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Review

Why and when to refer patients for vestibular evoked myogenic potentials: A critical review

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HIGHLIGHTS

- Cervical and ocular vestibular evoked myogenic potentials (cVEMPs, oVEMPs) in almost all cases of vestibular involvement can help both in diagnosis and in prognosis.
- More research is needed to understand the applications of both c- and oVEMPs, especially the latter.
- A significant percentage of published data have neglected one or more vital VEMP parameters.

ABSTRACT

Cervical and ocular vestibular evoked myogenic potentials (cVEMPs and oVEMPs respectively) are now used by an increasing number of laboratories to evaluate otolith inner ear function and their pathways through the central nervous system. However, the literature is incomplete or unclear as to what information both c- and oVEMPs can add beyond what a good clinical examination can provide, and what other paramedical tests can provide also, and the present review aims to clarify what is known so far. The following review will describe what is known with regards to both c- and oVEMPs and their use. MEDLINE (accessed by PubMed, years 1994–2018) was searched with the following string: ("vestibular evoked myogenic potentials" [all fields]). Only articles published in English were evaluated. Both c- and oVEMPs are useful not only for confirming the presence of superior semicircular canal dehiscence (SSCD), but also for confirming the presence of acoustic neuromas when MRI is not available, bilateral vestibulopathies, inferior vestibular neuritis and vestibular dysfunction in inherited neuropathies. Further work is required, especially with respect to oVEMPs. The usefulness of both c- and oVEMPs goes beyond the confirmation of SSCDs, and is useful in many clinical cases.

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1. Introduction

The vestibular evoked myogenic evoked potential (VEMP) is a short-latency vestibular reflex recorded either from the sternocleidomastoid muscle (cervical VEMPs, cVEMPs) or from the inferior oblique muscle (ocular VEMPs, oVEMPs) (Colebatch et al., 1994; Rosengren et al., 2005; Iwasaki et al., 2007; Todd et al., 2007). Stimulation methods used most frequently include air-conducted (AC) sound, bone-conducted vibration (BC), head taps and, more recently, impulsive lateral acceleration (Papathanasiou et al., 2014). The technical demands of galvanic stimulation have limited the application of this stimulus to date. Air-conducted sound relies on an intact middle ear apparatus, and it has been reported that even mild conductive hearing loss can result in abnormal VEMP responses. Using 500 Hz tone, a sound intensity of 120–140 dB SPL is needed to stimulate the otolithic end organs adequately (Papathanasiou et al., 2014). Bone-conducted vibration can be used in cases of conductive hearing loss, bypassing the outer and middle ear. Many studies have provided useful information by comparing air-conducted sound and bone-conducted vibration, as shown in this review, but the great majority of studies use the former as the only form of stimulation. BC is likely to have advantages over AC for oVEMPs (e.g. Rosengren et al., 2011) but is more demanding technically. There have been few consistent comparisons between AC and BC findings and it is possible this may provide additional information, given the mechanism of action of the two is likely to be different.

The reflexes are thought to reflect otolith function because animal studies have shown greater activation of otolith afferents by both air-conducted (AC) sound and bone-conducted (BC) vibration (Curthoys et al., 2006; Zhu et al., 2014). Semicircular canal afferents are also activated (Zhu et al., 2014), but they appear to be less sensitive in the normal ear, although there is disagreement about the relative degree of canal involvement. This evidence, combined with the results of animal experiments showing the otolith projections to the neck and eye muscles and human experiments showing the pattern of both c- and oVEMPs in normal volunteers and patients with selective vestibular lesions (Rosengren and Kingma, 2013), supports the view that the reflexes mainly reflect otolith function.

Cervical VEMPs originate predominantly in the saccule (Papathanasiou et al., 2014; Govender et al., 2015). In contrast, oVEMPs appear to be mainly utricular in origin and are transmitted along otolith afferents in the superior vestibular nerve (Govender et al., 2015). Until recently, the contribution of the two otolith organs in producing oVEMPs was the subject of debate (Colebatch, 2010; Papathanasiou 2012, 2013b; Todd, 2014). However, recent evidence using impulsive lateral acceleration in patients with superior vestibular neuritis, and using extracellular single neuron recordings in guinea pigs, has finally shown that oVEMPs are mainly utricular in origin, whatever the stimulus (Curthoys et al., 2014; Govender et al., 2015; Papathanasiou, 2015).

Both c- and oVEMPs have become an increasingly popular addition to the neuro-otology test battery. As more laboratories have published data on cVEMPs as a measure of vestibular function, a wide range of recording methods exist. Such variations in methodology and interpretation can be confusing to clinicians and limit potential comparisons of cVEMP data across laboratories. International guidelines on the clinical application of cVEMPs have recently been published to alleviate the problem associated with the heterogeneity of the methods used, in the hope that the technique would be understandable to more people (Papathanasiou et al., 2014). Because of recent developments with regards to the utricular origin of oVEMPs, as noted above, guidelines for oVEMPs are now in the process of being prepared.

While the guidelines for cVEMPs were welcomed as a methodological aid, colleagues interested in implementing both c- and oVEMP techniques in their practice frequently ask what the clinical purpose of both c- and oVEMPs really are. This prompted the writing of the present review, which will describe what is known with regards to the role of both c- and oVEMPs and their use in neuro-otology and neurology. Reviews have been published before with regards to both c- and oVEMPs, with the last comprehensive one having been published in 2016 (Colebatch et al., 2016). Since then and around this time, the clinical usefulness of both c- and oVEMPs has been investigated across many conditions, not only in Superior Semicircular Canal Syndrome (SSCD), but in many other cases where otolith involvement is suspected. More recently, a practice guideline has been published by the American Academy of Neurology (Fife et al., 2017). We do not agree that the usefulness of the VEMP in identifying vestibular disorders is confined to superior canal dehiscence (SCD). For instance, it has been shown that oVEMPs are more effective at identifying chronic unilateral vestibular loss than visual vertical measurements (subjective visual vertical during whole-body tilt or eccentric rotation) (Valko et al., 2011). The paper by the American Academy of Neurology does not investigate in a clinical-practical sense the possible usefulness of VEMPs in many neurological and otorhinolaryngological conditions, when this method is applied as an element of a whole battery of auxiliary vestibular tests. The lack of knowledge about the clinical usefulness of cVEMPs was recently reflected in a survey carried out amongst pediatric otorhinolaryngologists

(Dargie et al., 2014), where it was shown that 16% of respondents had not heard of a cVEMP testing, and the majority of the remaining survey participants were unsure about the clinical usefulness of cVEMPs.

The purpose of this review is to let the reader appreciate that both c- and oVEMPs provide useful information in patients with symptoms that may be attributed to the vestibular system, such as vertigo, dizziness and imbalance. Both c- and oVEMPs can help in the differential diagnosis, in tracking a disease process, or in assessing residual vestibular function. As both c- and oVEMPs reflect mainly otolith function, they can be used to assess the involvement of the otolith organs in patients with vestibular disorders and determine the extent of the otolith lesion. Alternative tests of otolith function either involve stimulation with linear acceleration, eccentric rotation or head tilt, which require sophisticated testing and recording equipment, or test the subjective visual perception of vertical or horizontal lines, which often recovers after the acute phase of vestibular illness due to central compensation. In contrast, both c- and oVEMPs are easier for both the tester and the patient and they typically remain abnormal after central compensation has occurred. Moreover, both c- and oVEMPs can be used to diagnose both chronic and acute otolith end organ and pathway (peripheral or central) dysfunction.

Most often, abnormal c- and oVEMPs point to peripheral vestibular problems, either in the labyrinth or along the vestibular nerve. These include Meniere's Disease (MD), vestibular schwannoma (VS), vestibular neuritis (VN), SSCD or labyrinthine stroke. The reflexes underlying both c- and oVEMPs, however, also depend upon central pathways through the brainstem, i.e. the vestibulocollic and vestibulo-ocular pathways for cVEMPs and oVEMPs respectively; thus test abnormalities can also be caused by disorders of the central nervous system associated with vertigo, such as stroke or multiple sclerosis (MS) within the brainstem. Both c- and oVEMPs are usually normal in disorders that do not produce obvious lesions, like vestibular migraine (VM), benign positioning vertigo (BPV) or chronic subjective dizziness (CSD). Both c- and oVEMP abnormalities are sometimes found in other 'non-vestibular' diseases, though mostly as an incidental finding.

A reduced amplitude or absent c- or oVEMP response indicates loss of otolith function or signal, or damage to the reflex pathway, or both. Like the caloric and head impulse tests, an abnormal result is usually not specific to a particular disease. There are two important exceptions to this: large reflexes with pathologically low thresholds are specific abnormalities associated with third window disorders, such as SSCD (Welgampola et al., 2008), and altered frequency tuning has been associated with MD (Rauch et al., 2004).

An abnormal c- or oVEMP is best interpreted in the context of the other audiometric and vestibular test results. The results of audiometric tests are particularly important when AC sound stimulation is used to evoke c- or oVEMPs, as conductive hearing loss will diminish the impact of the stimulus on the vestibule in the same way that it reduces the sound energy reaching the cochlea. The results of other vestibular tests are useful as they help gauge the likelihood that a c- or oVEMP abnormality indicates a true otolith lesion, and together with tests of canal function they can confirm the affected nerve divisions. Some normal subjects in the literature have been suggested to have abnormal c- or oVEMPs. It may be possible that this may be a result of not fully taking into account the patient's effort to contract target muscles, especially in the elderly. This has not been proven objectively in the literature as such, but this is something that needs to be kept in mind. There is increasing awareness of the fact that some cases may involve pure otolith organ dysfunction, originating either in the saccule or the utricle, and this is expanded in this review. Careful interpretation is especially required (a) in older patients and (b) if for cVEMPS a measure of the SCM muscle contraction is not available. Concerning this last point, it is important to note that cVEMPs can only be recorded during a tonic contraction of the SCM muscle and that cVEMP amplitude increases with increasing contraction strength. Failure to produce a sufficient and symmetric contraction can produce erroneous test abnormalities (e.g. McCaslin et al., 2013). Likewise, the oVEMP is contingent on up-gaze.

This review is restricted to the clinical significance of both c- and oVEMP testing. We will not describe c- or oVEMP methodology.

2. Methods

MEDLINE (accessed by PubMed, years 1994–2018 was searched with the following string: ("vestibular evoked myogenic potentials" [all fields]). Only articles published in English were evaluated. One author (ESP) independently screened all titles and abstracts of the articles identified by the searches to assess their eligibility, and excluded those that did not meet the selection criteria shown below.

Manuscripts were evaluated only if they fulfilled *all* of the following criteria:

(1) Papers whose main purpose was evaluation of the clinical application of either c- or oVEMPs or both. However, if possible clinical applications were included in the latter papers, these were included if determined to be relevant.
(2) Galvanic stimulation (GVS) was not the only method of stimulation used. This technique may be well suited to investigating central disorders of vestibular function but the technical demands have limited the application of this stimulus to date (Papathanasiou et al., 2014).
(3) The manuscript evaluated latencies or amplitudes both on a quantitative level. However, if the evaluation of only latencies was enough to draw valid clinically relevant conclusions from their studies, the paper was included in this review.
(4) Click or tone intensities that were sufficient for eliciting c- or oVEMPs (e.g. click intensities of more than 100 dB nHL).
(5) Papers that contained details of SCM contraction.
(6) Papers that analysed c- or oVEMP results separately, i.e. did not evaluate them only as part of a larger test battery. The c- or oVEMP or both parameters were clearly outlined and separate from the other paramedical examinations.

The exclusion criteria were:

(1) Papers whose sole aim was to determine the pathway of either c- or oVEMP.
(2) The only criterion for an abnormal VEMP was an absent response

3. Results of manuscript evaluations based on specific disease type

The search strategy yielded 1006 articles. After removing duplicates, reading abstracts and titles, and applying the above inclusion criteria, 107 were finally included in the analysis.

3.1. Diseases of the peripheral vestibular system

3.1.1. Superior semicircular canal dehiscence (SSCD)

Superior semicircular canal dehiscence is caused by a dehiscence of the bone overlying the superior semicircular canal and is characterized by sound or pressure induced vertigo and oscillopsia (Minor et al., 1998; Minor, 2000; Pfammatter et al., 2010), with pressure changes either in the middle ear or intracranially (Kanaan

et al., 2011), bone conduction hyperacusis and pulsatile tinnitus (Ward et al., 2017), autophony and hearing loss (Palma-Diaz et al., 2017). The temporal bone separates the superior canal from the middle cranial fossa or superior petrosal sinus, and a dehiscence creates novel pathways for sound transmission (Teixido et al., 2008), including heightened sensitivity to bone-conducted sound (Cox et al., 2003). Although the superior semicircular canal is directly involved pathoanatomically, the eye movements seen in this condition have been shown to be attributed not only to stimulation of the above canal but also to stimulation of the otoliths (Miller, 1961; Curthoys, 1987; Watson et al., 2000). With the dehiscence acting as a "third window", a path of lower impedance is created for the transmission of pressure and acoustic energy to the vestibule, leading to lower (both c- and o) VEMP thresholds and elevated amplitudes (Brantberg et al., 1999; Streubel et al., 2001; Minor, 2005; Chien et al., 2007; Songer and Rosowski, 2007; Welgampola et al., 2008; Janky et al., 2013). Conventional tests of vestibular function are often normal in cases of SSCD (e.g. caloric, tympanometry, acoustic reflexes and rotational testing), with only the demonstration of eye movements in response to loud tones being confirmatory of its presence (Watson et al., 2000). Routine neurologic (of the nervous system in general) and otologic (of the ear and related structures) examinations can be unremarkable. The cause of the dehiscence is usually unknown, although a potential genetic contribution has been proposed (Heidenreich et al., 2017).

It is important to note that abnormalities on computed tomography (CT) scans have been found frequently to be present bilaterally and thus also present in asymptomatic ears. Three to 9% of patients who have undergone CT imaging for other reasons have anatomic SSCD (defined as a dehiscent SSCD on CT-scan) (Niesten et al., 2012). It is therefore not possible to distinguish between frank dehiscence and an extremely thin plate of bone overlying the canal (Sequeira et al., 2011; Thabet, 2014), and a similar scan appearance can occasionally be seen in patients scanned for other reasons. Using both c- and oVEMPs therefore provides confirmation of the presence of SSCD. There is at least one publication that has reported VEMP abnormalities in the presence of near dehiscence (Ward et al., 2013), and so a frank dehiscence is not necessary.

Click-evoked thresholds with respect to cVEMPs are low for all affected ears and normal for unaffected ears (Colebatch et al., 1998; Brantberg et al., 1999; Halmagyi et al., 2003; Rosengren et al., 2008). If a cVEMP can be consistently elicited at 70 dB nHL, it suggests that the patient has SSCD (Halmagyi and Curthoys, 1999). Thresholds are important to measure with respect to cVEMPs (Rosengren et al., 2008), as their amplitudes, although larger than usual, do not always increase to above the upper limit of normal, nor is an interside amplitude difference useful. The amplitudes of oVEMPs related to the symptomatic ear clearly stand out above the range of normal values in any laboratory, and measuring thresholds is usually not necessary (Rosengren et al., 2008). This has been further confirmed with surgically-confirmed SSCD, where air-conducted oVEMPs (particularly using tone instead of click stimuli) have the largest amplitudes and less overlap with the normal population (Janky et al., 2013). Reflex hammer stimulation at Fz' is a poor stimulus for evaluating for SSCD when amplitude only is evaluated, both with respect to oVEMPs and cVEMPs. However, with regards to latency the oVEMP response with tendon hammer tap at Fz' is significantly prolonged compared to normal controls (Taylor et al., 2014). The size of the SSCD measured during surgery was not found to be correlated with the o- and cVEMP results (Janky et al., 2013). Both air-conducted cVEMPs and oVEMPs reveal reduced thresholds, whereas bone vibration does not follow a similar pattern and can have normal thresholds in SSCD (Welgampola et al., 2008).

Vestibular evoked myogenic potentials have been found to be useful in the differential diagnosis of otosclerosis and SSCD (Merchant et al., 2007), although sensitivity and specificity values were apparently not quoted in this manuscript. Such cases have also been noted by others and do not seem to be a rare problem (Streubel et al., 2001; Li et al., 2011). An air-bone gap on audiological testing can be due to a dehiscence either shunting sound away from the cochlea and/or increasing the inequality in the impedance between the scala tympani and the scala vestibuli. With such an air-bone gap, especially when bone conduction thresholds are close to zero or normal, VEMPs are useful to show an abnormally low threshold for the cVEMP and/or increased amplitude for the oVEMP. Performing VEMPs seem to be useful as a screening tool before the patient has a high resolution CT of the temporal bone. The performance of acoustic/stapedius reflex is also suggested as a confirmation of an ossicular etiology of conductive hearing loss before the patient goes to surgery (Zhou et al., 2012; Hong et al., 2015). However, this reflex test has been reported to be limited if the tympanic membrane is not intact (Zhou et al., 2012). In this latter study, 30–60% of patients did not have this reflex performed for this reason, but both c- and oVEMPs were performed in all of them, leading to absent responses in patients with middle ear pathologies. Therefore, performing both c- and oVEMPs is useful when the stapedius reflex cannot be performed for the above reasons.

Another very important point with respect to ruling out otosclerosis and the use of both c- and oVEMPs is that the latter should be absent if the former condition exists. In other words, the presence of both c- and oVEMPs rules out the presence of otosclerosis (Cox et al., 2003; Minor et al., 2003; Chien et al., 2011). The finding of a normal (c- and o-) VEMP would not be expected based on a middle ear cause of conductive hearing loss.

cVEMPs also seem to be diagnostically helpful when symptoms of SSCD are present in both ears or when the CT scan shows bilateral dehiscence in the roofs of the SSCs, and a patient may not always be able to report which ear is worse based on symptoms (Dournes et al., 2012; Niesten et al., 2013). As the worse ear is recommended to be repaired first, this ear has been found to have the lower cVEMP threshold (Dournes et al., 2012). Normalization of the cVEMP and oVEMP responses in the form of increased threshold and decreased amplitude is an indication of successful surgery (Welgampola et al., 2008; Rodgers et al., 2016). Recurrent symptoms following surgery revealed persistently low thresholds and large amplitude (both c- and o-) VEMP responses (Welgampola et al., 2008). Radiology, specifically CT, is not reliable for determining correct outcome, due to the radiotransparency of the materials used during surgery (Dournes et al., 2012).

Similar to SSCD, a lowered threshold cVEMP response was noted in a case of posterior semicircular canal dehiscence from a high riding jugular bulb in one study (Gubbels et al., 2013), but not in another (Lim et al., 2012). In both cases, neurotological testing was unremarkable. The former paper did not state whether the cVEMP result was used to confirm the presence of possible dehiscence seen on the CT, as is done with SSCD. In the latter, a low threshold oVEMP response confirmed the CT finding, and this has also been noted in a separate study (Aw et al., 2010). In this study (Lim et al., 2012), the figure depicting the oVEMP waveforms shows an accentuated waveform significantly larger than obtained from the contralateral side, and this accentuated response has also been noted elsewhere (Aw et al., 2010). It is interesting to note in this latter study that the accentuation involved predominantly the positivity of the response and not the negativity.

The presence of an oVEMP response at 4000 Hz (either air- or bone-conducted sound) has also been shown to indicate SSCD (Manzari et al., 2013), although we do not know yet if this is specific to SSCD or if it also appears in other inner ear disorders.

To summarize with respect to SSCD:

(1) Clinical Value: Highly desirable.
(2) CT cannot distinguish between frank dehiscence and an extremely thin plate of bone overlying the canal, and so both c- and oVEMPs can provide confirmation of the presence of SSCD (Niesten et al., 2012; Sequeira et al., 2011; Thabet, 2014).
(3) Normal cVEMP thresholds (and/or oVEMP amplitudes) suggest that the CT abnormalities have no functional effect (ie not SCD or near SCD).
(4) A pathological enlargement usually relates to oVEMP amplitudes.
(5) oVEMP amplitudes using air-conducted sound are large with respect to the symptomatic ear (Rosengren et al., 2008; Janky et al., 2013).
(6) cVEMPs show a reduced threshold with respect to the symptomatic ear (Colebatch et al., 1998; Brantberg et al., 1999; Halmagyi et al., 2003; Rosengren et al., 2008).
(7) Both c- and oVEMPs can show which dehiscence is likely to be symptomatic with bilateral CT findings, helping to plan surgery (Dournes et al., 2012; Niesten et al., 2013).
(8) Further studies are needed to determine the sensitivity and specificity of VEMPs in posterior canal dehiscence

3.1.2. Meniere's disease (MD)

Meniere's disease has generally been accepted to be associated with endolymphatic hydrops of the ear on histopathological studies, although whether the latter is the cause of the disease is another matter altogether, with its role in the pathogenesis and pathophysiology remaining elusive (Gulya and Schuknecht, 1982; Rauch et al., 1989; Shojaku et al., 2001; Gurkov et al., 2011; Lopez-Escamez et al., 2015). It is characterized by fluctuating hearing loss, tinnitus, aural fullness/pressure in the affected ear and rotatory vertigo (Minor et al., 2004; Ban et al., 2007; Miyashita et al., 2017). Although endolymphatic hydrops occurs most often in the cochlea, it can also occur in the saccule and less often in the utricle and semicircular canals (Fraysse et al., 1980; Schuknecht and Gulya, 1983; Okuno and Sando, 1987; Rauch, 2001; Young et al., 2003). Therefore, cVEMPs would be expected to be predominantly involved with respect to MD, followed by oVEMPs.

3.1.2.1. Differentiation between MD and normal controls. The combined use of cVEMPs (based on amplitude measurements alone) with electrocochleography, the glycerol test or the furosemide vestibulo-ocular reflex test increased the percentage of patients with one or more positive results in the above test battery (Shojaku et al., 2001). The latter authors concluded that the combined use of these four exams increased the incidence of abnormal results in patients with MD. Note that before the exam these patients had already been diagnosed based on criteria of the Meniere's Disease Research Committee of Japan and the criteria of the Japan Society for Equilibrium Research. A similar study with oVEMPs has not been done so far.

Related also to the early diagnosis of MD is the use of both c- and oVEMPs in detecting "Meniere-like" findings in the asymptomatic contralateral ear. Twenty-five to forty-eight percent of patients with unilateral MD had abnormal cVEMPs in the contralateral ear based on an increase in threshold, 45% based on a shift of best frequency away from 500 Hz towards 1 kHz, 27% with both, and also in the form of absent responses and prolonged p13 absolute latency (Ribeiro et al., 2005; Lin et al., 2006). Although a follow-up study was promised, to our knowledge it has not been published so far, and so we do not yet know if these patients will eventually develop MD in the contralateral ear. Temporal bone studies have not shown hydrops in the contralateral ear (Morita et al., 2009), although other studies showed pathological changes with respect to the stria vascularis (Kariya et al., 2007, 2009). Threshold values of cVEMPs were not found to be useful in the diagnosis of MD (differentiating between MD patients and controls) (Osei-Lah et al., 2008). Augmented amplitudes can be seen in early stages of MD (stage I and II) in a minority of cases (Young et al., 2003). Correlation of abnormal caloric responses with stage of disease was found not to be significant in the same paper.

The combined used of cVEMPs with pure tone audiometry may allow a more representative staging of MD than PTA (pure tone audiometry) alone, as MD not only affects the cochlea in the majority of cases, but also the saccule based on both latency and amplitude measurements (Paparella, 1985; Kim et al., 2013). Pure tone audiometry is considered positive when threshold is above 25–30 dB (for adults) across the speech spectrum (500–4000 Hz) (Fausti et al., 2005; Walker et al., 2013). Patients found to have milder forms of MD based on PTA (stages I and II), had different responses to cVEMPs, with some of the patients in each stage revealing absent cVEMP responses (Kim et al., 2013). It has also been shown that the above patients with abnormal cVEMP responses showed a decrease in hearing after about 20 months of follow-up. In the more severe stages (III and IV) where hearing had already deteriorated, the cVEMP response did not provide additional information about hearing outcomes. In other words, it could not predict a further deterioration in hearing function. The impact of vertigo was not examined in this study, and so information is not provided as to whether MD patients in stage I had a poorer quality of life when a cVEMP response was unobtainable. Whether patients with cVEMP present or absent should be subclassified into stages Ia or Ib, and how this would affect treatment decisions, may be considered. However, it appears that an abnormal cVEMP predicts deterioration of hearing, at least for MD stages I and II.

Frequency tuning changes, with the best VEMP responses (both c- and oVEMPs, predominantly cVEMPs) obtained shifting from a stimulus frequency of 500–1000 Hz and in response to an AC stimulus, is observed in MD patients (Rauch et al., 2004), with a specificity of 76% compared to BPPV and VM and unilateral VN (Murofushi et al., 2017a). However, why this shift in frequency preference occurs is only addressed on a few occasions in the VEMP literature, with the changes thought to be due to altered saccular motion mechanics arising from hydropic distention, at least with regards to cVEMPs (Rauch et al., 2004). The same frequency tuning changes have also been noted with oVEMPs (Singh and Barman, 2016). With regards to the frequency sensitivity itself, it is suggested that the phenomenon of frequency tuning in the vestibular end organs may be a result of an electrical resonance intrinsic to the hair cells (Ashmore, 1983; Todd et al., 2009). It is usually assessed by performing both c- and oVEMPs in the standard way, but separately using two different sound frequencies, namely 500 Hz and 1 kHz. Frequency tuning changes with oVEMPs have been observed in MD patients, with a shift from 500 Hz to 1000 Hz (Jerin et al., 2014). However, this latter paper included only patients with definite MD diagnosed through the use of hydrops MRI, and did not address the question when to use oVEMPs in the differential diagnosis of MD.

Age related changes in frequency tuning changes are known to exist, and so taking advantage of the above phenomenon needs to take age into account. Although in young individuals the best frequency to obtain both VEMPs is 500 Hz, a significant percentage (38–62%) of older individuals (greater than 60 years of age) showed the maximum amplitude at 1 kHz (Piker et al., 2013). A separate study found no frequency tuning above 60 years of age, revealing a flat threshold response curve (Janky and Shepard, 2009).

Asymmetric cVEMP abnormalities in patients with MD were also noted by Kingma and Wit with respect to amplitude (Kingma and Wit, 2011), but they stated that this feature was not useful

on an individual patient basis because of a large overlap with normal subjects. Although interside amplitude difference was found to be significant for patients with stable MD as a group compared to acute cases, this was not investigated at an individual patient level (Osei-Lah et al., 2008). Accordingly, in a separate study investigating cVEMPs on an individual level, no significant interside amplitude difference was found in patients with MD (Akkuzu et al., 2006), although this was described in another study in 96% of patients with definite MD (Seo et al., 2013b). This latter study also asserted that cVEMPs may be more sensitive than 3.0 tesla MRI after intratympanic gadolinium-diethylene-triamine pentaacetic acid injection. In 8 patients with normal hydrops MRI, only one patient had a normal cVEMP response in the affected ear.

One paper showed that it was possible to predict the onset of MD in patients who initially present with isolated episodes of vertigo alone (Lee et al., 2017). Abnormalities in cVEMPs, in the form of absent responses, prolonged absolute p13 latencies and interear amplitude difference were noted in a high percentage of patients that developed MD compared to patients that did not. oVEMPs did not show the same trend.

3.1.2.2. Differentiation between MD and vestibular migraine (VM). The tendency for the sound frequency to shift from 500 Hz to 1000 Hz (with both c- and oVEMP amplitudes higher in MD in response to 1 kHz) was also noted in VM by one study (Murofushi et al., 2009). However, these findings are not consistent between laboratories, especially with respect to MD (Papathanasiou, 2013a). In patients with MD, Taylor et al. (2011) described an asymmetry in the cVEMP amplitude ratio using 500 Hz tone air conducted sound stimulation. The authors found this asymmetry present in patients with MD, and less often found in patients with VM. In contrast, symmetric cVEMP abnormalities were frequent in patients with VM in the form of bilateral low amplitude responses (Baier et al., 2009). cVEMPs were determined to remain normal in children with VM (Brodsky et al., 2016a).

Taylor et al. showed that when auxiliary testing parameters are combined, results become more specific for either MD or VM, which allows to better differentiate between the two (Taylor et al., 2011). In other words, an amplitude asymmetry in the cVEMP responses using 500 Hz air-conducted sound becomes more sensitive and specific when this finding is combined with a better cVEMP response (in terms of amplitude) at 1 kHz compared to 500 Hz (0.5/1 kHz frequency ratio) and with an abnormally asymmetric caloric test (Taylor et al., 2011).

No significant differentiation by bone conducted oVEMPs between MD and VM was found in a separate study (Taylor et al., 2011), which confirmed the weakness of oVEMPs in differentiating the two disorders.

There are increasing reports in the literature of patients with episodic vertigo without migraine or auditory symptoms, and these patients have been described as having recurrent peripheral vestibulopathy (RPV, Attye et al., 2015; Murofushi et al., 2017b). The cause of this entity is still not known, but one study found a frequency shift with respect to cVEMPs, similar to what is encountered in MD (Murofushi et al., 2017b), suggestive of the fact that RPV may be vestibular endolymphatic hydrops. However, this requires further confirmation.

To summarize with respect to MD:

(1) Clinical value: Highly desirable.
(2) Abnormal cVEMPs can predict loss of hearing in mild forms of MD (stage I and II, based on PTA testing) (Kim et al., 2013).
(3) Asymmetrical cVEMP responses, when combined with a frequency shift from 500 Hz to 1 kHz and an asymmetrical caloric response, favours the presence of MD rather than VM (Taylor et al., 2011).
(4) Using cVEMPs can help in the diagnosis of patients with recurrent peripheral vestibulopathy, and its use suggests the presence of a vestibular endolymphatic hydrops (Attye et al., 2015; Murofushi et al., 2017b).
(5) Research with respect to MD has focused on cVEMPs predominantly, with work still remaining to be done with respect to oVEMPs. These can include:
   a. Comparing oVEMPs with electrocochleography, the glycerol test and the furosemide vestibulo-ocular reflex test.
   b. oVEMP recordings in the asymptomatic ear.
   c. Using oVEMPs as well in the representative staging of MD, and in looking for residual vestibular function after selective vestibular neurectomy.

To summarize with respect to VM:

1. Clinical value: Highly desirable.
2. cVEMPs reveal symmetrical findings, either normal bilaterally or bilateral low amplitude responses.
3. Frequency tuning is not usually noted with respect to both c- and oVEMPs.
4. Diverse findings are evident with respect to VM. Further investigations are needed as to why this should be (perhaps due to mixed or separate peripheral or central aetiologies as noted in the text).

3.1.3. Vestibular schwannoma

Vestibular schwannomas (VS), also called acoustic neuroma, is a benign and the most common tumor in the internal auditory canal arising from the Schwann cell sheath, and usually originates from the distal neurilemmal portion of the vestibular nerve near the porus at or close to the neurilemmal (Schwann cell)-glial junction (Sterkers et al., 1987; Tos et al., 1992; Moffat et al., 1993; Samii and Matthies, 1997; Lustig et al., 1998; Friedman et al., 2001; Day et al., 2008; Ushio et al., 2009b; Huang et al., 2013; Lin et al., 2014). It is usually a sporadic, unilateral tumor (Neff et al., 2006; Huang et al., 2013). It is therefore expected that both c- and oVEMPs will be affected, not only in terms of the origin of the VS but also due to compression from the neighbouring tumor if either the superior or inferior division is selectively involved. In the majority of cases, both c- and oVEMPs were found to be less sensitive than or as sensitive as some other paramedical examinations (based on amplitude and absence of response, latency not measured) in the diagnosis of VS, such as the caloric test (Ushio et al., 2009a; Piras et al., 2012) and brainstem auditory evoked potentials (BAEPs), using cVEMPs and based on amplitude measurement only (Murofushi et al., 1998; Ushio et al., 2001; Yavuz et al., 2014). Vestibular evoked myogenic potentials are also not a diagnostic replacement for MRI (Lachowska et al., 2018). However, both VEMPs were discovered to be more sensitive to schwannomas exceeding 14 mm diameter than the head impulse test based on both latency and amplitude (Taylor et al., 2015). cVEMP testing and subjective test of visual horizontal or vertical did not differ significantly in the percentage of abnormalities, based on the presence or absence of a response only (Ushio et al., 2008). VEMPs (either c- or o-) were unable to differentiate between a cerebellopontine angle meningioma and a VS based on both latency and amplitude (Hu et al., 2009; Su et al., 2013). In some cases, however, the BAEP and caloric tests were normal despite abnormal cVEMPs based on the presence or absence of a response only (Murofushi et al., 1998; Matsuzaki et al., 1999; Ushio et al., 2001) or oVEMPs and cVEMPs abnormal despite normal caloric results based on latency and amplitude or amplitude only (Halmagyi and Curthoys, 1999; Huang et al., 2012; Piras et al., 2012). Therefore, both c- and oVEMPs are useful as part of an auxiliary test battery (electrophysiology, head impulse test and calorics). This is

especially important in cases where MRI is not immediately available for economic reasons based on the presence or absence of a response only (Matsuzaki et al., 1999). Clearly, VS can selectively impair the saccular, auditory or horizontal semicircular canal afferents (Patko et al., 2003).

Information provided by BAEP and cVEMPs together may be useful for a surgeon by providing information on the relationship of cochlear and vestibular nerves in the internal auditory canal based on presence of absence of cVEMP or amplitude measurement only (Chen et al., 2002; Yavuz et al., 2014). The origin of the tumor may be a factor in hearing outcome (Brackmann et al., 2000; Jacob et al., 2007), and so determining from which nerve the tumor originates (superior or inferior vestibular nerve) may be important (He et al., 2016). However, one study has shown no correlation between the cVEMP (based on amplitude measurement and presence or absence of response only), the calorics and BAEP abnormalities and the nerve origin (Ushio et al., 2009b). Although attractive in theory, no study has investigated so far whether combined results of BAEPs and both c- and oVEMPs altered the way surgery was performed. The combined use of oVEMPs and cVEMPs can predict the size of the VS. When both are normal based on both latency and amplitude, the tumor is expected to be less than 2 cm in size, and so vestibular function is expected to be preserved in these cases after stereotactic radiosurgery (Lin et al., 2014). Absent caloric and cVEMP responses may indicate tumor size greater than 2.5 cm (Day et al., 2008). The use of both VEMPs has confirmed that preservation of the superior and inferior nerves can be achieved after Cyberknife radiosurgery based on both latency and amplitude (Lin et al., 2013), and so are useful for determining the residual function of the vestibular nerves after surgery (Chen et al., 2002). No studies have been done to determine if the results of both c- and oVEMPs or other paramedical examinations can give information about tumor progression. The above studies (Day et al., 2008; Ushio et al. 2009b; Lin et al., 2013, 2014; Yavuz et al., 2014; He et al., 2016) have shown size of the tumor at the time of the study.

Vestibular schwannomas are known to cause endolymphatic hydrops (Roosli et al., 2012), involving either the cochlea or vestibule, predominantly the latter (Naganawa et al., 2011), probably via ischemia of the cochlea due to compression, and could potentially cause the above VEMP abnormalities, rather than by direct involvement of the vestibular nerve. However, to our knowledge no study has been done comparing both c- and oVEMPs with the presence of endolymphatic hydrops due to the presence of VS.

Prehabilitation is a pretreatment plan of vestibular exercises before a planned vestibular lesion, which in this case is possible vestibular loss following surgery (Magnusson et al., 2011). However, to our knowledge no study exists relating this to both c- and oVEMPs and VS management.

To summarize with respect to VS:

(1) Clinical value: Useful.
(2) When MRI access is easy, both c- and oVEMPs do not appear to provide additional information in the presurgical workup (Lachowska et al., 2018).
(3) When MRI is not immediately available, the performance of an auxillary test battery (both c- and oVEMPs, head impulse test and calorics) provides important information with regards to vestibular function. Both c- and oVEMPs are a diagnostic clue together with the other exams and are analysed as a complementary exam, and not in place of the others (Murofushi et al., 1998; Halmagyi and Curthoys, 1999; Matsuzaki et al., 1999; Ushio et al., 2001; Huang et al., 2012; Piras et al., 2012 Taylor et al., 2015).
(4) The majority of studies looked at cVEMPs only in the absence of oVEMPS, most likely due to the later discovery of the latter. Future studies looking at oVEMPs also can help determine:
    (i) the sensitivity of oVEMPs compared to BAEPs and the subjective test of visual horizontal or vertical, and
    (ii) correlation with the nerve origin of the VS.
(5) No studies so far have investigated in detail if such an axillary test battery can predict the outcome of surgery, and such studies are needed. Nor have studies been done with respect to predicting tumor progression.
(6) However, following surgery, both c- and oVEMPs are useful in determining the presence of residual vestibular nerve function (Chen et al., 2002; Lin et al., 2013).
(7) A significant percentage of the studies found did not analyse latency, which underestimated the sensitivity of both c- and oVEMPs, specifically with respect to investigating:
    (i) the sensitivity of both c- and oVEMPs compared to other paramedical examinations, and
    (ii) determining the nerve origin of the VS.

3.1.4. Cochlear implantation

Cochlear implantation is used for the rehabilitation of patients with postlingual deafness (Wagner et al., 2010). But surgical complications can include dizziness disorders (Enticott et al., 2006; Basta et al., 2008; Melvin et al., 2009), with implantation found to cause injury to vestibular end organs, predominantly the saccule (Tien and Linthicum, 2002; Nordfalk et al., 2014). Therefore both c- and oVEMPs are expected to be involved in at least some cases. Some papers could not be reliably evaluated, for example, as the only criterion for an abnormal VEMP (both c- and o-) was an absent response (Wagner et al., 2010; Nordfalk et al., 2014; Janky and Givens, 2015; Thierry et al., 2015), which significantly underestimated the sensitivity of the VEMP examination. Also, the sound intensity was too low (e.g. 74 dB nHL, Devroede et al., 2016), and muscle contraction was not monitored for the cVEMP to reduce interindividual variability (Janky and Givens, 2015; Xu et al., 2015, 2017). In the latter papers (Xu et al., 2015, 2017), only oVEMP recordings were considered as they are not reliant on muscle monitoring. In the normal structure group, with normal middle and inner ear structures on CT, the response rate was 65% (Xu et al., 2017). In the abnormal structure group, with evidence of large vestibular aqueduct syndrome or Mondini dysplasia on CT, the response rate was 100%, with the latter showing significantly lower thresholds and larger amplitudes compared to the normal structure group. oVEMP response rate has been reported to decrease by 12.9% following surgery, even with the device off (Xu et al., 2015), involving elevation of threshold, decrease of amplitude and prolonged latencies. At least with regards to corrected amplitudes, cVEMPs were reported or shown in the results table to remain unchanged following implantation (Tsukada et al., 2013; Ajalloueyan et al., 2017). However, cVEMP deterioration (details were not provided) are reported in two separate studies by the same group in 62%-86% of cases following surgery (Krause et al., 2010; Louza et al., 2015). However, this did not correlate with the presence or absence of vertigo symptoms. A separate study also found deterioration of cVEMP responses following surgery and with the cochlear implant turned on rather than off, based on response rate, latency and amplitude (Demirhan et al., 2016). Electrode insertion depth was found not to be correlated with alterations in measurable cVEMP results (Louza et al., 2015).

To summarize with respect to cochlear implants:

1. Clinical value: Difficult to determine based on the available literature, and more studies with good methodology is needed.

2. Poor methodology is noted in a significant number of papers. Therefore, repeat studies with good methodology are worthwhile.
3. Accentuated oVEMP responses noted in abnormal structure cases, suggestive of easier sound access to the inner ear.
4. Decreased oVEMP responses following surgery
5. Poor cVEMP documentation with regards to effects following surgery. However, one study found deterioration. Further studies are needed to confirm this.

3.1.5. Vestibular neuritis (VN)

Vestibular neuritis is characterized by acute prolonged vertigo (≥24 h to over several days), nausea and vomiting without other parallel auditory or neurological symptoms and signs, commonly attributed to viral infection of the (usually superior) vestibular nerve (Dix and Hallpike, 1952; Baloh et al., 1996; Strupp and Brandt, 1999; Kim et al., 2008; Taylor et al., 2016). Prolonged oVEMP responses, found at six days after symptom onset, have been shown to be predictive of good outcome after 6 months of oral prednisone treatment (Adamec et al., 2014) in cases of superior VN, correlating with an increase in oVEMP amplitude. Those with poor outcome, that is those oVEMP responses that did not show prolonged latencies but only showed reduced amplitude, were found to have chronic white matter supratentorial lesions present on brain MRI. This analysis of the effects of selective parameter (latency versus amplitude) involvement is an important one and should be followed by other similar studies also.

Abnormal cVEMPs (based on the presence or absence of a response and amplitude) have also been shown to predict a slow recovery of the subjective visual horizontal in patients with vestibular neurolabyrinthitis or neuritis (Murofushi et al., 2007). The subjective visual horizontal is an additional measure of static utricular/superior nerve function (Halmagyi and Curthoys, 1999; Taylor et al., 2016). Therefore, this latter finding may represent that class of VN that involves both divisions of the vestibular nerve (see below), as the cVEMP is a measure of saccular/inferior nerve function. A separate study in children and adolescents (age < 19 years old), diagnosed with vestibular neuritis (11 out of a total of 301 patients over a 2.5 year period) revealed normal cVEMPs in all cases examined (6 in total performed the cVEMP, Brodsky et al., 2016b). This latter study may indicate that inferior VN does not occur in children, but the sample size is small, and more studies are needed.

Vestibular neuritis usually affects the superior vestibular nerve, and manifests with a mixed horizontal-torsional nystagmus beating away from the lesion side (Halmagyi et al., 2010). The diagnosis of isolated inferior VN is difficult as the usual signs are absent (Kim and Kim, 2012). The symptoms may erroneously be ascribed to a central pathology if there is no scrutinized evaluation for the inferior vestibular nerve (Newman-Toker et al., 2008). Such patients can show normal caloric responses initially. Also, if a laboratory does not have a quantitative head impulse test system, determining abnormalities of the posterior canal using the bedside HIT may be difficult if the involved saccades are covert (Kim and Kim, 2012). Inferior vestibular neuritis accounts for only 1.3% of total vestibular neuritis and labyrinthitis cases. Patients can present with normal neurological and neurotological examination, no spontaneous or gaze evoked nystagmus, normal CT and MRI scans, and yet present with an abnormal cVEMP. A series of 811 patients, found such a pattern in 4.9% of patients based on both amplitude and latency (Iwasaki et al., 2005). Of these, 20% could not be diagnosed as having a specific disease entity already recognized during the period at which the study was done.

The combined use of both c- and oVEMPs (evaluating both latency and amplitude) and the video head impulse test may allow to divide VN into four different types, namely entire VN, superior VN, inferior VN and ampullary VN (Walther and Blodow, 2013; Magliulo et al., 2015; Taylor et al., 2016). The diagnosis of ampullary VN is possible in the presence of normal hearing, absence of responses on caloric testing or abnormal video head impulse test, and bilateral preservation of c- and oVEMPs (Magliulo et al., 2012). Worst 1 year follow up outcome appear to be those patients with superior VN and ampullary VN. This follows on from a previous proposal to divide VN into types beyond simply superior and inferior VN, namely also including inflammation of the entire vestibular ganglion (Halmagyi et al., 2010). In deciding what examinations to carry out in order not to miss patients with an abnormal outcome, it is recommended that a minimum of three tests inclusive of the HC vHIT, PC vHIT and the cVEMP be performed to reduce as much as possible the presence of false negatives (Taylor et al., 2016). oVEMPs is not included in this battery as the HC vHIT produced more positive and relevant results than the oVEMP in cases that involved the superior vestibular nerve.

Vestibular neuritis can rarely manifest in the form of bilateral but sequential peripheral vestibulopathy (Young et al., 2016). In this reported case, total neuritis appeared in one ear, resolved, than later appeared in the contralateral ear. Both VEMPs were useful (when measuring both latency and amplitude) as part of a noninvasive test battery (with audiometry and VHIT) to differentiate this cause from other causes of acute vestibular syndrome.

Summary of findings:

1. Clinical value: Desirable
2. Prolonged oVEMP responses are a good prognostic sign in superior VN (Adamec et al., 2014).
3. Abnormal cVEMPs predict slow recovery of the subjective visual horizontal (Murofushi et al., 2007).
4. cVEMPs can be used to confirm the presence of inferior VN with normal caloric responses (Iwasaki et al., 2005; Newman-Toker et al., 2008; Kim and Kim, 2012).
5. cVEMPs as part of a vestibular test battery is recommended to detect all VN types (Halmagyi et al., 2010).
6. More studies in children using both c- and oVEMPs are needed.

3.1.6. Bilateral vestibulopathy (BV)

Bilateral vestibular failure or vestibulopathy is a disorder of both vestibular labyrinths or eighth cranial nerves (Brandt, 1996), with the cause remaining unknown in 20–50% of patients (Baloh et al., 1989; Fujimoto et al., 2009). When the cause is known or probably known, BV is usually due to ototoxic aminoglycosides, MD and meningitis (Zingler et al., 2007), bilateral tumors, neuropathies, sequential vestibular neuritis or autoimmune (Schuler et al., 2003). A genetic cause is also known to exist (Jen et al., 2004). It can be part of a cerebellar syndrome (e.g. CANVAS, Szmulewicz et al., 2011) or other neurodegenerative disorders (Agrawal et al., 2013). Patients with BV usually have impairment of the vestibulo-ocular reflex on clinical examination using the head impulse test (Bronstein, 2003) and impaired smooth compensatory movements (Migliaccio et al., 2004), and unsteadiness of gait and oscillopsia during movement of the head and body (Strupp et al., 2003; Deutschlander et al., 2005), worse in the dark (Wiest et al., 2001). There is absent or significantly affected nystagmic responses to both caloric stimulation and during chair rotation (Zingler et al., 2008). Both c- and oVEMPs can also be abnormal in these cases. oVEMPs are particularly likely to be affected in cases of aminoglycoside toxicity (compared to bilateral MD) when only amplitude is evaluated (Agrawal et al., 2013). Abnormal cVEMPs have not been shown to be related to any specific cause when both latency and amplitude were evaluated (Zingler et al., 2008; Agrawal et al., 2013).

Although an abnormal caloric test is usually seen in BV, it has been found that caloric responses may remain normal and that

bilateral vestibular deficits are revealed only with abnormal cVEMPs when both latency and amplitude were evaluated (Fujimoto et al., 2009). The diagnosis of BV requires bilaterally impaired or absent function of the vestibulo-ocular reflex (VOR) bilaterally, revealed using the head impulse test (HIT), the video-HIT (vHIT) technique or the caloric testing (Strupp et al., 2017). However, the possibility of a subtype of idiopathic BV affecting the inferior vestibular nerves only may be considered although further studies are needed before this can be accepted as part of the diagnostic criteria (Fujimoto et al., 2009).

Some studies have indicated that a BV can be mistaken for a unilateral vestibulopathy if cVEMPs are not performed. Of 1560 patients in one study who had both caloric tests and cVEMPs performed, 43 (2.8% of all patients) were found to have dissociated BV, i.e. unilateral abnormal caloric responses with contralateral abnormal cVEMPs when both latency and amplitude are evaluated (Fujimoto et al., 2012). Another study found a higher percentage (24.24%) of such cases based on both latency and amplitude (Serra et al., 2012).

With regards to future research, there is still very little we know about BV, with the etiology remaining unclear in more than 50% of cases (Strupp et al., 2016) although neurodegeneration is assumed. One can analyse the VEMP parameters, specifically amplitude versus latency, to determine which is more affected as the latter parameter tends to indicate central nervous system involvement. It may be possible to correlate both c- and oVEMP findings with response to therapy and to provide a prognosis with regards to treatment options on a specific combinations of both c- and oVEMP or other paramedical results.

To summarize with respect to bilateral vestibulopathies:

(1) Clinical value: Desirable.
(2) Although abnormal oVEMPs may be more common in cases of aminoglycoside toxicity, it cannot be considered diagnostic and as a way of determining the etiology of the vestibulopathy between bilateral MD and aminoglycoside toxicity (Agrawal et al., 2013), especially with respect to the fact that aminoglycoside use will be expected to be documented. More studies are needed in this regard.
(3) cVEMPs can be useful in the diagnosis of BV if the caloric response is abnormal only on one side, but further studies are needed with regards to this (Fujimoto et al., 2012; Serra et al., 2012).

3.1.7. Benign paroxysmal positional vertigo (BPPV)

Benign paroxysmal positional vertigo is a clinical syndrome characterized by recurrent, brief episodes of severe vertigo and rotatory nystagmus, precipitated by specific positions of the head relative to gravity (Van der Velde, 1999; Abbott et al., 2016). The symptoms are triggered by the act of moving the head to a new position, rather than by maintaining the head in a particular position or posture (von Brevern et al., 2015). The nystagmus may be torsional, vertical or horizontal, and is characterized by findings such as latency, crescendo and decrescendo, transience, reversibility and fatigability (Korres and Balatsouras, 2004; Balatsouras, 2012). It is one of the most common causes of vertigo and is due to the presence of otoconial debris within the semicircular canals, usually the posterior canal (Bronstein, 2005), either by way of canalothiasis or cupulolithiasis (Hall et al., 1979; Brandt and Steddin, 1993; Xu et al., 2016). Detachment of otoliths from the macula of the utricle is the suspected pathogenesis (Parnes and McClure, 1992; Welling et al., 1997; Akkuzu et al., 2006), with evidence involving the macule of the saccule also available (Parnes and McClure, 1992; Welling et al., 1997; Hong et al., 2008). Less commonly, the problem is found in one of the horizontal semicircular canals, and even more rarely in the anterior canals (Soto-Varela et al., 2013). As there are reports, as listed above, that indicate pathophysiological alterations of the macula of the utricle and saccule, both c- and oVEMPs are expected to be altered in BPPV also. Another possible cause of BPPV is following head surgery that involves some form of bone percussion or hyperextension of the neck, such as Le Fort I osteotomy for the correction of midfacial deformities (Deniz et al., 2017). Benign paroxysmal positional vertigo can be diagnosed clinically with the help of the provocation manoeuvres and the clinical assessment of the positional nystagmus (Bronstein, 2005). Therefore, both c- and oVEMPs and other auxiliary tests are not necessary for its diagnosis, even though cVEMPs have been found to be abnormal in 30% of diagnosed cases, predominantly showing prolonged latencies (Akkuzu et al., 2006). In contrast, oVEMPs with air-conducted sound have been found to be abnormal in 84.5% of diagnosed cases, this time in terms of interside amplitude differences rather than latencies (Seo et al., 2013a, 2013b). A prior superior vestibular neuritis can result in BPPV, which in turn may also result in a high percentage of oVEMP abnormalities, but this latter paper appears to have ruled this out. This would therefore suggest that the utricle is more dysfunctional compared to the saccule in this condition, which would make sense assuming that the otoliths that end up in the semicircular canals would originate from this neighbouring otolith organ. This has also been shown in a separate study using estimation of the subjective visual vertical and analysis of the torsional otolith-ocular reflex (von Brevern et al., 2006). But then again, it implies that the utricle is selectively involved compared to the saccule independent of its position. Further investigation would need to determine why the utricular macular is more prone to losing its otoliths compared to that of the saccule. The presence or absence of an abnormal cVEMP could not predict the number of canalith repositioning manoeuvres required before the Hallpike provocation manoeuvre normalized (Akkuzu et al., 2006). No significant difference was noted between the improvement in positional vertigo after the Epley canalith repositioning manoeuvre and the results of the posttreatment oVEMPs with air-conducted sound (Seo et al., 2013a). However, one study has showed that a decreased interaural amplitude ratio at the affected side may be prognostic of BPPV not resolving after a single repositioning manoeuvre (Chang et al., 2017). Other studies are therefore needed to confirm this latter finding. Therefore, based on the relevant literature, both c- and oVEMPs are not useful in the diagnosis of patients with BPPV. However, they may be useful in predicting resistance to canalith repositioning manoeuvres.

To summarize with respect to BPPV:

(1) Clinical value: Potentially useful with respect to determining refractory cases, but more research is needed.
(2) The use of both c- and oVEMPs is not recommended for the diagnosis of patients with BPPV.
(3) More confirmatory work is needed to confirm if both c- and oVEMPs can be used to predict resistance to canalith repositioning manoeuvres, especially with regards to the interaural amplitude ratio (Chang et al., 2017).

3.1.8. Sudden deafness

Sudden hearing loss (SHL) is defined as any sensorineural hearing loss (SNHL) $\geq$ 30 dB affecting at least 3 consecutive frequencies, showing a rapid decline, and occurring within 3 days (National Institute of Health, 2008; Stachler et al., 2012; Oiticica et al., 2013; Chen and Young, 2016), with possible causes including viral infection, ischaemia, autoimmune reaction or inner ear membrane rupture (Simmons, 1968; Schuknecht et al., 1973; Gussen, 1976; McCabe, 1979; Belal, 1980; Liu et al., 2017). Loss of both auditory and vestibular function is considered severe, whereas the mildest form is confined to the cochlea (You et al., 2014). As

the cochlea and vestibule are located very close to each other, the pathology of idiopathic sudden hearing loss is expected to involve both VEMPs (Stamatiou et al., 2009), clinically resulting in vertigo in some cases (Liu et al., 2017).

Studies concerning obtainable cVEMPs could not be evaluated due to absent EMG control (e.g. Stamatiou et al., 2009, Liu et al., 2017), making amplitude measurements unreliable. Absent VEMP responses (both c- and o-) were found in all patients examined with bilateral SHL (Chen and Young, 2016), but in this specific study no direct comparison in these patients was done with other paramedical examinations. Using absent or low amplitude responses as a criterion, oVEMPs were found to be abnormal in 76.2% of patients with vertigo and 42.9% of patients without vertigo (Liu et al., 2017). Calorics were also performed in these patients, but in this specific study no statistical comparison was done. However, there was no statistical correlation between the presence of vertigo and abnormal oVEMP result. oVEMP test results, and also caloric test results, were found to be significantly associated with hearing outcome, with absent oVEMPs tending to predict unchanged hearing recovery following treatment with plasma expander followed by oral medication (You et al., 2014). In fact, combining the oVEMP and caloric test results suggested a stronger predictor of hearing recovery. Corresponding results from caloric testing per patient (in patients with unobtainable oVEMPs) was not available for our analysis, as the results were analysed on a group and not individual basis.

Summary of findings:

(1) Clinical value: useful
(2) A few studies could not be evaluated due to poor EMG control with regards to cVEMPs. More studies are needed with good cVEMP protocol, namely controlling for SCM contraction.
(3) Normal oVEMPs, together with normal caloric responses, can predict hearing recovery after therapy, although the oVEMP result was not found to correlate with the presence of vertigo (You et al., 2014). This study included patients with and without vertigo.

3.1.9. Inherited and acquired neuropathies

Vestibular rehabilitation has been shown to improve postural stability in patients with vestibulopathy (Hilier and McDonnell, 2011). Therefore, it is important to demonstrate whether postural instability is not only caused by sensory ataxia due to proprioceptive sensory loss, but could also be caused by parallel vestibular impairment (Poretti et al., 2013). It has been shown that the neuropathic process in patients with Charcot-Marie-Tooth disease frequently involve the vestibular nerve when investigated using cVEMPs (92.3% of patients), with 30.8% of patients showing abnormal cVEMPs in the presence of a normal quantitative head-impulse test.

cVEMP responses have been shown to be unaffected by non-insulin dependent diabetes mellitus, with or without polyneuropathy (Bektas et al., 2008). However, although it has been noted in this publication during the discussion that diabetes mellitus (DM) patients occasionally have vestibular symptoms, as has been noted elsewhere (Perez et al., 2001), none of the studied patients in the above study by Bektas et al. had any such symptoms.

Familial dysautonomia has been found to be associated with abnormal c- and oVEMPs in the form of lower amplitudes and delayed responses (Gutierrez et al., 2017).

To summarize:

(1) Clinical value: useful
(2) Patients with CMT disease have a high prevalence of vestibular neuropathy.
(3) Publications are needed to investigate the possible benefit of vestibular rehabilitation in such patients.
(4) No studies with oVEMPs are available at the time of writing and are needed.

3.1.10. Idiopathic otolithic vertigo

Patients exist that present with episodic tilting or translational sensations in the pitch plane, without any other vestibular symptoms (Murofushi et al., 2013). These patients have normal caloric responses, with predominantly abnormal air-conducted cVEMPs, but also presenting to a lesser extent with abnormal air-conducted oVEMPs. The above symptoms, together with the above abnormal VEMP results and normal calorics may represent a specific clinical entity that affects only the otolithic end organs. This entity may also explain the following cases where only oVEMPs were found to be abnormal. One publication reported on 31 patients over a 5 year period at a tertiary academic referral centre (Pelosi et al., 2013), but the total number of patients that had been investigated was not reported. These patients exhibited a greater preponderance of postural instability and swaying/rocking sensation. Another study with similar findings described patients with lateral tilt sensation, including sensations of being pulled or pushed laterally (Murofushi et al., 2012).

To summarize with respect to idiopathic otolithic vertigo:

1. Clinical value: highly desirable.
2. Episodes of tilt or translation sensations, with VEMPs as the only abnormal examination, may represent an otolith specific disorder.
3. Performing VEMPs in patients with these symptoms is therefore very important, as other paramedical examinations will remain normal.
4. More examples of such cases need to be published.

3.1.11. Delayed endolymphatic hydrops

Delayed endolymphatic hydrops (DEH) manifests as recurrent vertigo following severe unilateral or bilateral hearing loss (Gu et al., 1984) after several years or decades (Cho et al., 2013). Ipsilateral DEH refers to one ear with profound hearing loss whereas contralateral DEH also shows fluctuating hearing loss in the opposite (better) ear (Lin and Young, 2012). As hydrops has been noted in the vestibules in diagnosed cases of DEH (Nonoyama et al., 2014), both c- and oVEMPs are expected to be involved. Glycerol cVEMPs has been found to be clinically useful in this disease, especially in the ipsilateral type based on corrected amplitude measurements (Shojaku et al., 2001), where profound hearing loss exists on the same side as the endolymphatic hydrops. In these cases, it is not possible to perform the glycerol test with pure tone audiometry and electrocochleography. Delayed endolymphatic hydrops showed more abnormalities with 3D fluid-attenuated inversion recovery MRI after transtympanic gadolinium injection (84%) than with cVEMPs when both latency and amplitude were measured (52%) (Gu et al., 2014). However, this study was performed on patients already diagnosed by other means. Also the literature with regards to this condition is limited, and more studies are needed to determine the usefulness or not of VEMPs (both c- and o).

To summarize with respect to DEH:

i. Clinical value: desirable
ii. Glycerol cVEMPs are useful in cases of profound peripheral hearing loss where pure tone audiometry and electrocochleography cannot be performed.
iii. More studies are needed with respect to diagnosis.
iv. Experience with oVEMPs is needed.

3.1.12. VEMPs as the only examination (bedside or paramedical) that can be performed to evaluate the vestibular system

Both c- and oVEMPs are very useful in cases where the presence of congenital nystagmus does not allow one to evaluate spontaneous nystagmus, head shaking nystagmus, vibratory-induced nystagmus or positional and positioning nystagmus, or perform calorics and the head impulse test, and it becomes the only way to objectively evaluate the vestibular nervous system (Manzari et al., 2012).

To summarize:

(1) Clinical value: Highly desirable
(2) VEMPs may be the only paramedical examination that can be performed when background ocular abnormalities co-exist.
(3) More examples of such cases need to be published.

3.2. Diseases of the central vestibular system

Several studies have indicated that prolonged VEMP latencies are observed more frequently in these disorders, compared with peripheral vestibular disorders (Shimizu et al., 2000; Murofushi et al., 2001). Therefore, such findings should alert the clinician to the possibility of central nervous system involvement.

Some of the studies comparing VEMPs (specifically cVEMPs) with MRI images, predominantly with regards to MS, fail to assess the upper cervical spinal cord (e.g. Bandini et al., 2004; Guven et al., 2014). For example, in the study by Bandini et al. (2004), in 20 patients with MS without clinical brainstem dysfunction, cVEMPs were abnormal in 4 patients with normal (brainstem) MRI. One study went as far as calling oVEMPs and cVEMPs "brainstem evoked potentials" (Ivankovic et al., 2013), which at least with regards to cVEMPs is not accurate. The cVEMP pathway is known to involve the spinal accessory motor nucleus, which in the upper cervical spinal cord can reach as far down as the C5 level (Kiernan, 2005). This is not a trivial point, since the involvement of the upper cervical spinal cord is not rare in MS (Bellenberg et al., 2013; Miraldi et al., 2013; Valsasina et al., 2013).

Increase in p13 latency appear to be more specific to central pathway dysfunction than to peripheral vestibular dysfunction, when comparing patients with Meniere disease, vestibular neuritis, acoustic neuroma and MS (Murofushi et al., 2001). The only major exception concerned acoustic neuromas, where patients with large tumours showed prolonged latencies. Amplitude changes are not as specific, as such changes can be seen in both central (Kim et al., 2014) and peripheral pathways (Zingler et al., 2008) with slight predominance for the latter.

It is known that patients with MS, a chronic demyelinating disease involving the white matter of the central nervous system (Gazioglu and Boz, 2012), report symptoms related to the vestibular system on a frequent basis (Alpini et al., 2004). Clearly, many of the above demyelinating lesions can involve the vestibular pathway in the central nervous system, including the brainstem and cervical spinal cord. Many papers were published on the use of evoked potentials including both c- and oVEMPs in patients with suspected MS (Bandini et al., 2004; Gazioglu and Boz, 2012). It is clear from the work that is published so far, especially with respect to both c- and oVEMPs, that they have only a limited role in making the diagnosis of MS. In the majority of cases, the presence of an abnormal MRI is more frequent than abnormal VEMPs (both c- and o-). For example, 59.4% of cases with abnormal brainstem MRI showed abnormal VEMPs based on both latency and amplitude measurements. In another series, brainstem lesions were evident on MRI in 43.8% of cases, oVEMP abnormalities were evident in 37.5% and cVEMP abnormalities in 31.0 % of cases based only on corrected amplitude measurements (Ivankovic et al., 2013). In another study, cVEMPs were found to be abnormal in 53.3% of cases based on latency only (this publication did not normalize their amplitude data based on muscle contraction and so this parameter could not be evaluated) (Sartucci and Logi, 2002). BAEPs have also been found to be better correlated with brainstem and/or cerebellar signs than cVEMPs based on both latency and amplitude (Versino et al., 2002). However, the BAEP correlation with brainstem involvement was still not good (Habek, 2013; Gabelic et al., 2015). But when the parameters of both oVEMPs and cVEMPs are combined in the form of a VEMP score (Gabelic et al., 2015), there is good correlation of this score with brainstem symptoms and signs and with disability.

Despite its limited role in MS diagnosis, both c- and oVEMPs still have a role in assessing patients with MS. For example, is a demyelinating lesion demonstrated with MRI responsible for new onset vestibular symptoms in an individual MS patient? A good example is illustrated by a case of a patient who presented with primary position upbeat nystagmus (Adamec et al., 2012). The MRI showed a demyelinating lesion in the lower medulla. Yet, not only were the cVEMPs but also the oVEMPs abnormal. Although not shown in this study, BAEPs were normal (Ivan Adamec, personal communication). Treatment with intravenous methylprednisolone resolved the patient's paraparesis but not the nystagmus. The latter may be due to loss of central adaptation of the vestibulo-ocular system caused by diffuse brainstem damage as indicated by the oVEMP (although perhaps not so diffuse as the BAEPs were normal). VEMPs in this patient could explain the persistence of the nystagmus despite treatment. Just having a BAEP available would have missed the realization of this phenomenon.

Apart from MS, evidence of brainstem dysfunction, with respect to both o- and cVEMPs, was noted in patients with idiopathic Parkinson's disease and atypical Parkinsonism, revealed as prolonged latencies as well as unobtainable responses (Venhovens et al., 2016). Pisa syndrome, characterized by a persistent lateral trunk flexion, was significantly associated with a bilateral absence of cVEMP responses (Di Lazzaro et al., 2018). Using cVEMPs also helped to determine the presence of different pathways related to either administration of dopa versus subthalamic stimulation (Potter-Nerger et al., 2012).

Other studies have failed to provide a significant advantage of both c- and oVEMPs over MRI in the evaluation of brainstem lesions (Itoh et al., 2001; Ahn et al., 2011). cVEMPs also have no advantage over the bedside examination in the evaluation of cerebellar ataxias (Kirchner et al., 2011). However, cVEMPs have been used to determine the pathophysiology of certain disorders that have still not been clarified. Attention deficit hyperactivity disorder is characterized by the presence of inattention, hyperactivity and impulsivity, and has been associated with abnormal gait and balance (Isaac et al., 2017). Low amplitude cVEMPs have been found to be significantly associated with ADHD, with a negative correlation shown with the sensory processing measure, possibly suggestive of brainstem dysfunction in this disorder. Bilaterally delayed cVEMPs, with normal oVEMPs, was used to conclude that unsteady gait in a 5-year-old with a bilateral anterior inferior cerebellar artery loop encroaching the internal auditory canal bilaterally was most likely caused by impaired perfusion of the lower brainstem (Wen et al., 2017). After giving medication to improve perfusion, the child improved together with normalization of the cVEMP response.

To summarize:

(1) Clinical value: Useful.
(2) It is important to keep in mind that the upper cervical spinal cord is part of the cVEMP pathway.
(3) VEMPs are not useful in the diagnosis of MS, and a combined VEMP score shows good correlation with disability.

Table 1
Conditions where VEMPs have been examined rarely.

Condition examined	Reference	VEMP used[1]	Findings	Comments[2]
Myelomeningocele	Picciotti et al. (2012)	c	The majority of cases showed normal cVEMPs	*No attempt was made to correlate cVEMP findings with vestibular symptoms*
HIV/AIDs	Heinze et al. (2014)	c	The majority of cases showed abnormal cVEMPs	*No attempt was made to correlate cVEMP findings with vestibular symptoms*
Auditory neuropathy	Sheykholeslami et al. (2000)	c	cVEMPs were abnormal in all patients	cVEMPs do not add new information to other paraclinical examinations, e.g. calorics
Superfical siderosis	Ushio et al. (2006)	c	Abnormal cVEMP in single case	cVEMPs do not add new information to other paraclinical examinations
Chronic noise-induced hearing loss.	Wang and Young (2007)	c	Half the patients showed abnormal cVEMPs	*Results not correlated with vestibular symptoms. Can VEMPs predict hearing return?*
Otosclerosis	Saka et al. (2012), Tramontani et al. (2014), Kuei-You and Young (2015)	c/o	About 50% of patients showed abnormal bone-conducted cVEMPs, 33–90% of these withcVEMPs, 33–90% of these with vestibular symptoms. oVEMPs were 84% abnormal with vestibular symptoms	VEMPs did not contribute clinically when already with diagnosis
Children with cancer on aminoglycoside therapy	Chen et al. (2013)	c	A cumulative dose of 1,200 mg/kg carries a 50% risk of cochlear/vestibular toxicity	**Good indication of follow-up for vestibular damage with such therapy**
Sensorineural hearing loss	Singh et al. (2012)	c	cVEMP amplitudes were lower compared to controls	No added information was provided
Early stage Amyotrophic Lateral Sclerosis	Kilic et al. (2012)	c	cVEMPs were normal	Not helpful for ruling in brainstem dysfunction
Benign Positional Vertigo	Chang and Young (2007)	c	Predominantly normal	cVEMPs not sensitive enough for objective vestibular evaluation
Myasthenia Gravis	Valko et al. (2016)	o	Decrement has been noted in patients with MG	Further comparative studies are needed

[1] c = cVEMPs, o = oVEMPs.
[2] Comments in italics show where further work is needed. Comments in bold demonstrate a definite use for VEMPs.

(4) On an individual patient basis, VEMPs can provide useful information with regards to the origin of vestibular symptoms and their presence with respect to therapy.
(5) cVEMPs have no advantage over the bedside examination in the evaluation of cerebellar ataxias.
(6) Similar studies with respect to oVEMPs need to be done with respect to CNS disorders in general.
(7) VEMPs can be useful in determining the cause of certain pathologies and their mechanisms.
(8) Prolonged VEMP latencies (cVEMPs and/or oVEMPs) should alert the clinician to the possibility of central nervous system involvement.

3.3. Diseases of the central and peripheral vestibular system that have been examined rarely

Both c- and oVEMPs have been investigated in other conditions, the findings of which are summarized in Table 1. These conditions are not discussed in the main text, particularly where no clinical value for c- or oVEMPs has been shown. There are many instances shown where both c- and oVEMPs could be useful, but more studies are required. Both c- and oVEMPs used during aminoglycoside therapy are clearly useful. It is clear also from this table that oVEMPs have not been used yet in the listed conditions, and this needs to be investigated in future studies.

4. Conclusion

From this review, we can conclude that the usefulness of both c- and oVEMPs goes beyond the confirmation of SSCD, and is useful in many clinical cases, which are summarized in Table 2. Based on this information, one can conclude that both c- and oVEMPs should be performed in *almost all* cases, in which the vestibular system is suspected or confirmed to be involved, both in the central and peripheral nervous systems, not only with respect to diagnosis, but also with respect to explaining the effects of therapy (with the exception of a few conditions listed below where both c- and oVEMPs clearly have no value). This impression is not new but applies to all other evoked potentials, such as visual, brainstem auditory and somatosensory.

Much work is left to be done, which is expected to add further importance to the use of both c- and oVEMPs in clinical practice. With regards to rare diseases, the recommended research is highlighted in Table 1. However, with respect to the more commonly studied diseases, the following conclusions can be drawn.

4.1. Areas in the authors' view where research is still needed:

(1) To determine if the results of electrophysiology (including both c- and oVEMPs) and calorics can affect how surgery is carried out with regards to VS and if this can determine patient outcome.
(2) To determine if an abnormal cVEMP with the glycerol test, after a single untreated episode of vertigo or dizziness, can predict the development of MD.
(3) To determine if patients with unilateral symptomatic MD, with abnormal cVEMP responses in the contralateral asymptomatic ear, will develop MD also in the contralateral ear.
(4) To determine if MD patients in disease stage I or II have a poorer quality of life when the cVEMP is abnormal. Does or can an abnormal cVEMP affect treatment options?
(5) To determine if both c- and oVEMPs are needed to confirm the presence of posterior semicircular canal dehiscence revealed by a CT scan.

Table 2
Conditions where VEMPs have been found to be useful.

Condition	VEMP[1]	Usefulness
Acoustic neuroma	o/c	Evaluation as part of an electrophysiological battery, especially where MRI is not immediately available
Bilateral vestibulopathy	o	A low amplitude response can suggest aminoglycoside toxicity as the cause
	c	Important in diagnosis, as in some cases calorics can be normal without neurotological signs
Meniere's Disease/Vestibular Migraine	c	Useful in the differential diagnosis when combined with calorics and stimulus frequency response
Meniere's Disease	c	An abnormal VEMP in stages I and II can predict deterioration in hearing after 20 months
SSCD	c/o	Either VEMP (although oVEMP better) to confirm its presence
Posterior SCD	o	Accentuated response, useful where neurotological examination is unremarkable
Air-bone gap on pure-tone audiometry	c/o	To clarify the etiology, ruling out SSCD rather than other causes such as otosclerosis
Inferior vestibular neuritis	c	May be the only manifestation where all other examinations, including neurotological, are normal
Postural instability	o	oVEMPs can be the only abnormal examination in some cases
Idiopathic otolithic vertigo	c/o	May be a distinct clinical entity where only VEMPs are abnormal
MS	c/o	Performing VEMPs prior to treatment can help explain why some symptoms improve, despite MRI findings
Congenital nystagmus	c/o	VEMPs can sometimes be the only way of evaluating the vestibular system
Peripheral neuropathies	c/o	With a view of introducing vestibular rehabilitation

[1] c = cVEMPs, o = oVEMPs.

(6) To determine if oVEMPs can predict good outcome after oral prednisone treatment in cases of superior vestibular neuritis.
(7) To determine if both c- and oVEMPs can predict the evolution of MS, including conversion to other forms.
(8) To determine the effectiveness of treatment of MS using both c- and oVEMPs.
(9) The use of interside amplitude difference as a way of determining resistance of BPPV to canalith repositioning manoeuvres needs more studies to be confirmed.
(10) More studies are needed with regards to the diagnosis of DEH, together with experience with oVEMPs.
(11) VEMPs have a potential role in explaining the presence of vestibular symptoms with respect to otosclerosis, but more studies are needed.
(12) There have been few consistent comparisons between AC and BC findings and it is possible this may provide additional information, given the mechanism of action of the two is likely to be different.

The reporting of both c- and oVEMP findings can be improved significantly. Many papers fail to directly compare the results of both c- and oVEMPs with bedside evaluation of the vestibular system, including the head impulse test, evaluation of smooth pursuit, and the evaluation of both spontaneous and gaze-evoked nystagmus in the presence or absence of visual fixation. This makes it difficult to convince otorhinolaryngologists or neurotologists familiar and experienced with these techniques to adopt both c- and oVEMPs in their patient evaluation. Also, although some studies have hinted at the possibility of using both c- and oVEMPs to select specific treatments (e.g. Serra et al., 2012), this has not been done up to now. Stimulus selection is important, particularly for the oVEMP due to the higher thresholds of utricular afferents to AC sound.

Many papers failed to publish both c- and oVEMP waveforms, which is important to determine the quality of their recordings. It is imperative that both c- and oVEMPs be recorded against an unrectified background to prevent distortion of the positive and negative phases of the waveform. They should clearly stand out from the background muscle activity, and not be superimposed by 60 Hz or 50 Hz line frequency artefact. If the VEMP amplitude (both c- and o-) is low compared to the background EMG amplitude, then the superimposition of two or more trials should be performed. VEMP waveforms should be published in the future.

Also, many studies using cVEMPs fail to control for muscle contraction and have underestimated the power of this specific paramedical examination. Amplitude was therefore unreliable in these studies, and this has added significance when one takes into account that amplitude is usually affected in peripheral vestibular disorders.

4.2. VEMPs have no current proven clinical value

4.2.1. Vestibular disorders

(1) In differentiating a cerebellopontine angle meningioma from a VS.
(2) In BPPV, for diagnosis.
(3) Hearing loss cannot be predicted in the more severe stages of MD (III and IV) where hearing is already compromised (in contrast to stages I and II where prediction is possible).
(4) In the evaluation of auditory neuropathy and pediatric cases of sensorineural hearing loss.

4.2.2. Non-vestibular disorders

(5) In the evaluation of cerebellar ataxias or of brainstem lesions, myelomeningocele, and early stage amyotrophic lateral sclerosis.

Declaration of Competing Interest

None.

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