RESEARCH

DETERMINATION OF NUTRITIONAL STATUS IN CHILDREN WITH CYSTIC FIBROSIS

Abstract

Cystic fibrosis is a disease that affects quality of life and life span as a result of mutation in the transmembrane conductivity regulator gene. The aim of this study is to determine the nutritional status of 2–18-year-old children with cystic fibrosis.75 children with cystic fibrosis between the ages of 2-18 were included in the study. The data were obtained online with the "Sociodemographic Form", "Nutrition Evaluation Form" and "24-Hour Food Consumption Record". Dietary quality, energy intake, macro and micronutrient intakes of the participants were determined by taking a 24-hour food consumption record. IBM Statistical Package for Social Sciences Version 24 statistical program and BEBIS 8.2 program were used to analyze the data obtained through the questionnaire.

In this study 32 girls and 43 boys were included. It was determined that the daily energy intake of the girls was 1368 ± 394 kcal, and the boys were 1496 ± 494 kcal. When the z-score values of the participants are examined, the z-score value according to the height is minimum -4.16, maximum 3.05; It has been determined that the z-score value for weight is minimum -3.79 and maximum 2.60. The mean z-score of the participants for height was -0.25; The mean z score according to weight was determined as -0.40. The average weight of the boys is 30 kg, and the girls are 28.4 kg. More studies are needed to determine nutritional status, new nutritional therapies, and supplemental doses in children with cystic fibrosis.

Keywords: Cystic fibrosis, nutrition, nutritional status

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KİSTİK FİBROZLU ÇOCUKLARDA BESLENME DURUMUNUN BELİRLENMESİ

Öz:

Kistik fibrozis, transmembran iletkenliğini düzenleyici gende meydana gelen mutasyon sonucu yaşam kalitesini ve yaşam süresini etkileyen bir hastalıktır. Bu çalışmanın amacı 2-18 yaş arası kistik fibrozisli çocukların beslenme durumlarının değerlendirilmesidir. Çalışmaya 2-18 yaş arası kistik fibrozlu 75 çocuk dahil edilmiştir.Çocuklara "Sosyodemografik Form", "Beslenme Değerlendirme Formu" ve "24 Saatlik Besin Tüketim Kaydı" online olarak uygulanmıştır. Katılımcıların diyet kalitesi, enerji alımları, makro ve mikro besin alımları 24 saatlik besin tüketim kaydı alınarak belirlenmiştir. Anket aracılığıyla elde edilen verilerin analizinde ve değerlendirilmesinde IBM Statistical Package for Social Sciences Version 24 istatistik programı ve BEBİS 8.2 programı kullanılmıştır.

Çalışmaya 32 kız ve 43 erkek dahil edilmiştir. Kızların günlük enerji alımlarının 1368 ± 394 kkal, erkeklerin ise 1496 ± 494 kkal olduğu belirlenmiştir. Katılımcıların z-puanı değerleri incelendiğinde boylarına göre z-puanı

değeri en az -4.16, en fazla 3.05; ağırlık için z-skoru değerinin en az -3,79 ve en fazla 2,60 olduğu belirlenmiştir. Katılımcıların boy için ortalama z puanı -0,25; ağırlığa göre ortalama z skoru -0.40 olarak belirlenmiştir. Erkeklerin ortalama ağırlığı 30 kg, kızların ağırlığı ise 28,4 kg'dır. Kistik fibrozlu çocuklarda beslenme durumunu, yeni beslenme tedavilerini ve ek dozları belirlemek için daha fazla çalışmaya ihtiyaç vardır.

Anahtar Kelimeler: Kistik fibrozis, beslenme, beslenme durumu

1. Introduction

Cystic fibrosis (CF) is a disease that affects the quality of life and duration of life as a result of mutation in the cystic fibrosis transmembrane conductivity regulator (CFTR) gene (1-3). Absence, deficiency, or structural and functional abnormalities of the CFTR protein lead to mucosal hyperconcentration in the respiratory, digestive, and reproductive systems and malabsorption of chloride and sodium in the sweat glands (4).

The overall prevalence of CF is thought to be 1/2500 live births in Europe (2;5). As a result of a study conducted in Turkey, it was determined that approximately 7 out of 100 children who applied to the hospital in our country with complaints of lung infection and malnutrition were diagnosed with cystic fibrosis (6).

CFTR protein allows chloride to pass through mucus-producing cells, followed by water and the mucus thins. Defective CFTR results in thick and sticky mucus that blocks pathways, leading to serious lung infections, particularly pseudomonas (7;8). It is recommended that diagnoses associated with CFTR mutations be made by evaluating CFTR function with sweat chloride test in all individuals, from newborn to adult (9). The diagnosis of CF is made within four weeks by sweat testing and/or genetic mutation analysis. Early diagnosis through newborn screening is very important especially in terms of growth-development and nutrition (2).

CF patients usually present to the clinic with complaints of chronic cough, shortness of breath and wheezing. As the disease progresses, patients develop bronchiectasis, a chronic condition of dilatation of the bronchi that develops as a result of recurrent lung infections (10).

A study of impaired gut microbiota in children with cystic fibrosis found that children with CF had significantly different gut microbiota when compared to a healthy control group. In the study, a decrease in the levels of eubacterium rectale, bacteroides uniformis, bacteroides vulgatus, bifidobacterium adolescentis, bifidobacterium catenulatum and faecalibacterium prausnitzii was found in children with CF (11).

Cystic fibrosis-related diabetes (CFRD) affects up to 50% of adult patients with CF. Patients with CFRD have a relative insulin deficiency and insulin loses its anabolic effects, causing a catabolic state and consequent significant weight loss. CFRD is associated with significant adverse effects on patients' health, quality of life, and life expectancy (12).

Regular measurements of weight and height in individuals with cystic fibrosis are very important for the prevention or early detection of malnutrition (2). Malnutrition is encountered in people with CF as a result of the combination of malnutrition, energy losses, high energy needs and inadequate nutritional intake (13). Metabolism of fat-soluble vitamins and fats is adversely affected due to pancreatic insufficiency in people with cystic fibrosis (1; 2; 14).

It is recommended that patients with CF meet 120 to 150% of the recommended daily energy intake to compensate for malabsorption and increased caloric needs (15). It is recommended to provide 40% of daily energy from fat (15), 15% from protein and the rest from carbohydrates (16).

Patients with CF, particularly those with pancreatic insufficiency, are at risk for fat-soluble vitamin deficiencies. Especially, fat-soluble vitamin deficiencies may be associated with worse clinical status (3). On the other hand, there is no problem in the absorption of water-soluble vitamins and there is no need for regular supplements (2). As a result of increased sweating, intestinal malabsorption, and chronic inflammation common in CF, patients may have higher-than-normal requirements for sodium, calcium, iron, zinc, and selenium (1).

In cases where nutritional status cannot be provided orally, the transition to enteral nutrition is important in controlling malnutrition and improving clinical symptoms (17; 2).

The aim of this study is to examine the clinical findings seen in individuals with cystic fibrosis, whose nutritional deficiency is common, to look at the nutritional status and to examine the effect of nutritional status on symptoms.

2. Material and Methods

2.1. Study Design and Ethical Considerations

This is a descriptive study. The study was carried out with 75 patients who were diagnosed with cystic fibrosis, between the ages of 2 and 18, registered in KifDer "Cystic Fibrosis Assistance and Solidarity Association", and who were eligible in terms of inclusion and exclusion criteria. This study was carried out between April - June 2022.

Ethical approval of our study was received by the Haliç University Non-Interventional Clinical Research Ethics Committee at the meeting dated 27.04.2022 (Haliç University Non-Interventional Clinical Research Ethics Committee, registration number 2022-83). It was carried out in accordance with the Declaration of Helsinki.

All patients and their family participating in the study were informed in writing about the content of the study and its method of application, participant rights, and the questionnaires to be used.

2.2. Data Collection Tools

The data were obtained online with the "Sociodemographic Form", "Nutrition Evaluation Form" and "24-Hour Food Consumption Record", which were created by the researcher and composed of questions about the sociodemographic data of the volunteers, the amount of food consumption for their nutritional status and their eating habits. The form and questionnaire specified for the purpose of collecting data were prepared online via "Google Forms" and sent to the participants via whatsapp and e-mail.

2.3. Instruments

2.3.1. Sociodemographic Form: It was prepared and presented to the participants in order to obtain information about the age, gender, education level and sibling status of the individuals participating in our study.

2.3.2. Nutrition Evaluation Form: It was prepared and presented to the participants in order to determine the appetite status of the individuals participating in the study, the amount of meals they consume, the amount of water, and the consumption of enteral products.

2.3.4. 24-Hour Food Consumption Record: It is a standard inquiry form that defines the 24-hour period as a standard (from 23:59 at midnight the previous day to 24:00 at midnight last night), facilitating the remembering of the foods and their amounts consumed in this process.

2.4. Evaluation of Data

IBM Statistical Package for Social Sciences Version 24 (SPSS inc, Chicago, IL, USA) statistical program and BEBIS 8.2 program were used to analyze the data.

Continuous variables are given as mean ± standard deviation, qualitative variables as numbers and percentages (%). Comparison of normally distributed bivariate data in terms of means was obtained with Independent Samples T Test, and for non-normally distributed data with Mann Whitney U Test. Comparisons in terms of means were analyzed using the One-Way Analysis of Variance Test for normally distributed data in more than two groups, and the Kruskal-Wallis Test for data from more than two groups that were not normally distributed. Statistical significance level was taken as $p < 0.05$.

3.Results

In this study, 75 patients participated, including 32 girls (42.6%) and 43 (57.4%) boys, between the ages of 2 and 18. The mean age of the individuals participating in the study was 8,900 ± 4,8935. It was determined that the average height of the individuals participating in our study was 128.87 ± 25.244 cm; their average weight was 29.5920 ± 14.94861 kg, and their birth weight average was 3107 ± 529.107 grams.

The average weight of the boys participating in the study is 30 kg, and that of the girls is 28.4 kg. While 29.3% (22 patients) of individuals aged 2-18 years who participated in our study were diagnosed with cystic fibrosis in heel blood screening, 70.7% were diagnosed as a result of tests and scans performed due to disease-specific symptoms in the future.

Individuals of 25.3% (19 patients) participating in our study were diagnosed with another disease in addition to cystic fibrosis disease. In addition to cystic fibrosis, 14 different diseases were diagnosed in the individuals participating in our study. The most frequently diagnosed disease was diabetes (5 patients).

Table 1: Evaluation of Participants' Status of Receiving Dietitian Support

		Enteral Products Use		
		Yes (n)	No (n)	Total
Dietitian Support	Yes(n)	27	18	45
	No(n)	8	22	30
	Total	35	40	75

Table 2: Evaluation of Participants' Food Consumption Records by Gender

	Gender	**Number(n)**	**X ± SD**	**p**
Energy (kcal)	Girl	32	1368± 394,58	0,211
	Boy	43	1496±494,00	
Protein (g)	Girl	32	53,41±15,84	**0,005**
	Boy	43	58,57± 26,50	
Fat (g)	Girl	32	62,41±22,41	0,274
	Boy	43	67,00±27,56	
Carbohydrate (g)	Girl	32	144,46± 54,33	0,707
	Boy	43	161,81±55,44	
Fiber (gr)	Girl	32	13,52± 5,63	0,110
	Boy	43	15,14± 8,49	
Polyunsaturated Fat (g)	Girl	32	11,59± 4,86	0,193
	Boy	43	13,16± 6,36	
SD, standard deviation; X, mean.				

When the energy intake of the participants was evaluated according to the gender parameter, no statistically significant difference was found (p=0.211). When the protein, carbohydrate and fat consumption amount of the participants were evaluated according to the gender parameter, a statistically significant difference was found in the amount of protein consumption between girls and boys (p=0.005), but there was no statistically significant difference in terms of carbohydrate (p=0.707) and fat (p=0.274) consumption. no significant difference was found (Table 2).

Table 3: Evaluation of Daily Vitamin and Mineral Intakes of Participants by Gender

	Gender	Number(n)	X ± SD	p
Vitamin A (µg)	Girl	32	939,73± 637,36	0,547
	Boy	43	977,91± 651,11	
Vitamin E (mg)	Girl	32	10,16± 4,67	**0,010**
	Boy	43	12,91± 7,53	
Sodium (mg)	Girl	32	2250±884,84	0,122
	Boy	43	2441±1316	
Iron (mg)	Girl	32	7,06±3,06	0,394
	Boy	43	7,43± 3,81	
Magnesium (mg)	Girl	32	186,92± 80,23	0,207
	Boy	43	200,03±104,21	
Zinc (mg)	Girl	32	7,05±3,07	**0,005**
	Boy	43	8,57±5,35	
Calcium (mg)	Girl	32	618,74±285,07	**0,026**
	Boy	43	636,19± 416,90	
SD, standard deviation; X, mean.				

Table 4: Evaluation of Participants' Z Score by Height and Weight

	Number(n)	Min	Max	X	SD
Z score by height	75	-4,16	3,05	-0,25	1,39108
Z score by weight	75	-3,79	2,60	-0,40	1,44970
Min-max, minimum-maximum; SD, standard deviation; X, mean					

Table 5: Evaluation of Participants' Z-Score Values According to Age Parameter

	Age	Number(n)	X	SD	p
Z score by height	2 – 10	49	0,056	1,37	0,804
	11 – 18	26	-0,84	1,24	
Z score by weight	2 – 10	49	0,15	1,18	0,107
	10 – 18	26	-1,44	1,33	
SD, standard deviation; X, mean					

4. Discussion

In this study, which aims to determine the nutritional status of children with cystic fibrosis between the ages of 2 and 18, the energy, carbohydrate, protein, fat, fiber, and polyunsaturated intake of the participants; the amount of vitamins and minerals they take with nutrition was examined and evaluated according to the gender parameter. As a result of this evaluation, a statistically significant difference was found only in daily protein intake and zinc intake. Cystic fibrosis is a hereditary disease that causes serious damage to many organs in the body and negatively affects life expectancy and quality (18).

Nutrition is an important component of cystic fibrosis treatment, and the cornerstone of treatment is a high-fat diet. However, adherence to dietary recommendations for cystic fibrosis appears burdensome for most children and adolescents. This leads to malnutrition, growth-development retardation, inadequate lung function and an increased risk of respiratory tract infections (19). In our study, in the evaluation of the z score for height according to age, the mean z score for children aged 2 to 10 was 0.0563 ± 1.37447, while the mean z score for children aged 11 to 18 was -0.8423 ± 1.24579. In the evaluation of the z score according to weight of the participants, the

mean z score for children aged 2-10 was 0.1500 ± 1.18621, while the mean z score for children aged 11-18 was -1.4431 ± 1.33970 and there is no statistically significant difference between the two age groups (p=0.107).In a cross-sectional study of 101 children and adolescents with cystic fibrosis in Brazil, Neri et al. reported adequate calorie and macronutrient consumption and adequate nutritional status in most patients. However, lower BMI z scores were observed in schoolchildren (5 to <10 years) and adolescents (≥10 years) (20). In another study, 76 children with CF in Greece, 9% of boys and 5% of girls were underweight (BMI Z score for age <-2.0) despite patients meeting or exceeding the recommended total energy intake (19). In our study, when the 24-hour food consumption record data of individuals between the ages of 2 and 18 and energy intakes by gender were examined, it was found that the average daily energy intake of girls was 1368,0450 ± 394,58258 kcal, and that of boys was 1496,8128 ± 494,00418 kcal no statistically significant difference is found between the two age groups (p=0.211).With 80 children with cystic fibrosis in 2018, it was determined that the daily energy intake was 3420 kcal/day in male participants and 2866 kcal/day in female participants (21). Poulimeneas et al., including 76 children with cystic fibrosis, when the energy intakes of children and adolescents were examined, it was determined that the daily energy intake of male participants (n= 32) was 2623 ± 654 kcal, and the energy intake of female participants (n=44) was 2486 ± 738 kcal (19). Cystic fibrosis is a disease that causes serious damage to many organs in the body and negatively affects the quality of life.Nutrition is an important component of cystic fibrosis treatment. Cystic fibrosis can cause malnutrition, growth and development retardation. For this reason, patients should be informed that sustainable dietary recommendations are provided by experts and that this adherence is an important factor in the quality and duration of life for most children and adolescents.

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Declaration of Interest

The authors declare that they have no conflicts of interest.

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