“Reality of living with familial mediterranean fever identity”: A phenomenological study

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ABSTRACT

Aims: This phenomenological study was carried out to explore and describe FMF patients’ (n=15) life experiences and coping behaviors.

Methods: This study was conducted in the rheumatology outpatient clinic of a training and research hospital. Data was collected using both a demographic data form and a semi-structured interview form using in-depth, face-to-face interviews and analyzed by Colaizzi’s phenomenological data analysis method.

Results: Data analysis brought out three categories; (1) Living with an unnamed disease, (2) Living with FMF identity, (3) Coping with FMF symptoms. Until being diagnosed with FMF, the patients have been had to live with the symptoms of an unnamed disease. After diagnosis, they try to adapt to FMF identity, which means unexpected recurrent attacks with physical effects, loss of social roles and functions, stigmatization and feelings of guilt. They try to continue their treatment and follow-up with the desire to maintain a healthy life and to cope with FMF.

Conclusions: This study presents how patients experience living with an unnamed disease before being diagnosed, living with FMF identity and coping with FMF symptoms. Findings demonstrated that most of the troubles patients lived were related to the pain during the attacks and resultant limitations. This study shows what really matter for FMF patients and how they try to compete with long-term distress.

Introduction

Familial Mediterranean Fever (FMF) is the most frequent autoinflammatory disease affecting more than 120,000 patients worldwide and more prevalent among the Mediterranean populations such as Jews, Turks, Arabs, Italians, and Armenians (1, 2). It is an inherited disorder characterized by recurrent febrile, inflammatory episodes (abdominal/joint/chest attacks, myalgia, erysipelas-like erythema, etc.), which starts typically in childhood or adolescence. FMF has two typical clinical periods; in inflammation (attack period) patients have an increased acutephase response and in remissions (attack free period) they usually return to normal state of healthy life (3). The attacks usually begin instantaneously, the frequency is quite variable and also the duration varies from days to weeks. It has been suggested that some triggering factor such as infection, trauma, psychological stress, exposure to cold, menstrual cycle, excessive physical activity and probably undetermined many others may initiate these attacks (4, 5, 6, 7). Although the interval between episodes is not predictable and may last days or months, the patients are healthy and functional between attacks during these intervals and typically live healthy (8). In short-term, the primary symptom the patients suffer is usually pain with febrile, inflammatory episodes, which enables the individual performing roles and functions in daily activities. In long-term FMF may cause liver, heart or renal failure because of amyloidosis. Currently, the treatment for FMF is a life-long use of the drug Colchicine, which if started early on, can both alleviate symptoms associated with recurrent attacks and prevent further complications like systemic amyloidosis (9, 10).

Although FMF patients experience the similar main characteristics of the disease, FMF experience is unique for each person. The duration, severity, and personal effects of symptoms are unpredictable and vary from patient to patient. Usually, along with fever, only one sign (peritonitis, arthritis, pleuritis, etc.) may comprise a single attack, although more than one symptom, any combination of clinical signs, may appear at a single attack. Besides, symptoms and the form of the disease manifestation may change in the course of the disease (11).

Therefore, perceptions of FMF experience, the attributed meaning to the disease, reactions, and coping behaviors might be affected by personal and cultural differences in FMF. There

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are many studies in the literature about the medical aspects of FMF, including treatment modalities, disease frequency and prevalence and a wide range of symptoms. Besides, qualitative studies are assessing the life experience of patients suffering from other rheumatic diseases such as scleroderma, ankylosing spondylitis (12, 13, 14). However, to the best of our knowledge, there is no study, which investigates how FMF influences the patients' lives, and coping behaviors. Qualitative studies give an opportunity for determining the experiences of a certain disease by giving the patients voice. So it has substantial importance to provide valuable information to health care professionals how patients experience illness's effects in their daily lives and how to engage in best care practices for patients (15). So the experiences of FMF patients shared in this study may guide to describe the nature and effects of attacks, develop strategies for coping with and giving care to the disease. In this present study, we aimed to investigate and describe FMF patients' life experiences using phenomenological approach.

Methods

Study Design, Sample, and Setting

This phenomenological study was conducted in the rheumatology outpatient clinic of a training and research hospital. In literature phenomenology is described as an approach, which focuses on the subjectivity of reality, continually pointing out the need to understand how humans view themselves and the world around them and it provides an opportunity to investigate the influence of illness on patients' lives (16). Beginning with experience and, through the narration of participants, of either a single shared incident or shared condition, phenomenology investigates the effects and perceptions of that experience (16). So it seems that phenomenological approach may be the best choice to explore and describe FMF patients' life experiences and coping behaviors.

For this purpose, patients with FMF who were being regularly followed-up at the rheumatology outpatient clinic of a university hospital were invited to participate in the study in consultation attendance. All patients who are meeting the inclusion criteria accepted to participate in the study. The inclusion criteria were: (i) fulfilling the classification criteria proposed for FMF by Livneh and colleagues (17); (ii) age ≥18 years; (iii) ability to speak and understand Turkish; (iv) willingness to participate and to share their experience related to FMF; (v) accepting the tape-recorded interview; and (vi) physically and cognitively able to communicate. Exclusion criteria included the following: having a diagnosis of a significant psychiatric disorder, cognitive impairment, comorbid terminal illness, or being clinically unstable. Fifteen participants of both genders were included in the study that was selected gradually according to consultation attendance.

The data were collected after the Institutional Ethics committee of the hospital permitted the study. Participants declared their willingness by written informed consent before the commencement of the study. A full explanation of the study aims and information about researcher's education, interests, personal goals and reasons for doing the research was given to all participants. They were assured that their identity would be kept confidential and informed that they could withdraw their consent at any time. The interviews were audiotaped after the participants confirmed the procedure.

Procedures

Data were collected using in depth, face-to-face interviews. The participants were interviewed in a private meeting room in the out-clinic space where only the participants and the interviewers were present to minimize interruption and ensure privacy. The interviews are two female nursing faculty members with Ph.D. of internal diseases nursing and mental health and psychiatric nursing who had qualitative research methods training courses in 2006 and 2013. The FMF patients did not know the interviewers before who are also the writers of this research; they met the face-to-face at the time of interviews. The interviews took approximately 45 minutes and were based on a questionnaire comprised of two separate sections. Section 1 posed questions including demographic features (i.e. age, sex, marital status, work status), the time of the disease onset (first attack), clinical features of the disease, compliance and response to colchicine and questions from the clinical data form including FMF-severity score-2 (F-SS-2) (18). Two male rheumatologists who are also the writers of this research are working in the rheumatology outpatient clinic of the university hospital and performed FMF-severity score which is used to assign FMF severity disease to 1 of 3 severity levels: mild, intermediate, or severe. Section 2 posed open-ended questions from semistructured interview form about FMF. The items in the interview form were based on the literature, theoretical background and the clinical knowledge and experiences of the investigators. The initial question of the questionnaire was: "What have you experienced regarding living with FMF? The question was followed up with others such as, "which conditions or circumstances have characteristically affected or impressed your experience with FMF?" Probing questions like, 'Tell me more about these' were used to encourage their speech without disturbing their own stories.

The interviews were recorded and transcribed verbatim. One of which was repeated as obtained data required for clarifying and the meanings of interviewee's expressions are confirmed regarding potential inconsistencies. The data collection process was ceased after a total of 15 interviews because the general saturation criterion was met which is described as an interruption of the inclusion of new participants occurred when the data obtained began to present redundancy or repetition (19, 20).

Data analyses

All interviews were transcribed from audiotapes into written Turkish. Two authors analyzed these transcriptions independently, as described by Colaizzi (21, Table 1).

Table 1. The steps in Colaizzi's phenomenological data analysis

1.Read all the participants' description of the phenomenon under study.
2.Extract significant statements that pertain directly to the phenomena.
3.Formulate the meaning of these significant statements.
4.Categorize the formulated meaning into clusters of themes.
5.Integrate findings into an exhaustive description of the phenomenon being studied.
6.Validate the exhaustive description by returning to some of the participants to ask them how it compares to their experiences.
7.Incorporate any changes offered by the participants into the final description of the essence of the phenomenon.

Table 2. Clinical and demographic features of patients (N = 15)

Characteristics	n (%)	mean ± SD
Age (years) (mean ± SD)		33.60 ± 6.37
Sex		
Male (M)	8 (53.3)	
Female (F)	7 (46.7)	
Marital status		
Married	11 (73.3)	
Single	4 (26.7)	
Work status		
Have regular job	12 (80.0)	
Unemployed	1 (6.7)	
Retired/left work due to illness	2 (13.3)	
Family history of FMF	11 (73.3)	
History of appendectomy	5 (33.3)	
History of penicillin use due to misdiagnosis of acute rheumatic fever	3 (20.0)	
Disease onset (years) (mean ± SD)		13.60±7.31
Age at diagnosis (years) (mean ± SD)		20.87±6.66
Disease duration (year) (mean ± SD)		20.00±9.26
Clinical findings*		
Fever	15 (100.0)	
Peritonitis	14 (93.3)	
Pleuritis	8 (53.3)	
Arthritis	7 (46.7)	
Arthralgia	8 (53.3)	
Myalgia	9 (60.0)	
Erysipelas-like erythema	5 (33.3)	
Orchitis	2 (13.3)	
Constipation	6 (40.0)	
Diarrhea	9 (60.0)	
Age at colchicine onset (year) (mean ± SD)		20.93±6.72
Regular colchicine use	14 (93.3)	
Patients with attacks despite colchicine	12 (80.0)	
Experience of colchicine side effects	12 (80.0)	
FMF-severity score (F-SS-2)		
Severe disease	10 (66.7)	
Moderate disease	2 (13.3)	
Mild disease	3 (20.0)	

* More than one symptom may be seen in one patient

The data were examined multiple times for the analyses and the analyses were repeated by more than one researcher to ensure reliability. No software was used to manage data. The significant statements about the phenomenon were extracted and defined as phrases and were organized into categories and clusters of themes, as described by Colaizzi (21). Three categories and five themes of responses emerged from the interviews, which represent the experiences of the participants (Table 4). Following the steps of Colaizzi, the themes and categories are re-turned to the eight participants who accepted to validate if the description compares their experience. The participants offered no changes for the final report, and they mentioned the study thoroughly explained their experiences. Data obtained from this research is reported according to Consolidated Criteria for Reporting Qualitative Research (COREQ) (22).

Results

Participants' Characteristics

The study population consisted of 15 participants (8 male). The mean age of the participants was 33.60±6.37 years. The mean age at the attack onset was 13.60±7.31, and the duration of the disease was 20.00±9.26 years. The clinical, demographic features and disease activity scores of participants with FMF are shown in Table 2.

Table 3. The frequency of attacks

	Frequency (per year)			
	0	1-10	11-20	>20
Attacks (average)	-	4 (26.7)	4 (26.7)	7 (46.7)
Fever (n=15)	-	6 (40.0)	3 (20.0)	6 (40.0)
Peritonitis (n=14)	1 (6.7)	3 (20.0)	4 (26.7)	7 (46.7)
Pleuritis (n=12)	3 (20.0)	8 (53.3)	3 (20.0)	1 (6.7)
Erysipelas-like erythema (n=5)	10 (66.7)	5 (33.3)	-	-
Arthritis (n=7)	8 (53.3)	6 (40.0)	1 (6.7)	-

Table 4. Categories and themes resulted from interviews

Category	Theme
Living with unnamed disease	
Living with FMF identity	Physical effects of the illness
	Loss of social role and function
	Unexpected and unwelcomed guest “Attack”.
	Stigmatization
	Feelings of guilt
Coping with FMF symptoms	Desire to maintain a healthy life
	Adherence to treatment and follow-up

Of participants, 46.7% stated that they experience more than 20 attacks/year, and the duration of the attacks ranges 3-5 days in 93.3%. The frequencies of attacks are shown in Table 3.

Life experiences of participants

The data obtained from the study demonstrated that the patients had been had to face a process that had begun before and continued after the time of diagnosis. In conjunction with this, three categories were determined. These categories and themes have been shown in Table 4.

Living with Unnamed Disease

The participants have been complaining that people including doctors and nurses know little about the symptoms of FMF, the disease cannot be diagnosed in time and confused with other conditions, and therefore the patients are exposed to unnecessary tests and treatments, even undergo to surgical procedures like appendectomy operation. Some of the participants stated that they had been felt worried about the recurrent, but unspecified signs and symptoms; and they had relieved when the disease was diagnosed, after a process that takes months, even years with full of uncertainty.

“I had my FMF diagnosis nearly ten years ago. I didn’t know what it was all about for years. I had had operations constantly. I had an appendectomy. I had another operation since they thought my intestines stick together. They performed another operation claiming that I have cysts. They were all irrelevant; it just turned out to be FMF. I used to have FMF attacks almost more than once a month. It was so frequent, more than 20 in a year.” (Participant Number: 4, F-SS: Severe; F; Age: 38).

Living with FMF Identity

After being diagnosed with FMF, although the patients get rid of the uncertainty that they suffer, continuity of the disease symptoms bring along many other troubles. The pain they experience during the attacks, limiting effect of FMF symptoms in daily activities, and the patients emphasized adversely affected social and occupational functionality as the significant issues related to the FMF symptoms. Almost all of the gathered data was mentioning about severe pain and daily life difficulties that patients experience during the attacks.

Physical effects of the illness: The participants have stated related with FMF that, the most significant issues that they have to face with, were the times during the attacks. The participants complain that along with the pain, they also suffer from the symptoms such as anorexia, fatigue, weakness, diarrhea or constipation. However, these were limited to the time during the attacks, afterward, they retrieve their pre-attack functionality. They also mention that the most debilitating symptom is the pain they feel in the chest, abdomen, and joints.

For describing the pain they suffered, the participants used some of the impressive expressions like;

“As if, somebody pulls off my intestines.”

“As if, they put my stomach into pincers and squeeze continuously.”

“As if there are lightning flashes inside me.”

“As if, a knife was stuck in my stomach.”

The participants used these sentences below to mention the duration and severity of pain they experience:

“I swear it is a pain, whole pain, nothing but the pain” (Participant Number: 9, F-SS: Moderate; M; Age: 36).

“During the attacks, my stomach bloats. I feel as if somebody punches my belly. I guess it is similar to women’s labor pain. Can somebody suffer any pain for three days every month?” (Participant Number: 14, F-SS: Moderate; M; Age: 29).

“The pain carves you inside like a drill. Or, it is like billions of cigarette put out on the stomach. Or, like an alien which lives inside you. A supernatural creature. I live and feel that my life is like hell because of pain.” (Participant Number: 3, F-SS: Severe; F; Age: 39).

Loss of social role and function: The physical effects of the attacks have an unfavorable impact on the social roles and functions of the patients. During the attacks, the patient couldn't maintain the social roles such as partnership, parenting, being a worker, student, etc. and they experience role loss. Consequently, these result in the feeling of failure, worthlessness, insufficiency, and finally they feel sorry. The participants describe the worsening of the sexual and romantic relations with their spouses:

"When you get married, there are certain promises that you give to your spouse. Sometimes I have difficulties in performing my duties as a wife, which causes disappointments. Therefore, occasionally I say yes and have sex despite my pain." (Participant Number: 7, F-SS: Severe; F; Age: 36).

As a parent, a mother who cannot take care of her child during the attacks said:

"My son is my beloved. I was having an attack, with severe pain, on the sofa. He came and said that he was hungry. He was only four. My pain was so severe that I could barely open my eyes and looked at him, but I could not even answer. I saw him opening the fridge, eating whatever meal he found there. This desperateness hurt me deeply. There was food in the fridge, but I couldn't heat and feed him…" (Participant Number: 7, F-SS: Severe; F; Age: 39).

Some of the participants told that the employer does not prefer them because sudden onset of an attack hinders to maintain their jobs, retain them from the position, and cause absenteeism in the workplace. Therefore, some of them have had to quit or change their jobs, and have difficulties in finding new ones.

One of the participants who live difficulties in maintaining his worker role because not going to the work during the attack periods, leave his or her responsibilities to the workmates, and being perceived as goldbricker has stated that:

"The tempo was intensive in my workplace. It was impossible for me to work with pain when I had FMF attacks. The doctors prescribed me bed rest for a few days. My colleagues said that I am getting off from work too much. I got fired three times, and I don't want to be fired again." (Participant Number: 8, F-SS: Severe; M; Age: 28).

Unexpected and unwelcomed guest "Attack": Along with the physical features of FMF that influences social role and functions considerably, another issue that the patients having difficulty in facing is the sudden and unexpected onset of the attack and being dependent for 3-4 days, until the ending of it. The uncertainty of the attack onset disturbs the regularity of life, and the accompanying pain decreases the quality of life. What will tomorrow bring or what will happen in any days to come? The uncertainty about whether they will have an attack or not, affects their life dramatically and increases their anxiety; and one of the participants expressed this issue as follows:

"The anxiety about having an attack anytime/anywhere disturbs me. Therefore I am always overstrung. For instance, if I am too tired or sleepless, oh no, an attack is around the corner. We will go on a holiday, the preparation process itself is a stressor, oh no, this stress may trigger an attack. At my brother's wedding, I had wondered whether I would have an attack or not, and unfortunately, I had. FMF attacks control my life, but I cannot control them at all. A person can control what she eats or how many hours she sleeps, but how can she control the stressors of life or repetition of the menstrual cycle every month?" (Participant Number: 8, F-SS: Severe; F; Age: 39).

Stigmatization: The patients stated that, since the cycle of getting sick/recovering is very fast, and after the attack, they seem entirely healthy, their condition perceived as unrealistic by their friends and relatives, and they considered themselves as stigmatized.

"Normally, I can do everything like anybody else; I am fully functional. I feel good from morning till noon, and then a severe pain comes suddenly, which makes me go to the emergency department. My friends or relatives say, "You seemed good just a few minutes ago. What happened to you in a while? What is wrong with you?" Or, you suffer pain for three days, insomuch that you writhe in pain. The fourth day you get better and functional. Then they say, "You were too bad, how come you recovered so fast?" That is the people's perspective. They don't believe in this disease, treat me as if I am playacting. Not only pain but also I have to fight with stigmatization…" (Participant Number: 7, F-SS: Severe; F; Age: 39).

Feelings of guilt: The patients worry about the genetic transition of FMF to their children. The patients whose children were diagnosed with FMF felt very guilty; and said that, if they foreknew their children would have FMF, they would never have a child, even get married:

"I took my daughter to a doctor, and he told me that she has FMF. I was shocked. I wish I did not get married, and she was never born. If I knew, would I get married? My child will experience just like me. The reason why this child will suffer is I. Therefore, I feel guilty, guilty, and guilty... (Participant Number: 6, F-SS: Severe; M; Age: 31).

Coping with FMF Symptoms

The way of coping behaviors of the patients widely differs. It varies from positive coping behaviors like taking personal precautions to survive a healthy life, to negative coping behaviors like not to use the drugs regularly.

The desire to maintain a healthy life: Most of the participants stated that despite the attacks and related problems, they endeavored hard to manage their health. The patients are trying to continue a healthy life. The patients struggle to take their health under control by avoiding factors such as stress, cold, sleeplessness, fatigue, nicotine, caffeine, sodas, that are considered as triggering factors for their attacks; besides they find additional solutions like quitting smoking, hiring a household assistant, taking support from family or friends.

"I usually do not prefer intercity busses when traveling. Because after traveling by bus, pain, swelling and redness occur at my leg joints. I have to choose either a fast or comfortable way of transportation to protect my joints. Because I should stop and have a rest any time I need. I have lots of things to do at home, cook, clean, iron; but I can't. When I am healthy, I can not do them not to be tired because tiredness triggers attacks, when I have attacks I cannot do them because of pain. Therefore, I had to employ an assistant at home to do those kinds of things. In my opinion, FMF is kind of disease for rich people." (Participant Number: 3, F-SS: Severe; F; Age: 39).

Adherence to treatment and monitoring: Some of the patients displayed positive behaviors such as taking medicines regularly, continuing follow-up visits, and getting psychological support when needed. Besides, they wanted to know how to manage their disease and, they requested help to learn about some issues such as how to control symptoms, drug side effects, etc. On the contrary to these positive behaviors, some of the patients behaved negatively, like ignoring the disease, not taking medications or taking irregularly, overusing of painkillers, social isolation and using herbal products with unknown ingredients. Suicidal ideation was the most ineffective coping behavior, and one-fifth of the participants stated that they considered or even attempted to suicide. They indicated the reasons for suicidal attempt as; “pain is really unendurable”, “not being understood enough by the people who are close to”, “being obliged to respond to the requests of the people around even during the attack period”, and “being devoid of attention and care that they need”.

“I did not take my pills for a year. Then my attacks started to visit me more often. I decided to take my pills, but this time I couldn’t manage to take regularly. I tried to relieve my pain with painkillers. It did not work. Some of my friends offered me some herbals or herbal products which of all also did not work. I was desperate. I felt myself in a trap. (Participant Number: 11, F-SS: Severe; F; Age: 44).

Discussion

In our study, we elaborate the life experiences and perspectives of the patients with FMF. From the onset of the symptoms to the time of diagnosis, the patients with FMF face with symptoms that could not be associated with a particular disease and meet many uncertainties. These symptoms occur unexpectedly and cause physical influences; social loses, stigmatization, and sense of guilt. The patients have to live with this unbearable FMF identity, and with the desire to manage their life healthily, they try to continue their treatment and follow-up and cope with the disease.

Participants’ characteristics features of the patients in this study showed that there were nearly seven years between the first FMF symptom patients’ experience and the diagnosis. Connecting to our qualitative data, this means the participants lived with symptoms for nearly seven years, seeking treatment for an unknown disease, having a large number of laboratory tests, being misdiagnosed with irrelevant disorders and using unrelated drugs till they met with a rheumatologist finally and diagnosed with FMF. As shown in Table 2, until they were diagnosed with FMF nearly one-third of the participants underwent unnecessary procedures such as appendectomy and one in five participants used penicillin due to misdiagnosis of acute rheumatic fever. In their study, Portincasa et al. introduces a list that includes both autoinflammatory syndromes (like Cyclic neutropenia, Behcet’s disease etc.) which require the differential diagnosis with FMF, and other surgical (like Pancreatitis, Cholecystitis, Appendicitis etc.) and systemic conditions (Systemic lupus erythematosus, HIV infection, Crystalinduced ar-thritis (gout, pseudogout), Inflammatory bowel diseases etc.) which could resemble the FMF attacks (23). The process that continues until the time of getting correct diagnosis and treatment is very long and quite worrisome. Although diagnosing the disease decrease the level of anxiety, the patients have emphasized that the symptoms they experienced during the attack periods were significantly influenced their lives. Among the physical effect of the illness, the most disturbing sign is the pain at joints, abdomen, and chest. The other symptoms of FMF include fever, fatigue, weakness, loss of appetite, weight loss, diarrhea (11). Consistent with the literature, all patients in our study emphasizes the severity of the pain (24) is the most disturbing symptom. Moreover, this pain, which considerably affects their life, is the epitome of the disease itself and causes limited functional ability and affects everyday activities (24). In attack periods, patients couldn’t manage to perform their roles like being a spouse, parent, worker, student and related functions. The extents of the role changes and loss that occur as a result of these attacks vary from person to person. Some of them think profession problems are the most significant, where-as others mention limitations in childcare are. Male patients especially state the difficulties that they lived in their professional life. They mentioned this situation as “the important issue is not the pain actually; it is rather not being able to do the job.” This statement demonstrates the deep sorrow they experienced about the disease. On the other hand, female patients complain about their incapability of both giving care for their children and sexual performance. These adverse effects lead to the marital conflicts, lose their hope and joy of living. A substantial group of the participants considers that not being able to plan even a single aspect of their life due to the uncertainty of the timing of the attacks as the most critical issue. Moreover since the risk of where and when the attacks will occur, fear due to the possibility of the emergence of the attacks at any given time, having to plan the life continuously following the course of the disease, lead to psychological effects like anxiety and impatience, to name a few. In the study conducted by Deger et al. (25), it has been demonstrated that depression and anxiety are significantly more common in patients with FMF, and FMF might adversely affect Health-Related Quality of Life.

The other major problems are not being understood by their acquaintances (family, friend, employer, etc.), being considered as unrealistic and accused of role-playing. The underlying reason of these perceptions is the nature of the disease, in other words, the patients seem to be healthy between the attacks, suddenly become severely ill, and recovered in 12-72 hours (26). To be a somewhat different and to have an unfamiliar chronic disease increases the person’s possibility of being stigmatized by other people (27). However, in the literature (12, 28), stigmatization usually is the case for the patients with apparent physical defects, on the contrary in our study, it is noteworthy that our participants claim that stigmatization occur due to the rapid change between being ill and healthy, and being physically normal during the attack-free periods of the disease.

The parents whose children had been diagnosed with FMF stated that, if they had the chance, to prevent facing of their children with this disease, instead of having children, they would prefer not to have a child, or even not getting married; besides they feel guilty about that. In their study, McAllister et al. demonstrated that parents have worry for transferring their genetic disorders to their children; and the feeling about transferring the defective genes to the children, and making them sick, lead to excessive guilt (29).

To minimize the unfavorable effects of FMF on the patients’ lives, they follow carefully the prodromal symptoms of the disease which has been described in the literature as a period, which might extend almost 24 hours before an attack, consists of discomfort and psychological distress (30). They also try to manage the circumstances, which are considered as triggering factors (dietary habits, physical conditions, stressors, etc.).

These precipitating factors given in the literature are viral infections, emotional stress, overexertion, trauma, fatty food, unusually high or low temperatures, and menses (1, 30, 31, 32). Managing these precipitating factors is essential for the patients to cope with the disease. There are some other coping behaviors which prove to be effective, such as adherence to the medications, and paying regular doctor visits; taking preventive measures (making physical arrangements for protecting their health, avoiding factors that are considered as triggering factors for their attacks); getting social or psychological support. On the other hand, the patient may use ineffective coping strategies including ignorance of the disease, not taking medications or taking irregularly, overusing of painkillers, using herbal products without shown beneficial effects. One of the most remarkable findings of our study was that one-fifth of the patients' group mentioned that they planned or even tried to commit suicide as a reaction to unbearable pain periods, not comprehended by family members, be compelled to respond others demands, being deprived of care they need. Pain is frequently encountered symptom in the most of rheumatologic diseases and one of the most disturbing signs that patients complain. Besides, concerning with the pain (33), in the literature, there are data related with unendurable nature and potential for causing suicidal tendencies in rheumatologic diseases (34).

It has been observed that patients, who receive active support from their family, show effective coping behaviors. It has been demonstrated in the study by Savelkoul et al. (24) that social support and effective coping strategies might influence the well-being of FMF patients, and passive coping make the social surroundings not supportive and decrease the subjective well-being.

All these findings emphasize that psychological effects of FMF should be handled carefully. It is not surprising that such biological and psychological effects are influencing their lives, also affects their family, friend, and social relationships. Although it has been suggested by Zadeh et al. (30) that FMF patients might live almost healthy life, in this study, we have demonstrated that, due to the unpredictable attacks, they live difficulties in not only carrying out scheduled or obligatory activities but also in performing their tasks as a parent, spouse or employee.

Finally, to manage the FMF patients properly, it should be taken into account that personal, interpersonal, social and cultural conditions constitute the patient's psychosocial adaptation to a particular disease. In this way, the health care providers can collaborate better with patients and achieve much better clinical outcomes.

Conclusions

This study presents how patients experience living with the unnamed disease before being diagnosed, living with FMF identity and coping with FMF symptoms. It has been demonstrated that most of the troubles they lived were related to the pain during the attacks and resultant limitations.

This study has particular importance since it presents the experiences of FMF patients from their perspectives, reveals a reality of FMF, which is almost impossible by using a simple visual analog scale and demonstrates a picture to health care providers to explain patients' experience. For healthcare providers, the analyses of this experience and subjective beliefs on the cause of the illness makes it clear that what the matter is for patients and how they try to compete with long-term distress. Besides, elucidating and improving the awareness among acquaintance and the family of the patients can make them understand the disease and lead to encourage them to provide further support.

There are strengths and limitations of this of phenomenological study. The phenomenology method used in this study gives an opportunity to understand the universal nature of FMF experience and which may provide a deeper understanding that permits the researchers to explore the big picture. It may contribute to the professionals to understand the patients' experiences better and may emphasize the need for psychosocial support for FMF patients. There are also some limitations. The findings are restricted with the patients' ability to articulate their thoughts and feelings about FMF experience; and the researchers' interpretation of the phenomenon. Like all phenomenological studies, the data obtained from this study cannot be generalized.

Relevance to clinical practice: The experiences of FMF patients shared in this study may guide the health care professionals to describe the nature and effects of attacks, develop strategies for giving care to the disease.

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Conflict of Interest

The authors declare that there is no conflict of interest.

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