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LIFE QUALITY OF ADULT PATIENTS WITH CYSTIC FIBROSIS

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ABSTRACT

Introduction: Cystic fibrosis is a genetic, multi-system chronic disease. The disease is caused by mutations in the *CFTR* gene. The incidence of the disease in Poland is estimated at 1 per 4400 births. In Poland in 2019, 1231 patients suffering from cystic fibrosis were registered.
Objective of the study: Analysis of patients' quality of life suffering from cystic fibrosis.
Material and methods: This cross-sectional study was conducted in a pulmonary ward between August and December 2022. The study group consisted of 52 adult patients suffering from cystic fibrosis. The mean age of the patients was 29.25 years (SD = 8.43). Twenty-three respondents received causal treatment. To collect the research material, a standardized Cystic Fibrosis Quality of Life (CFQoL) questionnaire was used, which covered number of domains: physical activity, social activity, treatment-related causes, respiratory system symptoms, emotional wellbeing, concerns for the future, interpersonal relations, body image, concerns about work/study, and personal data from a questionnaire filled in by the subject.
Results: The study showed that, in total, patients obtained an average of 51.24 points in the CFQoL questionnaire (SD = 20.18). Low quality of life was demonstrated in 23 patients. In the assessment of the quality of life, patients rated physical functioning the highest, and fears about the future and fears about work/study the lowest. There was no significant relationship between gender (p = 0.449), age (p = 0.362), education (p = 0.905), marital status (p = 0.084), professional status (p = 0.096), and the area of residence of the respondents (p = 0.122) and their overall quality of life. A significant relationship was found between the overall quality of life and the use of oxygen and causative drugs in patients' treatment.
Conclusions: Nearly half of patients with cystic fibrosis assess their quality of life as low. Patients who take causative drugs have a better quality of life. More frequent hospitalizations reduce the quality of life of patients.
Key words: quality of life, causal treatment, cystic fibrosis, CFQoL questionnaire.

INTRODUCTION

Cystic fibrosis is a chronic disease that impairs the function of many organs in the human body. It is a genetic disease that is inherited in an autosomal recessive manner [1]. This disease is caused by mutations in the *CFTR* (cystic fibrosis transmembrane conductance regulator) gene, which encodes the CFTR protein, which is a chloride channel located in cell membranes [2]. According to the Register of the European Society of Cystic Fibrosis, 1231 patients suffering from cystic fibrosis were registered (368 adults) in Poland in 2019 [3].

Cystic fibrosis is a life-shortening disease, leading to frequent infections and progressive systems and organs failure, including the lungs and pancreas. It is manifested by the retention of mucus in the respiratory tract and recurrent infections of the respiratory system. It is often accompanied by meconium obstruction in the neonatal period, deficiencies of vitamins A, D, E, and K, and the presence of diabetes and fertility disorders. The patient is subjected to time-consuming treatment regimens and frequent hospitalizations [4, 5].

Determining the level of quality of life among chronically ill people gives a chance to improve treatment or care. Living with a permanently accompanying disease, and what is more, with a terminal disease, particularly affects the mental state of patients, and consequently reduces their quality of life [6]. The meta-analysis of the data shows that the overall prevalence of anxiety and depression in cystic fibrosis patients is 26.22% and 14.13%, respectively [7]. Mental health variables were significantly associated with health-related quality of life in people with cystic fibrosis in a study by Cronly *et al.* [8]. Patients suffering from cystic fibrosis must face daily procedures supporting their respiratory system, such as physiotherapy and inhalations repeated several times a day. In the ad-

vanced stage of the disease, due to a decrease in lung function, they often do not have the strength to perform basic activities of everyday life, such as bathing, dressing, or preparing meals [9]. Recurrent infectious exacerbations of the respiratory system and frequent hospitalization exclude them from both social and professional life. The constantly accompanying symptoms of the disease effectively reduce their satisfaction with life [10]. The assessment of the quality of life in various aspects of functioning and the determination of their disorders allows us to provide patients with comprehensive, holistic care. An important element of assessing the functioning of chronically ill people is paying attention to their quality of life. Quality of life is defined by the World Health Organization (WHO) as the subjective perception of one's position by a human being in relation to the population, in which he lives, its ideas, expectations, and standards of life [11]. The most commonly used in medicine – health-related quality of life (HRQOL) – refers to the definition of health defined as not only the absence of disease, but also mental, physical, and spiritual balance. When analysing health-related quality of life, 4 dimensions of the patient's functioning are taken into account: physical condition, emotional state, social functioning and socio-economic conditions, and bodily sensations [12]. Considering the socio-economic aspect, the patient and his family are burdened with significant costs of treatment and rehabilitation [8, 13, 14]. According to the data, the monthly cost of treatment and rehabilitation of a cystic fibrosis patient is up to PLN 4000 [15].

Until recently, according to the recommendations developed by experts in 2017, treatment of cystic fibrosis consisted of reducing the symptoms resulting from the disease (symptomatic treatment) through the use of drugs that liquefy bronchial secretions, physiotherapy, supplementation of pancreatic enzymes and fat-soluble vitamins, and antibiotics [9]. There has been a breakthrough in therapy in recent years. Correctors and modulators of the CFTR protein have started to be used, which reduce the frequency of infectious exacerbations and improve spirometric indices [16]. In the opinion of scientists about the future of cystic fibrosis treatment in the next 15 years, therapies with CFTR modulators have the best chance of success [17]. In Poland, causal treatment has been refunded since 2020. From 1 March 2022 the refund covered a greater number of patients with cystic fibrosis [18].

This study is one of the first in which an attempt was made to assess the quality of life of patients with causal treatment.

The main aim of the study was to analyse the quality of life of adult patients suffering from cystic fibrosis. For the purposes of this paper, the following research questions were formulated:

1. What is the quality of life of adults with cystic fibrosis?
2. Is the quality of life of cystic fibrosis patients related to the social situation?
3. Is the quality of life of cystic fibrosis patients related to the number of hospitalizations?
4. Is the quality of life of cystic fibrosis patients related to the type of treatment used?

MATERIAL AND METHODS

The study included patients suffering from cystic fibrosis, hospitalized in one of the pulmonary wards in the period from August to December 2022. The consent of the facility management to conduct the study was obtained. The study was approved by the university bioethics committee No. KBE/285/2022.

Patients over 18 years of age were eligible for the study and gave their voluntary consent to fill in the questionnaire anonymously. The study was conducted in a group of 52 patients, 21 of whom were men and 31 women. The research tool was the CFQoL questionnaire, originally developed by Gee *et al.* [19], validated in a group of Polish patients by Dębska *et al.* [20]. The CFQoL questionnaire consists of 52 single-choice questions and consists of the following domains: physical functioning, social functioning, treatment issues, respiratory symptoms, emotional functioning, concerns about the future, interpersonal relationships, body image, and work/study concerns. Scores are calculated for each domain (range: 0-100 points) and then converted to a global score (range: 0-100 points). Getting less than 50 points for each domain and overall indicates a poor quality of life. Higher scores indicate better health-related quality of life.

In order to collect data on the patients' sociodemographic and clinical characteristics, an original questionnaire was developed, including the following: gender, age, education, area of residence, marital status, professional status, clinical form of cystic fibrosis, number of hospitalizations per year, current treatment, height, and body weight.

The χ^2 test was used to examine the statistical relationship between the analysed features. The obtained test results were subjected to statistical analysis using the χ^2 test for independent samples. In addition, Student's *t*-test for independent groups was used, which was used to verify the hypothesis about the equality of means of the examined variable in 2 populations. A 5% risk of inference error was assumed. A probability value $p < 0.05$ was considered statistically significant. The influence of one classification factor (divided into many levels) on the values of the measured measurable feature was tested by analysis of variance (ANOVA). The level of linear dependence between random variables was assessed using the Pearson correlation coefficient. The value of the correlation coefficient is in the closed range (–1, 1).

The greater its absolute value, the stronger the linear relationship between the variables. All calculations were performed using the SPSS Statistics 25.0 statistical package.

Table 1. Characteristics of the study group, sociodemographic and clinical variables (*N* = 52)

Sociodemographic/clinical variable	*n*
Gender	
Woman	31
Man	21
Education	
Primary/middle school	2
Vocational/sectoral	1
Secondary education/secondary vocational	31
Bachelor's/master's degree	18
Residence area	
City < 100,000 residents	8
City ≥ 100,000-500,000 inhabitants	12
City > 500,000 inhabitants	22
Village	10
Marital status	
Single	29
In relationship	23
Professional status	
Unemployed	29
Employed	23
Cystic fibrosis form*	
Abdominal	2
Mixed	35
Pulmonary	14
Treatment applied	
Inhalations	48
Antibiotic therapy	30
Rehabilitation	41
Oxygen therapy	7
Mucolytic drugs	34
Anti-inflammatory drugs	19
Anti-fungal drugs	4
Causal drugs	23
Lung transplantation	4
Vitamin supplements	38
Nutritional treatment – diet	33
Body weight	
Overweight	2
Underweight	10
Correct	38
Emaciation	1
Starvation	1

** Value does not add up to N = 52, 1 patient did not respond*

RESULTS

The study involved 31 women and 21 men. The age of the patients ranged from 18 to 57 years. The mean age of the patients was 29.25 years, SD = 8.43, median = 27. Thirty-one patients had secondary education, 18 patients had higher education, 42 patients lived in the city, and 10 patients lived in the country. Twenty-three patients were married or in a civil partnership, 23 patients were professionally active, and 16 were retired (Table 1).

The mixed form of the disease occurred in 35 patients and the pulmonary form in 14. The average number of hospitalizations per year in the study group was 2.06 (SD = 1.67). Inhalations were being used in 48 patients, rehabilitation in 41, vitamin supplementation in 38, mucolytic treatment in 34, and nutritional treatment in 33 patients. Twenty-three respondents received causal treatment (Table 1).

Good quality of life was demonstrated in 29 patients and poor in 23. The surveyed patients rated the highest quality of life in the domain of physical functioning M = 68.42, SD = 26.53, and the lowest in the domain of fear for the future M = 45.13, SD = 26.65 and job/study concerns M = 46.92, SD = 27.26 (Table 2).

Statistical analysis showed no significant relationship between the sex, age, education, and area of residence of the respondents and their quality of life. A relationship was found between the quality of life in the domain body image and the marital status of the respondents, which was on the verge of statistical significance (p = 0.052). In unmarried patients, the quality of life in this domain was lower. A significant relationship was found between the professional status of patients and their quality of life in the domain of fear of work/study (p = 0.001). In people who did not work, the quality of life in this domain was lower (Table 3).

The data show that the greater the number of hospitalizations reported by patients during the

Table 2. Characteristics of the study group, sociodemographic and clinical variables (*N* = 52)

Quality of life domains	M	SD	Me
Physical functioning	68.42	26.53	76.00
Functioning in society	63.56	27.66	72.50
Treatment issues	56.67	28.24	56.67
Respiratory symptoms	63.56	31.91	65.00
Emotional functioning	63.75	28.42	68.75
Concerns about the future	45.13	26.65	43.33
Interpersonal relations.	50.42	25.35	52.00
Body image	56.28	29.56	53.33
Work/study concerns	46.92	27.26	47.50
Total	51.34	20.18	51.00

M – mean, SD – standard deviation, Me – median

Table 3. Quality of life of cystic fibrosis patients depending on marital and professional status

Quality of life domains	Marital status										Professional status									
	Single			In relationship			Total				Non-working patients			Working patients			Total			
	M	*n*	SD	M	*n*	SD	M	*N*	SD	*t*	*p*	M	*n*	SD	M	*n*	SD	M	*N*	SD	*t*	*p*
Physical functioning	69.0	29	29.7	67.7	23	22.6	68.4	52	26.5	0.034	0.854	62.8	29	29.8	75.6	23	20.2	68.4	52	26.5	3.114	0.084
Functioning in society	63.1	29	32.8	64.1	23	20.0	63.6	52	27.7	0.017	0.896	62.9	29	31.2	64.3	23	23.1	63.6	52	27.7	0.033	0.857
Treatment issues	58.6	29	30.4	54.2	23	25.7	56.7	52	28.2	0.310	0.580	53.1	29	32.0	61.2	23	22.4	56.7	52	28.2	1.045	0.312
Respiratory symptoms	65.0	29	32.3	61.7	23	32.1	63.6	52	31.9	0.132	0.718	57.1	29	35.6	71.7	23	25.0	63.6	52	31.9	2.808	0.100
Emotional functioning	65.8	29	30.5	61.2	23	26.0	63.8	52	28.4	0.329	0.569	60.1	29	30.0	68.4	23	26.2	63.8	52	28.4	1.092	0.301
Future concerns	43.8	29	26.7	46.8	23	27.1	45.1	52	26.7	0.162	0.689	43.1	29	29.7	47.7	23	22.6	45.1	52	26.7	0.374	0.544
Interpersonal relations	46.6	29	25.9	55.2	23	24.4	50.4	52	25.4	1.489	0.228	44.6	29	28.5	57.8	23	18.8	50.4	52	25.4	3.703	0.060
Body image	48.5	29	29.9	66.1	23	267	56.3	52	29.6	4.882	0.052	51.7	29	29.6	62.0	23	29.2	56.3	52	29.6	1.576	0.215
Work/study concerns	42.8	29	26.6	52.2	23	27.7	46.9	52	27.3	1.546	0.219	36.6	29	25.7	60.0	23	23.7	46.9	52	27.3	1.428	0.001

N – number of observations, n – number of respondents, M – mean, SD – standard deviation, p – level of statistical significance, F – ANOVA test result, t – Student's t-test result

Table 4. Patients' quality of life of depending on the number of hospitalizations per year ($N = 49$)

Quality of life domains		Hospitalization number
Physical functioning	Pearson correlation	–0.546
	p	0.001
Functioning in society	Pearson correlation	–0.532
	p	0.001
Treatment issues	Pearson correlation	–0.515
	p	0.001
Respiratory symptoms	Pearson correlation	–0.375
	p	0.008
Emotional functioning	Pearson correlation	–0.413
	p	0.003
Concern about future	Pearson correlation	–0.304
	p	0.034
Interpersonal relations	Pearson correlation	–0.500
	p	0.001
Body image	Pearson correlation	–0.462
	p	0.001
Work/study concerns	Pearson correlation	–0.496
	p	0.001
General	Pearson correlation	–0.585
	p	0.001

p – level of statistical significance

year, the lower the quality of life they showed overall ($p = 0.001$) and in individual domains ($p \geq 0.005$) (Table 4).

Due to the clinical form of cystic fibrosis, the statistical analysis did not show a significant relationship between this variable and the quality of life of the respondents. A significant relationship was found between the use of oxygen therapy in patients' treatment and their overall quality of life ($p = 0.003$) and in 8 out of 9 domains. Patients treated with oxygen showed a lower quality of life overall. A significant relationship was found between the use of causative drugs by patients and their quality of life in general ($p = 0.001$) and in individual domains. No significant relationship was found only in the body image domain ($p = 0.154$) (Table 5).

Statistical analysis showed no significant relationship between the age, sex, education, professional, and marital status of the respondents and the applied

Table 5. Quality of life of cystic fibrosis patients depending on marital and professional status

Quality of life domains	Oxygen therapy						t	p	Causal treatment						t	p
	Yes (n = 7)		No (n = 43)		General (N = 50)				Yes (n = 23)		No (n = 28)		General(N = 51)			
	M	SD	M	SD	M	SD			M	SD	M	SD	M	SD		
Physical functioning	41.7	25.5	72.4	24.9	68.1	26.9	9.081	0.004	85.0	16.4	54.2	25.6	68.1	26.7	24.743	0.001
Functioning in society	35.0	29.6	68.0	24.7	63.4	27.7	10.196	0.002	77.8	17.8	50.7	28.4	62.9	27.6	15.832	0.001
Treatment issues	36.2	23.7	60.6	27.9	57.2	28.5	4.771	0.034	72.5	22.4	44.8	26.5	57.3	28.2	15.796	0.001
Respiratory symptoms	39.3	28.8	66.2	31.1	62.4	32.0	4.566	0.038	83.3	20.9	46.1	29.5	62.8	31.8	25.855	0.001
Emotional functioning	50.0	24.7	65.8	29.1	63.6	28.9	1.837	0.182	78.3	25.3	51.3	25.6	63.4	28.6	14.235	0.001
Concerns about the future	22.9	17.6	48.7	26.9	45.1	27.2	5.987	0.018	62.2	21.3	31.2	22.9	45.2	26.9	24.639	0.001
Interpersonal relations	30.9	13.9	54.3	25.5	51.0	25.5	5.581	0.022	60.6	22.4	43.1	25.0	51.0	25.2	6.745	0.012
Body image	29.5	16.7	61.7	29.1	57.2	29.8	8.055	0.007	63.2	29.0	51.2	29.8	56.6	29.8	2.097	0.154
Work/study concerns	24.3	19.5	50.2	27.2	46.6	27.6	5.847	0.019	59.1	25.3	37.1	25.6	47.1	27.5	9.420	0.003
General	30.9	17.9	54.7	19.2	51.3	20.6	9.438	0.003	64.1	15.5	40.8	17.9	51.3	20.4	24.253	0.001

N – number of observations, n – number of respondents, M – mean, SD – standard deviation, p – level of statistical significance, t – Student's t-test result

Table 6. Sociodemographic variables and the use of causal treatment

Sociodemographic/clinical variable	Casual treatment		χ^2	p
	No	Yes		
Gender				
Woman	19	12	0.949	0.245
Man	10	11		
Education				
Primary/middle school	2	0	3.27	0.352
Vocational/sectoral	1	0		
Secondary education/ secondary vocational	15	16		
Bachelor's/master's degree	11	7		
Residence area				
City < 100,000 inhabitants	6	2	9.27	0.026
City ≥ 100,000-500,000 inhabitants	8	4		
City > 500,000 inhabitants	7	15		
Village	8	2		
Professional status				
Unemployed	3	3	3.27	0.351
Working	10	13		
Pensioner/pensioner	11	5		
Studying/learning	5	2		
Marital status				
Bachelor/maiden	15	13	0.853	0.653
Divorced/divorced/ separated	1	0		
In relationship	13	10		
Age				
Up to 30 years	16	15	0.547	0.761
31-40 years	10	6		
Over 41	3	2		

p – level of statistical significance

causal treatment. Over 66% of respondents living in cities of ≥ 100,000-500,000 residents were not treated causally. On the other hand, over 68% of respondents living in a city of over 100,000 inhabitants were treated causally (Table 6).

DISCUSSION

The aim of this study was to analyse the quality of life of patients suffering from cystic fibrosis and to compare the patients' quality of life depending on their current treatment. In this study, patients overall assessed their quality of life lower than patients with cystic fibrosis after lung transplantation and in a stable disease state, but higher than patients requiring chronic oxygen therapy qualified for lung transplantation [21]. The review of studies shows that patients after bilateral or single lung transplantation

experience a better quality of life compared to people before transplantation [22]. In the study by Uchmanowicz *et al.*, 30 patients who were in the final phase of hospitalization due to exacerbation of the disease, in total and in individual domains, assessed their quality of life within 50 points [23]. The obtained results confirm the significant differentiation of the quality of life depending on the current clinical condition of the patient participating in the study and the progression of the disease.

This study and the results of studies by other authors showed that the domain of quality of life rated highest by patients was the domain of physical functioning [10, 23-28]. The physical functioning domain in this study referred to the sense of energy, the assessment of physical and motor fitness allowing patients to go about their affairs and activities of daily living, moving within the home environment, the ability to participate in sports, and in activities that the patient likes [20]. Health-related quality of life was directly related to physical fitness and, to a lesser extent, to self-reported physical activity in the study by Hebestreit *et al*. [29]. Another domain of quality of life rated best by patients in this study and in the results of other studies was the domain of social functioning [10, 24-26]. It should be noted, however, that these results are in the range of 60 points. In the range of up to 100 points difficulties in establishing social relationships, the feeling of being different and isolated may be associated with the accompanying symptoms and the need to undergo therapeutic and rehabilitation treatments many times a day [9]. Patients also perceive difficulties and limitations in the sphere of sexual functioning. As life expectancy improves, this aspect becomes an essential element of care. The review of the literature shows that in this group of patients there is a great need for education on sexual and procreative life, and therapeutic teams are not well prepared to advise their patients [30].

This study shows that one of the domains rated the lowest by patients is the domain of body image [10, 25]. According to the patients, they are too short and too thin, which may be related to malnutrition, a side effect of treatment, and malabsorption of nutrients [31]. The literature indicates that male cystic fibrosis patients are less satisfied from their body image, they want to weigh more and be more muscular, while women are more satisfied, but they want to be slimmer, which may result from cultural stereotypes.

Research results indicate that patients with cystic fibrosis have the lowest quality of life in the domain of fear for the future [10, 23, 25, 27, 32]. Patients' concerns about their future are associated with uncertainty about the possibility or necessity of receiving a lung transplant, worry that they will not be able to have children, and that the disease will shorten the patients' lives [20].

In this study and the study by Dębska *et al.*, patients assessed their quality of life as low in the domain of fear of work/study [10]. Taking into account sociodemographic variables, this study showed a significant relationship between the professional status of patients and their quality of life in the fear of work/study domain. In people who did not work, the quality of life in this domain was lower. Advances in the treatment of cystic fibrosis have significantly increased the life expectancy of patients, facilitating healthier lives and participation in working life. In this study, 23 patients were economically active. It is estimated that among European patients who are unemployed and not looking for work (18-30%), more than half indicated cystic fibrosis as the reason for stopping work or inactivity [33]. Working cystic fibrosis patients showed a higher "treatment burden" in study [34]. This seems to be independent of the clinical condition and the treatment used. These patients reported lower quality of life in the domains of interpersonal relations [25]. Both our study and those conducted by other authors showed the highest scores in the emotional functioning domains [24-26].

When analysing the patients' quality of life in this study in correlation with age, no significant relationship was found. In the study with the division of patients into people aged ≤ 25 and > 25 years, patients in the younger group, outside the domain of fear for the future, assessed their quality of life better than in the older group [23]. The decrease in self-esteem of the quality of life can be explained by the deterioration of physical parameters with the progression of the disease with age [32]. In a study determining changes in the quality of life in adult patients with cystic fibrosis over time, it was shown that older age was associated with reduced physical functioning. Women had significantly lower scores in physical functioning, vitality, and respiratory symptoms and higher scores in body image and weight [35]. Comparing different age groups, it was shown that older patients reported statistically higher scores in the physical functioning domain than younger patients. In the domain of interpersonal relations, patients under 33 years of age reported a statistically significantly worse quality of life. In addition, the analysis showed that older patients scored higher in the domain of fears related to work/study [25].

This study showed no relationship between the level of education and the patients' quality of life. In the study by Stofa *et al.*, patients who received higher education reported statistically higher scores compared to patients with other levels of education. Similar results were recorded in the domain of social functioning [25].

This study, similarly to the study by Stofa *et al.*, did not confirm the relationship between gender and the overall patients' quality of life and the quality of life in individual domains [25]. In the study by Uchmanowicz *et al.*, women outside the domain of interpersonal re-

lations rated their quality of life lower than men. However, these differences were not statistically significant [23]. A meta-analysis by Habib *et al.* showed that gender is an important predictor of quality of life in cystic fibrosis patients. Women achieved a lower quality of life in the following areas: physical functioning, emotional functioning, social functioning, and respiratory symptoms [32]. Women perceived themselves more negatively in the domain of fears about the future, interpersonal relationships, and fears related to work/study than men in the study by Tomaszek *et al.* and Gee *et al.* [24, 36]. Dębska *et al.* also showed differences in the assessment of the quality of life between women and men, although a statistically significant difference occurred only in the subscale of physical functioning [26].

It seems that having a partner may be associated with successful treatment of cystic fibrosis. In our study, on the verge of statistical significance, unmarried patients had lower quality of life in the body image domain. Having a partner was significantly associated with higher quality of life scores in the "emotions" dimension, but with lower scores in the "gastrointestinal symptoms" dimension according to the Cystic Fibrosis Questionnaire-Revised (CFQ-R) in the study by Ancel *et al.* [13].

Our data show that the greater the number of hospitalizations reported by patients during the year, the lower the quality of life they showed in general and in individual domains. This seems to be of great importance in terms of causal treatment. The use of CFTR modulators reduces the frequency of bronchopulmonary exacerbations, which in turn reduces the frequency of hospitalization in these patients [37]. This type of treatment also improves the patient's nutritional status through more efficient absorption of nutrients from food, which increases the patients' body weight [16].

In this study, 48 patients used inhalation for treatment. The analysis of the literature showed that the use of tobramycin in inhalation can improve the patients' quality of life, especially in the domain of physical functioning [38]. Similar conclusions were reached by Smith *et al.*, who showed that inhaled antibiotics can improve lung function and reduce the number of infectious exacerbations of cystic fibrosis [39].

Our research shows that patients taking causative drugs have a better quality of life. Causal treatment in Poland has been introduced relatively recently. In the literature review, there are no studies conducted in terms of the quality of life of patients who use treatment with CFTR modulators; therefore, it is difficult to compare the results of this study with studies by other authors. Undoubtedly, this is a topic that should be subjected to further research.

In previous studies on the quality of life, scientists unequivocally claimed that cystic fibrosis patients' quality of life is low. It seems that causal treatment and reduction of the number of hospitalizations may contribute to the increase in satisfaction with life. It seems that there is a need for continuous monitoring of cystic fibrosis patients' quality of life along with the introduction of new, or improvement of existing, treatment methods.

CONCLUSIONS

Nearly half of cystic fibrosis patients assess their quality of life as low.

Unmarried patients assess their quality of life lower in the fear for the body domain, while patients who do not work assess the quality of life lower in the fear for work/study domain.

More frequent hospitalizations reduce the cystic fibrosis patients' quality of life.

Patients taking causative drugs have a better quality of life.

Disclosure

The authors declare no conflict of interest.

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