Health Related Quality of Life of Children Aged 2-18 Years with Familial Mediterranean Fever

Ailevi Akdeniz Ateşi Olan 2-18 Yaş Arası Çocuklarda Sağlıkla İlişkili Yaşam Kalitesinin Değerlendirilmesi

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ABSTRACT

Objective: We aimed to evaluate both child-self reported and parents proxy reported health related quality of life of children with familial Mediterranean fever (FMF) aged between 2-18 years old and to compare the scores with their healthy peers.

Material and Methods: Fifty -one patients with FMF and fifty one gender and age matched controls were enrolled in the study. Children aged 8-18 years completed the PedsQL4.0 child form questionnaires. Parents of all cases completed the PedsQL4.0 parent form.

Results: Lower scores in the FMF group, with no statistical significance, was found between the FMF cases and healthy volunteers aged 8-18 years for the physical health, social health, school functioning, psychosocial total and scale total score of PEDsQL4.0 child self-report. The parent proxy report PedsQL4.0 scores of all patients with FMF were lower than the healthy group for physical, emotional, social, psycho-social, school functioning scores and scale total score but the difference was not statistically significant. A significant correlation was present between PedsQL4.0 child self-report and parent PedsQL4.0 report in all subscales except for social health total score. There was not a significant correlation between the demographic variables and clinical features of the patients and the child subscales of PedsQL4.0.

Conclusion: The scores of FMF cases from the scales were lower than those of the healthy volunteers from the questionnaires but this difference was not statistically significant. Considering the relatively small sample size of the study it is obvious that more comprehensive prospective studies are required on this subject.

Key Words: Children, Familial Mediterranean fever, Quality of life, Parent

ÖZ

Amaç: Bu çalışmada ailevi Akdeniz ateşi (AAA) olan 2-18 yaş arası çocuk hastaların ve ebeveynlerinin sağlıkla ilgili yaşam kalitesini değerlendirmek ve aynı yaş grubundaki sağlıklı çocuklar ile karşılaştırmak amaçlandı.

Gereç ve Yöntemler: Ailevi Akdeniz ateşi olan 51 çocuk ile yaş ve cins olarak eşleştirilmiş 51 sağlıklı gönüllü çalışmaya dahil edildi. Sekiz-18 yaş arası çocuklar PedsQL4.0 çocuk formunu doldururken, tüm yaş gruplarında ebeveynler PedsQL4.0 ebeveyn formunu doldurdu.

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Ethics Committee Approval / *Etik Kurul Onayı*: This study is conducted in accordance with the principles of the Declaration of Helsinki. The study protocol was approved by the Ethics Committee of Keçiören Training and Research Hospital (No: 10.12.2014/703).

Contribution of the Authors / Yazarların katkısı*: *YASAR DURMUS S: Planning methodology to reach the Conclusions, Taking responsibility in patient follow-up, collection of relevant biological materials, data management and reporting, execution of the experiments, Taking responsibility in necessary literature review for the study, Taking responsibility in the writing of the whole or important parts of the study. ***OZLU SG:*** Constructing the hypothesis or idea of research and/or article, Planning methodology to reach the Conclusions, Taking responsibility in patient follow-up, collection of relevant biological materials, data management and reporting, execution of the experiments, Taking responsibility in logical interpretation and conclusion of the results, Taking responsibility in the writing of the whole or important parts of the study. ***COP E:*** Constructing the hypothesis or idea of research and/or article, Planning methodology to reach the Conclusions, Organizing, supervising the course of progress and taking the responsibility of the research/study, Taking responsibility in logical interpretation and conclusion of the results, Taking responsibility in necessary literature review for the study, Taking responsibility in the writing of the whole or important parts of the study, Reviewing the article before submission scientifically besides spelling and grammar. ***BULBUL M:*** Organizing, supervising the course of progress and taking the responsibility of the research/study, Reviewing the article before submission scientifically besides spelling and grammar.

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Bulgular: Sekiz-18 yaş grubu çocuk formlarının değerlendirilmesinde, istatistiksel anlamlı olmamakla birlikte, fiziksel sağlık, sosyal sağlık, okul işlevselliği, psikososyal total skor ve ölçek toplam skoru açısından AAA olan çocukların puanlarının daha düşük olduğu saptandı.

Ebeveyn PedsQL4.0 değerlendirildiğinde ise tüm yaş gruplarında AAA olan çocukların ebeveynlerinde fiziksel, duygusal, sosyal, psikososyal, okul işlevselliği skorları ve ölçek toplam skorlarının sağlıklı çocukların aileleri ile karşılaştırıldığında daha düşük olduğu ancak istatistiksel anlamlı düzeye ulaşmadığı saptandı. Sosyal sağlık toplam skoru hariç tüm ölçeklerde AAA olan çocuklar ve ebeveynlerinin PedsQL4.0 değerlendirmeleri arasında anlamlı korelasyon olduğu saptandı. Demografik değişkenler ve klinik bulgular ile çocuk PedsQL4.0 arasında anlamlı ilişki saptanmadı.

Sonuç: Ailevi Akdeniz ateşi olan olgularda ölçek skorları istatistiksel anlamlı olmasa da sağlıklı kontrollere göre daha düşüktür. Çalışmamızda göreceli olarak az sayıda hasta olduğu göz önünde bulundurulduğunda bu alanda daha kapsamlı ve fazla sayıda hasta içeren çalışmalara ihtiyaç olduğu sonucuna varılmıştır.

Anahtar Sözcükler: Çocuklar, Ailevi Akdeniz Ateşi, Yaşam kalitesi, Ebeveyn

INTRODUCTION

Familial Mediterranean Fever (FMF) is a self-limiting autosomal recessive auto-inflammatory disease characterized by recurrent fever and episodes of inflammation in the serous membranes such as the peritoneum, pleura, pericardium, and synovia (1). The disease is especially common in certain populations such as the Turks, Armenians, Arabs, and Jews living in the Mediterranean and the Middle East (2).

The Tel Hashomer criteria, mostly based on clinical evaluation, are used in the diagnosis (3). The gene MEFV causing the disease has been described in 1997 at the 16p13.3 localization and to date 389 mutations and polymorphisms have been reported so far (4-6).

Colchicine has been the first choice of treatment for FMF patients since it had been discovered in 1972 (7, 8). It is effective in preventing attacks of FMF and preventing amyloidosis which is the most devastating complication of FMF (9).

The concept of quality of life (QoL) is often used in everyday life and also in the practice of medicine in recent years. It can briefly be defined as the individual's evaluation of its current situation using his/her own system of values and culture. Health-related QoL (HRQoL) is the perception of the effects created by a disease or its treatment on the patient (10). Health-related QoL studies are useful to understand disease and their effects on the individual, and to develop treatment plans and health policies (11).

The evaluation of QoL in rheumatic disorders is becoming increasingly important in recent years. Quality of life has been shown to be decreased in systemic rheumatic diseases such as systemic lupus erythematosus, rheumatoid arthritis, fibromyalgia and FMF in adult studies (12,13). Studies on HRQoL in children were first conducted in the 1980s. Health-related QoL is an important factor in terms of evaluating a disease and planning its treatment in children with rheumatological disorders (10,11). In FMF, conditions such as; recurrent characteristic of disease, retention from school, regular medical visits, having to use medications for life, affect the QoL (11). Buskila et al. (14) were the first to conduct QoL studies in adult FMF patients and found it decreased compared to healthy controls. Press et al. (15) reported that the QoL was also decreased in the parents of children with FMF. The QoL of pediatric FMF patients and their parents was first investigated by Makay et al. (16). They evaluated the QoL of children and adolescents with FMF aged 8-18 years and their parents in their study and detected that QoL scores of children aged between 8- 12 years with FMF were significantly lower than healthy peers for physical and psychosocial functioning for both children and parents.

In this study, different from previous studies, in addition to the QoL of children aged 8-18 years we also evaluated the QoL of children between 2-7 years old together with their families and identified the influencing factors. The aim of this study to evaluate both child-self reported and parents proxy reported HRQoL of children with FMF aged between 2- 18 years old and to compare the scores with their healthy peers and to determine the factors which affect the QoL of children with FMF.

PATIENTS and METHODS

Fifty-one children with FMF aged 2-18 years, followed-up at least six months and age-sex matched 51 healthy volunteers were included in this study who had been followed at Department of Pediatric Nephrology and Rheumatology of our hospital; between January 2015 and April 2015. FMF diagnosis was made according to Tel Hashomer criteria. Patients were diagnosed by using Yalçınkaya-Özen criteria after the year 2009. The majority of patients who had diagnosed with FMF were screened for the most common 12 MEFV mutations based on reverse hybridization with the CE/IVD-labeled FMF Strip Assay (Vienna Lab Diagnostics, Vienna, Austria).

In pediatric nephrology and rheumatology department of our hospital, patients with FMF continue to follow up quarterly. Healthy volunteers were picked up from children who were admitted to out-patients clinics of department of general pediatrics. They did not have any history of acute or chronic diseases.

Inclusion criteria: Patients who had diagnosed with FMF due to Tel Hashomer criteria between 2 to 18 years old, who do not have comorbid diseases and accepted to answer all questions of questionnaire were included in the study

Exclusion criteria: Patients who had diagnosed FMF and older than 18 years old, who had comorbid diseases or FMF attack at the time of the evaluation, did not answer the all questions of questionnaire were excluded from study.

Sociodemographic form was developed and applied by the authors to record characteristics of participants such as age and gender. The diagnosis duration, annual mean number of attacks, annual mean number of hospitalizations due to an attack, the dose of colchicine used, disease severity scores of 51 FMF cases and MEFV gene mutation were also recorded.

The duration of FMF was classified as 0-5 years, 5-10 years, and 10 years and higher. The mean annual number of attacks and the mean annual number of hospitalizations due to an attack were grouped as 0-1 low, 2-4 moderate and 5 and above frequent. The disease severity scores were graded as mild, moderate and severe using the method identified by Pras (17). Children aged 8-18 years completed the Quality of Life Scale for Children (PEDsQL) Child Form questionnaires. Parents of all cases completed the PEDsQL parent form.

We used the PedsQL4.0 scales developed by Varni et al. (18,19). Turkish reliability and validity were done by Üneri (20) (for the age of 2-7 years) and Memik (21) (for the age of 8-18 years). Physical health, emotional and social functioning and school functioning, all characteristics of health as identified by the World Health Organization, are queried with PedsQL. The PedsQL4.0 scale includes 21 items for 2-4 years and 23 items for all other age groups. Each item is scored between 0 and 100. An answer of 'never' receives 100 points, 'rarely' 75 points, 'sometimes' 50 points, 'often' 25 points and 'almost always' 0 (17). The scale total score (STS), physical health total score (PHTS) and psycho-social health total score (PSHTS; this is the mean computed sum of the items divided by the number of items answered in the Emotional (EHTS), Social (SHTS) and School functioning (SFTS) scales) are then calculated. Higher PedsQL4.0 total scores indicate better HRQoL.

This study is conducted in accordance with the principles of the Declaration of Helsinki. The study protocol was approved by the Ethics Committee of Keçiören Training and Research Hospital (No: 10.12.2014/703).

Statistics

SPSS 15.0 program (Statistical Package for Social Sciences, Chicago, SPSS Inc.) was used for statistical analysis. The normality of the distribution of numerical variables was determined via the Kolmogorov-Smirnov test. Numerical variables with normal distribution are shown as mean ± SD and those not normally distributed are shown as median. Frequency and (%) are used for categorical variables. G*Power v.3.0.10 (G*Power Franz Faul, Universitat, Kiel, Germany) was used to determine the sample size required for the study (22). Taking a sample of ≥ 102 cases (51 cases per group) was calculated to achieve 80% power with d=0.50 effect width, α=0.05 type 1 error, β=0.20 type 2 error (23).

Student's t test or Mann-Whitney U test (which was appropriate according to distribution of variables) was used to compare variables like age, gender, STS, PHTS, EHTS, SHTS, SFTS, and PSHTS scores between two groups. To measure the difference between the means of STS, PHTS, EHTS, SHTS, SFTS, and PSHTS scores of PEDsQL child form and of PEDsQL parent form, dependent sample t-test was done. Correlation analysis was done to evaluate the relation between STS, PHTS, EHTS, SHTS, SFTS, PSHTS scores and age, gender, duration of disease, mean annual number of attacks, mean annual number of hospitalizations due to an attack, dose of colchicine used, MEFV gene mutation, disease severity score of the FMF group. A p value of <0.05 was accepted as significant.

RESULTS

Fifty-one children with FMF and 51 healthy children were included in our study. Socio-demographic characteristics of the FMF cases including age, gender, diagnosis duration, mean annual number of attacks, mean annual number of hospitalizations due to an attack, dose of colchicine used and disease severity scores are summarized in Table I.

A mathematical difference, with no statistical significance, was found between the FMF cases and healthy volunteers aged 8-18 years for the PHTS, SHTS, SFTS, PSHTS and STS of PEDsQL4.0 child self-report. All of the caregivers of FMF patients had lower PHTS, EHTS, SFTS, PSHTS, STS, SHTS

Table I: Socio-Demographic features of participants.

Socio-Demographic features	FMF patients	Healthy volunteers	Statistics
Girls n (%)	25 (49)	24 (47.1)	p=0.84 X^2=0.39
Age, mean±SD (years)	11.2±4.1	11.7±3.8	p=0.48
Age at diagnosis,mean±SD (years)	6.3±3.5	-	-
Duration of disease, mean±SD (years)	4.9±3.7	-	-
Number of attacks in a year*		-	-
Mild	34 (66.7)		
Moderate	11 (21.6)		
Severe	6 (11.8)		
Number of hospitalizations*		-	-
Rare	48 (94.1)		
Moderate	2 (3.9)		
Frequent	1 (2)		
Dose of colchicine*		-	-
0.5 mg	16 (31.4)		
1 mg	26 (51)		
1.5 mg	8 (15.7)		
2 mg	1 (2)		
Disease severity scores*		-	-
Mild	37 (72.5)		
Moderate	14 (27.5)		

***n(%), **FMF:** familial Mediterranean fever, **SD:** Standard Deviation*

Table II: Comparisons of Quality of Life Scale for Children scores of healthy volunteers and familial Mediterranean fever patients (ages between 8-18 years) reports and their parent reports.

Comparison	FMF Patients (n=51)			Healthy Volunteers (n=51)			p
	Mean±SD	Median	Min-Max	Mean±SD	Median	Min-Max	
PedsQL children report							
PHTS	596.7±154.2	637.5	(300-800)	618.9±123.5	625	(250-800)	0.493
EHTS	379.6±90.9	387.5	(100-500)	370.9±80.2	375	(225-500)	0.664
SHTS	453.2±80.9	500	(200-500)	455.4±64.8	475	(200-500)	0.505
SFTS	333.5±89.7	325	(150-500)	366.8±72.4	375	(225-500)	0.081
PSHTS	333.5±89.7	1175	(105-1425)	1187.1±186.5	1225	(700-1450)	0.384
STS	1759.2±290.7	1850	(1050-2225)	1812.1±278	1850	(1200-2250)	0.426
PedsQL parent report							
PHTS	582.8±170.6	600	(250-800)	597.0±130.4	625	(275-775)	0.638
EHTS	341.1±87.4	350	(150-500)	353.9±94.6	375	(175-500)	0.482
SHTS	485.7±476.9	450	(200-500)	426.9±77.9	450	(175-500)	0.387
SFTS	339.2±102.2	350	(100-500)	458.2±586.1	375	(225-475)	0.156
PSHTS	1075.0±245.9	1125	(100-1475)	1139.2±246.3	1200	(100-1475)	0.190
STS	1675±338.3	1675	(925-2250)	1753.9±293.5	1750	(1150-2250)	0.217

PedsQL: *Quality of Life Scale for Children,* ***PHTS:*** *Physical health total score,* ***EHTS:*** *Emotional health total score,* ***SHTS:*** *social health total score,* ***SFTS:*** *School functioning total score,* ***PSHTS:*** *psycho-social health total score,* ***STS:*** *scale total score,* ***FMF:*** *familial Mediterranean fever,* ***SD:*** *Standard Deviation*

Table III: Intercorrelations of Quality of Life Scale for Children scale scores between familial Mediterranean fever patients and their caregivers; healthy volunteers and their caregivers.

PedsQL	FMF patients and their caregivers		Healthy volunteers and their caregivers	
	r	p	r	p
PHTS	0.631	<0.001*	0.606	<0.001*
EHTS	0.635	<0.001*	0.555	<0.001*
SHTS	0.278	<0.091	0.553	<0.001*
SFTS	0.696	<0.001*	0.694	<0.001*
PSHTS	0.457	<0.001*	0.644	<0.001*
STS	0.722	<0.001*	0.638	<0.001*

PedsQL: *Quality of Life Scale for Children,* ***PHTS:*** *Physical health total score,* ***EHTS:*** *Emotional health total score,* ***SHTS:*** *Social health total score,* ***SFTS:*** *School functioning total score,* ***PSHTS:*** *Psycho-social health total score,* ***STS:*** *Scale total score,* ***FMF:*** *familial Mediterranean fever Bold values are significant, *p<0.05*

Table IV: Correlation of the Quality of Life Scale for Children scores according to demographic features.

	PHTS	EHTS	SHTS	SFTS	PSHTS	STS
	p	p	p	p	p	p
Age	0.206	0.744	0.887	0.608	0.711	0.360
Gender	0.075	0.710	0.552	0.057	0.836	0.477
Age at diagnosis	0.899	0.077	0.913	0.415	0.182	0.565
Duration of disease	0.135	0.262	0.986	0.196	0.140	0.100
Number of attacks per year	0.111	0.069	0.973	0.432	0.413	0.470
Number of hospitalization per year	0.647	1.0	0.201	0.264	0.190	0.491
Dose of colchicine	0.075	0.955	0.800	0.130	0.605	0.397
MEFV mutation	0.679	0.150	0.476	0.978	0.496	0.639
Disease severity score	0.747	0.056	0.202	0.374	0.091	0.245

PHTS: *Physical health total score,* ***EHTS:*** *Emotional health total score,* ***SHTS:*** *Social health total score,* ***SFTS:*** *School functioning total score,****PSHTS:*** *Psychosocial health total score,* ***STS:*** *Scale total score,* ***MEFV:*** *Mediterranean Fever*

Table V: Summary of the studies about health related quality of life for patients with familial Mediterrenean fever in the literature.

Authors[ref], year	Study Group	Number of Cases	Assessment Methods	Results
Buskila et al.[13]	Adult FMF patients	102 FMF patients vs. 124 healthy controls	QOL Scale	The QOL of FMF patients were impaired
Press et al.[14]	Mothers of children with FMF	Mothers of 35 FMF children vs. 23 healthy controls	QOL Scale	The QOL of FMF mothers were impaired
Makay et al.[15]	Children with FMF	51 children with FMF vs. 81 healthy controls	PedsQL™ 4.0	The PedsQL children with FMF were impaired
Deger SM et al.[23]	Adult FMF patients	90 FMF patients vs. 67 healthy controls	SF-36	The SF-36 physical components scores of FMF patients SF-36 were impaired
Sahin S. et al[12]	Adult FMF patients	100 FMF patients vs. 100 healthy controls	SF-36	The all SF-36 scores of FMF patients were impaired
Giese A et al.[24]	Adult FMF patients	40 FMF patients from Turkey vs. 40 FMF patients from Germany vs.40 healthy controls from Germany	WHOQOL-BREF	The physical health QOL scores of all FMF patients were impaired
Düzçeker Y et al.[25]	Children with FMF, SLE	26 children with FMF, 25 children with SLE	HRQL	FMF patients global QL were the best

QOL: *Quality of Life,* ***SF-36:*** *Short Form-36,* ***WHOQOL-BREF:*** *World Health Organization Quality of Life Questionnaire-Short Form,* ***HRQL:*** *Health related quality of Life.*

of PEDsQL4.0 parent report compared to all healthy children's care givers, but the difference was not statistically significant Table II.

A statistically significant correlation was present between PEDsQL4.0 child self-report PHTS, EHTS, SFTS, PSHTS, STS of FMF patients in the 8-18 years age group and those of PEDsQL4.0 parent report of their caregivers, no such correlation was found for SHTS. There was a statistically significant positive correlation for between PEDsQL4.0 child self-report subscores of healthy controls in the 8-18 years age group and subscores of PEDsQL parent report of their caregivers Table III.

The PHTS, EHTS, SHTS, SFTS, PSHTS, STS of PEDsQL4.0 child self-report of FMF patients did not have a statistically significant correlation with age, gender, duration of FMF, mean number of attacks per year mean annual number of hospitalizations due to an attack, dose of colchicine used, MEFV gene mutation, and disease severity score Table IV.

DISCUSSION

The most important result of this study is that HRQoL of FMF patients with mild and modarate disease severity scores are not affected in comparison to healthy volunteer peers between 2-18 years of age. Our study is the first, to the best of our knowledge, evaluating the QoL in FMF cases between 2-8 years of age with questionnaires filled by the parents.

Studies evaluating the QoL in the pediatric cases are quite limited in the literature. Quality of life of FMF patients was first investigated in adults by Buskila et al. (14). in 1997 and shown to be lower than the healthy control group. Following studies on the QoL of adult FMF patients, supported the findings of study of Buskila et al. (12-14). The first relevant study about children with FMF was conducted by Press et al. and the QoL was found to be lower in mothers of children with FMF than in that of healthy children (15). Makay et al. (16) first evaluated the HRQoL of children 8-18 years of age. Summary of the studies about health related QoL for patients with familial Mediterranean fever in the literature are shown in Table V.

According to study of Makay et al. (16) the physical and psycho-social health scores for the 8-12 years age group and the physical health, emotional health and school functioning scores for the 13-18 years age group were significantly lower in the FMF patients than the healthy control group while the social health scores of the FMF cases in the 13-18 years age group were the same as the control group. In our study, physical health, psycho-social health, and school functioning scores of the 8-18 years age group were lower than in the control group but the difference was not statistically significant. Although the emotional health total score was interestingly found to be higher in the same age group, this difference also was not statistically significant. Additionally, no difference was found between all score types calculated from the questionnaires completed by the caregivers of the cases diagnosed with FMF from 2-18 years age groups. In our study the reason of no significance between HRQoL scores of FMF patients and controls is due to the fact that 66% of our patients annual attack rates were as low as 0-1. As seen in the study of Makay et al. (16) there were a negative correlation between number of attacks and HRQoL scores of patients (r=-0.571, p=0.000). This result as well supports our point of view.

While a statistically significant correlation was present for the physical health, emotional health, school functioning and psychological health scores obtained from the questionnaires completed by the group of FMF patients aged 8-18 years and from the person providing care to the same age group, no such relationship was found between the social health scores; this may reflect the difference between the family's perception of and adaptation to the chronic disease of their child and the child's perception of and adaptation to his/her own disease. Additionally families of the children aged between 8-18 years may not observe social relationship of their children at school closely so it was thought that they might have concerns about this issue.

Makay et al. (16) reported an inverse relationship between the scores of their patients and the disease severity score, annual number of attacks, and hospitalization frequency with the scale total scores decreasing as the disease severity score, annual number of attacks, and hospitalization frequency increased. In contrast to study of Makay et al. (16) we found no statistically significant relationship between the scores obtained from the questionnaires of the FMF cases in the 8-18 years age group and their age, gender, diagnosis duration, annual mean number of attacks, annual mean number of hospitalizations due to an attack, the dose of colchicine used, MEFV gene mutation, and disease severity score. The reason may be the higher annual number of attacks and hospitalization frequency and higher disease severity scores of the cases in the Makay et al. (16) study than our study.

Giese et al. (24) has reported that no statistically significant relationship between the disease severity score and the QoL of the adult patients. They have been stated that the possible reasons were the used QoL scale and the inadequacy of disease severity scoring (24). Similarly, in our study, the lack of a statistically significant relationship between disease severity scores and the QoL could be associated with the fact that a modified form of the Pras severity scoring has been used for study patients however Pras severity scoring has mainly been developed for adult patients (17).

In this study, there were no relationship between disease severity scores and PedsQoL. Disease severity scores were mild in 72.5% of our patients and moderate in 27.5% of them. None of the patients had severe disease severity score, therefore, this study has revealed that in the absence of relationship may be due to the mild and moderate disease severity scores in all of our patients.

Makay et al. (16) evaluated relationship between the PedsQoL scores of FMF patients and MEFV mutaions. They found no statistically significant difference (16). Similarly, no relationship was found between the mutation analysis results and PedsQoL in our study. In the study of Yalçınkaya et al. (25) about genotype-phenotype correlation in Turkish patients with FMF, they reported that the genotype can't always predict disease severity. They emphasized that environmental factors and genetic changes that are yet unknown can also affect the phenotype of the disease (25). This findings could explain the lack of the relationship between the genetic mutation and QoL in our study.

Limitations of this study are; the small sample size of patients and control group, patients without severe disease severity scores, patients attended from a specific geographic region and same rheumatology clinic. More comprehensive, multicentral studies which have large sample size of patients and evaluate HQoL for each disease severity score levels, in different geographic and ethnical regions are required on this subject.

In conclusion, the QoL in cases diagnosed with FMF was not found to be affected by socio-demographic factors in this study. Children with FMF agree with their parents about HRQoL apart from social health. Clinicians who follow children with FMF, may consider that there are limited studies about HRQoL in this field. It should be kept in mind when the disease severity score increases, the possibility of deterioration of HRQoL may arise and clinicians should be careful in the clinical evaluation process. Importance should be given to patient education to improve the quality of life in children with FMF and their parents.

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