RESEARCH ON THE QUALITY OF LIFE OF ADULT PATIENTS WITH CYSTIC FIBROSIS IN HUNGARY

*Tímea Tóth, Erzsébet Mák, Nóra Galló, István Szabolcs

Department of Dietetics and Nutrition Sciences, Faculty of Health Sciences, Semmelweis University, Budapest, Hungary
Head of Faculty: Professor Nagy Zoltán Zsolt, MD, PhD

Summary

Introduction. Cystic fibrosis (CF) is the most common autosomal recessive metabolic disease. All symptoms associated with the gastrointestinal system have adverse effects on the nutritional status, which, in turn, is closely related to the prognosis for the patient. Insufficient energy intake results in malnutrition, leading to worse respiratory function, and the early onset of comorbidities; it is accompanied with worse quality of life and a shortened lifespan.

Aim. The aim of our research was to assess the quality of life and nutritional status of adult patients with CF in Hungary and to determine the correlations between nutritional status, lung function and the quality of life. In Hungary, we are the first to study the relationship between nutritional status and the quality of life in adult CF patients.

Material and methods. 57 patients with CF from Hungary of both sexes were studied, all of them aged over 18. An assessment of the quality of life with a questionnaire was conducted among them. Their nutritional status was measured with the bioelectrical impedance analysis. The most recently measured FEV_1 (forced expiratory volume in 1 second) values for the patients were analyzed. In order to assess the quality of life, the validated Hungarian translation of the disease-specific Cystic Fibrosis Questionnaire-Revised (CFQ-R) – Teen/Adult version for ages 14 through adulthood – was used.

Results. The nutritional status is worse in female patients than in male. Undergoing lung transplantation did not have any impact on the nutritional status. However, we found significant differences in the FEV_1 values between the patients who underwent transplantation and those who did not.

No strong correlation was found between any parameters of the nutritional status, FEV_1 and the domains of the quality of life.

In the mean values of the quality of life domains, the highest score appeared in the eating domain, while the lowest one was found in the weight domain. We did not find any significant differences between the quality of life of male and female patients, except for the respiratory symptoms domain, where women's scores were lower. However, this result was not confirmed by FEV_1 data. Transplant patients' quality of life was higher than non-transplant ones in almost all domains, with 6 domains where statistically significant differences were found. The transplant CF patients, however, had lower scores in the weight and body image domains. CF patients with lower BMI had lower quality of life than their peers. Muscle mass percentage did not differ between groups. FEV_1 showed a statistically significant correlation with six of the CFQ-R domains.

Conclusions. Our research is the first disease-specific assessment of the quality of life among Hungarian adult patients with CF. In Hungary, we are the first to study the relationship between nutritional status and the quality of life in adult CF patients. We conclude that early dietetical interventions should be implemented to improve patients' condition and the quality of life.

Keywords: cystic fibrosis, mucoviscidosis, adult, quality of life, nutritional status

INTRODUCTION

Cystic fibrosis (hereinafter CF) is the most common autosomal recessive metabolic disease. It affects multiple organs and, therefore, can present with different symptoms, such as chronic respiratory disease, gastrointestinal symptoms, as well as changes in the pancreas, liver, biliary tract or reproductive tract. The disorder is characterized by the dysfunction of exocrine glands, which produce abnormally thick and sticky mucus. The phlegm congestion causes obstructive lesions in many vital organs, especially in the lungs and in the pancreas (1). The so-called CF-related diseases, such as diabetes mellitus (cystic fibrosis related diabetes – CFRD),

nasal polyposis, osteoporosis, chronic sinusitis or arthropathy, are likely to appear with ageing.

In 2012, there were 579 patients in the Hungarian registry of CF patients, including 261 women and 318 men. 248 out of these patients were adults. The proportion of adults among the patients has become significantly higher in the recent years due to improvements in therapy and easier access to transplantation. Mean age of CF patients accounted for 14.77 years of age in 2008 and rose to 16.54 years of age in 2012 (2). Due to the formerly relatively small adult population of the patients, it was not possible to conduct a proper analysis on them. Today, however, their increasing number enables to describe their problems with more accuracy.

All the symptoms related to the gastrointestinal system compromise the nutritional status. Since the nutritional status plays a significant role in the progression of cystic fibrosis, dietotherapy must be an essential part of the treatment (3, 4). Patients with good nutritional status are more likely to preserve or improve their health status, which also affects their quality of life (5). On the other hand, lower nutritional status increases the risk of death (6, 7). Therefore, watching for signs of malnutrition and promoting the development of an adequate nutritional status are crucial for optimal treatment of CF (5).

Due to the gastrointestinal symptoms, deficiencies are likely to be present in CF patients. To assess the patients' nutritional status, several parameters can be used. Body mass index (BMI) and body fat percentage accurately depict the nutritional status. Studies have shown that, thanks to easy calculation and precise results, BMI can be applied with good effects as a help in assessing CF patients. Bioelectrical impedance analysis is commonly used for estimating body fat percentage.

The best predictor of mortality rate in CF is the decrease in the lung function, as the exacerbation of a chronic lung disease remains the leading cause of death in CF (8). There is a direct correlation between the nutritional status, energy consumption and the functional capacity of skeletal and respiratory muscles. Patients with higher body mass index often have better lung function (7). Body weight and body mass index correlate with the FEV_1 value (forced expiratory volume in 1 second) (9).

In addition to physical, physiological and biochemical parameters, other factors must be taken into account when assessing the patient's health status, such as the individual's subjective perception of their own life, their physical and mental status, the ability to perform their everyday activities, their social perceptions, as well as the impact of all these factors on their health status. Quality of life tests can be applied specifically for this purpose.

Today there are standardised and validated methods available for the assessment of the quality of life, which include general and disease-specific questionnaires. The second ones are more sensitive and consequently, are able to detect smaller changes in the health status of a given group of patients, but they preclude the comparison of patients suffering from different diseases. The quality of life questionnaires can be further divided into index- and profile-type questionnaires. Index-type questionnaires present the quality of life as a list of scores, whereas profile-type questionnaires measure the quality of different life domains.

Among various data collecting methods, self-administered questionnaires are the most common one. They are easy to use, cost-efficient, as well as they are able to provide standardised measurements (10).

The Cystic Fibrosis Questionnaire-Revised (CFQ-R), which has a number of versions dedicated for specific age groups (children, adolescents and adults, parents), can be used specifically to measure the quality of life of patients with cystic fibrosis.

AIM

The aim of this study was to assess the quality of life in adult patients with CF in Hungary, as it had never been previously assessed. We aimed to collect the data about patients' nutritional status, lung function, as well as to identify correlations between BMI, body fat percentage, muscle percentage, FEV_1 and quality of life domains.

MATERIAL AND METHODS

Questionnaires were used to assess the quality of life in CF patients. Participants' nutritional parameters, including weight, height, BMI, body fat percentage and muscle percentage, were assessed. This paper analyses BMI, body fat percentage, muscle percentage, FEV_1 and quality of life only. We collected surveys from 57 patients over 18 years of age.

Personal data, including anthropometric measurements and questionnaires, were collected multiple times among outpatients and inpatients in the years 2013 and 2014.

Inclusion criteria were as follows: diagnosed cystic fibrosis, age over 18 years of age, appropriate cooperative skills, willingness to fill in the questionnaire as well as to provide anthropometric data and to complete the written consent form. Inadequate cooperation was considered an exclusion criterion.

Nutritional status and lung function

The patients' nutritional status was assessed with bioelectrical impedance devices, which enabled to obtain the patients' body weight, body fat percentage, muscle percentage and body mass index (after entering the height of the participant).

To obtain the indicators of the lung function, we collected the most recently measured FEV_1 values for each patient.

Quality of life

To assess the quality of life, we used the validated Hungarian translation of the disease specific Cystic Fibrosis Questionnaire-Revised (CFQ-R) – Teen/Adult version for ages 14 through adulthood (11, 12). This questionnaire consists of 50 separate questions, which group the aspects of the quality of life into 12 domains: physical functioning, emotional functioning, social functioning, body image, eating, treatment burden, respiratory and digestive symptoms, role fulfilment, weight, health perception and vitality. CFQ-R is a profile-type questionnaire. The scores for the subscales range from 0 to 100, where higher score indicates higher quality of life in the given domain. 54 out of 57 participants filled in the questionnaire, 41 underwent bioelectrical impedance analysis and 38 had a complete data file.

Statistical analysis

To analyse the data, we used the IBM SPSS Statistics 21.0 and Microsoft Office Excel, version 2010, software. Differences were considered statistically significant when p values were ≤ 0.05. We used Spearman's rank correlation coefficient (r) to express the strength of correlations. Correlation coefficients were defined as weak if $r < 0.3$, moderate if r ranged from 0.3 to 0.7 and strong if $r > 0.7$ (13). We used nonparametric tests to measure the alterations of the continuous variables and the Mann-Whitney test and Kruskal-Wallis test, depending on the size of the categories, to analyse the relationship between them. In order to analyse the relationship between categorical variables, we used the chi-square test. We have also applied the nonparametric Mann-Whitney or the Kruskal-Wallis tests to calculate and measure the factors that affect the nutritional status.

RESULTS

The average age of the patients was 28.25 (± 8.95) years of age – 28.46 (± 9.5) for men, and 28.07 (± 8.59) for women. The 54 evaluable questionnaires revealed that there were 10 transplant patients among the respondents, evenly divided between the sexes. The majority of the non-transplant patients were female.

Although the average body mass index (calculated from measured body weight and height) was in the normal range, the BMI of the majority of our patients remained in the lower normal range. The mean value of body fat percentage was in the sub-normal range in women. Muscle percentage was within the normal range for both sexes. The mean FEV_1 values were low for both sexes, as predictable in CF patients (tab. 1).

Nutritional status

52.6% of the participants (n = 30) had normal BMI, 43.9% (n = 25) – low BMI, and only 3.5% (n = 2) had high BMI. 59.3% of the male patients (n = 16) fell into the normal range, 33.3% (n = 9) had low BMI, while 7.4% (n = 2) had high BMI. 46.7% of the female patients (n = 14) had normal BMI, while low BMI values were found in 53.3% of female patients (n = 16). None of the female participants had high BMI score.

We obtained reliable results of bioelectrical impedance analysis for 41 out of 57 participants (71.93%). 35% of the male patients had low body fat percentage, 55% – normal body fat percentage, and 10% – high. The mean value for male patients accounted for 12.09% (± 5.34%) of body fat, and for female patients – 20.69% (±7. 82%). Both mean values are in the normal range (tab. 1). 52.4% of the female patients had low body fat percentage, 38.1% – normal, and 9.5% – high.

No significant correlation was found between any of the parameters of the nutritional status in the transplant patients. However, the FEV_1 mean value in transplant patients (72.11% ± 14.39%) was significantly higher than in the non-transplant patients (49.82% ± 24.62%; p = 0.009). As expected, transplant patients had better lung function.

Quality of life

After evaluating the 54 valid CFQ-Rs, we concluded that the highest mean score appeared in the eating domain, whereas the lowest – in the weight domain (tab. 2).

There was no strong correlation found between each of the quality of life domains.

BMI moderately correlated with the following quality of life domains: physical functioning (r = 0.424; p <

Tab. 1. Patient demographics and baseline characteristics

	Total			Male			Female			Reference values	
	Mean (SD)	Min	Max	Mean (SD)	Min	Max	Mean (SD)	Min	Max	Male	Female
Age	28.25 (8.95)	18.56	66.13	28.46 (9.5)	18.56	66.13	28.07 (8.59)	18.93	47.55		
Transplant patients; n (%)	10 (18.52)			5 (19.23)			5 (17.86)				
Non-transplant patients; n (%)	44 (81.48)			21 (80.77)			23 (82.14)				
BMI; kg/m² (n = 57)	19.41 (2.49)	15.13	25.94	20.28 (2.45)	16.60	25.94	18.62 (2.29)	15.13	23.49	18.5-24.99	
Body fat %; (n = 41)	16.49 (7.94)	5	36.1	12.09 (5.34)	5	21.5	20.69 (7.82)	10	36.1	8-19.9[14]	21-32.9[14]
Muscle %; (n = 41)	33.48 (8.11)	18.5	51.2	37.11 (9.45)	20.8	51.2	30.03 (4.59)	18.5	39.4	33.3-39.3[15]	24.3-30.3[15]
FEV1; %	54 (24.57)	16	102	53.96 (24.62)	16	102	58.33 (25.06)	16	96	80-120[16]	

CF – cystic fibrosis; BMI – Body Mass Index; FEV_1 – forced expiratory volume in 1 second
[14]Gallagher et al. 2000
[15]http://www.omron-healthcare.com/data/catalog/3/112/1/IM-HBF-510-E-04-08-08EN.pdf
[16]http://www.ttuhsc.edu/som/internalmedicine/pulm/education/pfts/pfts_normal_values.pdf?ref=binfind.com/web

0.01), body image (r = 0.656; p < 0.01), eating (r = 0.406; p < 0.01), health perception (r = 0.435; p< 0.01), vitality (r = 0.302; p < 0.05) and weight (r = 0.692; p < 0.01). Weak correlation was found between BMI value and the domains of respiratory symptoms (r = 0.271; p < 0.05) and roles fulfilment (r = 0.285; p < 0.05).

Moderate correlation was established between body fat percentage and the domains of emotional functioning (r = 0.352; p < 0.05), body image (r = 0.433; p < 0.01) and weight (r = 0.406; p < 0.05).

Moderate correlation was found between FEV_1 and the following domains: physical functioning (r = 0.646; p < 0.01), emotional functions (r = 0.301; p < 0.05), treatment burden (r = 0.400; p = 0.005), health perception (r = 0.515; p < 0.01), respiratory symptoms (r = 0.487; p < 0.01), roles fulfilment (r = 0.301; p < 0.05) and weight (r = 0.417; p < 0.05).

There was no significant difference between male and female patients' quality of life, with the exception of the respiratory symptoms domain, which presents with a lower score for female patients.

Transplantation has an important effect on patients' quality of life. We found significant differences between the groups of transplant and non-transplant patients in the following domains: physical functioning, social functioning, treatment burden, heath perception, respiratory function and vitality. In each of these domains, transplant patients demonstrated higher scores than the non-transplant ones.

In two domains, body image and weight, transplant patient scored lower than the non-transplant ones. However, the differences between the groups did not reach the level of statistical significance (fig. 1).

We found correlations between seven domains of the quality of life and body mass index. Patients with low BMI had lower scores in domains of physical functioning (p = 0.004), body image (p < 0.001), eating (p = 0.004)

health perception (p = 0.018), role fulfilment (p = 0.041), vitality (p = 0.015) and weight (p < 0.001). Patients with low body mass index had strikingly low values (< 40) in the body image and body weight domains.

There were significant differences between three groups according to the body fat percentage in the following quality of life questionnaire domains: physical functioning (p = 0.035), body image (p = 0.011), eating (p = 0.002), health perception (p = 0.016), roles fulfilment (p = 0.048) and weight (p = 0.004). The mean score of the weight domain was exceptionally low (16.67) in the group of patients with low body fat percentage.

We could not find any significant differences between the groups with different body muscle percentage in the domains of the quality of life.

After dividing the patients according to their FEV_1 values, which represent the lung function, we found significant differences between the groups in six domains: physical functioning (p < 0.001), treatment burden (p = 0.029), health perception (p = 0.002), respiratory symptoms (p = 0.012), roles fulfilment (p = 0.034) and weight (p = 0.007). It can be underlined that in five out of the six domains in which significant differences were found, patients with best lung function had best scores, with the exception of the role fulfilment domain, where patients with moderate lung function had highest values.

DISCUSSION

In this research, we measured not only the nutritional status of adult patients with CF, but also their quality of life. It is the first paper on Hungarian adult CF population aiming to examine the relationship of the nutritional status and the quality of life.

Nearly half of participants had low body mass index. On the other hand, the number of CF patients in the normal BMI range was higher than expected (tab. 1). The

Tab. 2. Patients' quality of life scores

|	N	Mean	SD	Minimum	Maximum	Percentile		
						25	50	75
Physical functions	54	66.13	28.28	0	100	52.08	70.83	88.54
Emotional functions	54	67.9	21.2	6.67	100	53.33	66.67	81.67
Social functions	54	67.7	16.41	22.22	100	55.56	66.67	79.17
Body image	54	53.5	28.07	0	100	22.22	55.56	77.78
Eating disorders	54	82.3	20.87	22.22	100	66.67	88.89	100
Burden of treatment	54	58.64	25.02	0	100	44.44	55.56	77.78
Health perceptions	54	54.32	26.14	0	100	33.33	55.56	69.44
Digestive symptoms	54	81.48	17.4	22.22	100	77.78	88.89	100
Respiratory symptoms	54	63.48	24	0	100	44.44	61.11	84.72
Role functions	54	72.07	24.07	0	100	58.33	83.33	91.67
Vitality	54	55.56	19.89	8.33	100	41.67	58.33	66.67
Weight	54	41.98	42.55	0	100	0	33.33	75
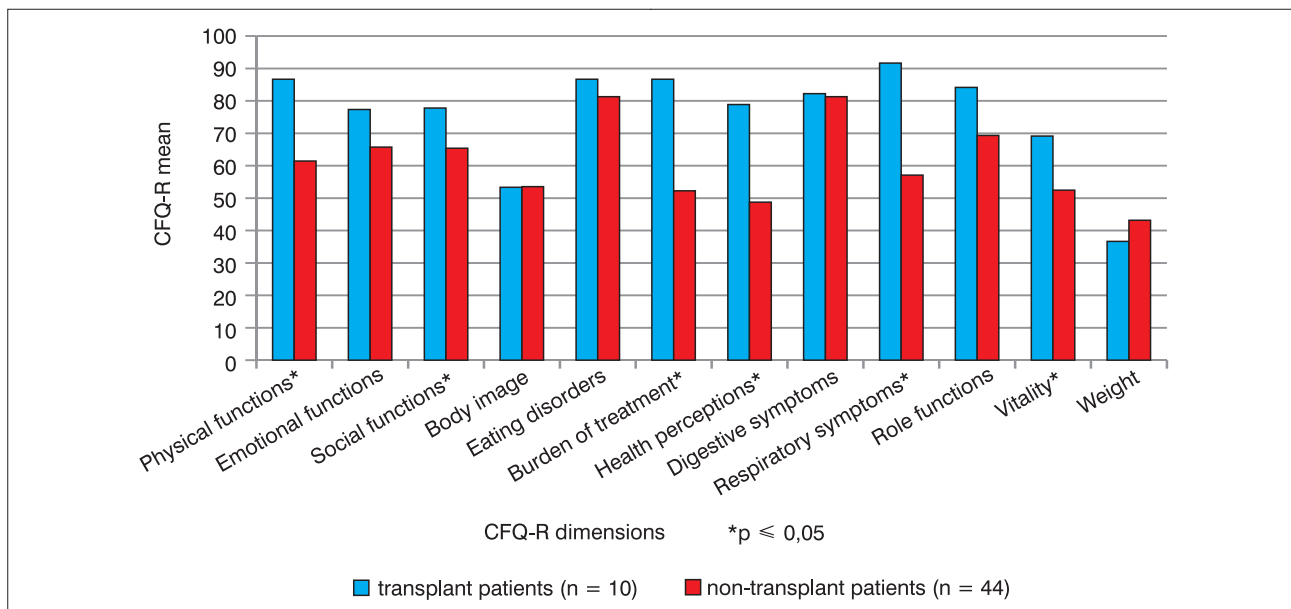
Fig. 1. Comparison of the quality of life scores regarding transplantation

successful application of hypercalorisation as well as having experience in the substitution of pancreatic enzymes in the adult patients may explain this phenomenon.

Low BMI was more prevalent among female patients. The body fat percentage of the majority of male participants was in the normal range, whereas a big group of women with low body fat percentage was found. These results suggest that women's nutritional status is worse than the nutritional status of men. We found contradictory results in other papers and only a part had results similar to ours (4, 6, 17).

What was unexpected, there was no difference in the nutritional status of the transplant and the non-transplant patients, contrary to the Hollander's test (18). While further investigation may be required to identify the underlying reasons, it is believed that the nutritional status is likely to be affected by several factors and undergoing transplantation is only one of them. Nevertheless, significant differences were found between the groups of the transplant and the non-transplant patients when it comes to the lung function.

As indicated by the quality of life scores, the biggest problem for the patients was body image and achieving desired body weight, as these were the domains with the lowest scores observed. The eating domain, also related to nutrition, had the highest scores of all the domains. International studies revealed similar results: according to the research conducted by Quittner, Riekert and Ashish, eating was the smallest problem for the examined CF patients (11, 19, 20). In the body image domain, the patient is asked about his perceived thinness, therefore, low scores suggest that patients consider themselves thin. However, this was not confirmed by the results of the nutritional status analysis, which may suggest that patients were not fully aware of their nutritional status, seeing it as worse than it actually was.

There was a significant difference in the lung function domain between sexes with women having lower scores, which does not comply with the mean FEV_1 values obtained, as female patients had higher FEV_1 mean values than men. Several studies have shown that women perceive their health condition as worse than men do, which may serve as an explanation for our findings. Gee et al., as well as Isabella Uchmanowicz, have also found differences between men's and women's quality of life, and concluded that women consider their quality of life inferior and judge their health condition more accurately (21, 22).

Transplantation has a powerful effect on patients' quality of life. As expected, the transplant patients scored higher in the quality of life questionnaire than non-transplant patients in almost all of the domains, and significant differences were found in six of them. However, they tend to have lower scores in the weight and body image domain. This means that transplant CF patients have poorer body image and see the need to gain weight as a major problem. All in all, while patients' quality of life does improve after the transplantation, it is not the case for the domains related to the nutritional status.

BMI and body fat percentage correlated with six domains of CFQ-R: physical functioning, body image, eating, health perception, role fulfilment and weight. We have also found a significant difference between the different BMI groups in the vitality domain. Patients with lower BMI had lower quality of life scores than their peers, which is in accordance with the research of Gee et al., in which low BMI correlated with the negative body image perception (23). No statistically significant correlations were found for the muscle body mass percentage in our study.

FEV_1 shown a statistically significant correlation with six of the CFQ-R domains: physical functioning, treatment burden, health perception, respiratory symptoms,

role fulfilment and vitality. Patients with lowest FEV_1 values had lowest quality of life in all of the domains, except for the digestive symptoms domain. Similarly, several foreign studies have demonstrated a moderate correlation between lung function and physical functioning, as well as the respiratory symptoms domain (11, 19, 23-27). This correlation can be detected not only with CFQ-R, since such a correlation between physical functioning and lung function was also established when using another quality of life questionnaire (CFQoL) (28, 29).

CONCLUSIONS

Our research is the first disease-specific quality of life assessment among Hungarian adult patients with CF. In Hungary, we are the first to study correlations between nutritional status data and the quality of life in adult CF patients.

Female patients had worse nutritional status parameters than men. Transplantation did not affect the nutritional status results of our patients.

Transplant patients had decisively better quality of life than their non-transplant peers, except for the body image and weight domains. CF patients with lower BMI had lower quality of life than their peers.

In conclusion, in spite of the fact that patients did not have eating problems and they did not have to force themselves to eat, they still had difficulties gaining weight. This suggests that the patients may not know what kind of diet is appropriate for them. Therefore, early dietetical interventions should be implemented to improve patients' condition and quality of life.

The research on the quality of life is necessary to understand the subjective side of the disease. In the future, longitudinal studies are required to provide a more accurate picture of the dynamics of the constantly changing quality of life, as well as to determine the efficacy of the interventions.

References

1. Csecsemő és Gyermekgyógyászati Szakmai Kollégium. (2013). A cystás fibrosis (Az Egészségügyi Minisztérium szakmai protokollja). http://www.pharmindex-online.hu/diagnosztika-es-terapia/terapias-iranyelvek/a-cystas-fibrosis-1893.html?print=1. 2. Hornyak A, Marsal G: CF register (CF regiszter. Betegregiszter elemzése a 2010-es adatok alapján). CF munkacsoport értekezlet. Budapest 2011. 3. Gordon CM, Anderson EJ, Herlyn K et al.: Nutrient Status of Adults with Cystic Fibrosis. Journal of the American Dietetic Association 2007; 107(12): 2114-2119. DOI: 10.1016/j.jada.2007.09.00. 4. Hollander FM, De Roos NM, De Vries JHM, Van Berkhout FT: Assessment of Nutritional Status in Adult Patients with Cystic Fibrosis: Whole-Body Bioimpedance vs Body Mass Index, Skinfolds, and Leg-to-Leg Bioimpedance. Journal of the American Dietetic Association 2005; 105(4): 549-555. DOI: 10.1016/j.jada.2005.01.03. 5. Abbott J, Morton AM, Musson H et al.: Nutritional status, perceived body image and eating behaviours in adults with cystic fibrosis. Clinical Nutrition 2007; 26: 91-99. DOI: 10.1016/j.clnu.2006.08.002. 6. Richardson I, Nyulasi I, Cameron K et al.: Nutritional Status of an Adult Cystic Fibrosis Population. Nutrition 2000; 16: 255-259. 7. Stallings VA, Stark LJ, Robinson KA et al.: Evidence-Based Practice Recommendations for Nutrition-Related Management of Children and Adults with Cystic Fibrosis and Pancreatic Insufficiency: Results of a Systematic Review. Journal of the American Dietetic Association 2008; 108(5): 832-839. DOI: 10.1016/j.jada.2008.02.020. 8. Schöni MH, Casaulta-Aebischer C: Nutrition and lung function in cystic fibrosis patients: review. Clinical Nutrition 2000; 19(2): 79-85. DOI: 10.1054/clnu.1999.0080. 9. Bell SC, Bowerman AR, Davies CA et al.: Nutrition in adults with cystic fibrosis. Clinical Nutrition 1998; 17(5): 211-212. 10. Kaló Z, Péntek M: Az életminőség mérése. [In:] Gulácsi L (szerk.): Egészség-gazdaságtan. Medicina, Budapest 2005: 161-189. 11. Quittner AL, Buu A, Messer MA et al.: Development and validation of cystic fibrosis questionnaire in the United States: A health-related quality of life measure for cystic fibrosis. Chest 2005; 128: 2347-2354. 12. Quittner AL: Measurement of quality of life in cystic fibrosis. Current Opinion in Pulmonary Medicine 1998; 4(6): 326-331. 13. Polgar S, Thomas S: Correlation. [In:] Polgar S, Thomas S (szerk.): Introduction to research in the health sciences. Churchill Livingstone, Melbourne 1995: 240-242. 14. Gallagher D, Heymsfield SB, Heo M et al.: Healthy percentage body fat ranges: an approach for developing guidelines based on body mass index. American Journal of Clinical Nutrition 2000; 72(3): 694-701. 15. http://www.omron-healthcare.com/data/catalog/3/112/1/IM-HBF-510-E-04-08-08-EN.pdf (access: 2015.07.21). 16. http://www.ttuhsc.edu/som/internalmedicine/pulm/education/pfts/pfts_normal_values.pdf?ref=binfind.com/web (access: 2015.07.21). 17. King S, Sekuloska Z, Gallo R et al.: Changes in the Nutritional Status of Victorian Adults with Cystic Fibrosis since 1997: Improvements in Males but not Females. AuSpen Conference, Sydney 2013. 18. Hollander FM, Van Pierre DD, De Roos NM et al.: Effects of nutritional status and dietetic interventions on survival in Cystic Fibrosis patients before and after lung transplantation. Journal of Cystic Fibrosis 2013, Advance online publication. DOI: 10.1016/j.jcf.2013.08.009. 19. Riekert KA, Bartlett SJ, Boyle MP et al.: The association between depression, lung function, and health-related quality of life among adults with cystic fibrosis. Chest 2007; 132(1): 231-237. 20. Ashish A, Shaw M, McShane J et al.: Health-related quality of life in Cystic Fibrosis patients infected with transmissible *Pseudomonas aeruginosa* strains: cohort study. JRSM Short Rep 2012; 3(2): 12. 21. Gee L, Abbott J, Hart A et al.: Association between clinical variables and quality of life in adults with cystic fibrosis. J Cyst Fibrosis 2005; 4(1): 59-66. DOI: 10.1016/j.jcf.2004.12.005. 22. Uchmanowicz I, Jankowska-Polanska B, Rosinczuk J, Wleklik M: Health-Related Quality of Life of Patients Suffering from Cystic Fibrosis. Adv Clin Exp Med 2015; 24(1): 147-152. DOI: 10.17219/acem/38147. 23. Gee L, Abbott J, Conway SP et al.: Quality of life in cystic fibrosis: the impact of gender, general health perceptions and disease severity. J Cyst Fibrosis 2003; 2: 206-213. 24. Sawicki GS, Rasouliyan L, McMullen AH et al.: Longitudinal assessment of health-related quality of life in an observational cohort of patients with cystic fibrosis. Pediatr Pulmonol 2011; 1: 36-44. 25. Quittner AL, Sawicki GS, McMullen A et al.: Psychometric evaluation of the Cystic Fibrosis Questionnaire-Revised in a national sample. Qual Life Res 2012; 21: 1267-1278. 26. Wenninger K, Aussage P, Wahn U, Staab D: The Revised German Cystic Fibrosis Questionnaire: Validation of a disease-specific health-related quality of life instrument. Qual Life Res 2003; 12: 77-85. 27. de Jong W, Kaptein AA, van der Schans CP et al.: Quality of life in patients with cystic fibrosis. Pediatr Pulmonol 1997; 23(2): 95-100. 28. Thomas C, Mitchell P, O'Rourke P, Wainwright C: Quality of life in children and adolescents with cystic fibrosis managed in both regional outreach and cystic fibrosis center settings in Queensland. J Pediatr 2006; 148(4): 508-516. DOI: 10.1016/j.jpeds.2005.11.040. 29. Abbott J, Hart A, Havermans T et al.: Measuring health-related quality of life in clinical trials in cystic fibrosis. J Cyst Fibros 2011; 10(2): 82-85.

Correspondence to:
*Tímea Tóth
Department of Dietetics and Nutrition Sciences Semmelweis University
17 Vas ut., H-1088, Budapest, Hungary
tel.: +36-30-214-9750
fax: +36-1-486-4830
e-mail: tothtimea@se-etk.hu