

ORIGINAL ARTICLE

Vestibular Evoked Myogenic Potential Responses in Behçet's Disease

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Objective: To evaluate the vestibular evoked myogenic potentials responses in patients with Behçet's disease.

Materials and Methods: Thirty-three patients (66 ears) with the diagnosis of Behçet's disease and 33 (66 ears) age and sex matched healthy volunteers were enrolled in the study. Logon type stimulus; 500 Hz frequency at an intensity of 120 dB HL with a 4/s stimulation rate was delivered to elicit the VEMP responses. The sternocleidomastoid muscle was chosen as the target to record the VEMPs. During the recording period subjects were in supine position with head elevation and simultaneous binaural acoustic stimulations were used.

Results: The response rate of p1n1 wave was 69.7 % for Behçet group and 89.4 % for the control group (X^2 , $p=0.009$). For Behçet group, the mean latencies of p1 and n1 were 13.7 ± 1.2 ms and 19.7 ± 1.4 ms, the mean amplitude of p1n1 wave was 12.2 ± 5.9 μ V. For control group, these values were 14.2 ± 1.3 ms; 20.8 ± 2.1 ms and 14.1 ± 6.1 μ V, respectively. Although there were no significant differences between the two groups with respect to p1 latency and p1n1 amplitude, n1 latencies were significantly shorter in Behçet group.

Conclusion: This preliminary report notify that in Behçet's disease, the VEMP response rate is lower and n1 latency was shorter than the healthy subjects.

Submitted : 01 September 2011

Accepted : 08 January 2012

Introduction

Behçet's disease (BD) bears the name of a Turkish dermatologist Hulusi Behçet who described the triad of recurrent oral and genital ulcers and uveitis in 1937^[1]. BD is a multisystem disease with an unknown cause in which an inflammatory perivascularitis can arise in almost any tissue. Definite diagnosis of BD is based on recurrent oral ulcerations together with at least two of the following disorders: eye lesions, genital ulcers, skin lesions, or pathergy^[2]. Since Behçet's original description, many musculoskeletal, gastrointestinal, urogenital, cardiac, cutaneous, and neurologic symptoms have been attributed to BD^[3].

BD is a chronic, multisystemic vasculitis with perivascular infiltration and affects many organs

including audio-vestibular system. It is a well-known fact that the hearing loss at high frequencies can be seen in Behçet's patients, especially in Neuro-Behçet's disease (nBD)^[4]. There is no correlation between audiologic findings and vestibular dysfunction in Behçet's patients. However, peripheral vestibular dysfunction and abnormal central vestibular findings have been reported in BD^[5]. The inflammatory process that leads to BD may be the cause of vestibular disturbances.

Vestibular Evoked Myogenic Potential (VEMP) has been described by Colebatch and Halmagyi in 1992. VEMP is a clinical test that assesses saccule and inferior vestibular nerve function^[6]. The VEMP is an

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inhibitory potential recorded from the sternocleidomastoid muscle (SCM) in response to intense sounds^[7]. VEMP is generated by activation of the saccular afferents and moving to the neurons of Scarpa's ganglion, through the inferior vestibular nerve, lateral or inferior vestibular nucleus, and medial or lateral vestibulospinal tract, and finally to the motor neurons of the SCM^[8,9]. This synaptic way is usually called sacculocollic reflex. Damage or lesions on any part of this synaptic way can cause impairment of the VEMP recording. For example, multiple sclerosis, vestibular neuritis, brainstem lesion (Wallenberg's syndrome) and stroke can disturb the VEMP responses^[9].

The aim of this study is to investigate the VEMP parameters in patients with BD.

Subjects and methods

Subjects

Thirty-three patients (66 ears) with the diagnosis of BD (Behçet group) and 33 (66 ears) age and sex matched healthy volunteers (control group) were enrolled in the study. The diagnosis of BD was relied on criteria of the International Study Group for Behçet's disease^[10]. Otoscopic examination was performed for all subjects and they were also evaluated with pure tone audiometry, tympanometry and stapedial reflexes on the first visit to document the possible otologic pathologies. BD group was not classified according to presence or absence of vestibular symptoms. Presence of conductive hearing loss is an exclusion criteria for Behçet group. A detailed history was taken to exclude the presence of otologic diseases and vertigo in control group. The hearing thresholds of all subjects were <25 dB HL at the frequencies of 250, 500, 1000, 2000, 4000 and 8000 Hz in control group.

The study protocol was approved by our Institutional Review Board and a written informed consent was obtained from all subjects.

VEMP recording

The SCM was chosen as the target to record the VEMPs. Surface EMG activity was recorded with Epic-Plus evoked acoustic potentials system (Labat

S.r.l. Mestre, Italy). The recording electrode was placed on the middle of the ipsilateral clavicle, the reference electrode was placed on the middle third of the ipsilateral SCM, and ground electrode was placed on the center of sternal manubrium. Attention was paid to place bilateral electrodes on symmetrical sites. The EMG signal was amplified and bandpass-filtered (10–1500 Hz). Logon type stimulus with a 500 Hz frequency was delivered at an intensity of 120 dB HL with a 4/s stimulation rate. Recordings were obtained averaging 200 stimuli and two traces from each test were obtained to assess reproducibility. Figures 1 and 2 show images of a Behçet patient's and normal subject's records.

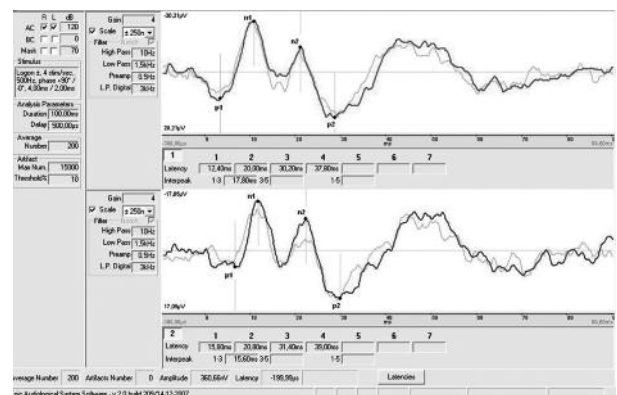


Figure 1. An example of Behçet's patient VEMP recording. Upper trace is the right ear and lower trace is the left ear.

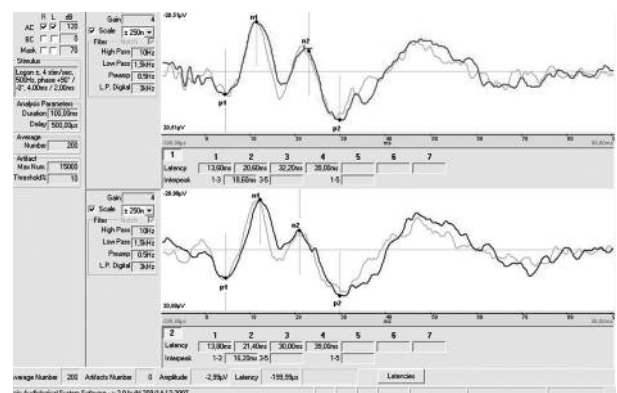


Figure 2. An example of normal subject VEMP recording. Upper trace is the right ear and lower trace is the left ear.

Data analysis

The initial positive-negative polarity of waveform with peaks termed p1 and n1 was used to determine the

presence or absence of the VEMP response. The latency of each peak (p1, n1), and peak to peak amplitude (p1– n1 amplitude) were measured. These parameters were compared between the two groups. All the statistical analyses were performed by using SPSS 18.0 for windows statistical program package and a p-level of 0.05 was considered to be the limit of significance.

Results

There were no otologic disease and history of vertigo in the control group and hearing thresholds were normal in all subjects. Although otoscopic examination was normal in all Behçet group patients, bilateral symmetrical moderate sensorineural hearing loss was detected in one patient and mild bilateral symmetrical high frequency sensorineural hearing loss were detected in other 3 patients. Typical VEMP responses were obtained in these 4 patients. The mean age was 36.2 ± 8.3 (range: 20-53) for Behçet group and 36.7 ± 8.5 (range: 19-53) for the control group. M/F ratio was 17/16 for Behçet group and 16/17 for the control group. There were no statistical differences between two groups with respect to age and sex. The response rate of p1n1 wave was 69.7% for Behçet group, and 89.4% for control group (Table 1). The difference was statistically significant (chi-square test, $p=0.009$). For Behçet group, the mean latencies of p1 and n1 were 13.7 ± 1.2 ms and 19.7 ± 1.4 ms, the mean amplitude of p1n1 wave was 12.2 ± 5.9 μ V. For control group, these values were 14.2 ± 1.3 ms; 20.8 ± 2.1 ms and 14.1 ± 6.1 μ V, respectively. The differences were insignificant for p1 latency and p1n1 amplitude. However, there was a significant difference between two groups with respect to n1 latency (student's t test, $p=0.003$). n1 latency of Behçet group was slightly shorter than the control group (Table 2). When we compare the VEMP results between right ears and left ears of Behçet group, there were no statistically differences between the two groups for VEMP parameters.

Table 1. VEMP response rates in Behçet and control groups

Group	p1n1 response (%)	
	Positive	Negative
Behçet	69.7	30.3
Control	89.4	10.6
Chi-square (p-value)	0.009	

Table 2. p1 and n1 latencies and p1-n1 amplitudes of Behçet and control groups.

	p1 latency (ms) Mean \pm S.D.	n1 latency (ms) Mean \pm S.D.	p1n1 amplitude (μ V) Mean \pm S.D.
Behçet n: 46	13.7 ± 1.2	19.7 ± 1.4	12.2 ± 5.9
Control n:59	14.2 ± 1.3	20.8 ± 2.1	14.1 ± 6.1
P	0.055	0.003	0.107

Student's t test.

Discussion

Behçet's syndrome is a systemic relapsing inflammatory disease with an unknown etiology and characterized by chronic multisystem vascular inflammatory disease that involves many organs ^[1]. The frequency of neurologic involvement in BD shows a high degree of variation in different series (2.5%-49%)^[12].

The central nervous system (CNS) is the major target of neurological involvement in BD. It was generally accepted that there are two categories of CNS involvement in BD; parenchymal and non-parenchymal involvement. Brainstem manifestations are the most common presentation of nBD. Pathologic findings of parenchymal CNS involvement include perivascular cuffing with lymphocytes or neutrophils and rarely eosinophils. There are demyelination with vasculitis, multifocal necrosis, and glial proliferation ^[1].

Audiovestibular disturbances can be presented as tinnitus, dizziness and hearing loss in BD ^[13]. Alajouanine et al. (1961) described gaze paretic nystagmus and hearing loss in BD in 1961 ^[14]. Following this report, the incidence of hearing loss and

vestibular involvement in BD has been reported with a wide variation between 12% - 80% [4,13-15]. In our study, we found that; four Behçet's patients had sensorineural hearing loss. They were nBD patients, but VEMP responses were obtained in all of them.

Normal VEMP responses are characterized by biphasic (positive-negative) waves, usually labeled "p" (for positive) and "n" (for negative) for each peaks. The first biphasic complex is usually called as p1-n1, and the second one is called as n2-p2. The response rate of p1-n1 was reported between 70% and 100% in healthy subjects depending on the stimulus pattern, stimulus intensity and test positions [16-20]. Erbek et al.(2008) investigated VEMP responses in patients with BD previously [21]. They obtained VEMP responses in all patients. They reported that p1 and n1 latencies were significantly prolonged in Behçet's patients. However, our findings are contradictory to Erbek et al's findings. In our study, the response rates of p1-n1 were 69.7% and 89.4% for Behçet group and normal group, respectively. VEMP response rate was significantly lower in BD than control group. There was no significant difference between two groups in respect to p1 latency and p1-n1 amplitude. But we found that n1 latency was slightly shorter in Behçet group.

There was not enough study in the literature to conclude VEMP parameters in BD. Our preliminary report shows that there are some certain changes in VEMP responses in BD. VEMP response rate was significantly lower in BD than control group. Inflammatory process in BD may affect the sacculocollic reflex pathway and may diminish the VEMP responses in some BD patients. It was also found that n1 latency was slightly shorter in BD group. Although the difference was statistically significant, we are not sure that if it can imply any clinical importance. We have also no idea about the causes of this n1 latency shortening. As a conclusion; this study points that, VEMP testing can be used as a diagnostic tool for the evaluation of Behçet's patients. However, we need further studies with large populations to classify the VEMP characteristics in BD.

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