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Cervical vestibular evoked myogenic potential and high-frequency audiometry results in Behçet's disease

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Abstract

Background: Hearing is commonly impaired in Behçet's disease (BD). Also, vestibular abnormalities as well as cochlear function affection have been found. The inflammatory process which leads to BD may be the cause of vestibular frequency audiometry abnormalities in those patients. The aim of this study is to assess hearing and vestibulo-colic reflex in Behçet patients using convential pure-tone audiometry (cPTA), high-frequency audiometry (HFA), and cervical vestibular evoked myogenic potential (cVEMP).

Results: This study included 25 patients with BD (group I), whose age ranged from 21 to 45 years, with a mean age of 34.24 ± 7.07 years. The control group (group II) comprised 20 normal adults whose ages are between 21 and 45 years, with a mean age of 30.8 ± 8.02 years. The measured cVEMP parameters which include the mean peak latency of P13 and the mean peak latency of N23 were statistically significantly different between the patients suffering from BD and the control groups. All BD patients had high-frequency sensorineural hearing loss (SNHL) starting from 4 up to 12.5 kHz. The mean thresholds of the audiometry were significantly higher in the diseased group.

Conclusion: cVEMP testing can be used for the evaluation of the vestibulo-colic reflex in BD patients. The occurrence of hearing loss especially in high-frequency regions is a common finding in patients suffering from BD, and a HFA may be a valuable tool in early exploration of hearing loss in patients with BD.

Keywords: Behçet's disease, Cervical VEMP, Hearing loss, High-frequency audiometry

Background

Behçet's disease (BD) is an immune-mediated vasculitis characterized by being a systemic disease and was first described by Hulusi Behçet, who first recognized and reported in 1937 a triad of symptoms consisting of recurrent eye inflammation, oral ulcers, and genital ulcers [1]. The presentation of disease is not limited to these three signs; it also involves many organs, including the central nervous system, gastrointestinal tract, lungs, skin, veins, and arteries [2]. BD presents worldwide, although the highest incidence is in the Mediterranean, Middle East, and Far East [3]. BD is predominant in males with a male to female ratio of 7:1 [4].

Hearing loss at high frequencies can be seen in BD patients, especially in neuro-Behçet's disease [5]. Hearing deficit in BD has been examined in several studies. Sensorineural hearing loss (SNHL) occurs in 24 to 55% of cases [5, 6]. Peripheral and central vestibular systems affections were found in 20–40% of patients with Behçet's disease [6–8].

Kulahli et al. [9] reported that there was no correlation between hearing impairment and vestibular abnormalities in Behçet's patients. However, both peripheral and central vestibular abnormalities have been found in BD [10–13]. Also, cochlear function is affected in BD with a higher incidence than vestibular system affection [13].

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Inner ear dysfunction in BD could be explained by the presence of vasculitis or neuropathy occurring on top of chronic inflammation process associated with BD [14].

Cervical vestibular evoked myogenic potential (cVEMP) has been described by Colebatch and Halmagyi [15]. cVEMP is a useful tool that assesses saccule and inferior vestibular nerve function [15]. cVEMP is an inhibitory potential in response to intense sounds [16]. 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 vestibulo-spinal tract and finally to the motor neurons of the sterno-cleido-mastoid muscle [17, 18].

This study aimed to assess hearing and vestibulo-colic reflex in Behçet patients using convential pure-tone audiometry (cPTA), high-frequency audiometry (HFA), and cervical vestibular evoked myogenic potential (cVEMP) in patients suffering from Behçet's disease.

Methods

The study is a case-control one that was conducted on 25 patients suffering from BD (group I). All the patients fulfilled the International Criteria of BD [19]. The purpose of this study was explained to all participants and an informed consent was obtained. The study protocol was approved by the medical ethics committee. All procedures performed were in accordance with the Helsinki Declaration ethical standards.

Exclusion criteria

None of the patients in the study or the control groups had any past history of hearing loss, ear infection, ear trauma, acoustic trauma, or usage of ototoxic drugs. Also, associated systemic diseases that can affect the hearing were excluded as infectious diseases (e.g., mumps, CMV), granulomatous diseases (e.g., sarcoidosis, Wegner's granulomatosis), other autoimmune diseases (e.g., Rheumatoid arthritis, Cogan's syndrome, polyarteritis nodosa, relapsing polychondritis), and diseases of the bone (e.g., Paget's disease).

All subjects who participated in this study were subjected to the following:

1. *Full history taking*
2. *Otological examination*
3. *Audiological evaluation (cPTA)* using clinical audiometer (Otometrics, Madsen, model Obiter 922, Denmark), in a sound-treated room (Amplisilence model E), and a TDH 39 earphones calibrated according to the ISO *standard*. Tonal audiometry in the frequency range 0.25–8 kHz and higher frequencies at 10 and 12.5 kHz were tested. Speech audiometry including speech reception threshold (SRT) using Arabic spondee words [20] and word discrimination score (WDS), using Arabic phonetically balanced (PB) words [21].

The cPTA for air conduction thresholds at 500, 1000, and 2000 Hz is used to classify the degree of hearing loss in each ear, as shown in Table 1 [22].

4. *Immittancemetry* was done using Otometrics (Madsen, Zodiac901, Denmark), calibrated according to the ISO *standard*, using single-component, single-frequency tympanometry with a probe tone of 226 Hz. Testing of the acoustic reflex threshold was done for ipsilateral and contralateral elicited reflexes, using pure tones at 500, 1000, 2000, and 4000 Hz.
5. *Cervical VEMP (cVEMP)* was performed using evoked potential: Interacoustics Eclipse "EP15".

Surface electrodes were placed as follows :The active (positive) electrode is placed on both upper 2/3 of sternocleidomastoid muscles right and then left, the inverting (negative) electrodes are placed on the upper sternum (suprasternal notch), and the remaining electrode (the ground one) is placed on middle of the forehead. Two repeatable results were recorded for each condition. Subjects were given 30 to 60 s to relax between each recording to avoid fatigue. During recording, the subject was instructed to raise his head and tilt it away from the stimulated side throughout the test to ensure good muscle tone. cVEMP responses were obtained by acoustic stimulation using short tone burst at frequency 500 Hz of a rarefaction polarity; 4 ms rise/fall time and 2 ms plateau, Blackman ramp was used for stimulation. The intensity used was 95 dBHL presented through TDH39 headphones. Stimulus repetition rate was 5.1 pulses per second. The accepted numbers of responses were 200 sweeps. VEMP responses were judged as either present or absent according to the presence or absence of P13-N23 biphasic response. The latencies of peaks p13 and n23, and p13–n23 peak to peak amplitude and inter-aural amplitude difference (IAAD) ratio

Table 1 Classification of degree of hearing loss

Degree of hearing loss	Hearing loss range (dB HL)
Normal	−10 to 15
Slight	16 to 25
Mild	26 to 40
Moderate	41 to 55
Moderately severe	56 to 70
Severe	71 to 90
Profound	91+

were measured. The IAAD was evaluated as follows: [(Ar − Al)/(Ar + Al) × 100], where Ar indicates the P13/N23 peak to peak amplitude on the right side, Al that on the left side, and [Ar − Al] the absolute value of (Ar − Al). IAAD was chosen rather than absolute amplitude to abolish the inter-subject differences because of tonic EMG activities [23, 24].

Statistical methods

All statistical calculations were done using computer programs Microsoft Excel (Microsoft Corporation, NY, USA) and SPSS (Statistical Package for the Social Science) statistical programs (SPSS Inc., Chicago, IL, USA). Data were statistically described in terms of mean ± standard deviation (± SD), in terms of range or in terms of frequencies (the number of cases), and in terms of percentages if possible. Descriptive statistics were done for quantitative data as minimum and maximum of the range and the mean ± SD (standard deviation) for the quantitative parametric data, while it was done for qualitative data as number and percentage. Inferential analyses were done for quantitative variables using unpaired t test in cases of two dependent groups with parametric data for comparing categorical data chi square test (χ^2) was performed. The Mann-Whitney method was used to compare the two unrelated groups. Correlation coefficients were calculated using Pearson correlation analysis. p values not more than 0.05 was considered statistical significant.

Results

The present study was carried out on 25 patients with BD whose age ranged from 21 to 45 years, with a mean age of 34.24 ± 7.07 years. They comprised 10 females (40%) and 15 males (60%). And the control group (group II) comprised 20 normal adults whose ages vary between 21 and 45 years, with a mean age of 30.8 ± 8.02 years. They comprised 10 females (50%) and 10 males (50%). There were no statistically significant difference in the results between the two groups as regards age and gender. The mean of the duration of the disease in the group suffering from BD was 9.44 ± 7.93 (vary from 1 to 35 years).

The clinical findings in BD patients are presented in Table 2: all patients had recurrent aphthous oral ulceration. Also, genital ulceration was found in 21 (84%) patients, eye lesions in 25 (100%), cutaneous lesions in 20 (80%), neurologic manifestations in 6 (24%), gastrointestinal manifestations in 5 (20%), vascular manifestations in 25 (100%), and arthritis in 7 (28%). Complaint of hearing loss was found in 7 (28%) and dizziness attacks in 9 (36%) patients. Abnormal cVEMP was observed in 23 (92%) patients and abnormal high-frequency audiometry result was present in all patients.

Table 2 Clinical characteristics and VEMP and PTA abnormalities of BD patients

Manifestations in descending order of frequency	Patients, no.*(%)
Oral ulcer	25 (100)
Eye manifestations	25 (100)
Vascular manifestations	25 (100)
High-frequency audiometry (HFA) abnormality	25 (100)
Abnormal VEMP	23 (92)
Genital ulcer	21 (84)
Skin manifestations	20 (80)
Dizziness	9 (36)
Arthritis	7 (28)
Hearing loss	7 (28)
Gastrointestinal manifestations	5 (20)
Neurologic manifestations	4 (16)

*NB, numbers are not mutually exclusive as some patients had multiple complaints and manifestations

All patients suffering from BD had high-frequency sensorineural hearing loss (SNHL) starting from 4 up to 12.5 kHz ranging in degree from mild to severe (Figs. 1 and 2). As regards the right ear, 5 (20%) patients had mild, 7 (28%) had moderate, 11 (44%) had moderately severe, and 2 (8%) had severe SNHL. As regards the left ear, 5 (20%) patients had mild, 3 (12%) had moderate, 12 (48%) had moderately severe, and 5 (20%) had severe SNHL. Only 3 (12%) BD patients demonstrated bilateral mild SNHL at cPTA and HFA. There were significant differences between Behçet's patients and controls with regard to cPTA ($p < 0.05$) in RT and LT ears at all tested frequencies except at 1.2 kHz in LT ears.

In this study, cVEMP was absent bilaterally in 8 patients (32%) and absent unilaterally in 3 patients (12%): 2 (8%) Rt and 1 (4%) Lt. So in 44%, IAAD could not be calculated. The remaining 56% of BD patients with bilaterally preserved cVEMP response (number = 14) BD patients. IAAD ratio was abnormal (prolonged) in 1/14 BD patients (7.14%), and normal in 13/14 (92.86%) patients.

The mean of the latencies of P13 and N23 were found to be delayed in patients suffering from BD when comparing them to the controls. These differences were found to be statistically significant. The mean P13-N23 amplitudes were higher in controls than BD patients and this difference was statistically significant. As regards the mean of the IAAD ratio, there were no statistically significant differences between the 2 groups (Table 3).

There was a statistically significant positive correlation between cPTA and the age at all tested frequencies except at LT 250 Hz, 500 Hz, 1.8 kHz, and RT 12.5 kHz. However, the negative correlation between age and cVEMP N23 latency in the Lt ear only was found to be statistically significant. We did not find any statistical significant correlation between cVEMP amplitude and the age (Table 4).

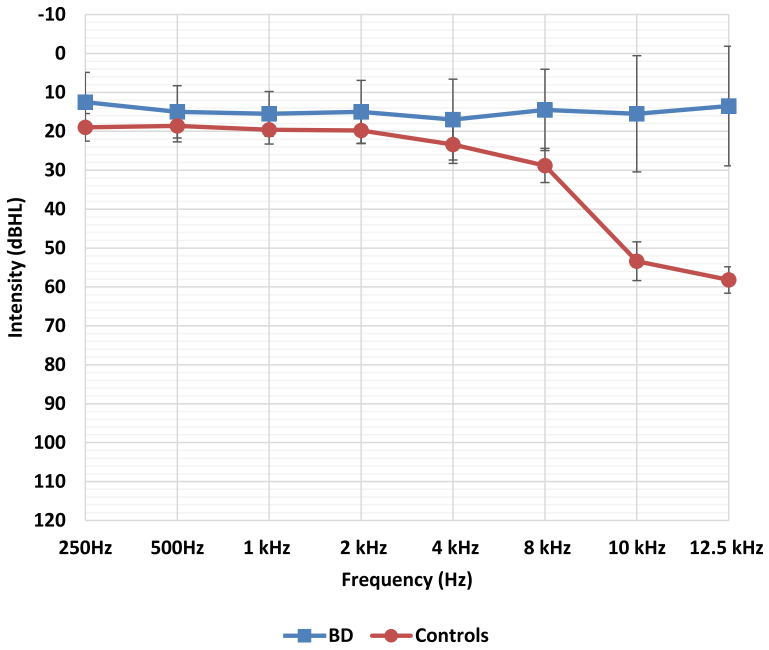

Fig. 1 PTA of BD patients and controls in the RT ears

We did not find any statistically significant difference between BD patients group with disease duration > 5 years and < 5 years regarding PTA results (Table 5).

There were no statistically significant differences between the group of the BD patients with disease duration > 5 years and < 5 years regarding cVEMP results (Table 6) except in N23.

There was no statistically significant difference between BD patients with and without neurological manifestations regarding cVEMP results (Table 7).

There was no statistically significant difference between BD patients with and without dizziness as regards cVEMP results (Table 8).

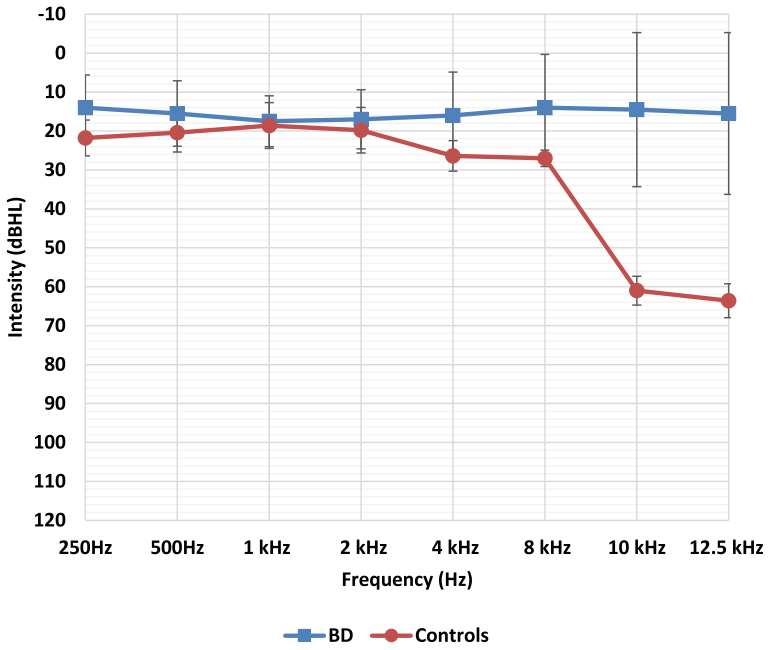

Fig. 2 PTA of BD patients and controls in the LT ears

Table 3 Comparison between BD patients and the controls regarding cVEMP results

VEMP parameters		Controls (n = 20)				Patients (n = 25)				P value
		Mean	SD	Min	Max	Mean	SD	Min	Max	
P13 latency(ms)	Rt	13.6	1.2	12	15.2	17.2	1.7	14.1	20.1	< 0.001
	Lt	14.2	1.2	12.9	16	17.13	1.7	13.8	20	< 0.001
N23 latency(ms)	Rt	22.23	1.1	20.8	23.8	25.3	1.94	21	29.2	< 0.001
	Lt	22.5	1.9	19.3	26.5	25.4	2	20.9	28.3	0.001
P13-N23 amplitude(μv)	Rt	62.24	34.1	33.2	152.4	36	14.1	11	62.4	0.007
	Lt	67.2	17.5	49.8	100.2	41.4	18.1	19.7	82.3	0.002
Inter-aural amplitude difference ratio		18.2	13.1	1	33	15.1	13.6	0.4	46.7	0.586

Discussion

Behçet's disease is a chronic, multi-systemic generalized vasculitis that may affect many organs [25]. Hearing impairment exists in autoimmune diseases and usually occurred at high frequencies [26, 27]. Several studies reported that hearing loss affects about 12 to 80% of patients with BD [28–30].

Table 4 Correlation between age of BD with PTA and VEMP results

		Age (years)	
		R	P value
PTA 250 Hz	RT	0.422	0.036*
	LT	0.219	0.292
PTA 500 Hz	RT	0.373	0.066
	LT	0.168	0.422
PTA 1KHz	RT	0.516	0.008**
	LT	0.129	0.538
PTA 2KHz	RT	0.537	0.006**
	LT	0.575	0.003**
PTA 4KHz	RT	0.612	0.001**
	LT	0.581	0.002**
PTA 8KHz	RT	0.408	0.043*
	LT	0.386	0.057
PTA 10KHz	RT	0.485	0.014*
	LT	0.444	0.026*
PTA 12.5KHz	RT	0.297	0.149
	LT	0.433	0.030*
VEMP P13 latency	RT	0.128	0.614
	LT	− 0.341	0.206
VEMP N23 latency	RT	− 0.171	0.496
	LT	− 0.681	0.005*
VEMP P13-N23 Amplitude	RT	− 0.265	0.288
	LT	− 0.362	0.185

*significant p-value
**highly significant p-value

In the present study, hearing loss was found in 7 patients (28%). All BD patients had high-frequency sensorineural hearing loss (SNHL) starting from 4 up to 12.5 kHz ranging in degree from mild to severe. Only 3 (12%) BD patients demonstrated bilateral mild SNHL at cPTA and HFA.

Bakhshaee et al. [31] and Erdinç et al. [5] reported that hearing loss was the fourth most common clinical manifestation, respectively. Also, Gemignani et al. reported that hearing loss was the fifth most common clinical manifestation of BD [13].

Bayraktar et al. observed sensorineural hearing loss in 9.3% of 32 patients with Behçet's disease, evaluated with audiometry, tympanometry, and acoustic reflex tests and all patients were bilateral. They also found SNHL to be the fifth most common symptom in their study [32].

The present study agreed with that of Bayram et al. who mentioned that the number of patients suffering from SNHL was significantly higher in the group of patients than the control group both in cPTA and HFA. In HFA, seven patients were detected to have SNHL who were in normal limits in cPTA [33].

Bakhshae et al. reported also hearing loss with down sloping configuration at high frequencies in 60% of the BD group by using high-frequency audiometry [31]. Süslü et al. stated that in a study performed with PTA including high frequencies (250–16,000 Hz), SNHL in BD patients begin at high frequencies, instead of SNHL in routine frequencies of the speech [34]. Kemal et al. found that 34.5% of BD patients had a SNHL particularly at the higher frequencies. The mean hearing thresholds were higher in the BD group than in the controls at all frequencies, but only the differences at 1 kHz and were statistically significant [35].

Also, Nada et al. demonstrated that hearing impairment is definitely present in BD patients evidenced by PTA, even in the presence of normal hearing sensitivity. They assessed 30 BD patients using pure-tone audiometry (PTA), otoacoustic emissions (TEOAEs, DPOAE), auditory brainstem response test (ABR), and cortical auditory evoked potentials (tone and speech CAEPs) and

Table 5 PTA results in BD patients with disease duration > 5 years and BD patients with disease duration < 5 years

PTA		Patients with disease duration > 5 years (n = 18)				Patients with disease duration < 5 years (n = 5–2)				t value	P value
		Mean	SD	Min	Max	Mean	SD	Min	Max		
250 Hz	RT	19.20	8.60	5.00	40.00	18.61	4.80	10.00	25.00	− 0.171	0.865
	LT	22.50	9.00	10.00	40.00	20.00	7.10	10.00	30.00	− 0.660	0.516
500 Hz	RT	19.70	7.20	10.00	40.00	15.70	4.50	10.00	25.00	− 1.368	0.185
	LT	20.80	9.30	10.00	40.00	19.30	6.11	15.00	30.00	− 0.406	0.688
1KHz	RT	19.20	5.20	10.00	30.00	20.70	7.31	10.00	30.00	0.595	0.558
	LT	20.00	6.40	10.00	35.00	15.00	5.80	10.00	25.00	− 1.794	0.086
2KHz	RT	19.70	6.50	10.00	35.00	20.00	11.90	10.00	45.00	0.075	0.941
	LT	20.30	5.31	10.00	30.00	18.60	12.11	10.00	45.00	− 0.498	0.623
4KHz	RT	22.50	7.90	10.00	40.00	25.70	15.70	15.00	60.00	0.687	0.499
	LT	27.50	9.60	10.00	50.00	23.61	14.90	10.00	55.00	− 0.786	0.440
8KHz	RT	28.30	10.80	15.00	60.00	30.00	10.00	20.00	50.00	0.352	0.728
	LT	27.80	13.00	15.00	50.00	25.00	16.31	10.00	55.00	− 0.448	0.659
10KHz	RT	53.60	13.90	30.00	85.00	52.90	18.50	30.00	80.00	− 0.111	0.912
	LT	60.00	20.40	25.00	95.00	63.60	19.50	40.00	95.00	0.398	0.694
12.5KHz	RT	57.80	15.20	35.00	90.00	59.30	16.90	40.00	90.00	0.216	0.831
	LT	65.00	20.90	30.00	90.00	60.00	21.60	30.00	100.00	− 0.532	0.600

found that the highest abnormality of CAEP latencies elicited by 500 Hz and 1000 Hz as well as speech stimuli (da and ga) among BD patients was delayed P1 and N1 waves at 80 dB with greater bilateral affection, with significant differences between patients and controls, denoting that BD patients had a sub-clinical cochlear pathology which was not affected by disease activity or different organ affection [36].

Regarding cVEMP results, in this study, cVEMP was absent bilaterally in 8 patients (32%) and absent unilaterally in 3 patients (12%): 2 (8%) Rt and 1 (4%) Lt. The remaining 14 patients (56%) had bilaterally preserved cVEMP response. The mean latencies of P13 and N23 were found to be statistically significantly delayed in BD group when compared to the controls. The mean P13-N23 amplitudes were statistically significantly higher in controls than BD patients. But we did not find any statistically significant differences between both groups as regards the mean of the IAAD ratio. In 44%, IAAD could not be calculated. In the remaining 56% of BD patients, IAAD ratio was abnormal (prolonged) in 1/14 of BD patients (7.14%), and normal in 13/14 (92.86%) of patients.

In agreement with our results, Erbek et al. investigated cVEMP findings in patients with BD. They reported that the latencies of P13 and N23 were significantly delayed in the Behçet's group [37].

Bayram et al. in their study on 30 BD patients and 30 controls reported that 5 patients had delayed cVEMP, while two patients had decreased cVEMP and three patients had absent cVEMP responses at all. But they did not find any significant difference between the BD group and the control groups (p =

Table 6 Comparison between BD patients with disease duration > 5 years and < 5 years regarding VEMP results

VEMP parameters		Patients with disease duration > 5 years (n = 18)				Patients with disease duration < 5 years (n = 5 Rt, 4 Lt)				t value	P value
		Mean	SD	Min	Max	Mean	SD	Min	Max		
P13 latency (ms)	Rt	16.70	1.80	14.10	20.00	18.41	1.11	17.10	20.10	2.005	0.062
	Lt	17.00	1.90	13.81	20.00	17.60	0.50	17.00	18.00	0.624	0.543
N23 latency (ms)	Rt	24.60	1.80	21.00	27.11	26.90	1.40	25.80	29.20	2.536	0.022*
	Lt	24.70	2.40	20.11	27.00	26.90	1.00	25.90	28.30	1.750	0.104
P13-N23 amplitude (μv)	Rt	36.87	13.80	11.00	61.80	36.60	16.40	18.90	62.40	− 0.033	0.974
	Lt	40.50	16.0	10.90	69.70	41.80	29.00	19.70	82.30	0.115	0.910
Inter-aural amplitude difference ratio		17.71	14.81	0.40	46.70	8.51	7.61	2.00	16.30	− 1.157	0.270

*significant p-value

Table 7 Comparison between BD patients with and without neurological manifestations regarding VEMP results

VEMP parameters		Patients with neurological Manifestations (n = 4)				Patients without neurological Manifestations (n = 21)				t value	P value
		Mean	SD	Min	Max	Mean	SD	Min	Max		
P13 latency (ms)	Rt	17.40	1.90	15.30	18.80	17.10	1.80	14.10	20.10	0.232	0.820
	Lt	16.80	0.90	16.10	17.40	17.20	1.70	13.80	20.00	− 0.228	0.733
N23 latency (ms)	Rt	25.90	0.10	25.80	26.00	25.11	2.10	21.00	29.20	0.656	0.521
	Lt	26.50	0.80	25.90	27.00	25.11	2.50	20.10	28.30	0.764	0.470
P13-N23 amplitude (μv)	Rt	42.50	3.80	39.60	46.80	35.70	15.20	11.00	62.40	0.745	0.456
	Lt	43.30	0.10	43.20	43.30	40.51	20.60	10.90	82.30	0.186	0.855
Inter-aural amplitude difference ratio		3.00	1.40	2.00	4.00	17.10	13.70	0.40	46.70	− 1.413	0.183

0.06) as regards to the mean latencies of wave p13-n23 response. IAAD values were found to be similar in cVEMP between the two groups [33].

In comparison to the current study, Baylr et al. found that the rates of the response of P13N23 were 69.7% and 89.4% for Behçet group and normal group, respectively. cVEMP response rates were found to be significantly lower in the patients group than the control group. However, they did not find any significant difference between both groups as regards P13 latency and P13N23 amplitude. Also, they found that N23 latency was slightly shorter in the Behçet group. They explained this by that inflammatory process that occurs in BD may disrupt the sacculo-collic reflex pathway and decrease the cVEMP responses in some BD patients [38].

The multi-systemic inflammatory process may affect the vestibular system in patients suffering from BD ranging from 20 to 40% [5, 8].The common cochlear artery and anterior vestibular artery are the main branches of the labyrinthine artery and can be selectively involved by immunologically mediated inflammation [39].

In the present study, 9 patients (36%) suffered from dizziness attacks. However, there were no statistically significant difference between patients suffer from BD with dizziness manifestations or between BD patients with neurological symptoms regarding cVEMP results. This is possibly due to the occurrence of central compensation for the dizziness, with the cVEMP test abnormality remaining as an objective finding of sacculocollic reflex abnormality in patients with BD. Or this may reflect an occurrence of sub-clinical hydrops in some BD patients with BD causing inner ear saccular dysfunction.

Ahmed in his study showed that dizziness, which denotes vestibular involvement, was the fourth most common presentation among the different clinical presentations of BD patients, more common than cochlear involvement presented by hearing loss. This could be explained by multiple causes of dizziness including peripheral and central causes of dizziness in BD [14]. Furthermore, there was no correlation between the audio-vestibular involvement and the other organs involved which could be explained by the multifocal nature of the disease process and different blood supply of the inner ear components [14].

Shabana et al. showed that BD patients had a subclinical vestibular hydrops through the electrocochleography testing and this was not influenced by activity of the disease or different organ affection but was influenced by duration of the disease [40].

In this study, results have shown that as age increased, cPTA thresholds were worse and this was statistically significant at all tested frequencies except at LT 250 Hz,

Table 8 Comparison between BD patients with and without dizziness regarding VEMP results

VEMP parameters		Patients with dizziness (n = 9)				Patients without dizziness (n = 16)				t value	P value
		Mean	SD	Min	Max	Mean	SD	Min	Max		
P13 latency (ms)	Rt	18.0	2.31	14.10	20.11	16.90	1.50	14.80	20.00	1.242	0.232
	Lt	16.40	1.51	15.10	18.00	17.30	1.70	13.80	20.00	− 0.871	0.400
N23 latency (ms)	Rt	26.10	2.40	22.50	29.20	24.90	1.70	21.00	27.00	1.134	0.274
	Lt	23.20	3.60	20.10	27.10	25.80	1.80	20.90	28.30	− 1.840	0.089
P13-N23 amplitude (μv)	Rt	34.10	20.60	11.00	62.40	37.80	11.60	18.90	61.80	− 0.490	0.631
	Lt	43.40	36.10	10.90	82.30	40.20	14.90	19.71	69.70	0.253	0.805
Inter-aural amplitude difference ratio		21.40	10.70	13.80	29.00	14.10	14.11	0.41	46.71	0.693	0.501

500 Hz, 1.8 kHz, and RT 12.5 kHz. As age increased, cVEMPN23 latency in the Lt ear was significantly decreased, but age was not statistically significantly correlated to cVEMP amplitude.

But regarding the duration of BD, patients with disease duration > 5 years showed comparable PTA results to those with disease duration < 5 years as well as cVEMP results except that those with disease duration < 5 years had a statistically significant prolonged N23 in the Rt ear only. This reflects that the disease duration does not adversely affect the audio-vestibular manifestations.

In comparison to results of the present study, Brama and Fainaru stated that higher hearing thresholds occurred more commonly in older patients with a longer duration of the disease [39].

In contrast, Gemignani et al. [13] did not find any relation between the age or the disease duration and affection of the inner ear. Kemal et al. [35] found that there were not any statistically significant relationship between duration of the disease and inner ear involvement. Also, Ahmed [14] reported that neither age of patients nor duration of the disease had any correlation with audio-vestibular involvement.

Conclusion

Autoimmune vasculitis in Behçet's patients can affect the inner ear causing high-frequency hearing loss. Autoimmune vasculitis can also affect the sacculo-colic reflex in Behçet's patients, resulting in cVEMP test abnormalities irrespective of dizziness complaint or neurological manifestations. So high-frequency audiometry may help in the early detection of hearing loss in patients suffering from Behçet's disease together with the conventional pure-tone audiometry. cVEMP test can be used as a clinical tool for the evaluation of the sacculo-colic reflex Behçet's patients.

Abbreviations

BD: Behçet's disease; cVEMP: Cervical vestibular evoked myogenic potential; dBHL: Decibel hearing level; EMG: Electromyogram; HFA: High-frequency audiometry; IAAD: Inter-aural amplitude difference; PB: Phonetically balanced; cPTA: Convential pure-tone audiometry; SNHL: Sensorineural hearing loss; SPSS: Statistical Package for the Social Science; SRT: Speech reception threshold; WDS: Word discrimination score

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Authors' contributions

All authors contributed to the study conception and design. Material preparation and data collection were performed by NMAB and AMAZ. Analysis and interpretation of the data were performed by RAK, MMH, and AOD. The first draft of the manuscript was written by RAK. MMH, AOD, NMAB, and AMAZ commented on previous versions of the manuscript. All authors read and approved the final manuscript. All authors agreed to be personally accountable for their own contributions and to ensure that questions related to the accuracy or integrity of any part of the work are appropriately investigated, resolved, and documented in the literature.

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Availability of data and materials

Data is available upon request.

Ethics approval and consent to participate

The purpose of this study was explained to all participants and an informed verbal consent was obtained. The study protocol was approved by the medical ethics committee of Rheumatoloy and Rehabilitation department; however, the committee's reference number is not available. All procedures performed were in accordance with the Helsinki Declaration ethical standards.

Consent for publication

Consent has been taken by all participants according to the tenets of the Declaration of Helsinki and respecting the patient's privacy.

Competing interests

The authors have no competing interests to declare.

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