Bilateral Vestibulopathy (BVP):

A Vestibular Physician's Deep Review of Pathophysiology, Diagnosis, and Management

Vestibular Medicine for Vestibular Physicians

Peripheral Vestibular Pathology — Module 2.4

Australian Dizziness Clinics | www.AustralianDizzinessClinics.com
Version 1.0 | May 2026

How to Use This Review

This literature review forms part of the Vestibular Medicine for Vestibular Physicians series published by the Australian Dizziness Clinics Education Hub. It is written for vestibular physicians, neuro-otologists, advanced ENT trainees, and vestibular physiotherapists working at the deep end of peripheral vestibular practice, where a working command of mechanism, criteria, and atypical presentations is expected rather than optional.

The review is dense by design — intended as a 30–40 minute deep read or a desktop reference. It is supported by an A4 clinician cheat sheet, short-form clinician videos, audio episodes, and a patient information leaflet within the same Education Hub module.

Callout Box Guide

□ **Key Point:** Foundational concepts and summary statements that anchor the core clinical content of each section.

□ **Clinical Insight:** Clinically relevant observations for direct application in assessment and management.

□ **Clinical Pearl:** High-yield memorable clinical points — the take-home messages most likely to change practice.

□ **Important:** Red flags, atypical presentations, and critical safety points requiring escalation or imaging.

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I. Introduction and Epidemiology

Historical Background

Bilateral vestibulopathy (BVP) — historically termed bilateral vestibular failure (BVF), Dandy's syndrome, or bilateral labyrinthine loss — refers to a clinically defined syndrome of chronic bilateral reduction or absence of vestibular end-organ or nerve function [1]. The condition's clinical recognition dates to the late 19th century. In 1882, William James noted vestibular dysfunction in deaf-mute individuals, associating labyrinthine damage with imbalance [3]. Robert Bárány, in 1907, documented bilaterally reduced caloric responses in similar patients, laying the foundation for objective vestibular assessment [3]. The most influential early description came from neurosurgeon Walter Dandy in 1941, who observed oscillopsia and severe gait instability in patients after deliberate bilateral vestibular nerve section for intractable Ménière's disease [3,5].

Over subsequent decades, the understanding of BVP expanded substantially. In 1965, oscillopsia during head movement was formally identified as a defining symptom of bilateral vestibular loss [3]. Baloh et al. defined idiopathic bilateral vestibulopathy in 1989 [38], characterising patients with chronic imbalance and oscillopsia in the absence of other neurological deficits [1]. A pivotal neuroscience contribution came in 2005, when Brandt and colleagues demonstrated hippocampal atrophy and spatial memory impairment in patients with chronic bilateral vestibular loss, revealing far-reaching cognitive consequences of bilateral deafferentation [7]. The CANVAS syndrome (Cerebellar Ataxia, Neuropathy, and Vestibular Areflexia Syndrome) was formally described in 2011, widening the clinical spectrum of BVP to include multimodal neurodegenerative presentations [8]. Current consensus terminology, adopted by the Bárány Society Classification Committee in 2017, standardises the term bilateral vestibulopathy as the preferred designation [1].

□ **Key Point:** The current preferred term is bilateral vestibulopathy (BVP), replacing older terms such as 'bilateral vestibular failure' or 'Dandy's syndrome'. The 2017 Bárány Society criteria provide the internationally recognised diagnostic standard [1].

Incidence and Prevalence — General Population

Bilateral vestibulopathy is a relatively rare disorder, though likely under-recognised in clinical practice. The 2013 U.S. National Health Interview Survey analysis by Ward et al. estimated the prevalence of bilateral vestibular hypofunction at approximately 28 per 100,000 adults, with some population-based studies reporting figures up to 81 per 100,000 [2,3,32]. This wide range reflects heterogeneity in diagnostic criteria and variable access to quantitative vestibular testing. Prevalence increases with age: one study of adults over 65 years identified bilateral vestibular [43] impairment meeting diagnostic criteria in approximately 9% [13]. Sex distribution is roughly equal, with some analyses suggesting marginally higher prevalence in women and in Hispanic populations compared to non-Hispanic groups [2,9].

In specialised dizziness and neuro-otology clinics, BVP constitutes approximately 1–4% of all patients evaluated [4,6]. Propper et al. (2022) identified 103 confirmed cases over a 10-year period among approximately 11,000 patients at a tertiary Dutch dizziness centre — representing approximately 0.9–1% of the cohort [4]. High-volume vestibular physicians report bilateral hypofunction in up to 5% of their patient panels, reflecting the referral enrichment seen in tertiary academic settings [22].

Paediatric Epidemiology

BVP is rare in children, typically accompanying congenital or syndromic conditions. A review of 832 children referred for balance disorders identified 26 cases of bilateral vestibular hypofunction — approximately 3% of that cohort [12]. Approximately 80% of affected children have co-existing bilateral sensorineural hearing loss, reflecting shared cochleo-vestibular aetiology (Usher syndrome, Mondini dysplasia, congenital rubella, meningitis sequelae) [12]. Unrecognised BVP in childhood manifests as delayed motor milestones, including late independent walking (often beyond 18 months), and chronic difficulty with balance and sports participation.

Table 1. Epidemiology of Bilateral Vestibulopathy by Population Subgroup.

Population	Estimated Prevalence	Key Features / Notes
General adult population	28–81 per 100,000	Likely under-recognised; wide range reflects diagnostic variability

		[2,3]
Adults >65 years	~9% meet criteria	Overlap with presbyvestibulopathy; age-adjusted norms required [13]
Specialised dizziness clinic	1–4% of patients	Tertiary referral enrichment; up to 5% in high-volume vestibular practices [4,6,22]
Paediatric balance clinic	~3% of referrals	80% have co-existing SNHL; congenital/syndromic aetiology predominates [12]
CANVAS-associated BVP	Up to 10–20% of idiopathic BVP	RFC1 biallelic repeat expansion; neuropathy and cerebellar ataxia co-present [8,14]

Aetiology and Causative Mechanisms

The aetiology of BVP is heterogeneous and determines management priorities. The most common identifiable cause in adults is aminoglycoside ototoxicity, followed by genetic mutations and idiopathic causes; in children, genetic and post-infectious aetiologies dominate. Approximately 30–50% of adult cases remain idiopathic after thorough investigation. CANVAS syndrome (biallelic RFC1 AAGGG intronic repeat expansion) is increasingly recognised and warrants dedicated genetic workup in adults with co-existing cerebellar ataxia or peripheral neuropathy [35,36]. Table 2 summarises the major aetiological categories with estimated prevalence proportions in adult and paediatric cohorts.

Table 2. Aetiology of Bilateral Vestibulopathy by Category — Adults and Paediatric.

Category	Adults	Paediatric	% Adult	% Paediatric
Ototoxic / Drug-Induced	Aminoglycosides (gentamicin, streptomycin); cisplatin; high-dose loop diuretics; salicylates; anti-epileptics; heavy alcohol; industrial solvents [20–23].	Aminoglycosides in NICU (neonatal sepsis); chemotherapy for childhood cancer; usually bilateral hearing loss co-present [6,21].	~15%	~20%
Genetic (Hereditary)	DFNA9 (COCH gene), DFNA11, RFC1 (CANVAS); autosomal dominant progressive vestibulocochlear loss in midlife [24].	Usher syndrome type 1, CHARGE, Waardenburg, Alport, EVA, Turner syndrome [25,26]; delayed motor milestones.	~15–20%	~30%
Infectious	Bilateral vestibular neuritis; sequential Ménière's disease; neurosyphilis; Lyme disease; bilateral herpes zoster oticus [5,7].	Post-meningitic labyrinthitis ossificans (leading childhood cause) [29,30]; congenital CMV; bilateral otitis interna [31].	~10–15%	~30%
Autoimmune	Autoimmune inner-ear disease (AIED); Cogan's syndrome; SLE, Sjögren's, vasculitis [32,33].	Juvenile autoimmune disorders rare; Bickerstaff encephalitis may mimic BVP.	~3–5%	<5%
Degenerative / Neurologic	CANVAS syndrome (RFC1 biallelic AAGGG expansion); spinocerebellar ataxias; MSA; presbyvestibulopathy [4,5,35,36].	Ataxia-telangiectasia; other neurodegenerative conditions — usually other deficits dominate.	~3–5%	<1%
Tumour (bilateral)	Neurofibromatosis type 2 (NF2) — bilateral vestibular schwannomas; bilateral metastatic/leukaemic	NF2 in teenage years; leukaemic infiltration exceptional.	~2–3%	<1%

	infiltration (rare) [5].			
Trauma / Iatrogenic	Sequential labyrinthectomies or vestibular neurectomies; bilateral temporal-bone fracture; barotrauma [1,2].	Severe head injury; bilateral cochlear implant trauma [39].	~3–5%	~5%
Idiopathic	No identifiable cause in 30–50% of adults [5,14]; possible microvascular ischaemia or unrecognised toxicity; vestibular migraine proposed as risk factor [42].	True idiopathic very rare — most traceable to genetic or post-infectious aetiology.	~30–50%	<5%

□ Key Point: Approximately 30–50% of adult BVP cases are idiopathic. The most important identifiable causes are aminoglycoside toxicity, CANVAS syndrome (RFC1 mutation), and genetic disorders (e.g. DFNA9). Aetiology-directed workup — including RFC1 genetic testing and full medication/ototoxin history — is mandatory in all new BVP diagnoses and directly guides management and prognosis.

II. Pathophysiology — VOR Failure, Sensory Substitution, and Cortical Impact

The Bilateral VOR Deficit: Final Common Pathway

The vestibulo-ocular reflex (VOR) generates compensatory eye movements precisely equal and opposite to head rotation, maintaining stable retinal images during movement [1,2,5]. In BVP, bilateral loss of afferent input from the semicircular canals and otolith organs eliminates this reflex, producing retinal slip during every head movement — the mechanistic basis of oscillopsia [5,15]. The vestibulo-spinal reflex, which drives rapid postural muscle activation in response to unexpected head motion, is simultaneously disrupted, generating the characteristic broad-based gait and marked increase in postural sway [1,2,6]. Unlike unilateral vestibular loss, in which the intact contralateral labyrinth partially substitutes, bilateral loss offers no residual vestibular drive — compensation depends entirely on non-vestibular sensory systems [2,5].

□ **Key Point:** In bilateral vestibulopathy, the VOR is abolished bilaterally. The resulting gaze instability (oscillopsia) and postural instability are not amenable to vestibular compensation — the brain can only substitute with visual and proprioceptive inputs, which are insufficient under dynamic conditions [1,2,5].

Sensory Substitution and Central Plasticity

The central nervous system attempts to compensate through sensory reweighting and neural plasticity [2,5,6]. Visual and somatosensory inputs become progressively up-weighted, explaining why BVP patients perform disproportionately worse in low-light environments or on compliant surfaces where these alternative cues are degraded [1,2]. The cervico-ocular reflex and optokinetic reflex [35] partially substitute for the absent VOR at low head velocities [5]. Some patients develop predictive saccadic strategies — 'covert saccades' — that partially compensate for absent reflexive eye movement at high velocities, a phenomenon detectable on video head impulse testing as refixation saccades with apparently normalised VOR gain on bedside examination [1,6].

Crucially, these compensatory mechanisms are incomplete and velocity-dependent. At slow head velocities, visual substitution may provide near-adequate gaze stabilisation, but at velocities above approximately 2 Hz — encountered during normal walking, running, or vehicle travel — the visual system cannot respond quickly enough to eliminate retinal slip [5,15]. This is why many BVP patients report being relatively functional at rest yet profoundly impaired during locomotion, particularly on uneven terrain or in visually complex environments [2].

Cellular and Molecular Mechanisms

The final common pathway of BVP is loss of vestibular hair cell or primary vestibular neuron function on both sides [5,6]. Aminoglycoside antibiotics (particularly gentamicin and tobramycin) destroy type I and

type II hair cells in the crista ampullaris and macula via oxidative stress mechanisms — producing a dose-dependent, cumulative cochleo-vestibular toxicity that affects the basal cochlear turn and peripheral vestibular apparatus preferentially [5,16,17]. Cisplatin and other platinum-based chemotherapeutics produce similar ototoxic cochleo-vestibular damage. In autoimmune inner ear disease, circulating antibodies and cellular immune effectors target endolabyrinthine structures, causing progressive bilateral dysfunction [6,18]. Genetic causes such as COCH mutations (DFNA9) produce progressive fibrocyte degeneration and endolymph ionic dysregulation leading to mid-life onset bilateral vestibular and cochlear loss [14,19].

□ **Clinical Insight:** Aminoglycoside vestibulotoxicity may occur with cumulative systemic doses well within 'safe' therapeutic ranges, particularly in patients with renal impairment who accumulate higher perilymph concentrations. Baseline and periodic vestibular monitoring (vHIT) during prolonged aminoglycoside courses is strongly recommended but rarely performed in clinical practice [16,17].

Hippocampal Atrophy and Cognitive Impact

Beyond the reflexive motor consequences of bilateral deafferentation, accumulating evidence establishes that the vestibular system contributes substantially to spatial navigation and hippocampal function [7,20]. Brandt et al. (2005) demonstrated bilateral hippocampal volume reduction and impaired spatial memory in patients with chronic BVP compared to age-matched controls — changes proportional to the duration of vestibular loss [7]. Rodent bilateral labyrinthectomy models produce hippocampal cell loss and impaired place-cell firing, providing a mechanistic underpinning for the clinical observations [20]. Patients frequently report spatial disorientation, difficulty navigating familiar environments, and impaired spatial [33] memory — manifestations of hippocampal dysfunction that add substantially to the functional disability of BVP beyond the motor deficits alone [7,21].

III. Clinical Features and Examination Findings

Core Symptom Triad

The defining clinical syndrome of BVP comprises three interrelated features: (1) postural imbalance and gait unsteadiness, (2) oscillopsia during head movement or locomotion, and (3) absence of acute rotatory vertigo [1,2]. This triad — chronic unsteadiness without vertigo, combined with movement-dependent visual disturbance — distinguishes BVP from most episodic vestibular disorders and from central balance impairment [1,6].

Postural imbalance is typically worst in low-light environments or on compliant or uneven surfaces where the two remaining sensory inputs — vision and proprioception — are simultaneously degraded [1,2]. Patients adopt a cautious, broad-based gait, lose the ability to tandem walk (heel-to-toe), and may require wall-touching or a walking aid for stability at night [2,6]. The 'supermarket effect' — overwhelming unsteadiness and spatial disorientation in busy visual environments — is a characteristic complaint that reflects the dependence on clear visual cues for balance maintenance [2].

Figure 1. Oscillopsia in Bilateral Vestibulopathy *— Schematic representation of retinal slip and visual instability during head movement in bilateral vestibulopathy. With absent VOR, compensatory eye movements fail during locomotion, producing bouncing or blurred visual field perception — the hallmark symptom of bilateral vestibular loss.*

Source: Australian Dizziness Clinics — educational image for clinical illustration.

Oscillopsia

Oscillopsia — the perception of environmental oscillation or blurring during head movement — is the hallmark visual symptom of BVP [1,5]. Patients report that stationary objects appear to bounce, judder, or blur when walking or when the head is in motion, yet remain stable during complete head stillness. This dissociation between motion and rest, combined with the absence of spontaneous nystagmus or positional provocation, is diagnostically characteristic [1,2]. In severe cases, oscillopsia is elicited by minimal motion — including the head bounce associated with each walking step ('pedestrian oscillopsia') or even transmitted cardiac pulsations [5,15].

□ **Clinical Pearl:** The combination of (1) oscillopsia that appears only during head movement, (2) marked worsening of imbalance with eyes closed or in darkness, and (3) no spontaneous vertigo or nystagmus constitutes the clinical fingerprint of bilateral vestibulopathy. If all three are present, the diagnosis should be confirmed with quantitative vestibular testing [1,2].

Absence of Vertigo and Nystagmus

True rotatory vertigo is typically absent in established BVP because symmetric bilateral loss produces no tonic imbalance of vestibular activity — the driver of the sensation of rotation [1,2]. Spontaneous nystagmus is also absent in pure BVP for the same reason. Patients who present with episodic vertigo in the context of BVP usually have an underlying episodic cause driving the original bilateral loss (e.g. bilateral Ménière's disease, autoimmune inner ear disease) — the vestibular loss in these cases is the end-result of repeated episodic destruction rather than the acute presentation [2,6].

Examination Findings

On structured vestibular examination, the bilaterally abnormal video head impulse test (vHIT) is the most specific bedside finding [1,6]. Horizontal head impulses to both sides produce corrective catch-up saccades — overt saccades in severe loss, covert saccades (visible only on video-oculography) in partial or well-compensated loss. A bilaterally abnormal bedside HIT combined with the characteristic history is sufficient to diagnose probable BVP even without quantitative laboratory testing [1]. Dynamic visual acuity testing — comparing visual acuity during horizontal head oscillation against the static baseline — demonstrates a significant drop of two or more Snellen lines in BVP, quantifying the functional impact of the VOR deficit on gaze stabilisation [1,6,36,37]. Romberg test is reliably positive: patients sway dramatically or fall with eyes closed on foam, reflecting loss of the vestibular contribution to upright stance control. Tandem gait is severely impaired [42]. Neurological examination is otherwise normal in isolated BVP — cerebellar signs, pyramidal features, or peripheral neuropathy signal an alternative or co-existing diagnosis such as CANVAS [1,8].

□ **Important:** If cerebellar ataxia, gait ataxia out of proportion to VOR deficit, sensory neuropathy, or MRI evidence of cerebellar vermian atrophy accompanies bilateral vestibular areflexia, consider CANVAS (RFC1 biallelic expansion). RFC1 genetic testing should be performed before attributing BVP to idiopathic degeneration in patients with these additional findings [8,14].

IV. Diagnostic Criteria — Bárány Society 2017 Consensus

Definite Bilateral Vestibulopathy

The Bárány Society Classification Committee published the internationally adopted diagnostic criteria for bilateral vestibulopathy in 2017 [1]. Definite bilateral vestibulopathy requires the co-presence of: (1) a characteristic chronic clinical syndrome and (2) objective laboratory evidence of reduced bilateral VOR function, with (3) no alternative diagnosis better accounting for the findings [1,2].

The clinical syndrome criterion requires chronic postural instability during standing or walking persisting for at least 3 months, plus at least one of: (a) movement-induced oscillopsia or blurred vision during head movements or walking, or (b) worsening of unsteadiness in darkness or on uneven ground [1]. The requirement for chronicity distinguishes BVP from acute bilateral vestibular injury. The absence of symptoms at complete rest distinguishes it from most central balance disorders [1,2].

Laboratory criteria for reduced bilateral VOR function require abnormality on at least one of: (a) video head impulse test (vHIT) — bilateral horizontal VOR gain below 0.6 for both leftward and rightward head impulses; (b) bithermal caloric testing — summed maximal slow-phase velocity below 6°/s for each ear individually; or (c) sinusoidal rotational chair testing — VOR gain below 0.1 at 0.1 Hz with phase lead above 68° or time constant below 5 seconds [1]. These thresholds were selected to achieve diagnostic specificity while acknowledging that individual tests have different frequency sensitivities — calorics probe very low frequencies, vHIT targets high frequencies, and rotational chair spans the mid-frequency range [1,6].

Table 3. Bárány Society 2017 Diagnostic Criteria for Bilateral Vestibulopathy.

Criterion	Requirement	Threshold / Definition
Symptom duration	Chronic (≥3 months)	Postural instability during standing or walking
Core symptom (≥1 required)	Oscillopsia OR dark/uneven worsening	Movement-induced blurred vision; OR marked worsening in darkness or on uneven ground
Absent at rest	No dizziness when seated or supine	Static conditions symptom-free
vHIT criterion	Bilateral horizontal gain <0.6	Both left and right head impulse directions; high-frequency VOR
Caloric criterion	Each ear summed SPV <6°/s	Bithermal caloric; total per ear below 6°/s; low-frequency VOR
Rotational chair criterion	Gain <0.1 at 0.1 Hz; phase lead >68° or Tc <5 s	Mid-frequency sinusoidal rotation
Probable BVP (bedside)	Abnormal bilateral bedside HIT	If lab testing unavailable; catch-up saccades bilaterally
Exclusion	No alternative diagnosis	Central, functional, or other peripheral cause excluded

Probable Bilateral Vestibulopathy and Otolith Subtypes

The 2017 criteria define probable bilateral vestibulopathy as the characteristic chronic syndrome [46] with bilaterally abnormal bedside head impulse tests in the absence of available or completed laboratory testing [1]. This category facilitates appropriate clinical management when full quantitative testing is pending or inaccessible. A diagnostically important subtype involves selective otolith dysfunction with preserved canal VOR: these patients have absent or severely reduced bilateral cVEMPs and oVEMPs alongside normal calorics and vHIT, corresponding to isolated saccular and utricular loss [1]. The clinical presentation is similar — oscillopsia and disequilibrium — reflecting the role of otolith-driven VOR in vertical and translational stabilisation. This otolith-selective subtype is especially relevant in evaluating older patients with unexplained falls, in whom canal-selective tests alone may miss significant vestibular loss [1,23].

□ **Key Point:** The Bárány Society 2017 criteria are the only internationally validated diagnostic standard for BVP. All three laboratory modalities (vHIT, calorics, rotational chair) assess different VOR frequency ranges and may disagree — meeting any one criterion with the appropriate clinical syndrome is sufficient for a definite diagnosis [1].

V. Investigations: Vestibular Function Testing, Imaging, and Aetiological Workup

Video Head Impulse Test (vHIT)

vHIT is the primary quantitative bedside-to-laboratory investigation for BVP and is central to the 2017 diagnostic criteria [1,6]. Using high-speed infrared video-oculography, vHIT measures VOR gain (eye velocity/head velocity) during brief, unpredictable head impulses at velocities of 150–300°/s, probing the high-frequency performance (3–7 Hz) of each semicircular canal individually [1,6]. In BVP, horizontal canal gains are bilaterally reduced, typically below 0.6 and often approaching 0.1–0.3 in complete areflexia, with corrective saccades visible on the velocity trace [1]. The three-canal vHIT also quantifies vertical canal (anterior and posterior) function, frequently demonstrating pan-canal deficits in advanced disease [24].

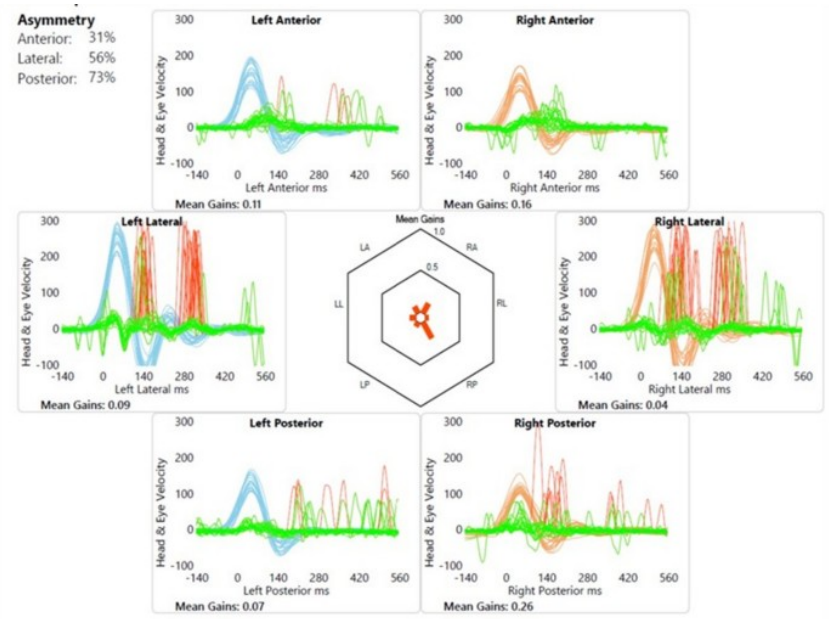

Figure 2. Video Head Impulse Test — Bilateral Vestibular Failure Pattern *— Representative vHIT trace showing severely reduced bilateral horizontal VOR gain with overt corrective catch-up saccades in a patient with bilateral vestibulopathy. Left and right gains are both well below the 0.6 threshold, confirming bilateral high-frequency canal loss.*

Source: Australian Dizziness Clinics — educational image from clinical vHIT database.

Caloric Stimulation

Bithermal caloric testing probes the low-frequency function of the horizontal semicircular canal by generating convection-driven endolymph flow through thermal gradients. In BVP, both ears demonstrate markedly reduced or absent slow-phase velocity responses, with the Bárány Society threshold of less than 6°/s per ear providing the diagnostic criterion for bilateral canal hypofunction [1]. Caloric testing retains unique clinical value in quantifying residual function in severely hyposensitive labyrinths: a patient with apparent bilateral vHIT areflexia may demonstrate small but measurable caloric responses, reflecting preserved low-frequency function at the frequency-selectivity limit of vHIT [6]. The complementary frequency coverage of vHIT (high frequency) and calorics (low frequency) makes their combined use the gold standard vestibular function assessment protocol [1,6,24].

Rotational Chair Testing

Sinusoidal rotational chair testing at 0.1 Hz produces a VOR gain below 0.1, a phase lead above 68°, or a time constant below 5 seconds in BVP [1]. The rotational chair is particularly valuable in three scenarios: (1) when calorics are technically unreliable (e.g. canal atresia, tympanic perforation), (2) when calorics demonstrate apparent bilateral loss but residual symmetric function is suspected, and (3) when tracking disease progression in longitudinal follow-up [6,24]. Step velocity testing and frequency-sweep sinusoidal rotation provide additional characterisation of VOR dynamics across the clinically relevant range of 0.01–2 Hz.

Vestibular Evoked Myogenic Potentials (VEMPs)

Cervical VEMPs (cVEMPs, assessing saccular function via the inferior vestibular nerve) and ocular VEMPs (oVEMPs, assessing utricular function via the superior vestibular nerve) characterise the otolith

contribution to the vestibular deficit [1,23]. In many BVP patients, both cVEMPs and oVEMPs are bilaterally absent or severely reduced, reflecting pan-labyrinthine loss [1]. Crucially, VEMPs are required for identifying the otolith-selective BVP subtype, in which canal function (vHIT and calorics) is preserved but bilateral VEMP absence confirms significant saccular and utricular dysfunction [1]. Age-related VEMP amplitude attenuation must be factored when interpreting responses in patients over 60, using published age-normalised reference ranges [23].

Table 4. Key Investigations in Bilateral Vestibulopathy: Findings and Clinical Significance.

Investigation	Finding in BVP	Clinical Significance / Notes
vHIT (horizontal)	Bilateral gain <0.6 with catch-up saccades	High-frequency canal function; first-line quantitative test [1,6]
vHIT (vertical)	Reduced anterior and posterior canal gains	Pan-canal involvement in advanced disease [24]
Caloric testing	SPV <6°/s each ear	Low-frequency canal function; complementary to vHIT [1,6]
Rotational chair	Gain <0.1 at 0.1 Hz; phase lead >68°	Mid-frequency; useful when calorics technically unreliable [1,24]
cVEMP / oVEMP	Bilaterally absent or severely reduced	Otolith function; required to identify otolith-selective subtype [1,23]
Dynamic visual acuity	≥2 Snellen lines drop during head oscillation	Quantifies functional oscillopsia severity; tracks rehabilitation [6]
Audiometry	Variable; SNHL present in many aetiologies	Guides aetiology; SNHL present in ototoxic, Ménière, genetic causes
MRI brain/IAC	Usually normal in idiopathic BVP	Exclude bilateral schwannoma (NF2), cerebellar degeneration, CANVAS atrophy [8]
RFC1 genetic testing	AAGGG biallelic expansion in CANVAS	Indicated if neuropathy, cerebellar ataxia, or unexplained areflexia [8,14]
Autoimmune bloods	ANA, ANCA, ESR, CRP (variable)	If autoimmune inner ear disease suspected; Cogan syndrome work-up [18]

Imaging and Neurological Investigations

MRI of the brain with internal auditory canal views and gadolinium enhancement should be performed in all patients with confirmed BVP to exclude bilateral vestibular schwannomas (a defining feature of neurofibromatosis type 2) and to identify cerebellar or brainstem pathology [8,25]. CANVAS-related cerebellar vermian atrophy may be identified on MRI. Gadolinium enhancement of the membranous labyrinth on delayed post-contrast 3D-FLAIR sequences may reveal bilateral endolymphatic hydrops in bilateral Ménière's disease or autoimmune labyrinthitis [25]. Nerve conduction studies and electromyography are indicated when clinical examination raises suspicion for sensorimotor neuropathy, given the CANVAS association and the frequency of peripheral neuropathy as a confounding or contributing cause of imbalance in BVP patients [8,14].

□ **Important:** RFC1 biallelic AAGGG intronic repeat expansion testing should be performed in all patients with unexplained bilateral vestibular areflexia, particularly if accompanied by peripheral neuropathy, cerebellar ataxia, or positive family history. RFC1-CANVAS may represent up to 10–20% of previously labelled idiopathic BVP cases [8,14].

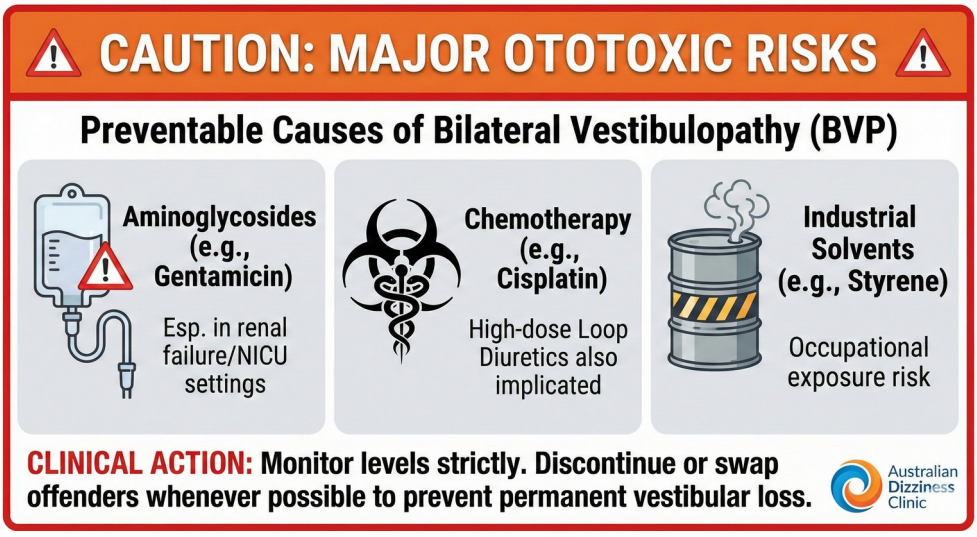

Figure 3. Common Vestibulotoxic Agents *— Infographic illustrating the principal classes of vestibulotoxic drugs associated with bilateral vestibulopathy. Aminoglycosides (particularly gentamicin, tobramycin) and platinum-based chemotherapeutics are the most prevalent ototoxic causes. Loop diuretics potentiate aminoglycoside toxicity when co-prescribed.*

Source: Australian Dizziness Clinics — educational infographic.

VI. Differential Diagnosis

Bilateral vestibulopathy must be distinguished from a range of disorders producing chronic imbalance, oscillopsia, or unsteadiness. The key diagnostic discriminators are the presence of oscillopsia (absent in most non-vestibular causes), the bilateral pattern of VOR loss on quantitative testing, and the characteristic worsening in darkness [1,2,6].

Cerebellar Ataxias

Degenerative or immune-mediated cerebellar disorders — including spinocerebellar ataxias (SCA3, SCA6, SCA7, SCA27B), autoimmune cerebellar degeneration, and alcoholic cerebellar vermian degeneration — produce gait ataxia that superficially resembles BVP [1,8]. Key distinguishing features: dysmetria on finger-nose and heel-shin testing, gaze-evoked nystagmus, impaired smooth pursuit, and ataxia that persists even when seated (unlike BVP, which improves with seated rest) [45]. Downbeat nystagmus syndrome (floccular cerebellar dysfunction) is a particularly important differential, as it produces oscillopsia and imbalance; the spontaneous downbeat nystagmus in primary gaze and preserved VOR distinguish it from BVP [1]. Vestibular testing will be normal in pure cerebellar ataxia.

Peripheral Sensory Neuropathy (Sensory Ataxia)

Severe proprioceptive deafferentation from peripheral polyneuropathy (diabetic, B12 deficiency, paraneoplastic) or dorsal column lesions produces positive Romberg sign and marked dark-worsening of imbalance, mimicking BVP [1,2]. The critical distinguishing feature is the absence of true oscillopsia in pure sensory neuropathy — VOR is intact, so gaze remains stable during head movement [1]. Neurological examination reveals impaired joint position sense, absent ankle reflexes, and reduced vibration sense; vestibular testing is normal. In CANVAS, both peripheral vestibular loss and sensory neuropathy co-exist, producing compound instability that is more severe than either alone [8,14].

Persistent Postural-Perceptual Dizziness (PPPD)

PPPD is characterised by chronic non-spinning dizziness and unsteadiness exacerbated by upright posture, complex visual environments, and motion — features that superficially overlap with BVP [1,26]. However, PPPD patients do not exhibit true oscillopsia (VOR is intact), and quantitative vestibular testing is normal by definition [26]. PPPD often follows an acute vestibular event (vestibular neuritis, BPPV) and has significant anxiety co-morbidity. Dynamic visual acuity is preserved in PPPD despite subjective visual sensitivity. The 'supermarket effect' occurs in both — but in PPPD it reflects visuo-vestibular mismatch hypersensitivity, not true VOR failure [26].

Presbyvestibulopathy

Age-related bilateral vestibular hypofunction — presbyvestibulopathy — was recently defined by the Bárány Society using slightly relaxed thresholds (age ≥60, bilateral vHIT gain 0.6–0.8 or reduced calorics not meeting the full BVP cut-offs) [27]. The boundary between presbyvestibulopathy and BVP is a continuum: older patients with bilateral hypofunction below diagnostic thresholds may nonetheless have significant functional impairment and benefit from rehabilitation. Age-adjusted normative data are essential to avoid both over-diagnosis in healthy elderly subjects and under-diagnosis in those with pathological bilateral loss [27].

Table 5. Differential Diagnosis: Bilateral Vestibulopathy vs Key Mimickers.

Diagnosis	Oscillopsia	Dark Worsening	VOR (vHIT/caloric)	Distinguishing Feature
Bilateral vestibulopathy	Yes — with head movement	Yes	Bilaterally reduced/absent	Bárány criteria met; no acute vertigo [1]
Cerebellar ataxia	Only if downbeat nystagmus	Variable	Normal	Dysmetria; gaze-evoked nystagmus; MRI atrophy [8]
Sensory neuropathy	No	Yes (proprioceptive)	Normal	Absent ankle jerks; impaired JPS; no oscillopsia [2]
CANVAS	Yes	Yes (dual loss)	Bilaterally absent	Sensory neuropathy + cerebellar signs + RFC1 expansion [8,14]
PPPD	No (subjective)	Variable	Normal	Normal vestibular testing; anxiety; follows acute event [26]
Presbyvestibulopathy	Mild	Mild	Mildly reduced (0.6–0.8)	Age >60; below BVP threshold; age-adjusted norms [27]
Parkinson/MSA	No	No	Normal	Bradykinesia; rigidity; autonomic dysfunction [2]
Downbeat nystagmus syndrome	Yes	Variable	May be reduced	Spontaneous downbeat nystagmus in primary gaze [1]

VII. Medical Management and Cause-Directed Therapy

Principle: No Restorative Pharmacotherapy

There is currently no medication that restores vestibular hair cell function or regenerates vestibular neurons in humans [2,5,6]. Management is therefore organised around three principles: (1) cessation or treatment of the causative agent or process, (2) symptomatic management of co-morbidities and secondary consequences, and (3) functional rehabilitation. Vestibular suppressant medications — benzodiazepines, antihistamines, and anticholinergics — are contraindicated in chronic BVP as they impair central vestibular compensation and worsen both imbalance and cognitive function [2,6].

□ **Important:** Vestibular suppressants (prochlorperazine, meclizine, dimenhydrinate, benzodiazepines) are CONTRAINDICATED in bilateral vestibulopathy. They further suppress residual vestibular function and impair the central compensation mechanisms on which patients depend [2,6]. Review all current prescriptions and cease vestibular suppressants as the first management step.

Aminoglycoside and Ototoxic Drug Exposure

When ototoxic drug exposure is the identified aetiology, immediate cessation of the causative agent prevents further loss — though existing damage is permanent [5,16,17]. Substitution of aminoglycosides with less ototoxic alternatives (e.g. tobramycin to colistin, or gentamicin-based to azithromycin-based protocols where clinically appropriate) is strongly recommended. Baseline and monitoring audiometry and vHIT during prolonged aminoglycoside courses (cystic fibrosis, multidrug-resistant tuberculosis, Gram-negative septicaemia) can detect early vestibular loss and prompt dose adjustment or cessation [16,17]. Cisplatin vestibulotoxicity follows a cumulative dose-response curve; where oncological objectives permit, alternative platinum agents (carboplatin, oxaliplatin) may be considered, noting they have lower vestibulo-cochlear toxicity profiles [17].

Autoimmune Inner Ear Disease

Suspected autoimmune bilateral labyrinthopathy warrants early immunosuppressive therapy, as the window of reversibility is narrow [6,18]. High-dose oral corticosteroids (prednisolone 1 mg/kg/day, tapering over 4–6 weeks) are the first-line agent for autoimmune inner ear disease presenting with bilateral progressive vestibulo-cochlear loss [6,18]. In corticosteroid-responsive cases, maintenance with methotrexate or mycophenolate mofetil may be considered to prevent relapse and facilitate steroid sparing [18]. Cogan's syndrome — characterised by interstitial keratitis and rapidly progressive bilateral audiovestibular loss — requires urgent high-dose corticosteroid treatment to prevent permanent bilateral deafferentation [18]. Serological monitoring (ESR, CRP, ANA, ANCA) guides therapy duration. Response can be tracked with serial vHIT and audiometry.

Bilateral Ménière's Disease

Bilateral Ménière's disease presents a therapeutic paradox in end-stage BVP: intratympanic gentamicin, effective for ablating residual episodic vertigo, accelerates vestibular loss [44]. When the dominant clinical problem has shifted from episodic vertigo to chronic bilateral disequilibrium — indicating transition to burnt-out bilateral loss — aggressive ablative treatment of any remaining vestibular function may worsen the chronic syndrome [2,6]. In earlier, still-episodic bilateral Ménière's, conservative measures (low-sodium diet, betahistine, thiazide diuretics) may reduce attack frequency and delay progression [6]. Endolymphatic sac surgery offers a non-ablative option to reduce endolymphatic pressure without intentionally eliminating vestibular function.

Management of Associated Conditions

Anxiety and depression are prevalent in BVP patients and substantially worsen functional outcomes [41]. outcomes [2,21]. Low-dose selective serotonin reuptake inhibitors (SSRIs) — particularly sertraline and escitalopram — may improve balance confidence and reduce vestibular anxiety, in addition to their primary anxiolytic/antidepressant effects [6,21]. Unlike traditional vestibular suppressants, SSRIs do not impair vestibular compensation. Cognitive behavioural therapy targeting vestibular-specific anxiety and avoidance behaviours improves outcomes significantly when delivered alongside vestibular rehabilitation [21,26].

Table 6. Cause-Directed Management of Bilateral Vestibulopathy.

Aetiology	First-Line Intervention	Notes / Evidence Level
Aminoglycoside ototoxicity	Cease offending agent; substitute to safer antibiotic	Damage permanent once established; monitoring during treatment prevents complete loss [16,17]
Cisplatin / platinum chemo	Alternative platinum if oncologically appropriate	Cumulative dose-response; coordinate with oncology [17]
Autoimmune inner ear disease	High-dose prednisolone → methotrexate/MMF maintenance	Narrow reversibility window; early treatment critical [6,18]
Cogan's syndrome	Urgent high-dose corticosteroids; systemic vasculitis workup	Rapid progression; refer ophthalmology and rheumatology [18]
Bilateral Ménière's (episodic)	Low-sodium diet, betahistine, thiazide diuretic	Avoid ablative intratympanic gentamicin in established BVP [2,6]
CANVAS / RFC1	No disease-modifying therapy;	Genetic counselling; identify

	supportive and rehabilitation	affected family members [8,14]
DFNA9 (COCH mutation)	No current disease-modifying therapy	Progressive; gene therapy research ongoing [19]
Idiopathic / degenerative	Vestibular rehabilitation; fall prevention; cessation of vestibular suppressants	Largest subgroup; supportive management [1,2,6]

VIII. Vestibular Rehabilitation and Emerging Technologies

Vestibular Rehabilitation Therapy (VRT)

Vestibular rehabilitation therapy is the mainstay of functional management for BVP and is supported by the highest-quality evidence in the vestibular rehabilitation literature [9,28]. A Cochrane systematic review by Porciuncula et al. (2012) identified moderate-quality evidence of significant functional improvement in balance, gait, and dynamic visual acuity with supervised VRT, though outcomes do not achieve normal function [9]. The therapeutic targets are threefold: (1) maximising sensory substitution — training patients to effectively utilise residual visual [34] and proprioceptive inputs for balance and gaze stabilisation; (2) gaze stabilisation exercises — head movement tracking tasks that promote whatever residual VOR function remains and optimise visual predictive and pursuit strategies; and (3) progressive balance and gait challenge — foam surface standing, tandem walking, turning, stair navigation, and gradual introduction of dynamic tasks [9,28].

VRT for BVP is fundamentally different from VRT for unilateral vestibular hypofunction: habituation and adaptation are less relevant without a compensating intact side. The emphasis is on substitution and optimisation rather than central recalibration [9,28]. The optimal programme frequency and intensity has not been definitively established, but expert consensus recommends at least 1–2 formal supervised sessions per week for an initial 8–12 weeks, with a structured home exercise programme progressing to independent community ambulation goals [9].

Assistive Devices and Sensory Substitution Technologies

When VRT alone is insufficient, assistive technologies can meaningfully supplement balance control. Vibrotactile sensory substitution devices — worn at the trunk, abdomen, or tongue — translate vestibular-equivalent tilt and motion signals from inertial measurement units into tactile stimulation patterns, providing a non-visual balance cue [29]. Clinical trials of vibrotactile feedback devices have demonstrated reductions in postural sway and patient-reported improvement in balance confidence, though widespread clinical adoption is limited by cost and device availability [29]. Weighted walking frames, cane use in low-light conditions, and night-lighting modifications reduce fall risk in daily life and should be routinely prescribed.

Vestibular Implant — Emerging Intervention

The vestibular implant — a neural prosthetic device that delivers electrical stimulation directly to the ampullary nerves modulated by gyroscopic inertial measurement — represents the most significant emerging intervention for severe BVP [10,30]. Modelled on cochlear implant technology, the device detects angular head velocity and encodes it as patterned electrical pulses to one or more semicircular canal nerves. Early clinical trials in patients with bilateral vestibular areflexia have demonstrated measurable restoration of head-movement-driven eye movements (prosthetic VOR), improvements in postural stability, and reduction in oscillopsia [10,30]. Della Santina et al. (2023) demonstrated vestibular implant efficacy even in patients with bilateral vestibular hypofunction exceeding 20 years in duration [10]. Currently approved for compassionate and experimental use in several international centres, vestibular implantation is anticipated to move towards formal regulatory approval within the next 5–10 years [10,30].

□ **Clinical Insight:** Vestibular implant candidacy criteria currently require: (1) confirmed bilateral vestibular areflexia (VOR gain approaching zero bilaterally), (2) significant functional disability despite maximal conventional rehabilitation, and (3) absence of cochlear implant precluding electrode array placement [10,30]. Refer appropriate candidates to research-active neuro-otology centres.

IX. Prognosis, Outcomes, and Special Populations

Vestibular Function Recovery

Spontaneous vestibular function recovery in established BVP is rare [1,2,6]. Once hair cells are destroyed by ototoxicity, infection, or degeneration, structural restoration does not occur in humans without intervention. The rare exceptions are autoimmune inner ear disease (if treated early in the inflammatory phase) and bilateral Ménière's disease in which a brief functional hypofunction episode partially resolves [6,18]. In most cases, vestibular testing performed at 6, 12, and 24 months will show stable bilateral hypofunction or gradual progression in hereditary or degenerative aetiologies. Any observed functional improvement over time is attributable to central adaptation rather than peripheral regeneration [1,2].

Functional Outcomes with Rehabilitation

Despite persistent vestibular deficit, most BVP patients achieve meaningful functional improvement with structured rehabilitation [9,28]. Objective measures — postural sway, Timed Up-and-Go (TUG), Dynamic Gait Index (DGI), and dynamic visual acuity — typically improve significantly over the first 6–18 months after diagnosis and rehabilitation commencement, then plateau at a functional level below healthy controls but substantially better than untreated baseline [9]. Patient-reported outcomes on the Dizziness Handicap Inventory (DHI) and vestibular-specific quality of life instruments document significant reductions in handicap score. The long-term trajectory in idiopathic or ototoxic BVP is of relative stability once vestibular loss is established — functional decline over years typically reflects ageing of the compensatory systems (visual acuity, lower-limb proprioception, muscle strength) rather than further vestibular loss [2,11].

Falls and Morbidity

Fall risk is substantially elevated in BVP [2,11]. Prospective data demonstrate fall rates of 2–4 times those of age-matched controls, with a corresponding increase in fracture risk — particularly hip and Colles fractures in older patients [2]. Fall-related injury can produce catastrophic secondary morbidity in an already-disabled population, precipitating loss of independent living. Fall prevention strategies — home environmental assessment, non-slip flooring, bathroom rails, adequate nocturnal lighting, cessation of evening sedating medications, and Tai Chi or balance training — constitute a mandatory component of BVP management [2,9,11].

Table 7. Functional Outcomes and Prognosis by Subtype and Aetiology.

Aetiology / Subtype	VOR Recovery	Functional Prognosis	Notes
Aminoglycoside ototoxicity	None (permanent)	Good functional improvement with VRT	Stable once drug ceased [5,16,17]
Autoimmune inner ear disease	Partial if treated early	Variable; relapse risk if immunosuppression withdrawn	Monitor with serial vHIT [6,18]
Bilateral Ménière's (burnt out)	None	Good rehab potential; vertigo ceases	Focus shifts to rehabilitation once episodic phase ends [2,6]
CANVAS / RFC1	None (progressive)	Progressive; triple sensory deafferentation	Cerebellar ataxia + neuropathy compound prognosis [8,14]
DFNA9 (COCH mutation)	None (progressive)	Moderate; predictable trajectory enables planning	Mid-life onset; slow progressive decline [19]
Idiopathic	Rare; <5%	Moderate to good with VRT; stable in most	Audit for RFC1 before idiopathic label [1,8]
Paediatric (congenital)	None (structural)	Good if early rehabilitation; significant brain plasticity	May achieve near-normal balance by adulthood [12]

Special Populations: CANVAS

CANVAS — caused by biallelic AAGGG intronic repeat expansions in the RFC1 gene — represents the single most important recent advance in BVP aetiology [8,14]. The syndrome produces a triple sensory deafferentation: bilateral vestibular areflexia, large-fibre sensory peripheral neuropathy (with Romberg sign and impaired proprioception), and cerebellar vermian degeneration (with gait and limb ataxia) [8,14]. Estimated to account for 10–20% of previously labelled [47] idiopathic BVP, CANVAS has an adult onset typically in the fifth to sixth decade, a slow progressive course, and an absence of effective disease-modifying treatment [8,14]. The identification of the RFC1 mutation has enabled familial testing and genetic counselling. Recent reports of multiple non-AAGGG RFC1 repeat motifs contributing to the CANVAS spectrum suggest that conventional repeat-primed PCR assays may miss some cases, and long-read sequencing may be required for complete characterisation [14].

X. Guidelines, Controversies, and Future Directions

Guideline Landscape

The 2017 Bárány Society diagnostic criteria for bilateral vestibulopathy [1,39] and the 2019 consensus document on presbyvestibulopathy [27] represent the only internationally validated diagnostic standards in this field as of 2026. No major specialty society (AAO-HNS, British Society of Audiology, ANZAN) has yet published clinical practice guidelines specifically addressing BVP management, though vestibular rehabilitation guidelines from the American Physical Therapy Association and the Cochrane vestibular review [9] address rehabilitation evidence. The absence of management guidelines reflects both the rarity of the condition and the limited randomised trial evidence, creating practice variation across centres in areas including aetiological workup protocols, vestibular monitoring during ototoxic therapy, and rehabilitation intensity [1,9].

Diagnostic Threshold Controversy

The selection of diagnostic thresholds — vHIT gain <0.6, caloric SPV <6°/s per ear — is a subject of ongoing debate [1,27]. These stringent thresholds prioritise specificity over sensitivity, potentially excluding patients with partial bilateral hypofunction who nonetheless experience significant functional disability. Advocates for relaxed thresholds argue for graded classification (complete BVP, partial BVP, presbyvestibulopathy) to enable management appropriate to the degree of deficit [27]. Age-adjusted normative values for all vestibular test modalities are critical but incompletely published, creating uncertainty in interpreting borderline results in patients over 65 [27]. The role of otolith testing (VEMPs) in the primary criteria remains contested — their absence from the core criteria may lead to under-diagnosis of the otolith-selective subtype [1].

RFC1 and the Revision of Idiopathic BVP

The 2019 discovery of RFC1 AAGGG biallelic repeat expansions as the cause of CANVAS has fundamentally altered the epidemiology of BVP [8,14]. What was previously labelled 'idiopathic' bilateral vestibulopathy may in substantial proportion be RFC1-CANVAS — raising the question of whether 'idiopathic BVP' remains a valid diagnostic category in patients who have not had RFC1 testing. Current expert opinion recommends RFC1 testing in all unexplained bilateral vestibular areflexia, particularly when accompanied by neuropathic features or cerebellar signs [8,14]. The expansion of the RFC1 variant spectrum (ACAGG, AAAGG, and other non-AAGGG motifs) means that standard assays may miss a proportion of pathogenic expansions, requiring updated genetic testing panels [14].

Hair Cell Regeneration — Biological Future

Gene therapy and hair cell regeneration represent the most transformative long-term research directions for BVP [31]. In non-mammalian vertebrates (fish, birds), vestibular hair cells regenerate spontaneously after damage — a capacity lost in mammalian phylogeny. Efforts to reactivate hair cell regeneration via Atoh1 (Math1) gene delivery, Wnt pathway agonism, and Notch pathway inhibition have demonstrated hair cell production in mammalian cochlear explants and in vivo murine models [31]. Human translation remains in early-phase investigation but represents the only approach that could achieve true vestibular function restoration. For defined genetic aetiologies (DFNA9-COCH, Usher syndrome), antisense oligonucleotide and AAV-mediated gene correction strategies are progressing towards clinical trial design [19,31].

Vestibular Implant — Clinical Horizon

As described in Section VIII, the vestibular implant is the most clinically advanced technology for severe BVP [10,30]. Ongoing multicentre trials are collecting the safety and efficacy data required for regulatory approval. Unresolved questions include optimal candidacy criteria, whether unilateral or bilateral implantation is preferable, long-term device performance and revision rates, and cost-effectiveness relative to intensive rehabilitation [10,30]. If regulatory approval proceeds in the next decade, vestibular implantation may become the standard of care for complete bilateral vestibular areflexia refractory to rehabilitation — a parallel trajectory to cochlear implantation for profound bilateral deafness [30].

□ **Clinical Pearl:** The two most important practice-changing advances in BVP of the past decade are: (1) RFC1 genetic testing, which reclassifies a substantial proportion of 'idiopathic' BVP to CANVAS, and (2) the vestibular implant, which offers the first realistic prospect of functional vestibular restoration in complete bilateral areflexia [8,10,14,30].

References

[1] Strupp M, Kim JS, Murofushi T, et al. Bilateral vestibulopathy: Diagnostic criteria — Consensus document of the Classification Committee of the Bárány Society. J Vestib Res. 2017;27(4):177–189.

[2] van Stiphout L, Szmulewicz DJ, Kingma H, Van De Berg R. Bilateral vestibulopathy: a clinical update and proposed diagnostic algorithm. Front Neurol. 2023;14:1308485.

[3] Brandt T, Dieterich M, Strupp M. Vertigo and Dizziness: Common Complaints. 2nd ed. London: Springer; 2013.

[4] Propper EJ, Koppelaar-van Eijsden HM, Schermer TR, Bruintjes TD. Bilateral vestibular hypofunction in a tertiary dizziness center: occurrence and etiology. J Int Adv Otol. 2022;18(4):327–333.

[5] Zingler VC, Cnyrim C, Jahn K, et al. Causative factors and epidemiology of bilateral vestibulopathy. Ann Neurol. 2007;61(6):524–532.

[6] Lucieer F, Vonk P, Guinand N, et al. Bilateral vestibular hypofunction: insights in etiologies, clinical subtypes, and diagnostics. Front Neurol. 2016;7:26.

[7] Brandt T, Schautzer F, Hamilton DA, et al. Vestibular loss causes hippocampal atrophy and impaired spatial memory in humans. Brain. 2005;128(Pt 11):2732–2741.

[8] Szmulewicz DJ, Waterston JA, Halmagyi GM, et al. Sensory neuropathy as part of the cerebellar ataxia, neuropathy, vestibular areflexia syndrome (CANVAS). Neurology. 2011;76(22):1903–1910.

[9] Porciuncula F, Johnson CC, Glickman LB. The effect of vestibular rehabilitation on adults with bilateral vestibular hypofunction: a systematic review. J Vestib Res. 2012;22(5–6):283–298.

[10] Della Santina CC, Migliaccio AA, Patel AH, et al. Vestibular implantation can work even after more than 20 years of bilateral vestibular hypofunction. Otol Neurotol. 2023;44(2):e152–e156.

[11] Guinand N, Boselie F, Guyot JP, Kingma H. Quality of life of patients with bilateral vestibulopathy. Ann Otol Rhinol Laryngol. 2012;121(7):471–477.

[12] Peng TH, Villarin C, Davin K, et al. Pediatric bilateral vestibular loss: an underrecognized condition. Otol Neurotol. 2021;42(1):60–66.

[13] Agrawal Y, Carey JP, Della Santina CC, Schubert MC, Minor LB. Disorders of balance and vestibular function in US adults: data from the National Health and Nutrition Examination Survey, 2001–2004. Arch Intern Med. 2009;169(10):938–944.

[14] Cortese A, Simone R, Sullivan R, et al. Biallelic expansion of an intronic repeat in RFC1 is a common cause of late-onset ataxia. Nat Genet. 2019;51(4):649–658.

[15] Halmagyi GM, Curthoys IS. A clinical sign of canal paresis. Arch Neurol. 1988;45(7):737–739.

[16] Fischel-Ghodsian N. Genetic factors in aminoglycoside toxicity. Ann N Y Acad Sci. 1999;884:99–109.

[17] Rizzi MD, Hirose K. Aminoglycoside ototoxicity. Curr Opin Otolaryngol Head Neck Surg. 2007;15(5):352–357.

[18] Bovo R, Ciorba A, Martini A. The diagnosis of autoimmune inner ear disease: evidence and critical pitfalls. Eur Arch Otorhinolaryngol. 2009;266(1):37–40.

[19] de Kok YJ, van der Maarel SM, Bitner-Glindzicz M, et al. Association between X-linked mixed deafness and mutations in the POU domain gene POU3F4. Science. 1995;267(5198):685–688.

[20] Stackman RW, Clark AS, Taube JS. Hippocampal spatial representations require vestibular input. Hippocampus. 2002;12(3):291–303.

[21] Edelman S, Mahoney AE, Cremer PD. Cognitive behavior therapy for chronic subjective dizziness: a randomized, controlled trial. Am J Otolaryngol. 2012;33(4):395–401.

[22] Szmulewicz DJ. Bilateral vestibulopathy: an underdiagnosed condition. Pract Neurol. 2020;20(4):271–272.

[23] Curthoys IS, Vulovic V. Vestibular primary afferent responses to sound and vibration in the guinea pig. Exp Brain Res. 2011;210(3–4):347–352.

[24] Mahringer A, Rambold HA. Caloric test and video-head-impulse: a study of vertigo/dizziness patients in a community hospital. Eur Arch Otorhinolaryngol. 2014;271(3):463–472.

[25] Naganawa S, Nakashima T. Visualization of endolymphatic hydrops with MR imaging in patients with Ménière's disease and related pathologies: current status of its methods and clinical significance. Jpn J Radiol. 2014;32(4):191–204.

[26] Staab JP, Eckhardt-Henn A, Horii A, et al. Diagnostic criteria for persistent postural-perceptual dizziness (PPPD): consensus document of the committee for the Classification of Vestibular Disorders of the Bárány Society. J Vestib Res. 2017;27(4):191–208.

[27] Iwasaki S, Egami N, Nishida T, Yamasoba T, Murofushi T. Presbyvestibulopathy: consensus document of the Classification Committee of the Bárány Society. J Vestib Res. 2021;31(4):255–271.

[28] Hall CD, Herdman SJ, Whitney SL, et al. Vestibular Rehabilitation for Peripheral Vestibular Hypofunction: An Updated Clinical Practice Guideline from the Academy of Neurologic Physical Therapy of the American Physical Therapy Association. J Neurol Phys Ther. 2022;46(2):118–177.

[29] Danilov Y, Tyler M. Brainport: an alternative input to the brain. J Integr Neurosci. 2005;4(4):537–550.

[30] Guinand N, Guyot JP, Kingma H, Kos MI, Pelizzone M. The video head impulse test to assess the efficacy of vestibular implants in humans. Front Neurol. 2012;3:119.

[31] Bermingham-McDonogh O, Reh TA. Regulated reprogramming in the regeneration of sensory receptor cells. Neuron. 2011;71(3):389–405.

[32] Ward BK, Agrawal Y, Hoffman HJ, Carey JP, Della Santina CC. Prevalence and impact of bilateral vestibular hypofunction: results from the 2008 US National Health Interview Survey. JAMA Otolaryngol Head Neck Surg. 2013;139(8):803–810.

[33] Hanes DA, McCollum G. Cognitive-vestibular interactions: a review of patient difficulties and possible mechanisms. J Vestib Res. 2006;16(3):75–91.

[34] Sprenger A, Wojak JF, Jandl NM, Helmchen C. Postural control in bilateral vestibular failure: its relation to visual, proprioceptive and higher-level cognitive factors. Front Neurol. 2017;8:ref531.

[35] Lacour M, Dutheil S, Tighilet B, Lopez C, Borel L. Tell me your vestibular deficit, and I'll tell you how you'll compensate. Ann N Y Acad Sci. 2009;1164:268–278.

[36] Herdman SJ, Hall CD, Schubert MC, Das VE, Tusa RJ. Recovery of dynamic visual acuity in bilateral vestibular hypofunction. Arch Otolaryngol Head Neck Surg. 2007;133(4):383–389.

[37] Herdman SJ, Schubert MC, Das VE, Tusa RJ. Recovery of dynamic visual acuity in unilateral vestibular hypofunction. Arch Otolaryngol Head Neck Surg. 2003;129(8):819–824.

[38] Baloh RW, Jacobson K, Honrubia V. Idiopathic bilateral vestibulopathy. Neurology. 1989;39(2):272–275.

[39] Strupp M, Dlugaiczyk J, Ertl-Wagner BB, Rujescu D, Westhofen M, Dieterich M. Vestibular disorders: diagnosis, new classification and treatment. Dtsch Arztebl Int. 2020;117(17):300–310.

[41] Bhatt JM, Bhattacharyya N, Lin HW. Relationships between vestibular disorders and anxiety: a population-based study. Otolaryngol Head Neck Surg. 2017;156(6):1073–1079.

[42] Honaker JA, Boismier TE, Shepard NP, Shepard NT. Fukuda stepping test: sensitivity and specificity. J Am Acad Audiol. 2009;20(5):311–314.

[43] van Leeuwen RB, Bruintjes TD. Dizziness in the elderly: diagnosing its causes in a multidisciplinary dizziness unit. Ear Nose Throat J. 2014;93(4–5):162–167.

[44] Carey JP, Minor LB, Peng GC, Della Santina CC, Cremer PD, Haslwanter T. Changes in the three-dimensional angular vestibulo-ocular reflex following intratympanic gentamicin for Ménière's disease. J Assoc Res Otolaryngol. 2002;3(4):430–443.

[45] Staab JP. Chronic subjective dizziness. Continuum (Minneap Minn). 2012;18(5 Neuro-otology):1118–1141.

[46] van de Berg R, van Tilburg M, Kingma H. Bilateral vestibular hypofunction: challenges in establishing the diagnosis in adults. ORL J Otorhinolaryngol Relat Spec. 2015;77(4):197–218.

[47] Szmulewicz DJ, McLean CA, MacDougall HG, Roberts L, Storey E, Halmagyi GM. CANVAS an update: clinical presentation, investigation and management. J Vestib Res. 2016;26(1–2):73–78.

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