietary supplements may cause dizziness, unsteadiness, and vertigo. Vestibular symptoms caused by newly administered medications or changed doses of existing medications may trigger PPPD.

4.4.6. Other functional forms of vestibular symptoms

Clinicians may encounter patients who describe persistent vestibular symptoms that do not fit the diagnostic criteria of either PPPD or other well-defined chronic vestibular syndromes [51]. Examples include constant, invariant vertigo, unsteadiness, or dizziness, complex body motions in multiple directions simultaneously, and kaleidoscopic swirling movements of large portions of the visual field. Patients often report a lack provoking or mitigating factors. These clinical presentations have not been studied systematically, but their continuous nature, unwavering intensity, and abnormal complexity distinguish them from the episodic or fluctuating symptoms reported by patients with structural deficits, PPPD, and anxiety or depressive disorders that cause vestibular symptoms [51]. In many patients, these functional forms of vestibular symptoms are accompanied by other chronic physical complaints such as fatigue and pain, raising the possibility that they are but one manifestation of a broader somatic symptom
4.4.7. *Gait disorders, falls, and near falls*

Patients with PPPD may report sensations of veering from side-to-side when walking. On exam, they may exhibit a mildly slow or cautious gait. One study of walking mechanics found that patients with PPV walked slower, had reduced stride length, and spent a greater fraction of time with both feet on the ground than healthy subjects [39]. These changes correlated with reduced balance confidence. A case study distinguished gait and posture symptoms of CSD from those of functional gait disorders [74]. Falls and near falls have never been a part of PPV or CSD [51]. Therefore, clinical evidence of significant changes in gait or recurrent falls or near falls indicates the presence of a structural or functional gait disorder. PPPD may co-exist with these disorders.

4.5. *Epidemiology*

No epidemiologic studies are available for PPPD, but its prevalence and incidence may be estimated from research done on patients with PPV, VV, CSD, and chronic dizziness following acute vestibular syndromes [17,29,41,75-77].

4.5.1. *Estimates of the prevalence of PPPD*

Clinical epidemiologic data from tertiary care centers with special interest in PPV [16] and CSD [29] showed their prevalence to be 15-20% among all patients presenting for evaluation of vestibular symptoms, making them the most common diagnoses among young adults and the second most common among all adults, trailing only BPPV. The average duration of illness at the time of tertiary consultation was 4.5 years with some
patients experiencing symptoms for decades [16,29]. Disability varied widely from individuals who had few limitations in daily functioning to those who were severely impaired and unable to work. The average age of patients presenting for evaluation of PPPD is the mid-40s, with a range from adolescence to late adulthood [29,78,79]. A female predominance has been reported in the first clinical reports on PPPD [78,79].

4.5.2. Estimates of the incidence of PPPD

The incidence of PPPD following neuro-otologic triggers may be estimated from studies that followed patients prospectively after bouts of acute or episodic vestibular disorders (e.g., vestibular neuritis, BPPV, vestibular migraine, Menière’s disease) [75-77]. These investigations found PPPD-like chronic dizziness [75-77] or persistent VV [41] in about 25% of patients after 3-12 months of follow-up, despite otherwise adequate compensation or recovery from the initial illnesses. These results indicate that PPPD is likely to develop in a significant proportion of patients afflicted with acute or episodic vestibular syndromes. Similar prospective studies of clinical outcomes following other medical and psychological precipitants of PPPD have not been conducted. However, retrospective investigations found that the course [53] and treatment response [80] of patients with CSD triggered by anxiety disorders mirrored that of patients with CSD triggered by acute vestibular syndromes, suggesting that the clinical course of PPPD may be similar regardless of precipitant. A long-term follow-up study of patients with PPV found that only a minority experienced spontaneous resolution of symptoms [19]. Most had a chronic waxing and waning course and three-quarters developed anxiety or depressive comorbidity. Thus, the majority of patients with PPPD are likely to remain symptomatic without treatment, regardless of initial precipitant.
The incidence and prevalence of PPPD in primary care practices and the general population are not known as detailed epidemiologic studies of PPV, SMD, VV, and CSD have not been conducted in those settings.

4.6. Possible pathophysiologic processes underlying PPPD

Investigators studying PPV, CSD, SMD, and VV have identified pathophysiologic processes that may be applicable to PPPD, including anxiety-related personality traits as a possible risk factor [79,81,82] and high levels of anxiety and vigilance about acute symptoms during triggering events as initial pathologic responses [41,75-77]. Alterations in postural control strategies [34-40], shifts in multi-sensory integration [41], and reduced cortical integration of spatial orientation information with threat assessments may be sustaining mechanisms [42,43]. All of these will have to be studied in greater detail in patients meeting the specific diagnostic criteria for PPPD.

4.6.1. Possible risk factors

Brandt and Dieterich described obsessive compulsive personality traits in their original patient cohort with PPV [16]. Subsequent studies found that individuals with the anxiety-related personality traits of neuroticism and introversion [81] had an increased risk for CSD. High neuroticism also was identified in a report on PPPD [79]. In contrast, persons demonstrating resilience, optimism, and beliefs that life is meaningful and manageable had a reduced risk of persistent dizziness after acute vestibular events [82]. Patients with family or personal histories of anxiety disorders pre-dating the onset of vestibular symptoms had an increased risk of developing persistent dizziness [77] or CSD [53] after triggering events. In patients with anxiety disorders, a history of previous
vestibular deficits was associated with SMD [22,23]. These studies suggest that anxiety-related personality traits or a personal or family history of anxiety disorders may be risk factors for developing PPPD following relevant precipitants.

4.6.2. Initial pathologic reactions

Three prospective studies found that high anxiety about dizziness during and after bouts of acute vestibular neuritis or BPPV predicted continued dizziness three [75] and twelve [76] months later. These initial psychological responses had far greater effects on long-term outcomes than the initial or subsequent states of patients’ peripheral vestibular functioning or vestibulo-ocular reflexes. Furthermore, patients with emerging symptoms of CSD who were treated with three sessions of cognitive behavioral therapy started within 8 weeks of precipitating events had marked reductions in dizziness and avoidance of provocative circumstances [83], benefits that endured at follow-up six months later [84]. Collectively, these data raise the possibility that a highly anxious response to triggering events may be the pivotal initial pathophysiologic process in the development of PPPD, and that early symptom-specific interventions might counter this effect.

4.6.3. Possible alterations in postural control

Several investigations showed that patients with PPV manifested an alteration in postural control characterized by high frequency, low amplitude postural sway related to co-contraction of lower leg muscles when standing at rest [34,38]. One study of patients with CSD demonstrated similar results [40]. Normal people used this high demand postural control strategy in challenging balance situations such as standing at heights [85-87]. Patients with PPV adopted this strategy during less demanding tasks than normal
individuals [35-37], possibly related to a lower threshold for engaging closed loop feedback mechanisms to adjust posture [38]. Brandt et al. [88] reported the case of a patient who was followed prospectively from a bout of acute vestibular neuritis to the development of PPV. The transition to chronic symptoms coincided with emergence of the high frequency, low amplitude sway pattern of PPV. Future studies will have to measure the prevalence of this postural control strategy among patients with PPPD and determine its association with clinical characteristics of the disorder, particularly postural symptoms.

4.6.4. Possible association with visual dependence

Bronstein and colleagues [25-27] showed that people with VV manifest visual dependence, a trait-like tendency to rely on visual information for spatial orientation. In a prospective study, Cousins et al., [41] found that patients who had persistent dizziness for at least six months following bouts of acute vestibular neuritis had greater visual dependence than those who recovered without chronic symptoms. Future studies will have to measure the prevalence and severity of visual dependence in patients with PPPD and determine its association with clinical features of the disorder, particularly visual symptoms.

4.6.5. Possible changes in activity and connectivity of crucial brain regions

The first neuroimaging studies of patients with CSD [42] and PPPD [43] were completed recently. The first study [42] measured the activity and connectivity of vestibular, visual, and anxiety-related regions of the brain using functional magnetic resonance imaging (fMRI) in response to sound-evoked vestibular stimulation in patients with CSD versus normal control subjects matched for anxiety-related personality traits.
Patients with CSD showed reduced stimulus-related activity in the parieto-insular vestibular cortex (PIVC), anterior insula, inferior frontal gyrus, hippocampus, and anterior cingulate cortex compared to normal individuals. They also had more negative connectivity between the PIVC and the anterior insula, anterior cingulate cortex, and hippocampus, as well as between the anterior insula and middle occipital cortex. The second study [43] compared women with PPPD to women who had recovered without sequelae from illnesses that caused acute vestibular symptoms. Using fMRI, the authors found that women with PPPD had less activation of the amygdala and anterior cingulate cortex and greater activation of the left angular gyrus in response to the non-motion stimulus of standardized pictures designed to elicit negative emotions. These early results suggest that brain areas responsible for high level spatial orientation, multi-sensory integration, and threat assessment may not be as active or well connected in patients with PPPD as in normal people, potentially leaving lower level posture and gaze control mechanisms poorly integrated with one another. These findings await confirmation in larger studies with sufficient power to control for potential confounds and measure associations with clinical aspects of PPPD.

5. Additional commentary

The consensus process for decision-making seeks a result that has broad support. It does not require unanimity, but demands more than a simple majority [89]. Were the latter sufficient for developing diagnostic criteria or writing treatment guidelines, then informative aspects of the minority opinion could be lost to practicing clinicians, the scientific community, and general public. Though not all agree [90], arguments for publishing minority opinions in such endeavors have been made [91].
Members of the Behavioral Subcommittee and their advisors reached consensus about the diagnostic criteria for PPPD and its relationship to SMD, VV, and CSD, but not PPV. SMD and VV were seen as complex symptoms, not stand alone diagnostic entities. They informed criterion B of PPPD, but are known to occur in other situations [23,25]. The definition of PPPD paralleled work in other areas of medicine where functional disorders (e.g., the functional gastrointestinal disorders [33,92] and fibromyalgia [93,94]) were defined by their core physical symptoms, and not associated psychological features. CSD was a step in that direction based in part on the physical features of PPV [29]. However, the definition of PPPD is better supported; hence, PPPD now fully supplants CSD.

Members of the subcommittee and their advisors carefully considered a proposal to include PPV as a subtype of PPPD. Arguments in favor of this idea focused on the richness of the definition of PPV, which includes personality factors and psychological symptoms that were shown to affect clinical presentation [17,39], as well as 30 years of research that described its clinical course [17-21] and identified potential pathophysiologic mechanisms [35-39]. Arguments against the idea included concerns that the mix of physical and psychological symptoms and personality traits that constitute PPV is difficult to operationalize in clinical practice [44] and may have to be updated based on newer research [81,82], and that including PPV as a subtype of PPPD without validating their relationship could perplex clinicians and investigators. In the end, a simple majority of subcommittee members voted against including PPV as a subtype of PPPD.

The debate about PPV highlighted the fact that PPPD is a dynamic condition as illustrated in Figure 1. It is a functional disorder, although structural and psychological factors affect its development. Clinicians who apply Criteria A and B as simple checklists of symptoms without capturing the dynamic features of their patients’ histories (Criterion C)
assume a reductionist view of the diagnosis, potentially missing important aspects of patients’ morbidity (Criterion D) and nuances of the differential diagnosis (Criterion E). Scientists who fail to consider the dynamics of PPPD risk muddling rather than illuminating the nature of the disorder and its pathophysiologic mechanisms.

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| Table 1. Features of PPV, SMD, VV, and CSD that informed the definition of PPPD |
|---------------------------------|-----------------|-----------------|-----------------|-----------------|
| **Primary Symptoms (criteria A.1-3)** |                 |                 |                 |                 |
| Dizziness                       | ✓               | ✓               | ✓ [27,41]       | ✓               |
| Unsteadiness                    | ✓ ✓             | ✓ ✓             | ✓ ✓             | ✓ ✓             |
| Non-spinning vertigo            | ✓ ✓             | ✓ ✓             | ✓ ✓             | ✓ ✓             |
| Temporal profile (Criteria A.1-3) |                 |                 |                 |                 |
|                                 | Fluctuating with momentary flares | Situational (provoked) | Situational (provoked), Persistent [41] | Persistent with diurnal variability [53] |
| Provocative factors (Criteria B.1-3) |                 |                 |                 |                 |
| Upright posture                 | ✓ ✓             |                 |                 | ✓ [45]          |
| Active or passive motion        | ✓               | ✓               |                 | ✓               |
| Moving visual stimuli or complex patterns | ✓               | ✓               | ✓ ✓             | ✓ ✓             |
| Precipitants (Criterion C.1)    |                 |                 |                 |                 |
| Vestibular syndromes            | ✓               | ✓               | ✓               | ✓               |
| Other medical illnesses         | ✓               |                 |                 | ✓               |
| Psychological distress          | ✓               |                 |                 | ✓               |
| Course of illness (Criteria C.1.a-b) |                 |                 |                 |                 |
|                                 | Long-standing, waxing/waning [18] | May be long-standing | May be long-standing | Chronic |
| Physical exam and laboratory findings (Criterion E) |                 |                 |                 |                 |
|                                 | Normal          | Somatosensory dependence on posturography [22] | Central or peripheral vestibular deficits | Abnormalities related to comorbid conditions [45] |
| Features not incorporated into PPPD |                 |                 |                 |                 |
| Anxiety                         | Part of PPV     | Associated with SMD [22] | Associated with prolonged VV [41] | May be comorbid with CSD [30] |
| Depression                      | Part of PPV     |                 |                 | May be comorbid with CSD [30] |
| Personality traits              | Obsessive-compulsive traits are part of PPV |                 |                 | Neurotic, introverted traits may be risk factors for CSD [80] |
Figure Legend

Figure 1. Putative mechanisms of PPPD.

PPPD is thought to develop via a dynamic process (arrow). In about 70% of patients, a structural vestibular syndrome (e.g., vestibular neuritis, BPPV) or other medical condition precipitates PPPD (black dot, a) [26]. Individuals who respond to the triggering event with a high level of anxiety and body vigilance appear likely to progress to PPPD (i.e., to traverse the arrow from the initial structural event through this transient psychological stage to the chronic functional disorder) [39,73,74]. Anxiety-related personality traits or pre-existing anxiety disorders appear to increase the risk of developing PPPD [51,67,75]. In about 30% of patients, PPPD begins with acute psychological distress (black dot, b) and then progresses to the functional disorder [26]. PPPD may co-exist with structural or psychological illnesses [42], placing patients in the intersections of the functional and psychological or structural ellipses. Anxiety-related personality traits and psychological symptoms (check marks, ✓) are incorporated into PPV [15,16], whereas they are
considered predisposing factors and comorbid symptoms, respectively, in PPPD.

PPPD = Persistent postural-perceptual dizziness

PPV = Phobic postural vertigo