European Journal of Pediatrics (2023) 182:3063–3073
https://doi.org/10.1007/s00431-023-04975-5

RESEARCH

The effect of education given to Turkish adolescents with Familial Mediterranean fever on anxiety, depression, and quality of life

Havva Kaçan[1] · **Gamze Kaş Alay**[2] · **Eren Yildiz**[3,4]

Received: 19 February 2023 / Revised: 4 April 2023 / Accepted: 6 April 2023 / Published online: 18 April 2023

Abstract

To examine the effects of individual education given to Turkish adolescents with Familial Mediterranean fever (FMF) on anxiety, depression, and quality of life. The randomized controlled experimental study was performed on 70 adolescents aged 12–18 years who were diagnosed as having FMF between October 2021 and April 2022 in Turkey. The disease management education was offered individually to adolescents in the intervention group with a booklet entitled "FMF is under my control;" no intervention was applied to adolescents in the control group. The training booklet was prepared by the researchers in a multidisciplinary team and was finalized by taking expert opinions. In the data collection process, a personal information form, the State-Trait Anxiety Inventory for Children (STAI-CH), the children's depression inventory (CDI), and the Pediatric Quality of Life Inventory (PedsQL) were used. After applying the scales specified in the pretest, individual training was given and the posttest was performed 2 months later using the same scales. After the education, there was a statistically significant decrease in the mean CDI score of the intervention group ($p < 0.05$), whereas there were statistically significant increases in mean scores obtained on PedsQL and its sub-scales ($p < 0.05$). However, the decrease in the mean STAI-CH score of the intervention group was not statistically significant ($p > 0.05$). There was no statistically significant difference in the mean STAI-CH, CDI, and PedsQL scores of the control group after the education ($p > 0.05$).

Conclusion: The effectiveness of the individual education program for adolescents with FMF in improving quality of life and reducing levels of depression within the scope of disease management has been confirmed. It is recommended that all health professionals working with children with FMF regularly provide individual or group-planned education programs.

What is Known:

- *The unpredictability of FMF attacks has a very negative effect on adolescents.*
- *Individual education programs on FMF focus on children with a holistic approach.*

What is New:

- *To the best of our knowledge, this study is the first study to evaluate disease management education given to adolescents with FMF.*
- *This is a pioneering study of the use of nurses in the education of adolescents with FMF and in fulfilling their educational roles.*

Keywords Familial Mediterranean fever · Anxiety · Depression · Quality of life · Education

---

Communicated by Peter de Winter.

✉ Eren Yildiz
eren70@gmail.com

Havva Kaçan
hkacan@kastamonu.edu.tr

Gamze Kaş Alay
gkas@kastamonu.edu.tr

[1] Department of Mental Health and Psychiatric Nursing, Department of Nursing, Faculty of Health Sciences, Kastamonu University, Kastamonu, Turkey

[2] Department of Child Health and Diseases Nursing, Department of Nursing, Faculty of Health Sciences, Kastamonu University, Kastamonu, Turkey

[3] Department of Pediatrics, Faculty of Medicine, Kastamonu University, Kastamonu, Turkey

[4] Kuzeykent Merkez Kampüsü, Kuzeykent Mah. Org. Atilla Ateş Paşa Cad, 37150 Kastamonu, Turkey

Introduction

Familial Mediterranean fever (FMF) is a chronic inflammatory disease characterized by acute attacks accompanied by recurrent fever, peritonitis, pleurisy, arthritis, or erysipelas-like skin findings [1]. FMF is the most common monogenic autoinflammatory disease and shows ethnic distribution. Its prevalence varies between 1/200–1/1000 among Turks, Arabs, Jews, and Armenians around the world [2, 3], with a ratio of 1/400–1/1000 in Turkish society, according to ethnicity. The incidence of the disease in the pediatric population in Turkey has been reported as 9.3/10,000 [4]. Due to migration among European countries in the twentieth century, it is seen in other populations in the world, albeit with a low prevalence [5].

The presence of chronic disease in childhood and adolescence is seen as an important risk factor for the emergence of mental disorders in the future [6]. Having a chronic disease affects the child's quality of life, academic success, and physical-mental development and brings anxiety and psychological problems. In addition to physical health, chronic diseases also have important effects on a child's emotions, nutrition, sleep patterns, behaviors, education, self-confidence, and social relationships [7]. Emotional and behavioral problems are defined in 10% of all children, and it is reported that this rate increases to 20% in children with chronic diseases [8]. Bakare et al. [9] reported that children with chronic diseases had difficulty controlling their emotions compared with their healthy peers.

Most patients experience their first attacks in early childhood [2–4, 10]. Adolescence is the period between childhood and adulthood and has its own particular features and problems. Nevertheless, adolescents, who are in a unique transformation and development process, have to struggle with chronic diseases such as FMF throughout their lives. Thus, together with FMF, adolescents experience social, psychological, developmental, and environmental problems in association with a variety of factors [11–13].

Mood changes [14], chronic pain, depression, and anxiety are frequently encountered psychiatric problems in children and adolescents with FMF [15, 16]. Makay et al. [17] found that children with FMF were more depressed than their healthy peers. Similarly, Durcan et al. [16] also determined that children with FMF had high levels of anxiety and depression. The lack of information about the disease process, uncertainty about when attacks will occur, and anxiety about the disease are sources of stress for the individual [2]. In addition, failing to attend school as a consequence of hospital follow-ups and attack symptoms, being obliged to use drugs regularly life-long, and experiencing emotional stress as a characteristic feature of adolescence can also affect the quality of life negatively. In the relevant literature, it is proposed that FMF, a restraining and chronic disease, increases susceptibility to depression and anxiety, and as a consequence of psychiatric problems associated with the decreased sleep quality, it negatively affects the quality of life in children and adolescents [13, 14, 18]. The reason for this situation is that, just as adolescents can perceive the chronic disease as the loss of autonomy and independence, they can experience mental effects because FMF causes recurrent attacks, which affect their daily lives negatively [13, 19].

However, healthcare professionals often focus on the medical dimension of the disease and show less interest in the patient's self-perception, adherence to treatment, and managing the effects of the disease. It is necessary to organize training programs that aim to meet the information needs of individuals with chronic diseases, to cope with their diseases effectively, and to monitor their lives and diseases [20]. The adolescent's education is important for effective FMF treatment and care.

Upon a review of studies performed on the topic, it was observed that giving individualized education on FMF to children/adolescents diagnosed with this disease was recommended [12–14]. Encouraging adolescents to adopt the proper self-care behaviors is a significant factor likely to contribute to the successful management of the disease, reducing the frequency and severity of attacks in adolescents with FMF, promoting supportive practices performed during attacks, increasing adherence to medical treatment and regular drug use, boosting the strength to cope with emotional change and stress, and ensuring the management of the disease can be achieved only with a planned educational program. In this context, promoting the success of the administered treatment and enhancing functional efficacy through education will assure that other health problems are avoided and the quality of life is improved [13]. As well as providing adolescents with a healthy lifestyle, an education program to be offered to adolescents about the management of FMF would help to control emotional stress and reduce mental problems experienced frequently in adolescence, such as depression and anxiety [14, 21–23].

For children with FMF to have a quality life by managing the disease appropriately, regular educational support should be provided. In studies where the educational role of health professionals is emphasized and their outputs are observed, the importance of providing holistic pediatric care within the framework of child and/or family-centered practices emerges. After a relevant literature review, it was found that there were descriptive studies about depression, anxiety, and quality of life in adolescents with FMF [14, 21–23]; however, there was no study performed aiming to ensure the physiologic and psychosocial control of FMF. This research was conducted to identify whether disease management education given individually to adolescents diagnosed with FMF had any effect on anxiety, depression, and quality of life.

Material and methods

Sample

This study was performed with the participation of adolescents with FMF and was designed as a pretest–posttest control group experimental research. The research population comprised adolescents aged 12–18 years who were diagnosed as having FMF and visited the pediatric outpatient clinics of Kastamonu Training and Research Hospital for follow-ups between October 2021 and April 2022 in Turkey. The research sample constituted 70 adolescents who satisfied the inclusion criteria designated for the research. The inclusion criteria were as follows: Being diagnosed as having FMF for a minimum of 1 year, being aged 12–18 years, being literate, having no communication problems, volunteering to participate in the research, and being a patient followed up in the above pediatrics outpatient clinic. Patients with any mental disorder or psychiatric diagnoses were excluded from the study because it would affect the data.

A power analysis was conducted to calculate the sample size for this research by using the G*Power 3.1 software package. The effect size was set as 0.80, at a high level relative to the one specified by Cohen [24] for the comparison of the means of independent samples. To ensure that the study would have a statistical power above 95%, the sample size was calculated as 70 patients (35 patients for each of the two study groups) at a significance level of 5% and with an effect size of 0.5 (df = 68; t = 1.668).

Randomization

Patients were randomly assigned to the two study groups using an online program [25]. Before entering the sample size into the above online randomization program, the first set was appointed to the intervention group and the second set was appointed to the control group after casting lots. To identify which of the two groups the participant adolescents would be in, numbers from 1 to 70 were non-repetitively entered into the online program. Next, using the online program, adolescents included in the sample were randomly assigned to the two groups.

Data collection ınstruments

A personal ınformation form, the State-Trait Anxiety Inventory for Children (STAI-CH), the children's depression ınventory (CDI), and the Pediatric Quality of Life Inventory (PedsQL) were used to collect research data.

Personal information form

This form was prepared by researchers in line with the relevant literature. The form comprised 27 questions designed to determine the adolescents' descriptive and disease and treatment-related characteristics [14, 21–23].

State-trait anxiety inventory for children

STAI-CH was developed by Spielberger [26]. It has two multiple-choice sub-scales for state anxiety (STAI-CH-S) and trait anxiety (STAI-CH-T), each of which has 20 items. Each item is rated as 1, 2, or 3, depending on the severity of the anxiety symptom. The minimum and maximum scores that can be obtained by a respondent from each sub-scale are 20 and 60 points, respectively. In the current research, relatively high Cronbach alpha coefficients were obtained for the state anxiety and trait anxiety sub-scales (0.765 and 0.803, respectively).

Children's depression inventory

The scale was developed by Kovacks et al. [27]. It has 27 items evaluating the child's last 2 weeks. Each item has three situational sentences rated as 0, 1, and 2, and a high score obtained from the scale shows that the respondent has high-level depression. The maximum score that can be obtained by a respondent from the scale is 54 points, and the cut-off point is set as 19 points. In the current research, a relatively high Cronbach alpha coefficient was obtained for the scale (0.823).

Pediatric quality of life inventory

This scale was developed by Varni et al. [28] for children aged 2–18 years to evaluate health-related quality of life. The scale is scored as per a 5-point Likert scale, and the total scale score, the physical health summary score, and the psychosocial health summary score are computed in the scoring process. The scale has a total of 23 items and four sub-scales, physical, emotional, social, and school functioning. The items are scored from 0 to 100 points, and a high score obtained by a respondent indicates that they have a high quality of life [28]. In the current research, a relatively high Cronbach alpha coefficient was obtained for the scale (0.817).

Procedure

Researchers prepared a booklet entitled "FMF is under my control" in light of relevant literature [2, 4, 29–36]. Expert opinions on this booklet were received in areas of pediatric nursing, psychiatric nursing, pediatric rheumatology, pediatrics, nutrition and dietetics, and physical education/sports, and a final version of the booklet was created. The final version of the booklet had the following four parts: (i) information about FMF, (ii) information about FMF treatment, (iii) information about the FMF attack periods (fever, pain, nutrition and constipation, and movement and exercise), and (iv) coping with anxiety and stress in the context of the FMF. Slide presentations were prepared in line with the developed training booklet. Individual training sessions lasted 30–45 min for each adolescent and their questions were answered after the training. A training booklet was given to those who completed the training.

The application stage of this study had two phases, pretest and post-test. In the pretest phase, a survey including the personal information form and scales was administered to both study groups. Face-to-face meetings with adolescents who volunteered to participate in the research were held during work hours of the outpatient clinic on weekdays. Participant adolescents could adhere to the education timetable because the education was given at times coinciding with adolescents' hospital follow-ups. In this regard, in the pretest phase, adolescents who were waiting for medical examinations at the outpatient clinic and who agreed to participate in the research were informed about the above survey and asked to complete it. While waiting for medical examinations or after the examination, adolescents in the intervention group received the disease management education offered individually in a vacant outpatient clinic room. No such intervention was given to adolescents in the control group. Next, appointments were made to meet with all participant adolescents once again after 2 months.

All adolescents who had appointments scheduled for 2 months after the first meeting the post-test phase were called by phone and invited to the outpatient clinic. All adolescents were asked to complete the survey again, including the personal information form, and the scales that were introduced in the first meeting. Upon the collection of the post-test research data, education was also given to adolescents in the control group using the study booklet so that they could also benefit from this education (Fig. 1).

Data analysis

The research data were analyzed using the Statistical Package for the Social Sciences (SPSS) for Windows 22.0 software package. In the data evaluation, descriptive statistics (number, percentage, mean, standard deviation) were used. Whether adolescents were homogeneously distributed across the two study groups was analyzed using the Chi-square test and *t*-test. Kurtosis and skewness statistics were examined to identify whether scores obtained by adolescents from the scales were normally distributed, and it was found that both statistics were within the acceptable range from $+1.5$ to -1.5 [37]. The independent samples *t*-test was used in the inter-group comparison of adolescents' mean scale scores, and the dependent samples *t*-test was used in the intra-group comparison of adolescents' mean scale scores. Cronbach's alpha coefficient was calculated to evaluate the reliability of scales used in the research. Significance was evaluated at a level of $p < 0.05$.

Ethics

Ethical endorsement for the research was obtained from the Clinical Trials Ethics Committee of the Faculty of Medicine of Kastamonu University of Turkey (Date: 25 August 2021, no: 2020-KAEK-143–106). Written permission was received for the study from the units where the research would be conducted. All adolescents included in the sample and their parents expressed their consent for participation in the research both verbally and in written format via an "informed consent form" before the research began.

Results

Socio-demographic characteristics of adolescents

The mean age of the patients was similar for both groups ($p > 0.05$). Of all adolescents in the intervention group, 57.1% were female, 54.3% were aged 15–18 years, 45.7% had two siblings, 34.3% had elementary school graduate mothers, 34.3% had high school graduate fathers, 40.0% had working mothers, 100% had working fathers, and 11.4% had parents who had a consanguineous marriage. Of all adolescents in the control group, 51.4% were female, 40.0% were aged 15–18 years, 34.3% had one sibling, 34.3% had elementary school graduate mothers, 31.4% had high school graduate fathers, 42.9% had working mothers, 94.3% had working fathers, and 20% had parents who had a consanguineous marriage. A comparison of the study and control groups in terms of their descriptive and socio-demographic characteristics (age, sex, number of siblings, mother's education level, father's education level, mother's employment status, father's employment status, having parents who had a consanguineous marriage) revealed no statistically significant difference between the groups ($p > 0.05$; Table 1).

FMF-related characteristics of adolescents

The mean age at which adolescents were diagnosed was similar for both groups ($p > 0.05$). Of all adolescents in the intervention group, 40.0% were diagnosed as having FMF after the age of 8 years, 25.7% had other diagnosed chronic diseases besides FMF, 31.4% regularly had FMF attacks, 54.3% said that they had FMF attacks in association with stress and fatigue, 31.4% performed certain attack mitigation practices in the FMF attack period, 5.7% had another disease associated with FMF, 68.6% regularly used FMF drugs, and 60.0% did not have adequate information about the consequences of not using their drugs regularly. Regarding the adolescents in the control group, 37.1% were diagnosed as having FMF after they were aged 8 years, 28.6% had other diagnosed chronic diseases besides FMF, 31.4% regularly had FMF attacks, 54.3% said that they had FMF attacks in association with stress and fatigue, 48.6% performed certain attack mitigation practices in the FMF attack period, 14.3% had another disease associated with FMF, 71.4% regularly used FMF drugs, and 57.1% did not have adequate information about

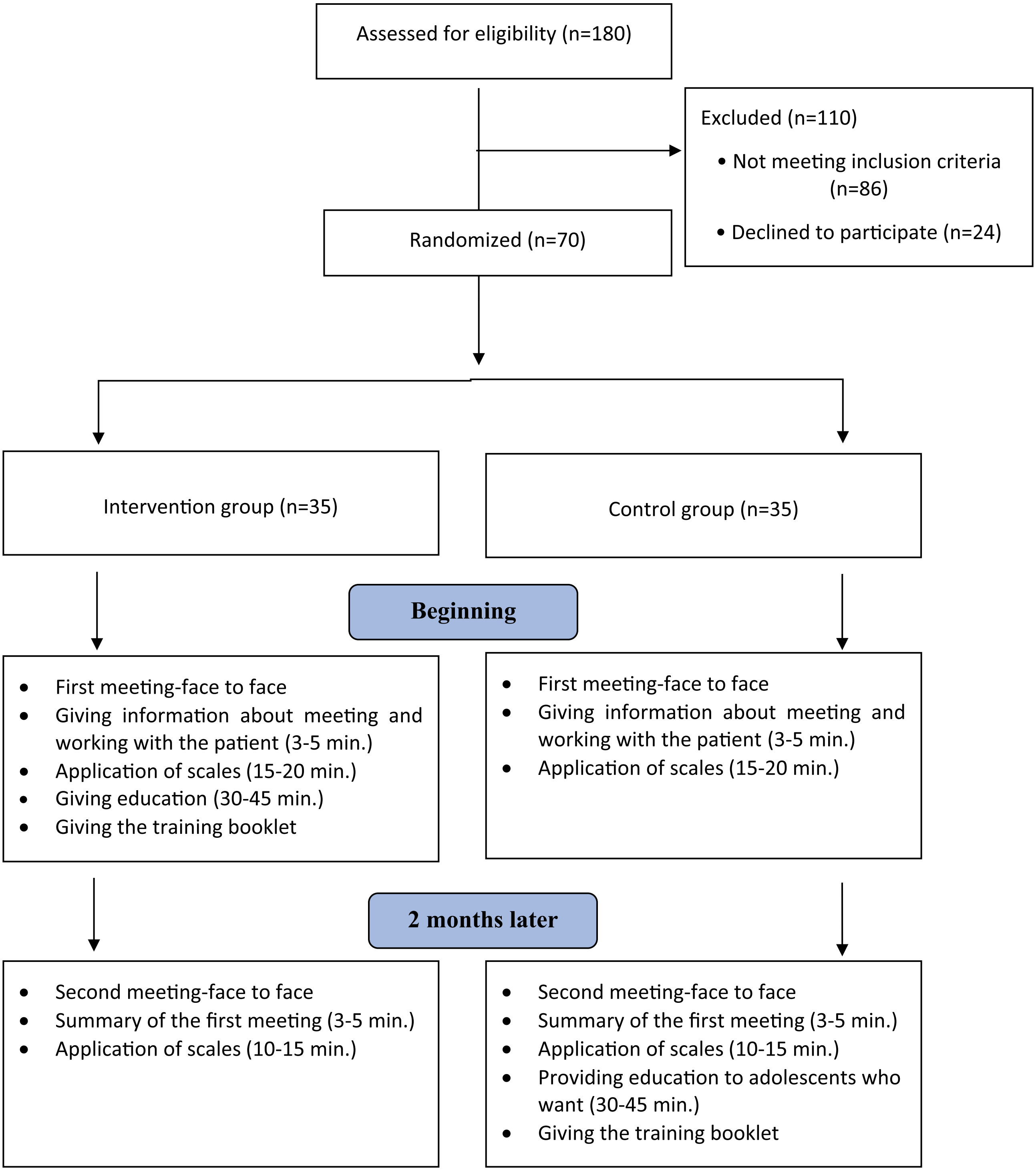

Fig. 1 Study flow chart

the consequences of not using their drugs regularly. Moreover, 65.7% of all adolescents in both intervention and control groups reported that they were worried about having FMF attacks and 54.3% stated that they had certain life activities inhibited by FMF (Table 2). It was determined that 25.7% of the intervention group and 28.6% of the control group had chronic diseases such as diabetes, asthma, and heart disease.

Table 1 Socio-demographic characteristics of adolescents in intervention and control groups ($n = 70$)

Characteristics		Intervention group ($n = 35$)	Control group ($n = 35$)	Total	Statistical analysis
Age (years)		**$\overline{X}$ ± SD** 14.46 ± 1.82	**$\overline{X}$ ± SD** 14.26 ± 1.66	**$\overline{X}$ ± SD** 14.36 ± 1.73	$t = 0.479$ $p = 0.633$
		***n* (%)**	***n* (%)**	***n* (%)**	**test/*p***
Age	12–14 years	16 (45.7)	21 (60.0)	37 (52.9)	$X^2 = 1.433$ $p = 0.169$
	15–18 years	19 (54.3)	14 (40.0)	33 (47.1)	
Gender	Male	15 (42.9)	17 (48.6)	32 (45.7)	$X^2 = 0.230$ $p = 0.405$
	Female	20 (57.1)	18 (51.4)	38 (54.3)	
Number of siblings	No sibling	3 (8.6)	1 (2.9)	4 (5.7)	$X^2 = 4.808$ $p = 0.186$
	1	5 (14.3)	12 (34.3)	17 (24.3)	
	2	16 (45.7)	11 (31.4)	27 (38.6)	
	> 3	11 (31.4)	11 (31.4)	22 (31.4)	
Mother's education level	Elementary school	12 (34.3)	12 (34.3)	24 (34.3)	$X^2 = 3.589$ $p = 0.309$
	Middle school	8 (22.9)	3 (8.6)	11 (15.7)	
	High school	7 (20.0)	12 (34.3)	19 (27.1)	
	University	8 (22.9)	8 (22.9)	16 (22.9)	
Father's education level	Elementary school	9 (25.7)	10 (28.6)	19 (27.1)	$X^2 = 0.240$ $p = 0.971$
	Middle school	7 (20.0)	6 (17.1)	13 (18.6)	
	High school	12 (34.3)	11 (31.4)	23 (32.9)	
	University	7 (20.0)	8 (22.9)	15 (21.4)	
Mother's employment status	Working	14 (40.0)	15 (42.9)	29 (41.4)	$X^2 = 0.059$ $p = 0.500$
	Not working	21 (60.0)	20 (57.1)	41 (58.6)	
Father's employment status	Working	35 (100.0)	33 (94.3)	68 (97.1)	$X^2 = 2.059$ $p = 0.246$
	Not working	0 (0.0)	2 (5.7)	2 (2.9)	
Having parents who had a consanguineous marriage	Yes	4 (11.4)	7 (20.0)	11 (15.7)	$X^2 = 0.971$ $p = 0.256$
	No	31 (88.6)	28 (80.0)	59 (84.3)	

X^2: Chi-square statistic $t = t$-test in independent groups

Breakdown of mean STAI-CH, CDI, and PedsQL phases of adolescent patients with FMF

The difference between the mean pretest and post-test STAI-CH-S and STAI-CH-T scores of the intervention group was not statistically significant ($t = 1.374$; $t = 1.982$; $p > 0.05$). The difference between the mean pretest and post-test STAI-CH-S and STAI-CH-T scores of the control group was not statistically significant ($t = 1.876$; $t = 1.541$; $p > 0.05$; Table 3).

Although the difference between the mean pretest and post-test CDI scores of the control group was not statistically significant ($t = 1.663$; $p > 0.05$), the difference between the mean pretest and post-test CDI scores of the intervention group was statistically significant ($t = 5.420$; $p < 0.001$; Table 3). The difference between the mean pretest and post-test PedsQL scores of the intervention group was statistically significant ($t = -7.900$; $p < 0.001$). However, the difference between the mean pretest and post-test PedsQL scores of the control group was not statistically significant ($t = -0.333$; $p > 0.05$; Table 3).

In intra-group and inter-group comparisons, there was no statistically significant difference in mean pretest and post-test STAI-CH-S and STAI-CH-T scores of both the intervention and control groups ($p > 0.05$), but there were statistically significant differences in mean pretest and post-test scores obtained by the intervention group from the CDI, the PedsQL, and PedsQL sub-scales ($p < 0.05$; Table 3).

Discussion

To the best of our knowledge, this study is the first study to evaluate disease management education given to adolescents with FMF. For this reason, our study results are discussed concerning the effectiveness of the training given in itself and other chronic diseases.

FMF can lead to emotional problems such as depression and anxiety in children/adolescents because it is a chronic disease starting at an early age and continues with attacks [13–15, 17, 18]. In our study, it was discerned that both the intervention and control groups had medium-level state

Table 2 FMF-related characteristics of adolescents in intervention and control groups ($n = 70$)

FMF-related characteristics		Intervention group ($n = 35$)	Control group ($n = 35$)	Total	Statistical analysis
Age of diagnosis (years)		$\overline{X} \pm SD$ 6.06 ± 3.38	$\overline{X} \pm SD$ 5.86 ± 3.06	$\overline{X} \pm SD$ 5.96 ± 3.21	$t = 0.259$ $p = 0.796$
		n (%)	n (%)	n (%)	test/p
Age when diagnosed with FMF	1–3 years	11 (31.4)	11 (31.4)	22 (31.4)	$X^2 = 0.085$ $p = 0.959$
	4–7 years	10 (28.6)	11 (31.4)	21 (30.0)	
	8 years or above	14 (40.0)	13 (37.1)	27 (38.6)	
Being diagnosed with other chronic diseases besides FMF	Yes[a]	9 (25.7)	10 (28.6)	19 (27.1)	$X^2 = 0.072$ $p = 0.500$
	No	26 (74.3)	25 (71.4)	51 (72.9)	
Playing sports regularly	Yes	12 (34.3)	8 (22.9)	20 (28.6)	$X^2 = 1.120$ $p = 0.214$
	No	23 (65.7)	27 (77.1)	50 (71.4)	
Receiving information about FMF	Yes	15 (42.9)	10 (28.6)	25 (35.7)	$X^2 = 1.556$ $p = 0.159$
	No	20 (57.1)	25 (71.4)	45 (64.3)	
Having information about FMF disease	Yes	19 (54.3)	25 (71.4)	44 (62.9)	$X^2 = 2.203$ $p = 0.108$
	No	16 (45.7)	10 (28.6)	26 (37.1)	
Taking measures regularly against FMF	Yes[b]	3 (8.6)	1 (2.9)	4 (5.7)	$X^2 = 1.061$ $p = 0.307$
	No	32 (91.4)	34 (97.1)	66 (94.3)	
Frequency of FMF attacks	Twice a month	4 (11.4)	3 (8.6)	7 (10.0)	$X^2 = 0.993$ $p = 0.911$
	Once a month	6 (17.1)	9 (25.7)	15 (21.4)	
	Once every 3 months	11 (31.4)	11 (31.4)	22 (31.4)	
	Once every 6 months	5 (14.3)	5 (14.3)	10 (14.3)	
	Once a year or less frequently	9 (25.7)	7 (20.0)	16 (22.9)	
Factors affecting the frequency of FMF attacks	Stress and fatigue	19 (54.3)	19 (54.3)	38 (54.3)	$X^2 = 4.043$ $p = 0.400$
	Menstruation and stress	4 (11.4)	2 (5.7)	6 (8.6)	
	Physical exercise and the long walk	5 (14.3)	6 (17.1)	11 (15.7)	
	Common cold	2 (5.7)	6 (17.1)	8 (11.4)	
	No factor	5 (14.3)	2 (5.7)	7 (10.0)	
Performing attack mitigation practices during the FMF attack period	Yes[c]	11 (31.4)	17 (48.6)	28 (40.0)	$X^2 = 2.143$ $p = .111$
	No	24 (68.6)	18 (51.4)	42 (60.0)	
Life activities inhibited by FMF	Yes[d]	19 (54.3)	19 (54.3)	38 (54.3)	$X^2 = .0000$ $p = 0.595$
	No	16 (45.7)	16 (45.7)	32 (45.7)	
Being worried about having FMF attacks	Yes	23 (65.7)	23 (65.7)	46 (65.7)	$X^2 = 0.000$ $p = 0.599$
	No	12 (34.3)	12 (34.3)	24 (34.3)	
Having other diseases associated with FMF	Yes[e]	2 (5.7)	5 (14.3)	7 (10.0)	$X^2 = 1.429$ $p = 0.214$
	No	33 (94.3)	30 (85.7)	63 (90.0)	
Using FMF drugs regularly	Yes	24 (68.6)	25 (71.4)	49 (70.0)	$X^2 = 0.068$ $p = 0.500$
	No	11 (31.4)	10 (28.6)	21 (30.0)	
Having adequate information about the consequences of not using FMF drugs regularly	Yes	14 (40.0)	15 (42.9)	29 (41.4)	$X^2 = 0.059$ $p = 0.500$
	No	21 (60.0)	20 (57.1)	41 (58.6)	
Symptoms exhibited in the FMF attack period	Fever and abdominal pain	13 (37.1)	18 (51.4)	31 (44.3)	
	Swollen joints, pain, fever, rashes	18 (51.4)	15 (42.9)	33 (47.1)	$X^2 = 3.079$ $p = 0.380$
	Nausea, vomiting, abdominal pain	2 (5.7)	0 (0.0)	2 (2.9)	
	Skin rashes	2 (5.7)	2 (5.7)	4 (5.7)	

X^2: Chi-square statistic $t = t$-test in independent groups

[a] diabetes, asthma, and heart disease

[b] avoiding stress and visiting health facilities regularly for controls

[C] applying an ice pack to joints, placing a hot water bag/hot towel on the abdomen, and taking a warm shower

[d] attending school, playing sports, and joining social activities

[e] appendicitis, urinary tract infection, and kidney disease

Table 3 Inter-group and intra-group comparisons of mean scores obtained by adolescents from the STAI-CH, CDI, and PedsQL and its subscales in pretest and post-test phases ($n = 70$)

	Intervention group (*n* = 35)		**Control group (*n* = 35)**			
Evaluations	$\overline{X}$	**SD**	$\overline{X}$	**SD**	t^a	p
Pretest STAI-CH-S	36.2	3.701	36.943	5.139	−0.694	0.49
Post-test STAI-CH-S	35.2	3.385	36.486	5.387	−1.196	0.237
t^b; p	1.374; 0.178		1.876; 0.069			
Pretest STAI-CH-T	41.571	3.665	40.686	4.993	0.846	0.4
Post-test STAI-CH-T	40.086	4.182	40.457	4.877	−0.342	0.733
t^b; p	1.982; 0.056		1.541; 0.133			
Pretest CDI	27.714	3.511	27.457	5.554	0.232	0.818
Post-test CDI	23.029	4.204	27.171	5.716	−3.454	0.001
t^b; p	5.420; **0.000**		1.663; 0.106			
Pretest PedsQL	49.441	11.69	54.087	15.969	−1.389	0.169
Post-test PedsQL	63.416	17.619	54.494	16.014	2.217	0.03
t^b; p	−7.900; **0.000**		−0.333; 0.741			
Pretest PedsQL Physical Functioning	44.286	20.202	49.429	2.275	−1.037	0.303
Post-test PedsQL Physical Functioning	57.857	21.722	48.989	19.525	1.796	0.077
t^b; p	−6.741; **0.000**		0.306; 0.761			
Pretest PedsQL Emotional Functioning	57.143	16.596	60.429	23.37	−0.678	0.5
Post-test PedsQL Emotional Functioning	69.286	22.398	61.429	23.812	1.422	0.16
t^b; p	−5.251; **0.000**		−1.268; 0.213			
Pretest PedsQL Social Functioning	59.571	21.052	64	23.571	−0.829	0.41
Post-test PedsQL Social Functioning	72.857	21.939	65.429	23.558	1.365	0.177
t^b; p	−4.211; **0.000**		−0.624; 0.537			
Pretest PedsQL School Functioning	39.857	21.091	45.286	20.613	−1.089	0.28
Post-test PedsQL School Functioning	57	19.258	45.857	20.237	2.36	0.021
t^b; p	−5.622; **0.000**		−1.276; 0.211			

statistically significant results are in bold

[a] the independent samples *t*-test

[b] the dependent samples *t*-test

anxiety and trait anxiety before the education was offered individually to the intervention group on the management of FMF. Although state and trait anxiety continued to remain at the medium level for the intervention group, there was a decrease in both after the disease management education, but there was no difference in state and trait anxiety levels of the control group in the post-test phase. There decrease in anxiety levels of the intervention group after the disease management education was not statistically significant, however.

Studies in the literature have shown that children who have frequent and severe attacks have higher levels of anxiety and depression than those who do not [14, 16, 17]. In our study, the frequency of attacks, the findings during the attacks, and the factors that triggered the attacks were similar in the children in both groups, and the severity of the disease could be interpreted as similar. On the other hand, in our study, both groups were determined as moderately anxious both in pretest and post-test assessments, and individual education did not affect the anxiety levels of the adolescents. We assume that this may have several different causes. The effectiveness of the individual training given was evaluated 2 months later, and no attempt was made to consolidate the training during this period. The effectiveness of the training likely decreased over time. Conversely, the lack of time to observe changes in anxiety may also be a factor. This demonstrated that long-term education and monitoring are necessary to influence anxiety levels. In addition, the fact that 65.7% of the adolescents in the intervention group were constantly worried about having an attack might not have made a difference in the post-test data. We think that various limitations of the self-reported STAI-CH, such as its length and complexity of use [38], may have affected the responses of adolescents.

Depressive disorders are seen in every period of childhood, but the incidence increases with increasing age. It is known that depressive symptoms are more common in the

15–19 years age group [39]. According to our study results, the mean depression scores of both groups with similar age ranges at the pretest stage were moderate and similar. However, the fact that the depression score average of the intervention group decreased significantly in the post-test assessment indicates that the individual education given to adolescents had a positive effect on depression. Individual education made a difference in the mental status of adolescents by enabling them to know the disease better and express their feelings and decreased their depression score averages. On the other hand, the improved quality of life of the adolescent may also have had a positive effect on depression scores. This result is important in terms of demonstrating the necessity of regular monitoring of the mental status of adolescents with FMF by health professionals [13, 14].

Unlike our study results, in a study examining the effect of an educational intervention in adolescents with type 1 diabetes, four sessions of individual and group training were given to an intervention group, and it was determined that the intervention did not affect the mean scores of the Child Depression Inventory [40]. Likewise, in the study by Sezer et al. [41], it was identified that the decrease in anxiety and depression levels of adolescents with chronic renal failure was not statistically significant, but the increase in their quality of life scores was statistically significant after receiving a group education. These results reveal the importance of using adaptation techniques in educational interventions for adolescents' coping with stress.

Poor quality of life is a known consequence of chronic disease in children. In numerous studies, it is stated that living with a chronic disease such as FMF, having acute attacks alongside FMF, using colchicine irregularly, having FMF attacks frequently, and exhibiting severe attack symptoms for a long time reduced the quality of life in children/adolescents by negatively affecting their school and daily lives [11, 12, 14, 17, 42]. As an example of this, in the study performed by Yıldırım et al. [13], which compared the quality of life in children with FMF and healthy children, it was discerned that the study group had a low quality of life. In another study, it was found that children with FMF had a lower quality of life than healthy children, but the difference between the two groups was not statistically significant [12]. Likewise, in the study by Makay et al. [17], it was stated that adolescents with FMF obtained lower mean scores from the quality of life scale and its sub-scales; however, this difference was not statistically significant. In their study with children with FMF, Yıldırım et al. [43] stated that school functioning was the most affected area among the sub-scores of the quality of life scale, and this was a result of missing school due to routine hospital visits and unplanned emergency room visits because of attacks, which affected their feelings about school. Similarly, in our study, school functionality was the most affected by the quality of life sub-scores. However, although the school functionality scores in the pre-test were quite low in both groups, they increased significantly in the intervention group after the training.

Among studies that explored the effect of education programs offered to children with a chronic disease, Kocaaslan and Kostak [44] found that the quality of life and disease self-efficacy levels increased after education was offered individually to children with asthma. In Ekici and Cimete's [45] study, asthmatic children and their families were trained in 45–90-min sessions at school and home, according to the "Living with Asthma" booklet, and it was concluded that the program was effective for asthma management. In the study by Gürhopur and Dalgıç [46], it was discerned that, after an education program prepared for children with epilepsy and their parents, the levels of knowledge about the disease and the mean quality of life score of children in the intervention group increased and their attack self-efficacy developed. Similarly, in another study conducted to evaluate the impact of patient education given to children with epilepsy on their quality of life and knowledge level, an educational guide was created that included medical issues such as the definition, classification, etiology, diagnosis, and treatment of epilepsy, as well as psychosocial difficulties experienced in epilepsy and ways of coping. According to the results of the study, the quality of life and the level of knowledge about the disease increased after the education was given using interactive education methods [20]. Kerimoğlu Yıldız et al. [47] compared a tablet game developed to evaluate symptom management and quality of life in children receiving chemotherapy and an educational booklet. As a result, it was determined that both interventions were effective in symptom management, but the quality of life was higher in the play group. This finding may be an indication that providing education programs to children supported by games and technological devices will create better results. The results of our study about the quality of life are parallel to the results of studies about chronic diseases in the relevant literature. Our findings show that adolescents with a relatively low quality of life before education have increased quality of life scores after education and that individual disease management education may offer an opportunity to improve their quality of life.

Strengths and limitations

The most significant strength of our study is that it is the first study to research whether disease management education given individually to adolescents with FMF affected anxiety, depression, and quality of life. The main limitations of this study are that the research results are limited to adolescents living in the province where the research was conducted and the long-term effect of the education on the management of the disease could not be evaluated. One of the limitations of our study is that the scores of the scales are not precise in distinguishing anxious and depressed individuals, rather than direct diagnostic interviews. Diagnosis cannot be made using scales, evaluation can only be made through scores. The results obtained in our study are based on adolescents' self-reports. On the other hand, the methodology of this study was designed as a randomized controlled experiment and no correlational findings were presented. Adolescents were reluctant to refill the personal information form in the post-test because the scales were large and long. For this reason, the characteristics section related to FMF could not be compared in the study.

Conclusion

This study is particularly relevant to health professionals working with children, adolescents, and their families. In particular, the finding that deterioration in the quality of life can be prevented using a brief intervention at minimal cost is encouraging. The following approaches are recommended. First, individual disease education should be an integral part of FMF management in all pediatric and rheumatology clinics, rather than a set of instructions given once at diagnosis. The education of children and adolescents with FMF should be supported by psychosocial interventions to improve coping skills and adaptation to the disease. In this respect, it is an important requirement to recognize the emotional changes or behavioral disorders of children/adolescents and to develop liaison services. Another issue is that regular and continuous training of health personnel (physicians, nurses, and psychologists) is necessary to raise awareness among adolescents about the importance of disease education, to motivate them for self-management, and to detect early psychological problems.

Acknowledgements The authors would like to thank parents who participated in this study.

Authors' contributions Havva Kaçan, Gamze Kaş Alay, Eren Yıldız contributed to the conception and design of the study. Havva Kaçan and Gamze Kaş Alay performed literature screening, drafted the manuscript. Eren Yıldız provided the list of children having FMF disease. Havva Kaçan, Gamze Kaş Alay collected and assembled data. Havva Kaçan, Gamze Kaş Alay, Eren Yıldız performed the analysis. All authors contributed to the manuscript revision and read and approved the submitted version.

Data availability The data that support the findings of this study are available from the corresponding author upon reasonable request. The data are not publicly available due to privacy or ethical restrictions.

Declarations

Ethics approval The ethical endorsement for the research was obtained from the Clinical Trials Ethics Committee of the Faculty of Medicine of Kastamonu University of Turkey (Date: 25 August 2021, no: 2020-KAEK-143–106). Before the research, all adolescents to be included in the sample and their parents expressed their consent for participation in the research both verbally and in written format via the "informed consent form."

Consent to participate Before the research, all adolescents to be included in the sample and their parents expressed their consent for participation in the research both verbally and in written format via the "informed consent form."

Conflict of interest The authors declare no competing interests.

References

1. Goldberg O, Levinsky Y, Peled O et al (2019) Age dependent safety and efficacy of colchicine treatment for Familial Mediterranean fever in children. Semin Arthritis Rheum 49(3):1–12
2. Aydin F, Özçakar ZB, Yalçınkaya F (2017) Çocuklarda ailesel akdeniz ateşi tanı ve tedavisi. Turkiye Klnikleri J Rheumatol Special Topics 10:46–54
3. Li J, Wang W, Zhong L et al (2019) Familial Mediterranean fever in chinese children: a case series. Front Pediatr 7:483. https://doi.org/10.3389/fped.2019.00483
4. Kısaaslan AP, Çiçek SÖ, Şahin N et al (2020) Diseases accompanying Familial Mediterranean fever: a single-centre experience. Bozok Med J 10(2):105–110
5. Alghamdi M (2017) Familial Mediterranean fever, review of the literature. Clin Rheumatol 36(8):1707–1713. https://doi.org/10.1007/s10067-017-3715-5
6. Erdoğan A, Karaman MG (2008) Kronik hastalığı olan çocuk ve ergenlerde ruhsal sorunların tanınması ve yönetilmesi. Anadolu Psikiyatri Dergisi 9:244–252
7. Çavuşoğlu H (2019) Kronik Hastalığı Olan Çocuk. In. Çocuk Sağlığı Hemşireliği. (13th edition). (volume 1). Ankara: Sistem Ofset Basımevi 113–128
8. Akkuş SY, Ayhan AB (2020) Kronik hastalığı olan çocukların davranışlarının ve yaşam kalitelerinin incelenmesi. Türkiye Çocuk Hast Derg/Turkish J Pediatr Dis 14:129–135
9. Bakare MO, Omigbodun OO, Kuteyi OB et al (2008) Psychological complications of childhood chronic physical illness in Nigerian children and their mothers: the implication for developing pediatric liaison services. Child Adolesc Psychiatry Ment Health 34:1–9
10. Turkish FMF Study Group (2005) Familial Mediterranean fever (FMF) in Turkey: results of a nationwide multicenter study. Medicine 84:1–11. https://doi.org/10.1097/01.md.0000152370.84628.0c
11. Durcan G, Barut K, Haslak F et al (2021) Evaluation of health-related quality of life in children and adolescents with Familial Mediterranean fever. Cerrahpaşa Med J 45:21–27. https://doi.org/10.5152/cjm.2021.21014
12. Durmuş SY, Ozlu SG, Cop E, Bulbul M (2022) Health related quality of life of children aged 2–18 years with Familial Mediterranean fever. Turk J Pediatr Dis 16(6):1–7. https://doi.org/10.12956/tchd.1013742
13. Yıldırım DG, Bakkaloğlu SA, Acar ŞS, Buyan N (2021) Parental knowledge about Familial Mediterranean fever: a cross-sectional study. Turk J Pediatr 63:1048–1055. https://doi.org/10.24953/turkjped.2021.06.013
14. Sönmez AÖ, Sönmez HE, Çakan M et al (2020) The evaluation of anxiety, depression and quality of life scores of children and adolescents with Familial Mediterranean fever. Rheumatol Int 40:757–763. https://doi.org/10.1007/s00296-020-04519-2
15. Güler T, Garip Y, Dörtbaş F, Dogan YP (2018) Quality of life in Turkish patients with Familial Mediterranean fever: association with fatigue, psychological status, disease severity and other clinical parameters. Egypt Rheumatol 40:117–121. https://doi.org/10.1016/j.ejr.2017.07.004

16. Durcan G, Yıldız M, Kadak MT et al (2020) Increased frequency of sleep problems in children and adolescents with Familial Mediterranean fever: the role of anxiety and depression. Int J Rheum Dis 23(10):1396–1403. https://doi.org/10.1111/1756-185X.13941
17. Makay B, Emiroğlu N, Ünsal E (2010) Depression and anxiety in children and adolescents with Familial Mediterranean fever. Clin Rheumatol 29:375–379. https://doi.org/10.1007/s10067-009-1330-9
18. Lidor I, Tiosano S, Sharif K et al (2021) The risk of depression and anxiety in patients with Familial Mediterranean fever – a cross-sectional study. J Affect Disord 292:695–699. https://doi.org/10.1016/j.jad.2021.05.113
19. Isaacs D, Sewell JR (2003) Children with chronic conditions. Med J Aust 179(5):235–236. https://doi.org/10.5694/j.1326-5377.2003.tb05524.x
20. Avcı Ö, Bayat M (2021) The Effect of education about epilepsy on quality of life and knowledge level of children: a mixed-method study. Turkiye Klinikleri J Nurs Sci 13(2):219–230
21. Alayli G, Durmus D, Ozkaya O et al (2011) Frequency of juvenile fibromyalgia syndrome in children with Familial Mediterranean fever: effects on depression and quality of life. Clin Exp Rheumatol 29(69):127–132
22. Bodur H, Ataman S, Rezvani A et al (2011) Quality of life and related variables in patients with ankylosing spondylitis. Qual Life Res 20:543–549. https://doi.org/10.1007/s11136-010-9771-9
23. Düzçeker Y, Kanbur NÖ, Demirkaya E et al (2014) Quality of life measures and psychiatric symptoms in adolescents with systemic lupus erythematosus and Familial Mediterranean fever. Int J Adolesc Med Health 26(4):541–549. https://doi.org/10.1515/ijamh-2013-0332
24. Cohen J (1998) Statistical power analysis for the behavioral sciences, 2nd edn. Lawrence Erlbaum, Hillsdale NJ
25. Research Randomizer. Available from https://www.randomizer.org/
26. Spielberger CD (1973) Manual for the state-trait anxiety inventory for children. Palo Alto; CA: consulting psychologists press
27. Kovacs M, Akiskal HS, Gatsonis C, Parrone PL (1994) Childhood onset dysthymic disorder: clinical features and prospective naturalistic outcome. Arch Gen Psychiatry 51:365–374. https://doi.org/10.1001/archpsyc.1994.03950050025003
28. Varni JW, Seid M, Kurtin PS (2001) PedsQL 4.0: reliability and validity of the pediatric quality of life inventory version 4.0 generic core scales in healthy and patient populations. Med Care 39(8):800–812. https://doi.org/10.1097/00005650-200108000-00006
29. Fidan T, Ertekin V, Sürücü İ (2011) Psychological correlates of children and adolescents with Familial Mediterranean fever. J Child Health Dis 54:204–210. https://doi.org/10.1007/s00296-010-1405-6
30. Kasapçopur Ö, Arısoy N (2006) Ailesel akdeniz ateşi ve diğer otoenflamatuar hastalıklar. Turkish Archives of Pediatrics 4:9–17
31. Migita K, Asano T, Sato S (2018) Familial Mediterranean fever: overview of pathogenesis, clinical features and management. Immunological Medicine 41(2):55–61. https://doi.org/10.1080/13497413.2018.1481579
32. Önder ENA, Ertan P (2020) Ailesel akdeniz ateşi. Celal Bayar Üniversitesi Sağlık Bilimleri Enstitüsü Dergisi 7(3):413–417
33. Özdoğan H, Uğurlu S (2019) Familial Mediterranean fever. La Presse Medicale 48:61–76. https://doi.org/10.1016/j.lpm.2018.08.014
34. Savaşer S, Yıldız S, Gözen D, Balcı S, Mutlu B, Çağlar S (2009) NANDA nursing diagnoses which selected for child health and disease nursing . In S. Savaşer, & S. Yıldız (Eds.), Child Health and Diseases Learning Guide for Nurses. İstanbul: İstanbul Tıp Kitabevi pp. 22–25.
35. Semin N, Sabaz MS (2018) Ailesel akdeniz ateşi hastalığı olan çocuğa hemşirelik yaklaşımı. Türkiye Klinikleri Hemşirelik Bilimleri Dergisi 11(1):81–89. https://doi.org/10.5336/nurses.2018-61196
36. Üstebay S, Üstebay DÜ, Yılmaz Y (2015) Ailevi Akdeniz Ateşi. J Acad Res Med 5:89–93
37. Tabachnick BG, Fidell LS (2013) Using multivariate statistics, 6th edn. Pearson, Boston, MA
38. Nilsson S, Buchholz M, Thunberg G (2012) Assessing children's anxiety using the modified short state-trait anxiety inventory and talking mats: a pilot study. Nurs Res Pract 2012:932570. https://doi.org/10.1155/2012/932570
39. Arslan Maden A, Örsdemir Hortu H, Üzüm Ö et al (2020) Beta talasemi minör tanılı ergenlerde anksiyete ve depresyon düzeylerinin değerlendirilmesi. İzmir Katip Çelebi Üniversitesi Sağlık Bilimleri Fakültesi Dergisi 5(3):277–280. Retrieved from https://dergipark.org.tr/en/pub/ikcusbfd/issue/57021/528507
40. Abolfotouh M, Kamal MM, El-Bourgy MD, Mohamed SG (2011) Quality of life and glycemic control in adolescents with type 1 diabetes and the impact of an education intervention. Int J Gen Med 4:141. https://doi.org/10.2147/IJGM.S16951
41. Sezer TS, Çavuşoğlu H, Düzova A (2021) Self-management program for adolescents with chronic kidney disease: a randomized controlled trial. J Ren Care 47(3):146–159. https://doi.org/10.1111/jorc.12357
42. Korkmaz C, Cansu DU, Cansu GB (2020) Familial Mediterranean fever: the molecular pathways from stress exposure to attacks. Rheumatology 59:3611–3621. https://doi.org/10.1093/rheumatology/keaa450
43. Yildirim DG, Bakkaloglu SA, SoysalAcar AS et al (2021) Evaluation of quality of life and its associations with clinical parameters in pediatric patients with Familial Mediterranean fever. North Clin Istanb 8(3):255–260
44. Kocaaslan EN, Kostak MA (2019) Effect of disease management education on the quality of life and self-efficacy levels of children with asthma. J Special Pediatr Nurs 24(2):e12241. https://doi.org/10.1111/jspn.12241
45. Ekici B, Cimete G (2015) Effects of an asthma training and monitoring program on children's disease management and quality of life. Turk Thorac J 16:158–165. https://doi.org/10.5152/ttd.2015.4869
46. Gürhopur FDT, Dalgiç AI (2018) The effect of a modular education program for children with epilepsy and their parents on disease management. Epilepsy Behav 78:210–218. https://doi.org/10.1016/j.yebeh.2017.07.048
47. Kerimoğlu Yıldız G, Yıldız S, Yoruk MA, Sevgen S (2022) A tablet game or training booklet? Two methods for evaluating symptom management and quality-of-life of children receiving chemotherapy. Eur J Oncol Nurs 61:102190. https://doi.org/10.1016/j.ejon.2022.102190