REVIEW

Peripheral vestibular disorders: an update

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Purpose of review
To provide an update on the most frequent peripheral vestibular disorders.

Recent findings
The on-going classification of vestibular disorders by the Bàràny Society represents major progress. The diagnosis of bilateral vestibulopathy (BVP) requires quantitative testing of vestibular function. 'Acute unilateral peripheral vestibulopathy' (AUPVP) is now preferred over 'vestibular neuritis.' Menière's disease is a set of disorders with a significant genetic contribution. The apogeotropic variant of hcBPPV and acBPPV can be distinguished from a central vestibular lesion. Vestibular paroxysmia is now an internationally accepted clinical entity. The diagnosis of SCDS is based on conclusive findings.

Summary
Diagnosis of BVP requires significantly reduced vestibular function. The clinical picture of AUPVP depends on how much the vestibular end organs or their innervation are affected. Menière's disease phenotype is a constellation of symptoms. Although diagnostic and therapeutic criteria for pc and hcBPPV are well defined, a number of less frequent and controversial are increasingly diagnosed and can be treated. Diagnosis of vestibular paroxysmia requires that a patient responds to treatment with a sodium channel blocker. The diagnosis of SCDS requires conclusive findings with various methods. There is still a great need for state-of-the-art randomized controlled trials in various vestibular disorders.

Keywords
acute unilateral peripheral vestibulopathy, benign paroxysmal positional vertigo, bilateral vestibulopathy, Menière's disease, superior canal dehiscence syndrome, vestibular paroxysmia

INTRODUCTION

Peripheral vestibular disorders are a group of diverse conditions that can manifest as acute, episodic, or persisting vestibular syndromes. This review will summarize the major and clinically relevant recent findings on (in the following order) bilateral vestibulopathy, acute unilateral peripheral vestibulopathy, Menière's disease, benign paroxysmal positional vertigo (BPPV), vestibular paroxysmia, and superior canal deshiscence syndrome.

BILATERAL VESTIBULOPATHY

Bilateral vestibulopathy (BVP) is a chronic vestibular syndrome with the leading symptom of unsteadiness when walking or standing, which worsens in darkness and/or on uneven ground, or during head motion. There are typically no symptoms while sitting or lying down under static conditions. Patients may describe head or body movement-induced blurred vision or oscillopsia. The syndrome was recently re-classified by the Classification Committee of the Bàràny Society [1▪]. For the diagnosis, the impaired vestibular function has to be quantified by laboratory testing, otherwise only the diagnosis of probable BVP can be made (Table 1).

According to a systematic review [88 studies included (41 clinical studies, 47 case reports)], the symptoms reported by patients with BVP were summarized as imbalance (91 and 86%), chronic dizziness (58 and 62%), oscillopsia (50 and 70%), and recurrent vertigo (33 and 67%) [2]. There were also

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KEY POINTS

- Bilateral vestibulopathy is now precisely defined. The diagnosis requires quantitative testing of vestibular function.
- Acute unilateral peripheral vestibulopathy is a diagnosis by exclusion of a central lesion. There is increasing evidence that is caused in most cases by Herpes simplex type 1.
- Clinical and 'omics' data in Menière's disease can facilitate patient stratification for personalized management.
- Diagnosing and treating different BPPV variants is fundamental to improve patients' quality of life and differentiate some uncommon forms from central vestibular disorders.
- Vestibular paroxysmia is now precisely defined. Other causes leading to similar symptoms have to be ruled out.
- High-resolution CT temporal scans in combination with VEMPs are considered the gold standard to confirm unilateral or bilateral SCDS.

Table 1. Diagnostic criteria for bilateral and probable bilateral vestibulopathy according to the Classification Committee of the Bàràny Society

Bilateral vestibulopathy
A: Chronic vestibular syndrome with the following symptoms
(1) Unsteadiness when walking or standing plus at least one of 2 or 3
(2) Movement-induced blurred vision or oscillopsia during walking or quick head/body movements and/or
(3) Worsening of unsteadiness in darkness and/or on uneven ground
B: No symptoms while sitting or lying down under static conditions
C: Bilaterally reduced or absent angular ~~VOR~~ function documented by:
bilaterally pathological horizontal angular VOR gain less than 0.6, measured by the video-HIT or scleral-coil technique and/or
reduced caloric response (sum of bithermal maximum peak SPV on each side $<6^{\circ}$/s) and/or
reduced horizontal angular VOR gain less than 0.1 upon sinusoidal stimulation on a rotatory chair (0.1 Hz, $V_{max} = 50^{\circ}$/s) and a phase lead greater than 68 degrees (time constant <5 sec).
D: Not better accounted for by another disease
Diagnostic criteria for probable bilateral vestibulopathy
A. Chronic vestibular syndrome with the following symptoms
(1) Unsteadiness when walking or standing plus at least one of 2 or 3
(2) Movement-induced blurred vision or oscillopsia during walking or quick head/body movements and/or
(3) Worsening of unsteadiness in darkness and/or on uneven ground
B. No symptoms while sitting or lying down under static conditions
C. Bilaterally pathological horizontal bedside head impulse test
D. Not better accounted for by another disease

Data from [1▪].

symptoms beyond direct vestibular deficits, such as limited social activities, depression, concentration and (spatial) memory impairment, and reduced quality of life in general. Even in patients with partial BVP, a significant decrease in gray matter in the mid-hippocampal (supported by a recent similar MRI study [3]) and posterior parahippocampal volume as well as functional changes with delayed spatial learning performance and higher spatial anxiety were found [4]. These findings are in line with animal experiments in BVP, which demonstrated a downregulation of hippocampal and striatal M1 muscarinic acetylcholine receptors that are involved in spatial learning and memory [5]. In a study with gait analysis on 55 patients, two significant predictive factors for falls in BVP were found: the presence of a concomitant peripheral neuropathy [odds ratio (OR) = 3.6) and an increase in temporal gait variability, especially at slow walking speeds (OR = 1.3) [6].

The impact of proprioceptive, visual, vestibular, and cognitive input on postural control was measured by posturography in patients with BVP [7]. The best predictors for the severity of BVP were standing on foam with eyes closed and even eyes open. This means that proprioceptive deprivation heavily destabilizes these patients even when visual control is provided. This is in accordance with the above-mentioned study on the risk of falls in BVP [6] as well as an experimental psychophysical evaluation with a reverse engineering approach, based on Bayesian inference principles, on sensory reweighting in BVP [8]. All in all, such a simple test (standing on foam) should be added to the bedside examination.

The various causes of BVP were studied in a retrospective case review on 154 patients [9]. The cause could be determined in 47%, a probable cause was found in 22% (with 20 different causes), and in 31%, it remained idiopathic. In the latter, the percentage of migraine was higher than in the non-idiopathic group (50 vs. 11%, $P < 0.001$). Among all patients, 23% had known autoimmune disorders in their medical history. In another case series of 126 patients with 'idiopathic' BVP, 15 patients (12%) had a history of a treatment with the antiarrhythmic drug amiodarone before the diagnosis of BVP [10▪]. Therefore, patients treated with this agent should be

informed about this side-effect and monitored for the development of BVP [11▪].

To evaluate the function of the three semicircular canals (SCCs), vHIT recordings of 109 patients with BVP because of aminoglycosides, Menière's disease, infectious inner-ear disorders, cerebellar-ataxia neuropathy vestibular areflexia syndrome (CANVAS), other causes, and of unknown origin ($n = 47$) were retrospectively analyzed [12]. Impaired function of the anterior SCC ($n = 86/218$) was found less often ($P < 0.001$) than impairment of the horizontal ($n = 186/218$) or posterior ($n = 194/218$) SCC. Preserved anterior SCC function was associated with aminoglycosides, Menière's disease, and BVL of unknown origin, but not inner-ear infections or CANVAS. The same group evaluated the involvement of the three SCC and the utriculus/sacculus in 101 patients [13▪]. On average [± standard deviation (SD)], the number of damaged sensors was 6.8 ± 2.2 out of 10. More sensors ($P < 0.001$) were impaired in patients with BVP because of aminoglycosides (8.1 ± 1.2) or inner-ear infections (8.7 ± 1.8) compared with Menière's disease (5.5 ± 1.5).

There is increasing evidence that central lesions or the combination of central and peripheral lesions with selective SCC involvement can mimic BVP, which is summarized in a recent article [14]: For instance, acute floccular lesions may lead to isolated bilateral horizontal SCC, whereas lesions of the vestibular nuclei can bilaterally affect horizontal and posterior SCC function only. In Wernicke encephalopathy, a specific pattern of both horizontal SCC involvement has been increasingly found by quantitative testing of SCC function, for instance, in 14 out of 14 cases with normal or minimal vertical canal impairment [15]. This is most likely related to enhanced vulnerability of the medial vestibular nuclei neurons because of thiamine deficiency.

Anterior inferior cerebellar artery infarction can lead to two types of deficits: ipsilateral deficits affecting all SCC in combination with a contralateral deficit of the horizontal SCC or a bilateral isolated horizontal SCC deficit [14]. In contrast, bilateral vestibulopathy in Gaucher is characterized by a loss of function of all SCC with minimal horizontal catch-up saccades. In genetic cerebellar ataxias and CANVAS, the function of all SCCs is reduced [14]. All in all, despite an analysis of the function of all SCC diagnosis of 'central VOR deficits' remains challenging, as BVP often does not involve all SCC.

The treatment of BVP is still challenging. Head-movement-emphasized rehabilitation led in two patients with BVP to an increase in dynamic vision because of enhanced VOR and saccadic compensation [16], findings, which have to be confirmed in a randomized controlled trial (RCT). A different approach is the use of noisy vestibular stimulation, which is based on stochastic resonance by adding an appropriate level of noise to the sensory system [17]. This improves dynamic walking stability, in particular, during slow walking (study on 13 patients with BVP) [18], increases gait speed (study on 12 patients) [19], and lowers the vestibular threshold to elicit balance-related reflexes that are required to adequately regulate postural equilibrium [20], which may be one of the underlying mechanisms.

Finally, the vestibular implant seems to be the ultimate therapy for BVP but is still at an experimental stage [21▪]. In three patients, a restoration of the angular VOR in a broad frequency range using motion-modulated electrical stimulation of the vestibular afferents was demonstrated [22]. In patients with residual vestibular function 'artificial' vestibular implant-input significantly influenced and could even counteract the response to residual 'natural' input [23]. In a single patient, it was demonstrated that using the 'translabyrinthine approach' with electrode insertion in the SCC is possible without acutely impairing hearing – as a proof-of-principle clinical investigation [24].

ACUTE UNILATERAL PERIPHERAL VESTIBULOPATHY ('VESTIBULAR NEURITIS')

Acute unilateral peripheral vestibulopathy (AUPV) is the third most common peripheral vestibular disorder, after BPPV and Menière's disease [25,26]. The term AUPV is to be preferred over 'vestibular neuritis' as it is unclear whether it is always an inflammation [27]. However, the most likely cause of AUV is viral, namely herpes simplex type 1, which is supported by a recent genome-wide association study linking the disease to single nucleotide variants of the host-factor for HSV 1 replication [28].

The clinical picture depends on the extent to which the vestibular end organs or their innervation are affected [29]. A reduced gain in the video head impulse test (VHIT) [30] and caloric testing paralysis/paresis confirm the diagnosis. Vestibular-evoked myogenic potentials are less helpful for the diagnosis but differentiate between an involvement of the superior and inferior vestibular nerves [31]. The VHIT also offers insights for prognosis, for example, a high amplitude and prevalence of overt saccades correlate with worse short-term (8 weeks) quality of life [32].

One must differentiate AUPV from acute central vestibular disorders (in particular, stroke) that can mimic AUPV and the 'HINTS plus' protocol, coupled with an emphasis on the timing and triggers of symptoms, is the most efficient clinical protocol

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[33]. Abnormalities in the clinical and quantitative head impulse test (bilateral reduced gain, increased gain, changes in vertical gain, cross-coupled vertical corrective saccades) also help to differentiate AUPV from central vestibular disorders [34].

Management of AUPV includes use of corticosteroids [27,35] and vestibular rehabilitation [36]. Vestibular suppressant drugs should only be used in the very early and symptomatic stages of the disorders and for a short term.

MENIÈRE'S DISEASE

Menière's disease is a complex syndrome defined by multiple episodes of spontaneous vertigo associated with low-to-medium frequency sensorineural hearing loss (SNHL) and fluctuating ear symptoms, such as hearing loss, tinnitus, or aural fullness that occur during the episodes [37▪]. Menière's disease is attributed to an accumulation of endolymph in the cochlear duct and it usually involves one ear (unilateral Menière's disease), but it may affect both ears (bilateral Menière's disease) from the onset of the disease or several years later [38]. The diagnosis of Menière's disease is based on the 2015 clinical criteria proposed by the International Classification Committee for Vestibular Disorders of the Barany Society [37▪], and the role of MRI in the diagnosis of Menière's disease is controversial, as endolymphatic hydrops can be found in the saccule in 10% of normal subjects and in 40% of patients with greater than 45 dB SNHL without any vestibular symptom [39]. Therefore, MRI cannot replace the diagnostic criteria of Menière's disease.

As the Menière's disease phenotype is a constellation of symptoms, clinical subtyping of Menière's disease patients may facilitate diagnosis and treatment [40]. The Meniere's Disease Consortium (a European initiative for large-scale clinical and genomic research in Menière's disease) has found five major clinical groups of patients with Menière's disease (Table 2). So, a cluster analysis using a few clinical variables, such as the onset of hearing, migraine, familial history, or a comorbid autoimmune disorder, can identify subgroups of patients with Menière's disease [38,41]. In patients with bilateral involvement, Menière's disease type 1 (which included 46% of patients) was defined by SNHL starting in one ear and involving the second ear in the following months or years but without migraine and autoimmune comorbidities. Menière's disease type 2 (17% of cases) was characterized by simultaneous onset of hearing loss in both ears without migraine or autoimmunity. Menière's disease type 3 (13%) included families with Menière's disease, and type 4 (12%) was associated with migraine in all cases. Menière's disease type 5 represented 11% and was associated with an autoimmune disease.

One thousand and seventy-three patients with unilateral Menière's disease were analyzed and some of the predictors, such as familial Menière's disease, migraine, and autoimmune disease, were found in patients with bilateral and unilateral involvement [41]. Unilateral Menière's disease type 1 was observed in 53% of cases and it included patients without a familial history of Menière's disease, migraine, or autoimmune comorbidity; Menière's disease type 2 was termed delayed Menière's disease and was a rare condition (8%) characterized by SNHL, which occurred before the vertigo episodes; familial Menière's disease or type 3 (13%) included

Table 2. Clinical groups of patients with Menière's disease

Unilateral Menière's disease	
Type 1	Sporadic Menière's disease (if concurrent migraine, autoimmune disease, or familial Menière's disease is observed, patients do not belong to this subgroup)
Type 2	Delayed Menière's disease (hearing loss precedes vertigo attacks by months or years)
Type 3	Familial Menière's disease (at least two patients in the first or second degree)
Type 4	Sporadic Menière's disease with migraine (temporal relationship not required)
Type 5	Sporadic Menière's disease plus an autoimmune disease
Bilateral Menière's disease	
Type 1	Unilateral hearing loss becomes bilateral
Type 2	Sporadic, simultaneous hearing loss (usually symmetric)
Type 3	Familial Menière's disease (most families have bilateral hearing loss, but unilateral patients may coexist in the same family)
Type 4	Sporadic Menière's disease with migraine
Type 5	Sporadic Menière's disease with an autoimmune disease

all familial cases of Menière's disease, although some patients in these families may show unilateral SNHL; Menière's disease type 4 (15%) was associated with migraine with or without aura; and Menière's disease type 5 (11%) was associated with a concurrent autoimmune disorder.

The syndrome is associated with several comorbidities including brain disorders, such as migraine [42–44], anxiety or depression [45,46], and autoimmune disorders, such rheumatoid arthritis or psoriasis [47,48]. Familial aggregation has been found in 6–9% of cases [49,50]. Most of these families show an autosomal dominant inheritance with variations in penetrance and expressivity (partial phenotype), leading to clinical differences in familial Menière's disease; rare allelic variants in a few genes, such as *COCH, DTNA, FAM136A, PRKCB, DPT* and *SEMA3D* genes, have been linked to familial Menière's disease, suggesting a genetic heterogeneity [51–53].

Several new pieces of evidence support an innate immune dysfunction in Menière's disease. First, the genetic marker rs4947296 was found to be associated with bilateral Menière's disease and this allelic variant is a trans-expression quantitative trait locus that regulates gene expression in the TWEAK/Fn14 pathway in lymphoid cells from Menière's disease patients [54▪▪]. So, although further studies should validate this finding, the Fn14 receptor and NF-κB are potential targets for drug therapy for carriers of the risk genotype in Menière's disease; second, a subset of Menière's disease patients have higher basal levels of proinflammatory cytokines, such as IL-1β, IL-6 and TNF-α, and exposure to *Aspergillus* and *Penicillium* spp. extracts may trigger additional TNF-α release and help to exacerbate the inflammatory response [55▪].

Evidence to support a treatment for Menière's disease is limited [56▪,57,58]. The European Academy of Otology & Neurotology (EAONO) Working Group on Vertigo has published a position statement paper to manage therapy in patients with Menière's disease [59]. The treatments include betahistine, diet recommendations, diuretics, intratympanic steroids, endolymphatic sac surgery, intratympanic gentamicin, labyrinthectomy, and vestibular neurectomy. However, there is an urgent need for randomized clinical trials in Menière's disease with a better selection of patients according to the clinical subtypes of Menière's disease.

BENIGN PAROXYSMAL POSITIONAL VERTIGO

BPPV is one of the most prevalent vestibular disorders, despite being largely underdiagnosed [25]. Screening techniques for BPPV either with tailored questionnaires [60] or 'smart' devices [61] have increased the accuracy of epidemiological estimates of its prevalence.

The diagnostic criteria for posterior canal and horizontal canal BPPV variants (pcBPPV, hcBPPV) are well defined [62]. hcBPPV can also show spontaneous horizontal nystagmus in the sitting position that reverses direction when putting the head forward ('bow and lean' nystagmus). These findings help to identify the pathological side when the intensity of the nystagmus does not change depending upon which ear is down (normally in the supine right or left head roll test the nystagmus is most intense when it is beating toward the pathological ear) [63].

Anterior canal (ac)BPPV [64], apogeotropic pcBPPV [65] and subjective BPPV without positional nystagmus [66] are controversial and there is no consensus on their diagnosis and treatment. The diagnosis of acBPPV is a challenge both in understanding its mechanism and in identifying the affected side. In acBPPV (and apogeotropic pcBPPV) the Dix-Hallpike and/or deep head-hanging maneuvers elicit downbeat nystagmus often with a torsional component. The reversal of nystagmus when resuming the sitting position is rarely observed, whereas generally symptoms are more severe on returning upright than in the Dix-Hallpike position. Recently, transient downbeat nystagmus on sitting up has been described just after an Epley maneuver in pcBPPV [67]. This finding is in accordance with the idea that positional downbeating nystagmus attributed to acBPPV may arise from otoconia displaced in the long arm of pc (close to common crus).

When pcBPPV and geotropic hcBPPV have their typical patterns, the cause is almost never a central vestibular lesion. Radiographic imaging and additional vestibular testing are not necessary in patients who meet the diagnostic criteria for BPPV in the absence of additional vestibular/neurological signs and/or symptoms inconsistent with BPPV [68].

On the other hand, one must distinguish the apogeotropic variant of hcBPPV from a central (usually vestibulocerebellum) lesion. The characteristics of spontaneous and positional nystagmus in peripheral and central disorders have been investigated. The main difference is represented by the greater intensity of nystagmus in the supine vs. the sitting position in hcBPPV [69▪]. Migraine may also present with horizontal direction changing positional nystagmus and vertigo [70]. acBPPV (positional downbeat nystagmus) has to be distinguished from central vestibular disorders [71].

The gold standard treatments for pcBPPV are the Epley and Semont maneuvers. Both these treatments are classified with Level 1 efficacy based on

evidence-based medicine [68], with an excellent success rate. The efficacy of treatment does not depend on the age of patients or preexisting neurological disorders [72].

The Gufoni maneuver for hcBPPV is the only treatment that has been shown to have Level 1 ~~efficacy~~. Other commonly used treatments for hcBPPV are the barbecue maneuver and forced prolonged position. A new quick maneuver was described for geotropic hcBPPV [73]. A new treatment strategy was developed for apogeotropic hcBPPV to treat otoconial debris both in the anterior arm of the semicircular canal and attached to the utricular side of the cupula [74]. In a preliminary study, another maneuver, associated with mastoid vibration of the affected ear with the head turned 135° to the lesion side in the supine position, was effective in treating apogeotropic hcBPPV [75].

Treating BPPV with vestibular suppressant medications (antihistamines and/or benzodiazepines) as well as postmaneuver restrictions of head movements or sleeping positions is not recommended [68]. In case of refractory BPPV or in special cases (severely obese ~~subjects~~), particle repositioning chairs may be used [76]. Unilateral surgical plugging of a semicircular canal may be indicated in severe refractory/recurrent BPPV, but only after a long follow-up and when there are no recurrences in different canals or when both sides are involved [77]. Complete recovery of patients should be defined by an absence of positional nystagmus and disappearance of symptoms during daily life activities [78]. Self-administered maneuvers in the treatment of recurrent BPPV may not be effective because recurrences may not be from the same canal (only 24% same side and same canal) [79]. Residual dizziness and anxiety are common findings in BPPV patients [80,81] and their specific treatment improves long-term quality of life.

VESTIBULAR PAROXYSMIA

The leading symptom of vestibular paroxysmia, a rare vestibular disorder, is recurrent spontaneous attacks of vertigo, typically lasting less than 1 min. This disease was re-classified [82] with two subtypes: vestibular paroxysmia and probable vestibular paroxysmia with the major difference being the response to carbamazepine/oxcarbazepine (Table 3).

The most likely pathomechanism is a neurovascular compression (NVC) of the rostroventral part of the eighth cranial nerve in the transition zone or proximal to the transition zone where the nerve is covered by oligodendrocytes, and therefore more vulnerable. The eighth nerve has a transition zone of 11 mm (e.g. 4 mm for the fifth cranial nerve and 2.5 mm for the facial nerve), with symptomatic NVC typically at the internal auditory canal, in particular when associated with nerve displacement and atrophy. [83] However, the role of MRI for the diagnosis of vestibular paroxysmia has still to be elucidated, in particular, as a high percentage of healthy subjects also show a neurovascular compression.

Table 3. Diagnostic criteria for vestibular paroxysmia according to the Classification Committee of the Bàràny Society

Vestibular paroxysmia (each point needs to be fulfilled)
At least 10 attacks of spontaneous spinning or nonspinning vertigo
Duration less than 1 min
Stereotyped phenomenology in a particular patient
Response to treatment with carbamazepine/oxcarbazepine
Not better accounted for by another diagnosis
Probable vestibular paroxysmia (each point needs to be fulfilled)
At least five attacks of spinning or nonspinning vertigo
Duration less than 5 min
Spontaneous occurrence or provoked by certain head movements
Stereotyped phenomenology in a particular patient
Not better accounted for by another diagnosis

Data from [82].

An analogous disease is 'typewriter tinnitus.' Two recent studies conclude that history-taking in terms of the psychoacoustic characteristics and the response to carbamazepine are more reliable diagnostic clues than are radiological or neurophysiological data [84,85], which is similar to the diagnostic criteria of vestibular paroxysmia.

In a well documented case with NVC of the right vestibular nerve, video-oculography revealed persistent left-beating nystagmus, which reversed every 47 s to right-beating nystagmus for 10 s. The periodicity of vertigo with paroxysmal nystagmus was explained by direct pulsatile compression with ephaptic discharges in the peripheral vestibular nerve or secondary central hyperactivity in the vestibular nuclei, which is induced and maintained by long-standing compression [86].

Various conditions can mimic vestibular paroxysmia, for instance a cerebellopontine angle meningioma [87] or even a lower brainstem melanocytoma [88], the latter leading to paroxysmal brainstem attacks. Therefore, contrast-enhanced magnetic resonance (MR) imaging of the brainstem and cerebellopontine angle should be performed in patients even with a typical history of vestibular paroxysmia.

The treatment of choice for neurovascular compression syndromes is so-called sodium channel blockers, such as carbamazepine (50–200 mg three

times daily) or oxcarbazepine (100–300 mg three times daily) [89] and the response supports the diagnosis ex juvantibus. For vestibular paroxysmia only one randomized double-blind, placebo-controlled trial has been reported so far: the Vestparoxy, a cross-over trial with oxcarbazepine [90▪]. The number of attacks during the observed days ratio was 0.53 [95% confidence interval (CI) 0.42–0.68, $P < 0.001$] under oxcarbazepine compared with placebo. However, the drop-out rate was 90% because of side-effects of the agent. Thus, there is a need for further RCTs using other sodium channel blockers, which are better tolerated, such as lacosamide.

SUPERIOR CANAL DESHISCENCE SYNDROME

Superior canal dehiscence syndrome (SCDS) is a rare disease caused by the loss of the bone overlying the superior semicircular canal leading to the creation of a 'mobile third window' in the inner ear ward [26,27,91▪,92]. SCDS may include auditory symptoms (autophony, bone-conducted hyperacusis, pulsatile tinnitus, and low-frequency hearing loss) and vestibular symptoms (Tullio phenomenon, Hennebert's sign, oscillopsia, vertigo, or chronic disequilibrium), most of which are triggered by loud sounds or pressure stimuli [92].

In addition to clinical symptoms, air-conducted ocular vestibular-evoked myogenic potentials are able to identify most dizzy subjects affected by SCDS, with high sensitivity and specificity, and they represent an ideal screening test for SCDS [93]. However, high-resolution CT temporal scans are considered the gold standard to confirm unilateral or bilateral SCDS [92].

Individuals with mild symptoms and SCDS may be managed conservatively with observation. However, if patients show incapacitating symptoms, surgical treatment is recommended. Currently, the middle fossa approach is considered the standard surgical procedure to repair a superior canal dehiscence, either plugging or resurfacing the canal [26]; however, transmastoid, endoscopic, or transcanal (to reinforce the round window) approaches have also been used [43,94]. Currently, there is no consensus on the best technique to treat SCDS and further studies are needed including outcome measures with assessment of the vestibular and auditory function before and after surgical treatment.

CONCLUSION

The diagnosis of BVP can now be precisely made. In terms of the cause, most cases remain even nowadays 'idiopathic.' In AUVP, there is need for more clinical trials, in particular to evaluate the effect of steroids and of drugs for symptomatic treatment and improvement of central compensation. Menière's disease is a set of various disorders of the inner ear associated with several comorbidities and the implementation of precision medicine will improve its clinical management. More RCTs are necessary for its treatment. BPPV is one of the most prevalent vestibular disorders and it accounts from a number of different variants that can be diagnosed and treated effectively. More RCTs are needed for vestibular paroxysmia. SDCS diagnosis must be confirmed by VEMPs and high-resolution CT before planning surgical therapy. Finally, for the treatment of Menière's disease and AUVP, there is a high need of RCTs.

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Conflicts of interest

M.S. is Joint Chief Editor of the Journal of Neurology, Editor in Chief of Frontiers of Neuro-otology and Section Editor of F1000. He has received speaker's honoraria from Abbott, Actelion, Auris Medical, Biogen, Eisai, Grünenthal, GSK, Henning Pharma, Interacoustics, Merck, MSD, Otometrics, Pierre-Fabre, TEVA, UCB. He is a shareholder of IntraBio. He acts as a consultant for Abbott, Actelion, AurisMedical, Heel, IntraBio, and Sensorion.

M.M. is Review Editor of Frontiers in Neuro-otology. He has received speaker honoraria from: Sensorion, CRS Amplifon and Sun Pharma limited. J.A.L.-E. is Associate Editor of Frontiers of Neuro-otology and Editorial Board member of Current Otolaryngology Reports and Scientific Reports.

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Papers of particular interest, published within the annual period of review, have been highlighted as:

- ▪ of special interest
- ▪▪ of outstanding interest

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