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REVIEW ARTICLE

Audiovestibular Test Battery Approach in Patients with Vertigo

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ABSTRACT

All audiological tests are site-of-lesion tests and are used as a part of a test battery. Pure tone audiometry gives estimate of peripheral hearing status. Speech audiometry, acoustic reflex testing and auditory brain stem response differentiate cochlear and retrocochlear sites of lesion. Addition of cervical vestibular evoked myogenic potentials (VEMP) and ocular VEMP to this battery can further differentiate between saccular *vs* utricular lesions and between lesions of inferior *vs* superior vestibular nerve. Overall, this battery of tests provides useful insights into the otoneurological diagnosis of vertigo. Few case examples are cited to elucidate the utility of audiovestibular test battery.

Keywords: Vestibular evoked myogenic potentials, Auditory brain stem response, Acoustic reflex testing, Speech audiometry.

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INTRODUCTION

Vertigo or sensation of rotation is a commonly encountered complaint in an ENT and audiological set up and may be associated with a myriad of clinical conditions ranging from a cochlear condition like Meniere's disease, vestibular conditions such as vestibular neuritis, benign paroxysmal positional vertigo (BPPV), labyrinthitis to retrocochlear pathologies such as vestibular schwannoma. The role of audiovestibular testing in these patients is to ascertain degree of associated hearing impairment and to identify the site of lesion (whether cochlear or retrocochlear) leading to the vertigo and other associated symptoms like hearing loss, tinnitus, etc. Retrocochlear pathology refers to site of lesion at the VIIIth cranial nerve, cerebellopontine angle (CPA), or root entry zone of the nerve into the brain stem.[1]

All audiovestibular tests are topological and provide information about site of lesion and not the type of lesion. Further, each test assesses different site in the audio-vestibular system and is based on different principle thus assessing the system in a unique perspective. Therefore audiological diagnosis is always based on a test battery approach. This is further justified by Jerger's cross-check principle which states that the results of a single test must be cross-checked by an independent test measure. When the results on independent tests correlate, it increases our confidence in the diagnosis and even when they do not agree, they further add to our understanding of the site and type of lesion.

The inclusion of tests in the battery depends on the sensitivity and specificity of the test in detecting the site of lesion. Following is a review of the contemporary audiological test battery used in evaluating patients with vertigo. This is followed by three case examples illustrating their utility.

Pure Tone Audiometry

Pure tone audiometry (PTA) is the most basic test in audiological assessment. Being a test of detection, it uses pure tones from 250 to 8000 Hz to estimate hearing thresholds. An audiogram is a plot of hearing thresholds (in dB hearing level) obtained using air-conduction and bone-conduction tones as a function of frequency. It thus provides ear-specific and frequency specific information about peripheral hearing status. It can indicate the degree of hearing loss, its type and configuration, thus being a starting point for conduction of further tests.

Patients with vertigo may present with different audiometric patterns upon the underlying pathology. Pure vestibular disorders may present with normal hearing sensitivity whereas those with labyrinthitis may show profound hearing loss. Meniere's disease may present with fluctuating low frequency sensorineural hearing loss (SNHL) in the initial stages when the loss accompanies the episodes of vertigo; later it may become a permanent flat SNHL. In these patients PTA is used as an outcome measure in glycerol testing for the diagnosis of the disease. Asymmetric, typically high frequency SN hearing loss is characteristically seen in RCP; however; it is not uncommon to witness an early lesion with normal hearing sensitivity.

Speech Audiometry

It involves use of standardized speech samples to estimate various measures of performance like the speech recognition threshold (SRT), word recognition score (WRS), performance intensity function (PI function), most comfortable loudness level (MCL) and uncomfortable loudness level (UCL).

SRT is the lowest level at which a person can identify the stimuli as speech 50% of the times it is presented and is routinely determined using spondees, i.e. bisyllabic words with equal stress on each syllable. SRT-PTA agreement is an important diagnostic tool wherein SRT poorer than PTA is associated with presence of RCP. WRS is the percentage of correctly identified monosyllabic words from standard word list and gives us an idea about the patient's ability to recognize speech at supra threshold levels. Poor WRS than would be expected based on audiometric configuration and severity is indicative of RCP. Further, poorer WRS with increase in the intensity of speech stimulus or roll-over phenomenon is diagnostic of RCP. Patients with cochlear pathologies show fair to good SRT-PTA agreement, fairly good WRS and reduced dynamic range (UCL-SRT) due to lowered UCL.

Acoustic Reflex Testing

It is an important test in the immittance test battery and plays an important role in the differentiation of cochlear lesions with RCP. It involves presentation of tonal/noise stimuli to elicit reflexive stapedial contraction. The resulting change in the ear's immittance is measured by the immittance meter. The acoustic reflex threshold (ART) refers to the lowest intensity of the stimulus needed to elicit the reflex. ART for individuals with normal hearing sensitivity is generally 60 dB SL, i.e. 70 to 100 dB HL.

Cochlear losses less than 70 dB HL will typically show presence of reflexes at normal hearing levels (70-100 dB HL) but at low sensation levels (< 60 dB SL) referred to as Metz test of recruitment. The degree of cochlear hearing loss affects the ART. The median ART sensation level decreases from 70 dB for persons with 20 dB SN loss to 25 dB for persons with 85 dB SN loss.[2]

Degree of cochlear loss	*Likelihood of presence of ART*
<60 dB	90%
85 dB	50%
100 dB	5 to 10%

Findings in RCP should also be considered in the light of degree of hearing loss. Absence of ART/elevated ART in persons with normal or near normal hearing sensitivity is strongly suggestive of retrocochlear site of lesion. AR is absent 30% of persons with eighth nerve lesions with normal hearing whereas in 70% of persons with eighth nerve lesions with a mild 30 dB hearing loss.[2] In persons with hearing loss greater than 75 to 80 dB, absence of reflex cannot be considered diagnostic of RCP thus leading to confounding results.

As the reflex is consensual, it is elicited in ipsilateral and contralateral conditions. Comparison of ipsilateral and contralateral reflex findings gives different reflex patterns thereby providing useful insights to the site of lesion decision.

Presence of acoustic reflex when stimulus presented to the ear with RCP is an exception rather than the rule. In such cases testing is done to find evidence of reflex decay: Relaxation of stapedius muscle during sustained presentation of reflex activator. Stimulus is presented at 10 dBSLre:ART for 10 sec at 500 and 1000 Hz. Following guidelines may be used for the interpretation of acoustic reflex decay testing:[3]

RD^{+++} Reflex amplitude declines ≥ 50% within 5 seconds at 500 and 1000 Hz is a positive indicator of 8th nerve lesion

RD^{++} Reflex amplitude declines ≥ 50% within 5 seconds at 1000 Hz but not at 500 Hz is a questionable indicator of 8th nerve lesion

RD^{+} Reflex amplitude declines <50% within 5 seconds at 500 and 1000 Hz is not indicative of eighth nerve lesion.

Auditory Brain Stem Response

Auditory brain stem response (ABR) is the result of synchronous neural activity of the 8th nerve and auditory brain stem, evoked in response to certain specific stimuli such as clicks. Stimuli are delivered to the ears, typically by means of earphones. Bioelectric activity, which may include the ABR, is picked up from the scalp and processed to enhance the response and minimize the background random EEG activity. The processed response, which is a graph of voltage *vs* time, is then visually assessed. Jewett and Williston[4] were the first to record systemically these responses from the scalp of human subjects to various stimulus and recording parameters and to suggest its clinical utility.

In normal adults, the ABR to click stimuli presented at relatively low rates (less than 10/s) at relatively high intensity levels (60 to 70 dB SL) consists of five to seven well-formed vertex-positive wave peaks, each designated consecutively with Roman numerals I to VII and separated in duration by approximately 1 msec. Judgement of such a response, accurate identification of various peaks and their measurements generally have very high reliability (cases with retrocochlear sites of lesion are a major exception which itself will be of diagnostic significance).

Features of any ABR can broadly be classified into two categories:

Quantitative

i. Response latency: Measured in milliseconds.

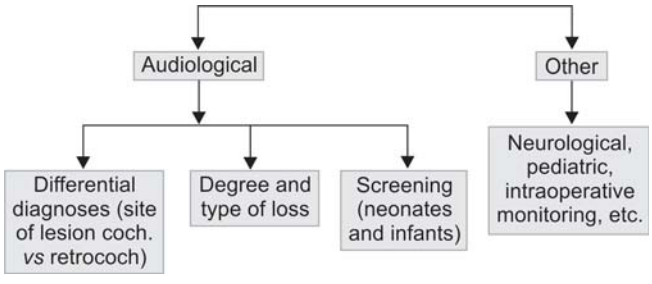
a. *Absolute latency*: The time period between stimulus onset and the peak of a measured response.
b. *Interpeak/interwave interval*: Time interval between two wavepeaks

ii. Response amplitude
a. *Absolute amplitude*: Height between the peak of one wave component to its following trough. It is measured in micro- or nanovolts.
b. *Relative amplitude ratio (RAR)*: The ratio of absolute amplitude of two wavepeaks.

Qualitative

Morphology of ABR refers to the visual appearance of the wave configuration.

The clinical applications of ABR can be summarized as under:



There are two prerequisites for a normal ABR:

- Adequate functioning of peripheral hearing mechanism
- Integrity of CANS up to perhaps mid brain structures.

If one of these is known to be intact, the status of the other can be inferred.

Jagadeesh and Gore[5] proposed several indices based on empirical considerations for the differentiation of cochlear *vs* retrocochlear sites of lesion as summarized below.

Sl no.	*Measure*	*Criterion*	*Remarks*
1.	Absolute latency of wave V	Delayed	High false-positive rate because wave V latency is delayed in many non-retrocochlear conditions, like: – Conductive hearing loss – Low intensity of stimulation – High stimulus repetition rates – High frequency hearing loss (esp. at 4 kHz > 50 dB HL: 0.1-0.15 ms increase for 10 dB increase in 4 kHz threshold >50 dB HL
2.	L-I function	I: Normal V: Delayed	– Time consuming – Inapplicable, if I is not identified
3.	ILD for V	> 0.4 ms	– Hit rate: >90% – High false-negative because of possibility of bilateral abnormal latency – False-positive due to interaural asymmetry
4.	IPL/IWI I -V	>N (mean = 4 ms)	– Perhaps the best – Inapplicable if I is not identified – False-positive: Notched audiogram
5.	BTT	>N (mean = 4.5 ms)	– Not very popular
6.	Morphology	Selective loss of waves; Grossly degraded morphology	– False positive: Subject movement, tension and equipment artefacts, electrode impedance mismatch, etc. – Very subjective
7.	Rate variations	Delayed latencies and degraded morphology	– Not popular – Time consuming – Extensive norms
8.	Amplitude ratio	Vamp <0.5 Iamp	– Highly variable – Least useful

Notes: L-I function: Latency-intensity function; ILD: Interaural latency difference; IPL/IWI: Interpeak latency/interwave interval; BTT: Brain stem transmission time: Interval between I and negative trough following V peak

ABR has been one of the sensitive tests in the diagnosis of acoustic tumors. Also using ABR, 'silent' plaques of multiple sclerosis were demonstrated,[6] brain stem death could be ascertained[7] and altered myelin development in childhood.[8] Currently the role of ABR is not as relevant for detection of such abnormalities because of the availability of imaging methods like magnetic imaging that can identify tumors measuring few mm. Sensitivity of ABR to detect tumors <1 cm is 53%.[9] Normal ABR may be seen in small size tumors as thay affect audibility of a narrow frequencies and a click evokes ABR from the gross response of the basilar membrane. Lutman[10] suggested that ABR may be considered primarily a screening test for acoustic tumors. According to him the preferred diagnostic tests for patients suspected of a CP angle tumor were high-resolution CT, or preferably MRI. If these tests are not readily available,

patients with normal ABR results might forego imaging studies. However, in such cases there is a risk that target cases may be missed. Stacked ABR may be used to increase the sensitivity of ABR to identify smaller lesions not picked up in conventional ABR.

Cervical Vestibular Evoked Myogenic Potentials

Cervical VEMP are short latency electromyograms (EMG) evoked by high level acoustic stimuli recorded from surface electrodes over tonically contracted SCM muscles.[11] These responses arise from the saccule as the response was preserved in patients with semicircular canal ablation due to streptomycin toxicity and in patients with Benign Paroxysmal Positional Vertigo (BPPV) but absent in patients with advanced Meniere's disease and in those who had undergone cochleosacculotomy.[12] Colebatch and Halmagyi[13] (1992) established a reliable procedure to record the myogenic potentials evoked by clicks. They recorded the potentials with surface electrodes placed on the sternocleidomastoid (SCM) muscle and with high quality EMG recording techniques; they showed the responses to be repeatable. They studied VEMP findings in three patients before and after selective sectioning of the vestibular nerve on one side and found absence of VEMP on ipsilateral side but VEMP were preserved in subjects with severe to profound hearing loss thus concluding that the vestibular afferents to be important in mediating VEMP. Studies on human participants and animal models reveal that VEMP is mediated by an ipsilateral reflex pathway.

VEMP is recorded from the SCM in either supine or sitting position. In supine position, the subject is asked to elevate his head thereby causing bilateral activation of VEMP whereas in sitting position, subject is asked to rotate his head to the side contralateral to acoustic stimulation causing unilateral activation of SCM. The noninverting electrode is placed at the center of the belly of SCM, inverting electrode is placed at the sternoclavicular junction and ground electrode is placed at the mastoid.[11,14,15] This electrode placement results in a biphasic waveform (Fig. 1). The first positive-negative complex is present in almost all normal participants as cited in published studies and occurs at latency of 13 and 23 ms respectively. This is referred to as P13-N23 or P1-N1 peak.[14] Latency of the P1-N1 complex is a stable measure and does not show much variation. However, there are variations in VEMP amplitudes ranging from a few microvolts to several hundred microvolts depending upon many factors like the stimuli used, intensity level, muscle tension, etc. hence it is advisable that clinic collect its own normative data for the protocol followed. Akin and Murnane[16] reported P1-N1 amplitudes of 13 to 178 micro volts for 100 dB nHL clicks and 15 to 337 micro volts for 500 Hz tone bursts at 90 dB nHL. Most studies use interaural amplitude difference/amplitude asymmetry ratio (AAR) for clinical interpretation where

$$AAR = 100 \text{ (amplitude left – amplitude right)/ (amplitude left + amplitude right)}$$

Ratios above 40% are found to be clinically significant.

VEMP results have been studied in various clinical conditions and a brief review of the same is presented.

VEMP and Meniere's syndrome (MD): VEMP is a useful tool in the assessment of patients with MD as saccule is a frequent site of endolymphatic hydrops. MD may lead to absence of P1-N1 complex on the affected side or may affect its amplitude; however latency of P1 and N1 is not affected by the hydrops.[17,18] Murofushi et al[17] showed that 51% of their 43 patients with MD showed abnormal amplitudes on their affected side with normal responses in 21 cases. Young et al[18] found that VEMP were normal in 24 ears, augmented in 3 ears, depressed in 6 ears and absent in 7 ears in 40 patients with MD. Latencies were not significantly delayed in any case. They also studied how different stages of MD affected VEMP results. VEMP were normal in 83% of stage I ears of MD indicating that the sacculocollic reflex retains normal velocity conduction in the earliest stages of MD. Augmented VEMP were explained by these authors as the dilatation of the saccule due to the hydrops, extending to press against the footplate of stapes thereby enhancing the sensitivity of

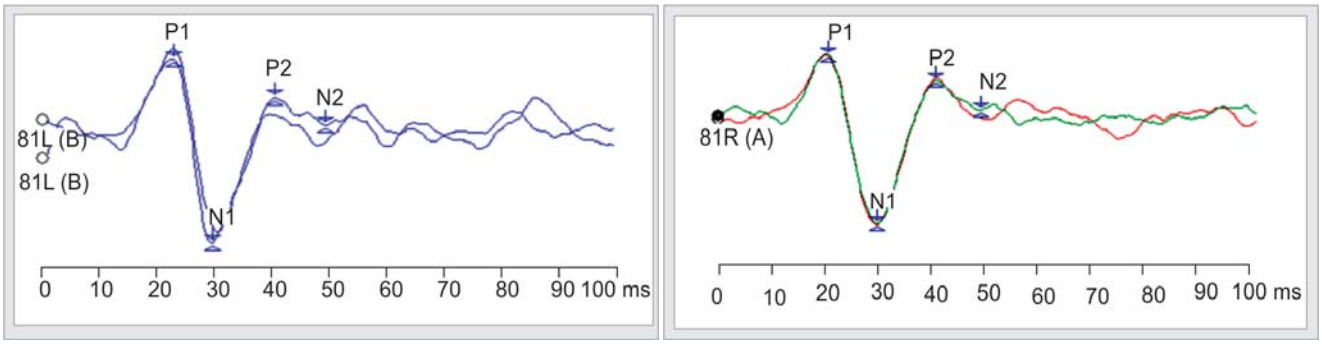


Fig. 1: cVEMP tracings in the right and left ears

saccular macula to loud sounds. A dilated saccule with atrophied saccular macula could be the explanation for depressed VEMP. Absent VEMP in stage IV of MD could result from the saccular wall collapsing onto the otolithic membrane. They demonstrated that the AAR of VEMP increased significantly with the stage of MD and was better correlated with disease stage than caloric testing.

Many investigators have used VEMP as a part of glycerol test wherein amplitudes of VEMP in MD patients were measured pre- and postglycerol intake.[19,20] Cianfrone, Gagliardi, Cuiuli and D'Amico[19] used distortion product otoacoustic emissions (DPOAE) and VEMP to monitor cochlea saccular performance in 29 patients with early MD pre- and postglycerol intake. After analysis of DPOAE and VEMP results, they divided their patients in 4 groups; first group had a postglycerol improvement with both measures suggestive of hydrops in both anterior and posterior parts of the labyrinth. In the second and third groups, there was an improvement in either one of the measures indicating that only one endolymphatic compartment was involved. In the last group all patients had positive glycerol test with DPOAE on one side and with VEMP on the other side. Thus, they highlighted the importance of both the tests in the early diagnosis of endolymphatic hydrops.

VEMP and Acoustic Neuroma

Since the neural pathway for VEMP involves the vestibular nerve, VEMP has been used in evaluation of nerve functioning in patients with acoustic neuroma (AN). Murofushi, Matsuzaki and Mizuno[21] reported abnormal VEMP in 80% of 17 patients with AN.[15] Patients had no VEMP while remaining two showed decreased amplitude. Another study by Murofushi et al[23] showed that 77% of 48 patients with AN had absent or decreased amplitude VEMP; 4 patients with large tumors showed prolonged P1 latency suggestive of lesion in vestibule-spinal tract.

Further, Wang, Hsu and Young[22] studied VEMP and Caloric responses in patients with neurofibromatosis type 2 (NF2) and found 71% ears exhibited abnormal caloric responses while only 14% ears displayed absent VEMP—a significant difference. This indicated that NF2 tumors originated from the superior vestibular nerve more often than the inferior vestibular nerve. Inferior vestibular nerve invasion is possible if the NF2 tumor is sufficiently large. Thus VEMP can serve as an indicator of the degree of NF2 infiltration without operation.

VEMP and Multiple Sclerosis

Shimizu, Murofushi, Sakurai and Halmagyi[23] tested 3 patients with multiple sclerosis (MS) using VEMP and found P1 latency to be significantly delayed. They attributed this delay to demyelination of either primary afferent axons at the root entry zone or the secondary vestibulospinal tracts and concluded that VEMP could be a useful clinical test to evaluate the function of vestibulospinal pathways. Tu and Young[24] evaluated a MS patient with ABR, caloric testing and VEMP and found all the tests to be useful in monitoring the audiovestibular functioning in MS. They speculated that absent VEMP implied a progressive demyelinating process causing severe damage to the myelin sheath while prolonged P1 latency was attributable to reduction in conduction velocity along the demyelinated fibers. Patko, Simo and Aranyi[25] conducted an extensive study in 30 patients with MS to determine the factors leading to abnormality of VEMP in MS patients. They found that the amplitude of VEMP was significantly lower and the latency of the P1 peak was longer in MS group as compared to the typical group. However, the sensitivity of VEMP in identification of MS was 39%, i.e. lower than the other evoked potentials. They stated that VEMP was able to demonstrate subclinical dysfunction as majority of their patients with abnormal VEMP were free of vertigo or other clinical signs of vestibular dysfunction. Majority of patients with abnormal VEMP had extensive morphological abnormality in the brain stem as shown on MRI; the correlation was statistically significant. Further the chances of VEMP abnormality was higher in advanced stages of the disease.

VEMP and Tullio Phenomenon

Tullio phenomenon refers to patients who have vestibular symptoms in response to acoustic stimulation such as vertigo, imbalance and discomfort. Colebatch, Rothwell, Bronstein and Ludman[26] tested seven patients with tullio phenomenon and showed that all patients had a reduced (better) threshold of click-evoked VEMP; threshold of $\leq$70 dB nHL could be used as a diagnostic criterion for Tullio effect. They thought that the better threshold could be due to increased effectiveness of transmission of sound energy to saccular receptors and this overactivation probably contributed to the vestibular symptoms experienced by these patients.

One of the conditions causing Tullio phenomenon is superior semicircular canal dehiscence (SCD), i.e. a defect of the bony capsule forming the roof of the superior semicircular canal. Watson, Halmagyi and Colebatch[27] studied high resolution CT scans and VEMP in four patients with Tullio phenomenon. VEMP thresholds were lower (better) for all affected ears and normal for three unaffected ears. All ears showed SCD on CT scan. They proposed that patients with Tullio phenomenon should undergo both HRCT and VEMP. If both reveal dehiscence, further

investigations, such as middle ear exploration to exclude stapes dislocation or a perilymph fistula are not required. Similar findings of lower VEMP thresholds are also reported in patients with perilymph fistula.

VEMP and Vestibular Neuritis

Vestibular neuritis is a frequent cause of vertigo diagnosed on clinical history and caloric testing which shows unilateral deficit. Superior vestibular nerve (SVN) is more frequently affected than inferior vestibular nerve (IVN), hence many researchers did not find VEMP a useful tool in the evaluation of patients with vestibular neuritis. However, if the IVN is affected, VEMP may reveal absence of response or significant AAR or prolonged latency.[28] Halmagyi and Colebatch[29] studied 22 patients with vestibular neuritis using VEMP and caloric testing. All patients had no caloric responses on the affected side indicating dysfunction of the lateral semicircular canals whereas VEMP showed reduced amplitudes in five patients, absent responses in 11 patients indicating involvement of the IVN; normal responses in six patients were seen. Thus, VEMP can help to localize lesion in cases of vestibular neuritis. Further, Murofushi et al[28] found that in patients with vestibular neuritis, presence or absence of VEMP was prognostic of BPPV occurrence. In 47 patients of acute vestibular neuritis, 10 developed subsequent BPPV of the posterior canal of the same side as the neuritis. All these patients had normal VEMP inspite of vestibular neuritis whereas BPPV did not occur in any of the 16 patients with absent VEMP responses. They concluded that if VEMP is absent after vestibular neuritis, BPPV is unlikely to develop as the IVN is affected in these cases.

Fujomoto, Murofushi, Chihara, Suzuki, Yamasha and Iwasaki[30] reported three patients with bilaterally absent VEMP in presence of normal caloric responses. They considered these cases to be a subtype of idiopathic bilateral vestibulopathy which had selectively affected the IVN and spared the SVN.

VEMP and Sudden Hearing Loss

Stamatiou, Gkorista, Xendlis, Riga and Korres[31] evaluated VEMP findings in 86 patients with unilateral idiopathic sudden hearing loss and compared results with caloric responses. They found that 30.2% patients showed abnormal VEMP while 52.3% had abnormal caloric responses. Combining results of these both tests gave a clearer picture of the vestibular involvement. Patients with normal findings on both tests had a lesion confined to the cochlea whereas those with abnormal findings on both tests had extensive lesions involving the cochlea, saccule and semicircular canals. Prevalence of vertigo as a clinical symptom was greater in patients with lesions of both saccule and semicircular canals.

Ocular Vestibular Evoked Myogenic Potentials

Rosengren, Todd and Colebatch[32] demonstrated that short latency electromyographic changes could be recorded from extraocular muscles in response to auditory stimulation and labelled them as ocular VEMP or oVEMP. The response is vestibular in origin and reflects contralateral otolith-ocular function.[33] The oVEMP waveform is characterized by an initial negative peak at 10 to 12 msec (N1) and a subsequent positive peak (P1) at 15 to 20 msec. It can be elicited using air conduction or bone conduction clicks or tonebursts as well as using forehead taps and galvanic stimulation. It is typically recorded from the inferior oblique muscle with active electrodes placed 1 cm infraorbitally beneath each eye and the reference electrodes placed directly beneath the active electrodes, i.e. 3 cm infraorbitally. The ground electrode is placed at FPz. The patient is asked to gaze upwards by 30° to activate the inferior oblique muscle.

The oVEMP response can be used as an diagnostic tool along with other vestibular tests as air conducted cVEMP represents the saccular and inferior vestibular nerve functioning whereas AC oVEMPs mainly help in evaluating utricular or superior vestibular nerve functioning.[34,35] The combination of oVEMP and cVEMP test can be used as screening tool in order to assess crossed vestibular ocular reflex and ipsilateral sacculocollic reflex in patients with unilateral vestibular hypofunction.[36] In patients with total unilateral vestibular neuritis in whom saccular function is intact but utricular functioning is affected, shows reduced n10 response.[37]

Lin, Hsu and Young[38] used a combined approach of oVEMP and cVEMP to test children with BPPV. Normal oVEMPs are obtained in BPPV patients as compared to abnormal cVEMPs thus, reflecting intact vestibulo-ocular reflex pathway that travels though upper brain stem. Hence, oVEMP helps in differentiating site of lesion to lower *vs* upper brain stem when compared with cVEMP that reflects retrolabyrinthine lesion along the sacculocollic pathway.

Patients with semicircular canal dehiscence (SCD) have significantly larger sound evoked vestibular reflexes. In patients with SCD the oVEMP amplitude is significantly large as compared to the normals with reduced n10 latencies.[39]

The oVEMP test also helps in detecting patients with Meniere's disease. Patients with Meniere's disease show lower amplitudes and higher thresholds as compared with

normal subjects. The frequency tuning also vary in these patients as compared with normals, i.e. 1000 Hz.[40]

In addition, oVEMP test can also differentiate between cerebellar and brain stem lesions. Abnormal oVEMPs are obtained in patients with cerebellar disorder which may indicate adjacent brain stem involvement.[41] Iwasaki et al[42] found that oVEMP response was reduced or absent with asymmetry in amplitude ratios in patients with vestibular schwannoma.

Thus, use of a comprehensive battery of above tests and caloric testing/ head-impulse test, can help to determine the integrity of the entire labyrinth noninvasively. PTA gives an estimate of peripheral hearing loss. Speech audiometry, acoustic reflex testing and ABR can differentiate cochlear *vs* retrochlear lesions causing SNHL. cVEMP can assess saccule and inferior vestibular nerve functioning whereas oVEMP is a diagnostic indicator of lesions of the utricle and superior vestibular nerve. Inclusion of these tests along with the more commonly used caloric testing/head-impulse test will lead to a holistic assessment of the audiovestibular system. The utility of the above-mentioned test battery can be elucidated using a few case examples as presented below:

Case 1

LK, a 28-year-old case was referred by neurology department for ABR. She was a known case of left cerebello-pontine angle tumor. She complained of decreased hearing and tinnitus in the left ear since 1 year. Complaints of headache, nausea and giddiness were also present.

Pure tone audiometry of the patient revealed:

Right ear: normal hearing sensitivity

Left ear: severe sensorineural hearing loss.

Click ABR was done:

Wave morphology was extremely degraded in the left ear (Fig. 2) with no identifiable waves even at maximum intensity and low repetition rates. Right ear shows well-formed early waves I, II and III but latter waves were reduced in amplitude and delayed. At high repetition rates, no waves were clearly defined.

Comments

Left ear: Patient has a severe sensorineural hearing loss in the left ear. In ABR testing, no peaks were identifiable even at maximum intensity of 100 dB nHL the left ear. This could be a result of the severity of peripheral hearing loss or due to presence of retrocochlear pathology—cerebellopontine angle tumor on the left. Thus, ABR alone is not a reliable tool for differential diagnosis of cochlear *vs* retrocochlear sites of lesion, if the hearing loss in that ear exceeds 65 to 70 dB HL. However, in cases of large mass lesions, contralateral effects on ABR may provide diagnostic information.

Right ear: Patient had normal hearing sensitivity in the right ear and no complaints. So a well-defined ABR with normal values was expected. However, the ABR showed an atypical morphology wherein the early waves (I-III) were robust and well-formed than waves IV and V that were relatively less well-defined and smaller in amplitude. Further, the early

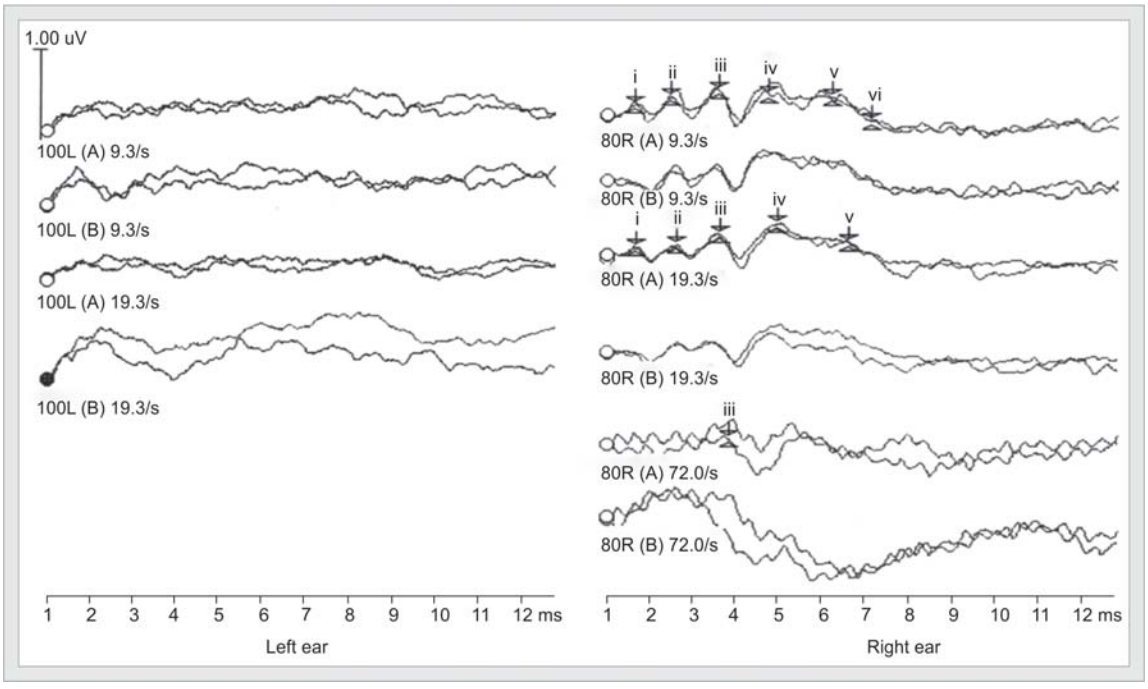


Fig. 2: ABR findings

waves showed normal absolute latencies and Interpeak latency intervals (IPL). But IPL V-III and therefore V-I were significantly prolonged. At higher repetition rate of 72/sec, the wave morphology showed complete degradation with absence of all the wave peaks.

These findings cannot be attributed to any pathology of the right ear. It is the contralateral effect of the cerebellopontine angle tumor on the left side. MRI of the patient revealed an extraaxial mass of the left cerebellopontine angle causing a mass effect in the form of rotation of the brain stem toward the right and effacement of fourth ventricle with resultant dilatation of bilateral lateral ventricles and the third ventricle. The pressure effects of the tumor at upper brain stem level near the fourth ventricle due to the rotation of the brain stem toward the right is responsible for the delayed latencies of latter waves and their reduced amplitudes.

Note that in this case the MRI was available prior to ABR testing. The case emphasizes the utility of ABR in cerebellopontine angle tumors large enough to cause pressure effects by brain stem displacement to the opposite side by virtue of significant latency and morphology changes in the latter waves. Complete absence of latter waves may also be seen in such cases. Latter waves are affected as their generators /sources are mainly the superior olivary complex and the Lateral Lemniscus located in the upper brain stem.

In this case, tone decay tests and immittance testing, especially reflex testing and reflex decay testing would help to corroborate the results. Otoacoustic emission (OAE) testing could also be undertaken.

Case 2

KR is a 22-year-old known case of acoustic schwannoma in the right internal auditory meatus. He presented with complaint of decreased hearing in right ear since 1 year and objective vertigo.

His pure tone audiometry revealed bilateral hearing sensitivity within normal limits.

MRI findings: A 4.5 mm intracanalicular mass seen in the right internal auditory meatus, probably acoustic schwannoma.

His click ABR was done (Fig. 3):

Wave morphology was good in the left ear but poor in the right ear. Only wave V could be identified in the right ear even at 80 dB nHL presented at a rate of 9.3/second.

The following are the absolute latencies and the Interpeak intervals based on the averaging of 4000 responses at 80 dB nHL for the right ear and 70 dB nHL for the left ear.

	Right			*Left*		
	9.3	19.3	72	9.3	19.3	72
I	—	—	—	1.77	1.85	1.90
III	—	—	—	3.92	3.98	4.03
V	7.4	7.42	8.22	5.78	6.08	6.25
I-III	—	—	—	2.15	2.13	2.13
V-III	—	—	—	1.85	2.10	2.22
V-I	—	—	—	4.00	4.23	4.35

Following points are to be noted:

- The absolute and interpeak latencies in the left ear are within normal limits

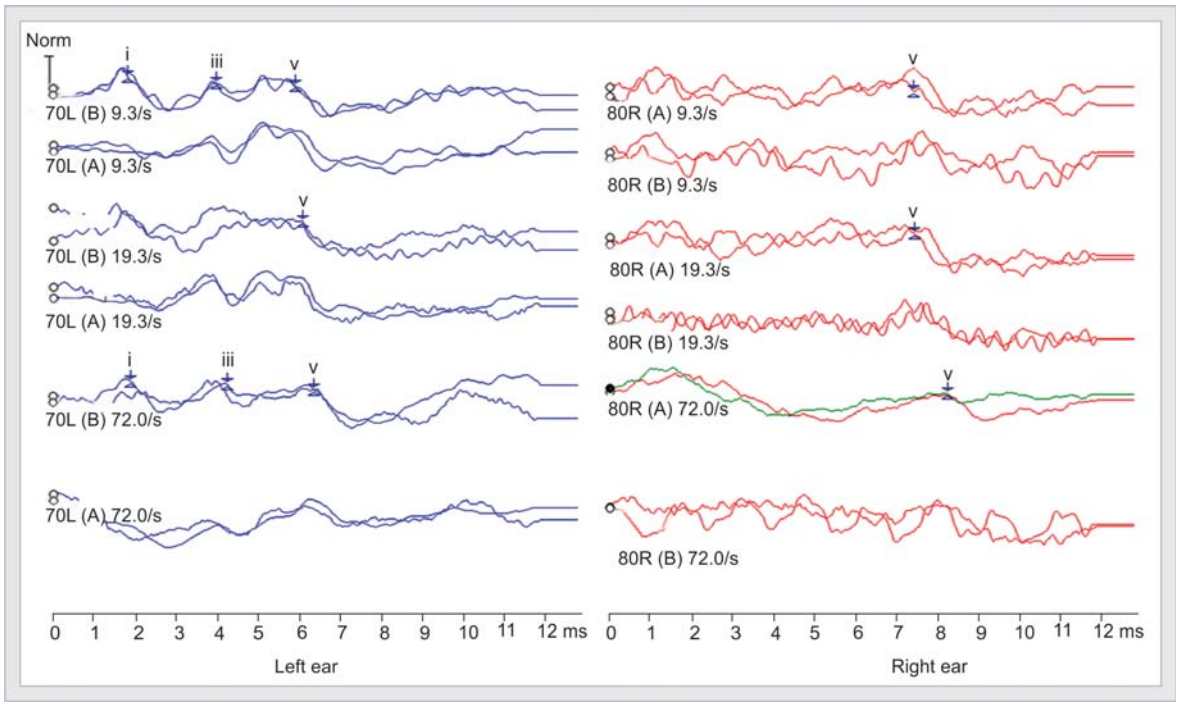


Fig. 3: ABR findings

- Absolute latency of wave V in the right ear is significantly prolonged as compared with the left ear leading to inter aural latency differences of 1.62, 1.34 and 1.98 ms at the repetition rates of 9.3/sec, 19.3/sec and 72/sec respectively. In the presence of symmetrical hearing, this finding is strongly indicative of retrocochlear pathology.

Cervical and Ocular Vestibular evoked myogenic responses (VEMP) were recorded to ascertain the integrity of the sacculocollic pathway and the vestibulo-ocular pathway respectively.

CERVICAL VEMP

cVEMP was recorded unilaterally using 81 dB nHL tone bursts presented via inserts and response was recorded from the sternocleidomastoid muscle (Fig. 4).

Wave morphology was good. P1 and N1 waves were recorded at normal latencies in both ears and amplitude of P1-N1 complex was within normal limits bilaterally. These findings indicate integrity of inferior vestibular nerves and the descending vestibule-spinal tracts as is expected in a small intracanalicular tumor.

OCULAR VEMP

oVEMP was recorded with 81 dB NHL tone bursts presented via inserts and reponses were recorded from the inferior oblique muscles.

Right ear amplitude (Fig. 5) is significantly reduced as compared to left ear, and latency of N1 is prolonged in the right ear, suggestive of compromised integrity of the vestibule-ocular reflex, in v/o MRI findings, it could suggest involvement of superior vestibular nerve.

Comments

Findings on ABR strongly suggestive of retrocochlear pathology on the right side consistent with presence of acoustic neuroma. This case highlights the use of ABR in early detection of acoustic tumors. Further, use of VEMP further can give us an idea of the branch of vestibular nerve involved tumor. Periodic monitoring of the patient with ABR and VEMP will help to determine the growth of the tumor and aid in the decision making for further management.

Case 3

MG, M/55 years reported with complaint of decreased hearing and tinnitus in the left ear since 2 to 3 years, episodic vertigo and vomiting. PTA revealed normal hearing sensitivity in the right ear and a moderate sensorineural hearing loss in the left ear. ABR revealed good waveform morphology with replicable waves bilaterally. The inter-peak latencies were also within normal limits thus suggestive of normal findings in the right ear and a cochlear site of lesion in the left ear. MRI testing was done to confirm

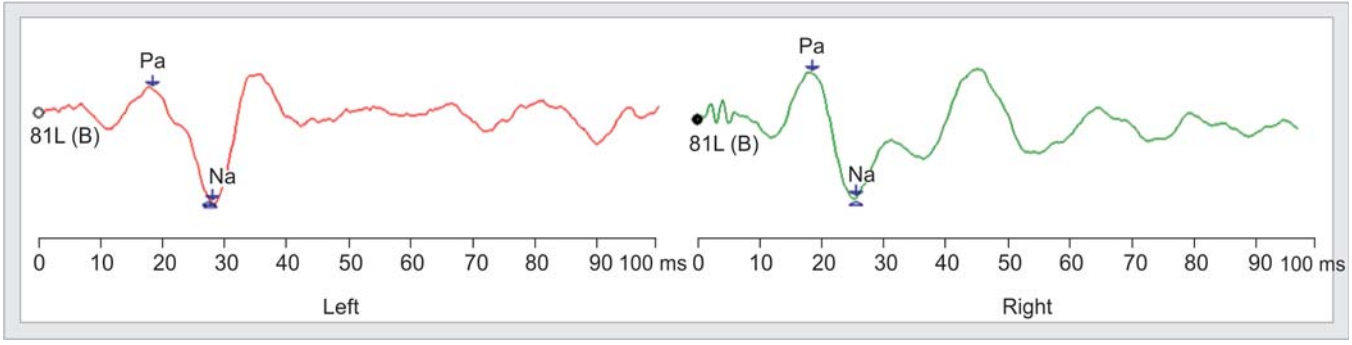


Fig. 4: cVEMP findings

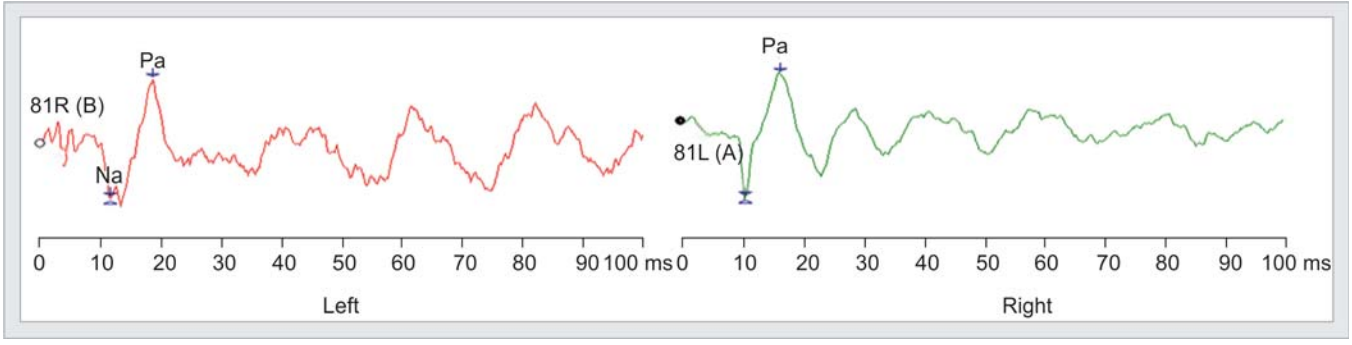


Fig. 5: oVEMP findings

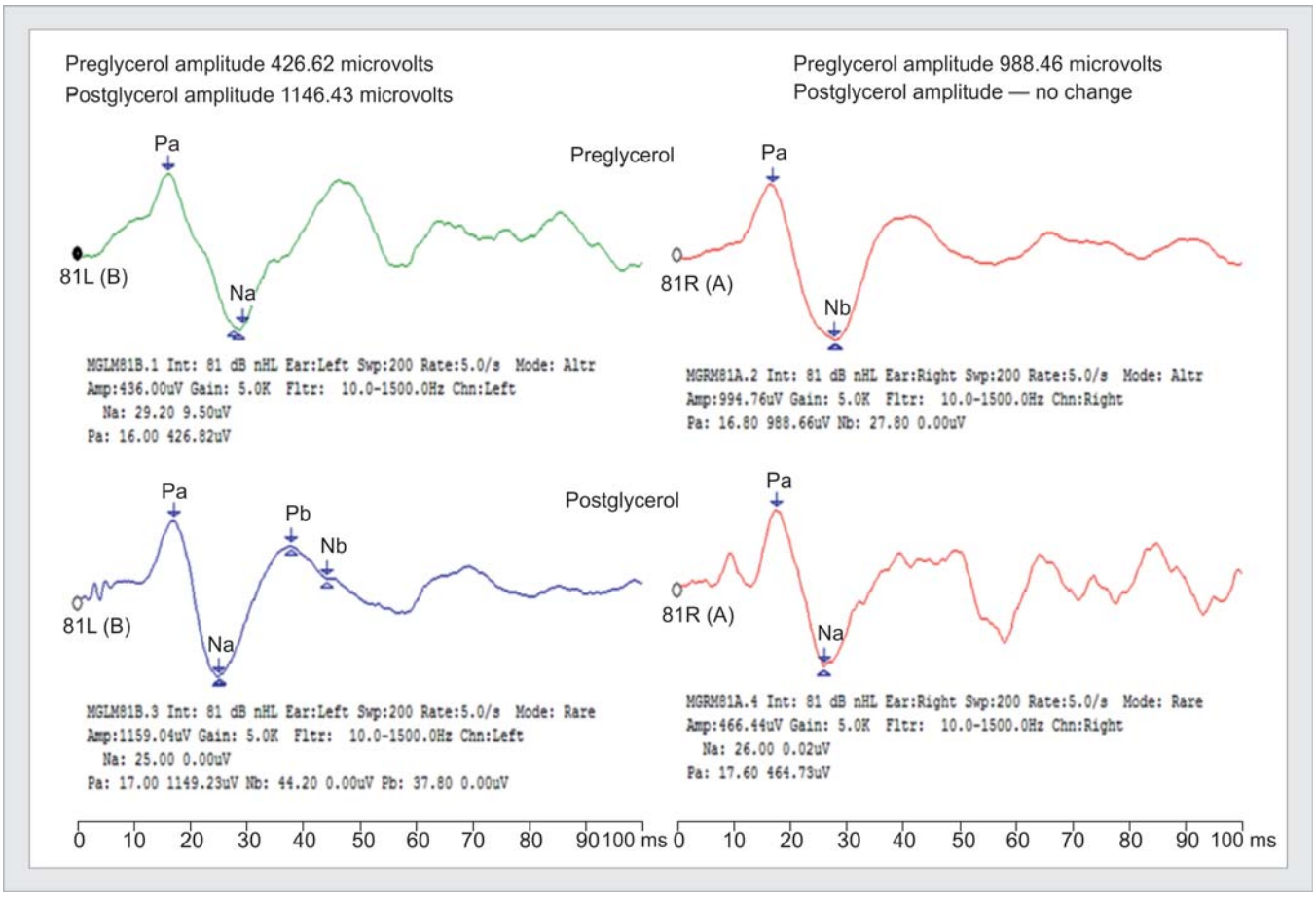


Fig. 6: cVEMP findings pre- and postglycerol

absence of space occupying lesion and no abnormality was detected.

Based on the above findings and case history, Meniere's disease was suspected and patient was undertaken for cVEMP testing. Following were the cVEMP amplitudes of the patient in two ears:

Right: 988 microvolts

Left: 426 microvolts

Thus, left ear had reduced amplitude as compared with the right ear as well as in comparison to age-specific norms. The AAR was 0.36 that is greater than the norm and was diagnostic of saccular lesion. To confirm the diagnosis, glycerol testing was done where in the patient was administered glycerol mixed with saline solution orally in a single dose of 1 ml per kg per body weight. PTA and cVEMP were repeated two hours postglycerol intake and these readings were compared with the preglycerol test results (Fig. 6).

As we can see, in the affected ear, i.e. left ear, the cVEMP amplitude showed an increase by more than 100% whereas no improvement was noted in the right ear. Thus, cVEMP can be used in the diagnosis of Meniere's disease and, in this case, denotes saccular hydrops.

Presence of hypoactive lateral semicircular canal was observed on the head impulse test. Overall, the findings suggest that the disease has affected the cochlea, saccule as well as semicircular canals thus helping to reach a holistic assessment of the labyrinth.

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