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Quality of life in patients with cystic fibrosis depending on the severity of the disease and method of its treatment

Ocena jakości życia chorych na mukowiscydozę z uwzględnieniem stopnia zaawansowania choroby i metod leczenia

Authors' Contribution:
A Study Design
B Data Collection
C Statistical Analysis
D Data Interpretation
E Manuscript Preparation
F Literature Search
G Funds Collection

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Summary

Introduction: Quality of life is a measure not only of disease severity and its functional impact (physical, psychological, and social functioning) but also the efficacy of novel therapies. The aim of this study was to analyze the quality of life in cystic fibrosis patients depending on the severity of the disease and methods of its treatment.

Material/Methods: The study included groups of cystic fibrosis patients: 1) after lung transplantation, 2) requiring chronic oxygen therapy, and 3) in stable clinical status. Forty-five men and women older than 18 years were enrolled. The participants were examined with the Polish version of the Cystic Fibrosis Quality of Life Questionnaire (CFQoL) adapted by Dębska & Mazurek.

Results: Patients from analyzed groups differed significantly in terms of their quality of life in most of the subscales included in CFQoL, but not in *Future Concerns* and *Interpersonal Relationships*.

Discussion: Although lung transplantation markedly improves the quality of life of patients with cystic fibrosis, they still experience problems with social functioning and future concerns.

Keywords: **cystic fibrosis • lung transplantation • quality of life**

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Abbreviations: **BI** – Body Image, **CC** – Career Concerns, **CFQoL** – Cystic Fibrosis Quality of Life Questionnaire, **CS** – Chest Symptoms, **EF** – Emotional Functioning, **FC** – Future Concerns, **IR** – Interpersonal Relationships, **ISHLT** – International Society for Heart and Lung Transplantation, **PF** – Physical Functioning, **SF** – Social Functioning, **TI** – Treatment Issues.

Introduction

Quality of life is a measure not only of disease severity and its functional impact (physical, psychological, and social functioning) but also the efficacy of novel therapies. The results of clinical observations suggest that lung transplantation is the only treatment modality which can improve the survival in patients with advanced cystic fibrosis [7,9]. In view of longer survival, the issues of improved physical, social, and psycho-emotional functioning become of vital importance.

A total of 67 single lung transplantations and 602 bilateral transplantations were carried out in the Eurotransplant member countries in 2012. The number of transplants was highest in Germany, Belgium, and Austria [18]. Every year, approximately 1,600 single or bilateral lung transplantations are carried out worldwide. Overall, 1.3 thousand lung transplantations were performed in individuals under 18 years of age during the last 15 years, most often between 12 and 17 years of age [10,20].

Lung transplantation in cystic fibrosis patients has been performed successfully for more than 20 years. According to the data gathered by the International Society for Heart and Lung Transplantation (ISHLT), cystic fibrosis is the third most frequent cause of lung transplantation in adults, which is reflected by constant improvement of survival in patients with this condition [21].

Unfortunately, only a few transplantations are performed every year in the majority of centers, which, in the experts' opinion, is reflected by an increased risk of mortality within one year after the procedure [10,12,15,20,21]. However, this situation is changing on a yearly basis, as dynamic progress in transplantology and the resultant increase in experience of relevant specialists leads to a decrease in perioperative mortality [2].

However, aside from the higher number of transplantations and decreased risk of perioperative morbidity, improved quality of life is also a prerequisite of therapeutic success in patients with cystic fibrosis, irrespective of their age, severity of the disease, and type of its treatment. Assessment of the quality of life in various functional areas and identification of potential dysfunction in role functioning will allow holistic individualized care to be offered to patients with cystic fibrosis. Only the situation in which a patient has a possibility of self-realization without the stigma of the disease and related risks can be considered complete therapeutic success.

The aim of this study was to analyze the quality of life in cystic fibrosis patients depending on the severity of the disease and methods of its treatment.

Materials and methods

The study was conducted at the Department of Pneumonology and Cystic Fibrosis of the Tuberculosis and Lung Diseases Institute in Rabka-Zdroj. Enrollment in the study was voluntary, and all the participants gave their written informed consent to participate in the project.

The study included patients with various stages of cystic fibrosis. Forty-five men and women older than 18 years were enrolled, and divided into three subgroups designated as A, B, and C. Subgroup A included 10 patients after lung transplantation. The time elapsed after the transplantation ranged from three months to three years and one month; three patients underwent transplantation within one year prior to the study. Subgroup B (n=15) comprised patients in poor clinical status, receiving chronic oxygen therapy and qualified for lung transplantation. Subgroup C (n=20) included patients in stable clinical status. The lack of significant limitations in physical activity and no necessity of oxygen therapy constituted inclusion criteria for this subgroup.

We used the Polish version of the Cystic Fibrosis Quality of Life Questionnaire (CFQoL), developed by Gee et al. [8] and adapted by Dębska & Mazurek [6]. The questionnaire comprises 54 questions grouped in the subscales of *Physical Functioning* (PF), *Social Functioning* (SF), *Treatment Issues* (TI), *Chest Symptoms* (CS), *Emotional Functioning* (EF), *Future Concerns* (FC), *Interpersonal Relationships* (IR), *Body Image* (BI), and *Career Concerns* (CC).

The results of each subscale were transformed into numeric values ranging between 0 and 100 points (with 100 points corresponding to the optimal quality of life). Answers to individual questions were expressed on a 6-item scale, where 1 corresponded to the lowest, and 6 to the highest score (except for question no. 6, which was scored in a reverse order). Raw scores of individual questions equal to or lower than 3 points and transformed score ≤50 points corresponded to poor quality of life.

The characteristics of the quality of life in various functional domains were presented as mean values and their standard deviations, medians, and interquartile ranges. Normal distribution of the analyzed parameters was verified with the Shapiro-Wilk test. The Kruskal-Wallis test and Dunn post-hoc test were used for comparison of the scores of three studied subgroups. Statistical significance of the tests was set at $p \leq 0.05$. The statistical analysis was conducted with Statistica 6.0 software (StatSoft, Tulsa, OK, United States).

Results

the average transformed scores of most subscales in patients after lung transplantation (subgroup A) exceeded 50. The global quality of life score in this group amounted to x=73.9±7.9. Participants from group A reported the highest quality of life (the highest average scores) in *Chest Symptoms* (x=95±10.1), *Treatment Issues* (x=95.3±8.9), and *Emotional Functioning* subscales (x=87.3±12.8). Also the quality of life in *Physical Functioning* and *Social Functioning* subscales was high, amounting to x=82.4±26.1 and 73.0±18.4, respectively.

In contrast, the lowest quality of life scores were documented in the case of *Career Concerns* (50.0±21.1), *Future Concerns* (56.0±11.8), *Body Image* (69.3±17.8), and *Interpersonal Relationships* (Table 1).

Both the average transformed global quality of life score and the average transformed scores of most subscales determined in the subgroup of patients awaiting lung transplantation (subgroup B) were lower than 50 points, pointing to low quality of life in this group. The only exceptions pertained to *Emotional Functioning* and *Future Concerns* subscales, the average scores of which were 55.0±17.6 and 50.0 ±20.6 points, respectively. The average global quality of life score equaled 42.6 ±12.42 points (Table 1).

The distribution of the average transformed quality of life scores in patients in stable clinical status who did not require oxygen therapy (subgroup C) was variable. The global quality of life score amounted to x=70.8±15.7 points. The highest average scores were documented in the case of *Physical Functioning* (x=84.2±15.46) and *Social Functioning* subscales (x=81.1±16.96). The average scores of the remaining subscales – *Chest Symptoms*, *Emotional Functioning*, *Treatment Issues*, *Career Concerns*, *Interpersonal Relationships*, and *Body Image* – ranged from x=76.61±5.7 to x=63.2±23.5. The lowest average score, being at the threshold of low quality of life, was recorded in the case of the *Future Concerns* subscale (x=51.9±19.2; Table 1).

The average scores of most subscales in the group of patients awaiting the transplantation (subgroup B) ranged between 32±15.4 and 55.0±17.6 points, and were significantly lower than in individuals after the transplantation (subgroup A, from 50.0±20.1 to 95.5±10.1 points) or those in stable clinical status (subgroup C, from 51.9±19.2 to 84.2±15.5 points). Only in the case of two subscales, *Future Concerns* and *Interpersonal Relationships*, did the intergroup differences prove insignificant ($p>0.05$). Finally, the average quality of life score in the *Treatment Issues* subscale was significantly higher in patients from subgroup A than in the participants from subgroups C and B (95.5±10.12 vs. 43.7±19.04 vs. 76.6±15.73 points, $p<0.001$; Table 2).

Discussion

our analysis of the quality of life in patients with various stages of cystic fibrosis unambiguously confirmed that patient's status constitutes the determinant of functioning in many areas. Our study confirmed good functioning of patients after transplantation in physical, social, and emotional spheres. Our participants were not bothered by questions related to treatment or the presence of respiratory symptoms. In contrast, they reported low quality of life regarding future concerns, work and education, and body image. In turn, the patients in the worst clinical status who were qualified for the transplantation presented the lowest quality of life in all

Table 1. Statistical characteristics of the quality of life scores (means ± standard deviations) in patients after lung transplantation (subgroup A), awaiting lung transplantation (subgroup B), and at a stable stage of the disease (subgroup C)

Parameter	Subgroup A (n=10)	Subgroup B (n=15)	Subgroup C (n=20)
Physical Functioning (PF)	90.6±8.5	34.1±22.0	84.2±15.5
Social Functioning (SF)	73.0±18.4	34.8±22.5	81.1±17.0
Treatment Issues (TI)	95.3±8.9	39.8±19.4	68.1±20.1
Chest Symptoms (CS)	95.5±10.1	43.7±19.0	76.6±15.7
Emotional Functioning (EF)	87.3±12.8	55.0±17.6	77.6±19.6
Future Concerns (FC)	56.0±11.8	50.0±20.6	51.9±19.2
Interpersonal Relationships (IR)	60.6±13.2	47.5±14.6	63.2±23.5
Body Image (BI)	69.3±17.8	41.7±23.8	64.6±23.2
Career Concerns (CC)	55.0±10.0	32.0±15.4	65.3±25.8
Global quality of life	73.9±7.9	42.6±12.4	70.8±15.7

Table 2. Comparison of the quality of life scores (medians, interquartile ranges) in patients after lung transplantation (subgroup A), awaiting lung transplantation (subgroup B), and at a stable stage of the disease (subgroup C)

Parameter	Subgroup A (n=10)	Subgroup B (n=15)	Subgroup C (n=20)	p-value
Physical Functioning (PF)	91 (82.4-100)	28 (22-54)	88 (76-94)	<0.0011
Social Functioning (SF)	77.5 (60-85)	32.4 (15-50)	85 (70-95)	<0.0012
Treatment Issues (TI)	100 (93.3-100)	37.3 (20-60)	66.7 (60-80)	<0.0013
Chest Symptoms (CS)	100 (100-100)	45 (30-55)	80 (65-85)	<0.0014
Emotional Functioning (EF)	91.3 (75-100)	52.5 (40-70)	77.5 (62.5-97.5)	0.0015
Future Concerns (FC)	55 (46.7-66.7)	40 (36.7-66.7)	50 (36.7-70)	0.469
Interpersonal Relationships (IR)	65 (52-70)	44 (36-58)	66 (44-78)	0.057
Body Image (BI)	66.7 (53.5-93.3)	40 (20-53.3)	66.7 (40-80)	0.0086
Career Concerns (CC)	55 (50-65)	30 (20-50)	70 (40-85)	0.0017
Global quality of life	74.2 (67.3-80.8)	38.8 (35.4-48.8)	73.1 (52.3-80.4)	<0.0018

[1]Group A vs. B: p<0.001, Group A vs. C: p>0.999, Group B vs. C: p<0.001; [2]Group A vs. B: p=0.009, Group A vs. C: p>0.999, Group B vs. C: p<0.001; [3]Group A vs. B: p<0.001, Group A vs. C: p=0.020, Group B vs. C: p=0.012; [4]Group A vs. B: p<0.001, Group A vs. C: p=0.066, Group B vs. C: p=0.002; [5]Group A vs. B: p=0.001, Group A vs. C: p=0.638, Group B vs. C: p=0.012; [6]Group A vs. B: p=0.023, Group A vs. C: p>0.999, Group B vs. C: p=0.023; [7]Group A vs. B: p=0.024, Group A vs. C: p>0.999, Group B vs. C: p<0.001; [8]Group A vs. B: p<0.001, Group A vs. C: p>0.999, Group B vs. C: p<0.001

functional spheres. It should be noted, however, that undergoing transplantation does not release the patient from anxiety and life concerns. It cannot be excluded that the constant, negative emotional tension of the patient can become an additional factor modulating the duration of survival after transplantation.

Other authors pointed to health-related quality of life as a prognostic factor of survival [1,21]. Havermans et al. [11] identified anxiety and depression as the sources of worse social and emotional functioning and disturbed body image.

Noticeably, our study revealed that both the patients in stable clinical status and those subjected to transplantation experienced problems with body image and interpersonal relationships. It cannot be excluded that the disease, namely cystic fibrosis, stigmatizes a patient in his/her own opinion. Lack of acceptance of body image, and self-imposition of barriers in social relationships can markedly hinder establishment of partner relationships, and generate negative emotional states. According to Besier et al. [3], patients with cystic fibrosis who stay in partner relationships show greater satisfaction with life and health than those who lack a partner. Satisfaction with family, sexual, and professional life in the group of patients with cystic fibrosis can positively affect the therapeutic outcome, irrespective of the treatment method. Unfortunately, the number of reports dealing with the problem in question is sparse.

Moreover, the results of other studies suggest that patients with cystic fibrosis are satisfied with the quality of their life and health after lung transplantation. They show the best functioning in the physical sphere, but some deficits exist in the social and environmental domains. Health and family were the highest values for this group of patients. Optimistic look into the future enables them to verify the purpose and sense of life, while gaining satisfaction from overcoming the disease and achieving previously defined objectives [5,13,14,19].

However, this does not exclude the risk of adverse effects, especially when the purpose and sense of life are lost, and in the presence of negative emotions (anxiety, depressive behavior, depression). They can generate suicidal behaviors, as revealed in the previous study by Cepuch et al. [4].

However, one cannot disagree with the opinions presented by Quinter et al. [16] and Singer et al. [17], according to whom the validation of existing tools and implementation of research findings during the care of patients constitute a chance for future studies, and thus also for therapeutic success in a wide sense. Identification of specific determinants of the quality of life at each stage of cystic fibrosis and its treatment, especially in longitudinal studies, still seems an open area of research. Assessment of the other functional domains of the patients, not necessarily from the perspective of the physical health domain, deserves particular attention.

The data published by Dębska et al. [5] suggest that comparison between the results of various studies is difficult if the quality of life was determined with different instruments; in this study we used the WHOQOL-BREF questionnaire, adapted by Wołowicka and Jaracz [22].

Our study confirmed the conclusion that the quality of life assessment is an important component of the therapeutic process, especially when the treatment raises hopes for better life. Therefore, in this study we focu-

sed on the quality of life assessment in a small group of patients with cystic fibrosis who had an opportunity to undergo lung transplantation.

In conclusion, our study revealed that patients after lung transplantation positively assess their quality of life with regards to physical and social functioning, but experience problems with body image, interpersonal relationships, and future concerns, including work and education. Individuals qualified for the transplantation presented the lowest scores in all domains of quality of life.

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