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Vestibular evoked myogenic potentials in pediatric patients with familial Mediterranean fever

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ABSTRACT

Objectives: We aimed to investigate whether the chronic autoinflammatory process in familial Mediterranean fever (FMF), which affects numerous systems, results in vestibular dysfunction in pediatric patients being followed up for diagnosis of FMF using VEMP recordings.
Methods: 30 patients (60 ears) diagnosed with FMF and 20 (40 ears) healthy volunteers were included in the study. Following routine ear, nose, and throat examination, transient-evoked otoacoustic emissions (TEOAE) and vestibular-evoked myogenic potential (VEMP) tests were performed.
Results: A total of 30 FMF pediatric patients (13 male, 17 female) and 20 controls (8 male, 12 female) were included in the study. The mean age of FMF patients was 12.13 ± 2.88 years, while that of the controls was 12.90 ± 2.80 years. All of the otoacoustic emission results of the patient and control groups were "pass VEMP recordings received in both ears of patients with FMF (60 ears) and both ears of controls (40 ears). There was no statistically significant difference for latencies or amplitudes for either patients or controls ($p > 0.05$).
Conclusion: In order to research the effect of FMF on vestibular functions, we measured VEMP. However, we did not detect alterations of VEMP in FMF patients.

1. Introduction

Familial Mediterranean fever (FMF), which is common in Turks, Jews, Arabs and Armenians, is characterized by recurrent fever alongside attacks of abdominal pain (peritonitis), arthritis and serositis such as pleuritis. It is an autosomal recessive inherited chronic systemic autoinflammatory disease [1,2]. It is the best-known and most prevalent hereditary periodic fever syndrome [3]. FMF patients have been demonstrated to have elevated serum levels of IL-1β and TNF-α during both attacks and attack-free periods. It has been demonstrated that, during treatment and even during remission, there is a still subclinical inflammation [4,5]. From this point of view, our patients with FMF were in remission when we aimed to find out whether the vestibular system in FMF patients was affected by inflammation. In Muckle–Wells syndrome, which is another periodic fever syndrome, progressive sensorineural hearing loss is observed in most patients. Although the cause of hearing loss in this syndrome is not clear, it was thought that elevated IL-1β could affect cochlear functions [6–8]. For this reason, supposing cochlear functions can also be affected in FMF patients in a similar manner, studies were conducted investigating auditory functions in this disease [9,10]. However, there are no data regarding the vestibular system in FMF.

While the vestibular system is the main component of the balance system, the visual and proprioceptive senses are contributive systems. The vestibuloocular, vestibulospinal, and vestibulocollic reflex arc between these systems contributes to maintaining balance. The vestibulocollic reflex is responsible for stabilization of the head in space; it acts especially on neck muscle groups. This reflex represents the otolithic function of the inner ear, especially the function of the saccule. For this reason, it is also called the sacculocollic reflex [11]. This reflex arc is formed by high intensity sound vibrating endolymph inside the saccule, and the resulting action potential creating electromyographic stimulus in the sternocleidomastoid muscle via the inferior vestibular nerve, lateral

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vestibular nucleus, and medial vestibulospinal tract in order [12]. Vestibular evoked myogenic potential (VEMP) is the test evaluating this reflex arc.

In this study, we aimed to investigate whether the chronic autoinflammatory process in FMF, which affects numerous systems, results in vestibular dysfunction in pediatric patients being followed up with a diagnosis of FMF using VEMP recordings.

2. Materials and methods

30 patients (60 ears) previously diagnosed with FMF in the Pediatrics unit of Gaziosmanpaşa University Faculty of Medicine, and 20 healthy volunteers (40 ears) were included in our study. FMF was diagnosed according to the criteria of Tel-Hashomer. Informed consent was obtained from all patients' parents, and the local ethics committee approved the study protocol. Exclusion criteria included diabetes mellitus, chronic renal insufficiency, epilepsy, hypothyroidism, any anatomical or functional problem of the external or middle ear, chronic otitis media, acute otitis media, otitis media with effusion, otological surgery, and systemic, metabolic, or autoimmune disease.

Following a complete physical and ear, nose, and throat examinations, transient evoked otoacoustic emissions (TEOAE) and vestibular-evoked myogenic potential (VEMP) tests were performed.

2.1. TEOAE

The hearing levels of both the patient and control groups were measured by transient evoked otoacoustic emission (TEOAE). We used the Madsen Capella system (GN Otometrics, Taastrup, Denmark) otoacoustic emission device. TEOAE results were considered "positive" if the overall value of the correlation percentage was $\geq$50%, and the S/N ratio was $\geq$3 dB at three frequencies [13]. The results were evaluated as "pass" and "refer" for every single ear.

2.2. VEMP

The sternocleidomastoid muscle was selected as the target for recording the vestibular evoked myogenic potential. Surface electrodes were placed on the following positions: active, on the upper one-third of the SCM; reference electrode, on the suprasternal notch; and ground electrode, on the forehead. Patients were positioned supine on a bed, and were instructed to elevate and turn their head contralaterally towards the ear being tested, to achieve maximal contraction of the sternocleidomastoid muscle. VEMP recordings were performed with an evoked potentials machine (Otometrics, ICS Chartr EP 200, Denmark). The acoustic stimuli had an intensity of 95 dB HL and a frequency of 5 Hz, were of 5 ms duration with 1 ms rise and fall, and were conducted to the ears monoaurally with an insertion-type earphone. Electromyographic responses from each side were amplified and bandpass-filtered (10 Hz to 3 kHz). The analysis time was 50 ms. Responses were acquired by calculating the average of 128 stimuli. Two trials were obtained for each test in order to confirm the accuracy of responses. After acoustic stimulation, the first positive myoelectrical peak was considered to be p13 and the first negative peak n23. The p13 and n23 latencies for the three trials were averaged to represent the latencies of each test. Latencies of the peak p13 and n23, interpeak p13–23 interval, and p13–n23 amplitude were measured.

The patient group consisted of 30 pediatric FMF patients. The diagnosis of FMF was made according to the Tel-Hashomer criteria: the presence of at least 1 of 4 major criteria, 2 of 5 minor criteria, 1 minor criterion plus 5 of 10 supportive criteria, or 4 of 5 specific supportive criteria [14]. The disease severity score (DSS) was calculated for each patient. The age of onset was divided into five degrees of severity, the frequency of febrile attacks into three degrees of severity, and arthritis into two degrees of severity (acute and protracted). Erysipelas-like erythema was given a score of 2 and amyloidosis a score of 3. The response to colchicine treatment was divided into four degrees of severity [15]. DSS $\leq$ 5 represents low disease activity and DSS $>$ 5 represents moderate to high disease activity. The patients were classified into two groups according to their DSS; patients with DSS $\leq$ 5 and those with DSS $>$ 5. 13 patients were evaluated as DSS $\leq$ 5, and 17 patients were evaluated as DSS $>$ 5. The patients in present study had no AA-type amyloidosis or chronic renal failure.

3. Statistical analyses

The Statistical Package for Social Sciences software (SPSS, version 20.0 for Windows; SPSS Inc., Chicago, Illinois, USA) was used to perform all analyses. Because the data showed homogeneous distribution, Student's *t*-test was used for comparison of two groups. One-way ANOVA test was used for comparison of more than two groups. All data were expressed as Mean $\pm$ SD. Statistical significance was set as $p < 0.05$.

4. Results

A total of 30 FMF pediatric patients (13 male, 17 female) and 20 controls (8 male, 12 female) were included in the study. The mean age of FMF was 12.13 $\pm$ 2.88 years, while that of the controls was 12.90 $\pm$ 2.80 years. No significant difference was found between FMF patients and the control group in terms of age ($p = 0.766$) and gender ($p = 0.641$). All of the otoacoustic emission results of the patient and control group were "pass" and, therefore, not evaluated in statistical analysis.

VEMP recordings were obtained in both ears of patients with FMF (60 ears) and of controls (40 ears). There was no statistically significant difference in latencies or amplitudes, for either patients or controls ($p > 0.05$). Table 1 presents the latencies and amplitudes for vestibular evoked myogenic potential responses obtained from all FMF patients and control subjects. Table 2 presents the latencies and amplitudes for vestibular evoked myogenic potential responses obtained from patients with DSS $\leq$ 5, those with DSS $>$ 5, and control subjects.

Table 1
Vestibular evoked myogenic potential parameters in FMF and control group (FMF: Familial Mediterranean fever).

	Groups	*n*	Mean	Standard deviation	*p* Value
Left latency p13	FMF	30	14.72	3.04	0.098
	Control	20	15.18	1.43	
Left latency n13	FMF	30	23.66	4.70	0.120
	Control	20	24.10	2.86	
Right latency p13	FMF	30	15.69	1.36	0.465
	Control	20	15.74	1.14	
Right lateny n13	FMF	30	24.71	2.74	0.130
	Control	20	23.88	1.87	
Left interpeak latency	FMF	30	8.93	3.14	0.800
	Control	20	8.99	2.80	
Right interpeak latency	FMF	30	8.38	1.16	0.472
	Control	20	8.17	1.05	
Left amplitude	FMF	30	468.30	192.40	0.144
	Control	20	451.21	221.92	
Right amplitude	FMF	30	492.35	200.61	0.641
	Control	20	516.68	211.92	