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Izabella Uchmanowicz[1, 2], Beata Jankowska-Polańska[1, 2], Marta Wleklik[2], Joanna Rosinczuk-Tonderys[3], Grażyna Dębska[4]

[1]Department of Clinical Nursing, Faculty of Health Science, Wroclaw Medical University
Head: Prof. K. Łoboz-Grudzień, MD, PhD
[2]Students' Scientific Group at the Internal Nursing Department, Faculty of Health Sciences, Medical University of Wroclaw
Tutor: I. Uchmanowicz, B. Jankowska-Polańska, MD
[3]Department of Nervous System Diseases, The Faculty of Health Science, Wroclaw Medical University
Head: J. Rosinczuk, MD
[4]Podhale Public School of Higher Vocational Education Speciality Nursing in Nowy Targ
Head: G. Dębska, MD

Health-related quality of life of patients with cystic fibrosis assessed by the SF-36 questionnaire

Jakość życia pacjentów z mukowiscydozą mierzona kwestionariuszem SF-36

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Abstract

Introduction: Cystic fibrosis (CF) is a genetic disorder, which is most common among Caucasians. There are about 100,000 people suffering from this disorder in the world, including 25 000 in Europe. Although the first mention of cystic fibrosis is thought to have occurred in 1595, recognition of the entire clinical spectrum of CF and the resultant development of contemporary knowledge occurred in the 20th century. In the past, CF was considered a fatal childhood disorder; however, contemporary statistical data shows that 50% of people with cystic fibrosis have a chance to live up to 30 years of age, and the lifespan of children born in the 1990s is projected to be at least 40 years. Consequently, the number of adults with cystic fibrosis is increasing, making it necessary for multidisciplinary actions aimed at the improvement of clinical management of the condition as well as minimizing the influence of CF and its treatment on the quality of continually extending life of patients. Since cystic fibrosis interferes with almost all important aspects of human functioning, quality of life (QoL) of individuals with CF should be constantly and closely monitored, thus allowing for consideration of their needs and providing an opportunity to modify the therapeutic approach if necessary.
The aim of this study was to visualize the QoL of people with cystic fibrosis, to identify differences in their assessment of QoL depending on sex and age, as well as to compare the QoL of patients with CF with a control group of people without CF.
Material and methods: The study was conducted among patients hospitalized at the Clinic of Pulmonology and Cystic Fibrosis at the Institute of Tuberculosis and Lung Diseases in Rabka-Zdrój from February to April 2012. The study encompassed 30 patients (19 women and 11 men), aged between 16 and 42 years, with acute episodes of bronchopulmonary disease. A Short Form-36 (SF-36) questionnaire was used to evaluate the health-related quality of life. The control group encompassed 30 healthy individuals with the same sex ratio and similar age ratio as in the group of patients.
Results: The analysis of quality of life with the use of the SF-36 questionnaire showed that in general the quality of life of the assessed patients was low. The patients scored on average below 50 in the following subscales: Role Physical (RP), General Health (GP), Vitality (VT), Social Functioning (SF), Physical Component Summary (PCS) and Mental Component Summary (MCS). In other subscales the results slightly exceeded 50.
In women, low quality of life was found in the following subscales: Role Physical, Bodily Pain (BP), General Health (GH), Vitality (VT), Social Functioning (SF), Physical Component Summary (PCS) and Mental Component Summary (MCS). In men, average results below 50 were found in the following subscales: General Health (GH) and Vitality (VT). Moreover, in all subscales, the quality of life assessed by women was lower than that reported by men.

Address for correspondence: Izabella Uchmanowicz, RN, MSN, Medical University of Wroclaw, Poland, Faculty of Health Sciences, ul. K. Bartla 5, 51–618 Wrocław, tel.: +48 71 784 18 05, +48 71 341 95 33, fax: +48 71 343 20 86; email: izabella.uchmanowicz@umed.wroc.pl
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Patients > 25 years assessed their quality of life as lower in all subscales except for the Role Emotional (RE) subscale. Statistically significant differences were identified with regards to Vitality (VT) and Mental Health (MH).
The analysis of the influence of CF patients' age on their quality of life, conducted with the use of the SF-36 questionnaire, showed that there were statistically significant correlations ($p < 0.05$) between age and the following domains: Physical Functioning (PF), Role Physical (RP), Bodily Pain (BP), Vitality (VT), Social Functioning (SF), Mental Health (MH) and Physical Component Summary (PCS).
The comparison between the QoL of patients with CF and those of a control group of individuals not suffering from CF demonstrated that there were statistically significant differences in all subscales. Patients' quality of life was significantly poorer than quality of life of individuals without CF.
Conclusions:
1. Patients with CF generally perceive their quality of life as low.
2. There are differences in the assessment of quality of life between women and men.
3. The older the patients, the worse their quality of life assessment in most subscales.
4. There are critical differences in the quality of life assessment between patients with CF and a control group encompassing individuals without CF. Patients with CF find their quality of life poorer.

Key words: cystic fibrosis, health-related quality of life, survey

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Streszczenie

Wstęp: Mukowiscydoza (CF) jest chorobą genetyczną, występującą najczęściej u osób populacji kaukaskiej. Na świecie choruje około 100 tysiąca osób, z czego w Europie liczba chorych wynosi około 25 000. Choć historia wiedzy o mukowiscydozie sięga najprawdopodobniej 1595 roku, to poznanie choroby mające wpływ na rozwój współczesnej wiedzy o CF dotyczy dopiero XX wieku. Jeszcze kilka lat temu uważana była za śmiertelną chorobę wieku dziecięcego, jednak aktualne dane statystyczne wskazują, że 50% chorych na mukowiscydozę ma szansę dożycia do 30. roku życia, a długość życia dzieci urodzonych w latach 90. XX wieku ma wynosić co najmniej 40 lat. Obserwuje się w związku z tym wzrost liczby chorych dorosłych na mukowiscydozę. Konieczne staje się zatem podejmowanie interdyscyplinarnych działań mających na celu zarówno poprawę skuteczności działań medycznych, jak i zminimalizowanie wpływu choroby i jej leczenia na jakość wydłużającego się życia chorych. Ingerencja mukowiscydozy we wszystkie najważniejsze obszary funkcjonowania rodzi potrzebę ciągłego, kompleksowego monitorowania jakości życia, co pozwoliłoby zwrócić szczególną uwagę na ich potrzeby, a także w razie konieczności umożliwiłoby modyfikacje podejścia terapeutycznego.
Celem było zobrazowanie jakości życia chorych na mukowiscydozę, wykazanie różnic w samoocenie jakości życia w zależności od płci i wieku oraz porównanie jakości życia chorych z grupą kontrolną osób bez mukowiscydozy.
Materiał i metody: Badanie przeprowadzono wśród pacjentów hospitalizowanych w Klinice Pneumonologii i Mukowiscydozy Instytutu Gruźlicy i Chorób Płuc w Rabce-Zdroju w okresie od lutego do kwietnia 2012 roku. Objęto nim 30 pacjentów (19 kobiet i 11 mężczyzn) z zaostrzeniem choroby oskrzelowo-płucnej w wieku 16–42 lat. W badaniu wykorzystano kwestionariusz ogólny SF-36 (*Short Form 36*) do oceny jakości życia uwarunkowanej stanem zdrowia. Grupa kontrolna obejmowała 30 zdrowych osób w podobnym wieku oraz dopasowanych pod względem rozkładu płci.
Wyniki: Analiza jakości życia kwestionariuszem ogólnym SF-36 wykazała, że chorzy generalnie nisko oceniają swoją jakość życia. Średnie wyniki poniżej 50 uzyskali w podskalach dotyczących: ograniczenia aktywności z powodu zdrowia fizycznego (RP), ogólnej oceny stanu zdrowia (GH), witalności (VT), funkcjonowania społecznego (SF), poziomie aktywności fizycznej (PCS), poziomie aktywności umysłowej (MCS). W pozostałych podskalach średnie wyniki nieznacznie przekroczyły 50.
Wśród kobiet niską jakość życia odnotowano w podskalach: RP — ograniczenia aktywności z powodu zdrowia fizycznego, BP — ograniczenia bólowe, GH — ogólnej ocenie stanu zdrowia, VT — witalności, SF — funkcjonowanie społeczne, PCS — poziomie aktywności fizycznej, MCS — poziomie aktywności umysłowej. Wśród mężczyzn średnie wyniki poniżej 50 odnotowano w podskalach: GH — ogólna ocena stanu zdrowia i VT — witalność. Ponadto kobiety oceniły swoją jakość życia gorzej niż mężczyźni we wszystkich podskalach.
Chorzy powyżej 25 lat gorzej ocenili swoją jakość życia we wszystkich podskalach, oprócz podskali dotyczącej ograniczenia aktywności z powodu problemów emocjonalnych (RE). Wykazano istnienie różnic statystycznych w zakresie VT — witalności i MH — zdrowia psychicznego.
Analiza wpływu wieku na jakość życia chorych na mukowiscydozę badana kwestionariuszem SF-36 wykazała, że istnieją zależności istotne statystycznie (przy $p < 0,05$) między wiekiem a domenami: PF — funkcjonowanie fizyczne, RP — ograniczenia aktywności z powodu zdrowia fizycznego, BP — dolegliwości bólowe, VT — witalność, SF — funkcjonowanie społeczne, MH — zdrowie psychiczne, PCS — poziom aktywności fizycznej.
Porównując jakość życia chorych na mukowiscydozę z grupą kontrolną osób bez mukowiscydozy, wykazano istnienie różnic statystycznych we wszystkich podskalach. Jakość życia osób chorych jest w znacznym stopniu gorsza niż jakość życia osób bez mukowiscydozy.

Wnioski:
1. Chorzy na mukowiscydozę generalnie nisko oceniają swoją jakość życia.
2. Istnieją różnice w ocenie jakości życia pomiędzy kobietami a mężczyznami.
3. Im starszy chory, tym gorsza jego jakość życia w większości podskal.
4. Istnieją duże odstępstwa w ocenie jakości życia chorych na mukowiscydozę, a grupą kontrolną osób bez mukowiscydozy.
5. Chorzy oceniają gorzej swoją jakość życia.

Słowa kluczowe: mukowiscydoza, jakość życia uwarunkowana stanem zdrowia, kwestionariusz

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Introduction

Cystic fibrosis (CF) is a genetic disorder, which is most common among Caucasians. It is estimated that 3–5% of the population is a carrier of one mutated allele. There are about 100,000 people suffering from this disorder in the world, including about 25,000 in Europe. On average, one in every 3200 newborns born alive is found to be affected by CF [1]. Although the first mention of cystic fibrosis dates back most probably to 1595, when an autopsy conducted in Leiden on an 11-year-old girl suggested the occurrence of this disorder [2], recognition of the entire clinical spectrum of CF, influencing the development of contemporary knowledge, occurred in the 20th century. In 1938, a pathologist by the name of Dorothy Andersen introduced the term *Cystic Fibrosis of the pancreas* based on autopsy results. In 1945, the term *mucoviscidosis* was coined by Faber, who noticed changes in other exocrine glands as well. In 1959, pilocarpine iontophoresis was introduced (Gibson and Cook), which is, to date, a definitive and effective diagnostic test. At the end of the 1980s, the CFTR gene was located and its structure, together with its most common mutation (delF508), was identified [3, 4]. Despite significant progress in medicine and relevant research, and although the disease aetiology is known, CF remains incurable and the treatment does not address the cause. Nonetheless, since CF is an example of a chronic and systemic disease of progressive course, it requires multispecialty symptomatic treatment aimed at delaying the disease's progress mainly in respiratory and digestive systems, as well as preventing complications, which have a significant influence on the length and quality of patients' lives [3, 5].

In the past, CF was considered a fatal childhood disorder. Contemporary statistical data, however, shows that 50% of individuals with cystic fibrosis have a chance to live to 30 years of age, and the lifespan of children born in the 1990s is projected to be at least 40 years. Consequently, the number of adults with cystic fibrosis is increasing. In 1998, adults constituted 18% of all patients registered in the Polish Register of Cystic Fibrosis, while in 2010 they accounted for 32.3% of all patients. The oldest registered patient was 59 years old [6–8]. Thus, multidisciplinary actions aimed at both the improvement of clinical management of the disorder as well as minimizing the impact of CF and its treatment on the quality of continually extending life of patients is necessary [7, 9]. Since cystic fibrosis interferes with almost all important aspects of human functioning, quality of life (QoL) of individuals with CF should be constantly and closely monitored; thus allowing for consideration of their needs and providing an opportunity to modify the therapeutic approach if necessary [9].

The main aim of the study was to assess the quality of life of people with CF using the SF-36 general questionnaire. Specific goals included identifying differences in self-assessment of the quality of life depending on sex and age, as well as comparing quality of life of CF patients with a control group encompassing individuals without CF.

Materal and methods

The study was conducted among patients hospitalized at the Clinic of Pulmonology and Cystic Fibrosis at the Institute of Tuberculosis and Lung Diseases in Rabka-Zdrój from February to April 2012. The study encompassed 30 patients (19 women and 11 men) aged between 16 and 42 years with acute episodes of bronchopulmonary disease.

Participation in the study was voluntary. All individuals taking part in the study were informed about the study aims and were ensured that complete anonymity would be preserved.

The participants of the study were divided into groups depending on sex and age (≤ 25 years and > 25 years).

The SF-36 questionnaire was used in the study to assess health-related quality of life. This

questionnaire included 36 questions divided into 8 categories: Physical Functioning (PF), Role Physical (RF), Bodily Pain (BP), Social functioning (SF), Mental Health (MH), Role Emotional (RE), Vitality (VT) and General Health Perceptions (GH). Physical Component Summary (PCS) is assessed by grouping all physical components (PF, RP, BP and VT) together; similarly, the Mental Component Summary (MCS) encompasses mental components, such as SF, RE, MH and GH. Each answer is assigned a certain number of points; the score obtained in a given category may equal from 0 to 100 points. It is widely accepted that the lower the score, the worse the quality of life.

Statistical analysis was carried out with the use of Statistica 10 PL package and Microsoft Office Excel 2007. Basic statistical parameters, i.e. arithmetic mean, median, and minimum and maximum standard deviation, were calculated. To compare individual groups, the non-parametric Mann-Whitney U test was employed. Correlations were measured based on Spearman's rank correlation coefficient; $p < 0.05$ was considered statistically significant.

The study was approved by the Bioethical Commission at Wroclaw Medical University (decision number: 320/2012).

Results

Statistical description of groups participating in the study

The study encompassed 30 patients with CF including 19 women and girls (63.3%) and 11 men and boys (36.7%). The mean age was 24.83 ± 6.98 years, 25.8 ± 7.27 years, and 23.5 ± 6.56 years in the case of the study group, women and men, respectively. 13 women and 7 men were aged ≤ 25 years; moreover, 6 women and 4 men were aged > 25 years (Table 1).

The control group encompassed 30 healthy individuals with the same sex ratio and similar age ratio as in the group of patients. The mean age was 24.93 ± 7 years, 26 ± 7 years, and 23.91 ± 6.81years in the case of all participants, women and men, respectively. 13 women and 6 men were aged ≤ 25 years; furthermore, the group aged > 25 included 6 women and 5 men (Table 2).

Quality of life assessment in patients with CF with the use of Short Form-36

The QoL of patients with CF assessed using SF-36 questionnaire was poor, scoring below 50 in the case of Role Physical, General Health, Vitality, Social Functioning, and Physical Component Summary and Mental Component Summary. The Role

Table 1. Statistical description of the group of patients with cystic fibrosis

Tabela 1. Statystyczny opis grupy pacjentów z mukowiscydozą

	Women and men			Women			Men		
Number of patients	n = 20 (66.7%)	n = 10 (33.3%)	n = 30 (100%)	n = 13	n = 6	n = 19 (63.3%)	n = 7	n = 4	n = 11 (36.7%)
Age bracket	≤ 25 y	>25 y	All	≤ 25 y	>25 y	All	≤ 25 y	>25 y	All
$\bar{X}$ ± SD	20.6 ± 2.7	33.3 ± 4.72	24.83 ± 6.98	21.4 ± 2.53	34.8 ± 5.12	25.8 ± 7.27	19.1 ± 2.54	31 ± 3.37	23.5 ± 6.56
Me	21	32.5	23	21	35.5	23	19	29.5	22
$\bar{X}_{min} \div \bar{X}_{max}$	16 ÷ 25	29 ÷ 42	16 ÷ 42	16 ÷ 25	29 ÷ 42	16 ÷ 42	16 ÷ 23	29 ÷ 36	16 ÷ 36

Table 2. Statistical description of the control group of individuals without cystic fibrosis

Tabela 2. Statystyczny opis grupy kontrolnej — pacjentów bez mukowiscydozy

	Women and men			Women			Men		
Number of patients	n = 19 (63.3%)	n = 11 (36.7%)	n = 30 (100%)	n = 13	n = 6	n = 19 (63.3%)	n = 6	n = 5	n = 11 (36.7%)
Age bracket	≤ 25 y	>25 y	All	≤ 25 y	>25 y	All	≤ 25 y	>25 y	All
$\bar{X}$ ± SD	20 ± 3	33 ± 5	24.93 ± 7	21 ± 2	35 ± 6	26 ± 7	19 ± 2	30 ± 4	23.91 ± 6.81
Me	20	32	23	21	36	23	19	29	22
$\bar{X}_{min} \div \bar{X}_{max}$	16 ÷ 25	26 ÷ 42	16 ÷ 42	16 ÷ 25	27 ÷ 42	16 ÷ 42	16 ÷ 22	26 ÷ 36	16 ÷ 36

Table 3. General assessment of the quality of life of patients with cystic fibrosis

Tabela 3. Ocena jakości życia pacjentów z mukowiscydozą

Subscales	PF	RP	BP	GH	VT	SF	RE	MH	PCS	MCS
$\bar{X} \pm SD$	61.2 ± ± 27.4	31.7 ± ± 40.4	55.0 ± ± 29.2	31.8 ± ± 19.5	45.2 ± ± 20.6	49.6 ± 28.5	53.3 ± ± 46.0	56.4 ± ± 17.7	48.3 ± ± 26.0	47.8 ± ± 21.8
Me	65.0	0.0	46.0	32.0	40.0	37.5	66.7	52.0	39.0	44.0

Domens of SF-36 Questionnaire: Physical Functioning (PF), Role Physical (RF), Bodily Pain (BP), Social functioning (SF), Mental Health (MH), Role Emotional (RE), Vitality (VT), and General Health Perceptions (GH). Physical Component Summary (PCS) Mental Component Summary (MCS)

Table. 4. Quality of life of patients with cystic fibrosis depending on sex

Tabela 4. Ocena jakości życia pacjentów z mukowiscydozą w zależności od płci

Subscales		PF	RP	BP	GH	VT	SF	RE	MH	PCS	MCS
Women	$\bar{X} \pm$	54.2 ±	18.4 ±	48.7 ±	31.2 ±	42.1 ±	43.4 ±	50.9 ±	54.9 ±	40.9 ±	45.1 ±
	± SD	± 27.4	± 31.0	± 26.7	± 15.2	± 17.4	± 24.8	± 47.6	± 17.7	± 21.9	± 19.3
	Me	55.0	0.0	41.0	32.0	40.0	37.5	66.7	48.0	35.3	42.5
Men	$\bar{X} \pm$	73.2 ±	54.5 ±	65.9 ±	32.9 ±	50.5 ±	60.2 ±	57.6 ±	58.9 ±	61.0 ±	52.4 ±
	± SD	± 24.0	± 45.9	± 31.5	± 26.2	± 25.1	± 32.5	± 44.9	± 18.1	± 28.7	± 26.0
	Me	80.0	75.0	64.0	35.0	55.0	50.0	66.7	56.0	69.8	56.3
Significance test	p	0.0612	0.0502	0.1752	0.9143	0.4259	0.0932	0.6985	0.5612	0.0813	0.3437

Domens of SF-36 Questionnaire: Physical Functioning (PF), Role Physical (RF), Bodily Pain (BP), Social functioning (SF), Domens of SF-36 Questionnaire: Physical Functioning (PF), Role Physical (RF), Bodily Pain (BP), Social functioning (SF), Mental Health (MH), Role Emotional (RE), Vitality (VT), and General Health Perceptions (GH). Physical Component Summary (PCS) Mental Component Summary (MCS)

Physical aspect of the quality of life was assessed by the patients with CF as the poorest (31.7 ± 40.4), while the Physical Functioning aspect of the quality of life was assigned the biggest number of points. Relevant data is presented in Table 3.

The study showed that in comparison to men, women assess their quality of life more negatively in all subscales. Women's scores exceeded 50 only in the following subscales: Physical Functioning, Role Emotional and Mental Health. Role Physical received the lowest score (18.4 ± 31), whereas Mental Health was given the highest number of points (54.9 ± 17.7). In men, only General Health scored less than 50 (32.9 ± 26.2); it was the most negatively assessed subscale. The highest result was observed in Physical Functioning. Despite differences in the assessment of QoL between men and women, no statistically significant differences were demonstrated. The results are presented in Table 4.

Except for Role Emotional, the QoL of patients with CF aged > 25 years was lower than that of patients aged ≤ 25 years. Only in the Role Emotional subscale did the patients aged > 25 years achieve results exceeding 50; the results achieved in other subscales point to poor QoL. The lowest result (20.0 ± 42.2) was observed in Role Physical. The group of patients aged ≤ 25 years achieved results indicating poor QoL (37.5 ± 39.3) in Role Physical and General Health. Only in two subscales, i.e. Vitality and Mental Health, statistically significant differences between patients aged ≤ 25 and > 25 years were observed (Table 5).

The analysis of the impact of age on QoL of patients with CF assessed with the SF-36 questionnaire showed that there were statistically significant correlations ($p < 0.05$) between age and the following domains: Physical Functioning, Role Physical, Bodily Pain, Vitality, Mental Health and Physical Component Summary. This means that the older a person is, the lower the QoL in these domains. No statistically significant correlations between age and the following domains: General Health, Role Emotional, and Mental Component Summary, were observed. Statistically significant data has been marked in red and presented in Table 6.

The comparison of QoL of patients with CF and the control group of individuals without CF

Data presented in Table 7 shows that the QoL of the controls exceeded 50 for each subscale of the SF-36 questionnaire. The control group achieved the highest QoL in Physical Functioning and the lowest in Vitality.

Table 5. Quality of life of patients with cystic fibrosis depending on age

Tabela 5. Ocena jakości życia pacjentów z mukowiscydozą w zależności od wieku

Subscales		PF	RP	BP	GH	VT	SF	RE	MH	PCS	MCS
≤ 25	$\bar{X}$ ±	67.0 ±	37.5 ±	57.6 ±	33.9 ±	50.5 ±	53.1 ±	51.7 ±	61.6 ±	53.2 ±	50.1 ±
	± SD	± 25.6	± 39.3	± 27.9	± 16.7	± 21.3	± 31.1	± 45.2	± 18.2	± 25.9	22.4
	Me	75.0	37.5	51.0	36.0	50.0	37.5	50.0	64.0	44.8	49.5
25	$\bar{X}$ ±	49.5 ±	20.0 ±	49.8 ±	27.8 ±	34.5 ±	42.5 ±	56.7 ±	46.0 ±	38.5 ±	43.2 ±
	± SD	± 28.4	± 42.2	± 32.6	± 24.6	± 14.6	± 22.2	± 49.8	± 11.2	± 24.7	± 21.0
	Me	50.0	0.0	36.5	26.0	27.5	43.8	83.3	44.0	29.3	42.1
Significance test	p	0.0903	0.2437	0.3909	0.2811	0.0408	0.4953	0.8430	0.0278	0.0713	0.4414

Domens of SF-36 Questionnaire: Physical Functioning (PF), Role Physical (RF), Bodily Pain (BP), Social functioning (SF), Mental Health (MH), Role Emotional (RE), Vitality (VT), and General Health Perceptions (GH). Physical Component Summary (PCS) Mental Component Summary (MCS)

Table 6. Correlation coefficient between subscales of the SF-36 questionnaire and age

Tabela 6. Współczynnik korelacji między oceną jakości życia w poszczególnych domenach kwestionariusza SF-36 a wiekiem ankietowanych chorych

Age	R. Spearman	–0.623	–0.401	–0.406	–0.311	–0.516	–0.455	–0.027	–0.490	–0.555	–0.339

Table 7. Overall QoL of the control group of individuals without cystic fibrosis

Tabela 7. Ogólna ocena jakości życia w grupie kontrolnej osób bez mukowiscydozy

Subscales	PF	RP	BP	GH	VT	SF	RE	MH	PCS	MCS
$\bar{X}$ ± SD	90.3 ±	85.0 ±	73.2 ±	74.8 ±	60.3 ±	76.3 ±	81.1 ±	69.3 ±	77.2 ±	75.4 ±
Me	± 14.4	± 25.9	± 22.0	± 18.4	± 18.5	± 24.6	± 32.4	± 17.7	± 13.7	± 18.5
	95.0	100.0	74.0	77.0	67.5	75.0	100.0	72.0	82.9	81.8

Domens of SF-36 Questionnaire: Physical Functioning (PF), Role Physical (RF), Bodily Pain (BP), Social functioning (SF), Mental Health (MH), Role Emotional (RE), Vitality (VT), and General Health Perceptions (GH). Physical Component Summary (PCS) Mental Component Summary (MCS)

Table 8. The comparison of individual domains of QoL of patients with cystic fibrosis and the control group of people without cystic fibrosis

Tabela 8. Porównanie oceny jakości życia między chorymi z mukowiscydozą a grupą kontrolną osób bez mukowiscydozy

Healthy individuals	$\bar{X}$ ±	90.3 ±	85.0 ±	73.2 ±	74.8 ±	60.3 ±	76.3 ±	81.1 ±	69.3 ±	77.2 ±	75.4 ±
	± SD	± 14.4	± 25.9	± 22.0	± 18.4	± 18.5	± 24.6	± 32.4	± 17.7	± 13.7	± 18.5
	Me	95.0	100.0	74.0	77.0	67.5	75.0	100.0	72.0	82.9	81.8
Patients	$\bar{X}$ ±	61.2 ±	31.7 ±	55.0 ±	31.8 ±	45.2 ±	49.6 ±	53.3 ±	56.4 ±	48.3 ±	47.8 ±
	± SD	± 27.4	± 40.4	± 29.2	± 19.5	± 20.6	± 28.5	± 46.0	± 17.7	± 26.0	± 21.8
	Me	65.0	0.0	46.0	32.0	40.0	37.5	66.7	52.0	39.0	44.0
Significance test	p	0.000005	0.000007	0.011228	0.000000	0.002624	0.000770	0.026078	0.006378	0.000054	0.000008

The comparison between the QoL of individuals with CF and the control group of individuals without CF using the SF-36 general questionnaire showed that the QoL of patients was significantly lower than the QoL of the control group in all subscales. All differences in this comparison were found to be statistically significant (Table 8).

Discussion

The study attempted to describe health-related QoL in patients with CF with the use of the SF-36 general questionnaire. Although, due to the unique course of CF, such investigations are carried out with specific questionnaires more

sensitive to changes in health condition, the selected tool allowed the assessment of QoL in the most important domains of functioning that could be significantly affected by the disease, and the performance of comparative analysis. It should be emphasised that self-assessment carried out with the use of questionnaires may change in time and under the influence of objective factors [10, 11]. The study proved that QoL assessed by the patients was low. Mean results obtained in all subscales were close to 50; the lowest were in General Health and Role Physical and the highest in Physical Functioning. These results are comparable to the studies by Abbot et al. and Britto et al. [12, 13] in which the lowest results were obtained in General Health. In the study conducted by Britto et al., similarly as in the cited studies, Physical Functioning scored the gratest number of points. It is worth noting that in this study the mean results in all subscales were lower than in the studies by Abbot et al. [12] and Britto et al. [13]. This may indicate that there are differences in QoL between the Polish group of patients and the group of patients from other countries, which needs further analysis. The studies carried out by Grochans et al. [14] also confirm that QoL of Polish patients is lower as her study results were similar to those obtained in this study. Similarly as in the cited analyses, the General Health subscale gained the fewest number of points, and Physical Functioning gained the most.

The analysis of women's and men's self-assessment of QoL shows that average female results were poorer than those of males. According to Gee et al. [15], there are differences in the assessment of QoL between men and women. Similarly, Arrington-Sanders et al. [16], who used *The Child Health Questionnaire* to evaluate QoL, demonstrated statistically significant differences in the assessment of QoL between adolescent boys and girls in the subscales concerning general health, physical functioning, mental health, self-respect and family relations. Grochans et al. [14] also mention the differences in QoL between women and men; nonetheless, a statistically significant difference was found only in the Physical Functioning subscale. It is believed that women may achieve lower QoL results in comparison to men due to personality-dependent differences.

We found a statistically significant difference with regards to Vitality and Mental Health in patients aged over 25 years; moreover, there was a correlation between QoL in most subscales and age. Negative values of correlation mean that the older the patients, the worse their QoL in a given subscale. In our study, the observed correlation showed that the older were the patients, the worse was their QoL in the following domains: Physical Functioning, Role Physical, Bodily Pain, Vitality, Social Functioning, Mental Health and level of physical activity. Decreased self-assessment of QoL with regards the physical components may be due to the age-associated progress of the disease. Notably, Włodarczyk et al. [17] demonstrated that the functional status significantly affects the emotional state of patients with chronic respiratory conditions.

The comparison of QoL of patients with CF and the control group of individuals without CF showed statistically significant differences in all subscales. Britto et al. [13] found smaller differences in the assessment of QoL than the authors of the present study; moreover, statistically significant differences did not occur in all subscales. While comparing the QoL of patients with CF with the QoL of patients with other chronic conditions of the respiratory system, Dębska et al. [14] observed that all mean values obtained in all subscales by the former group were lower than those of the latter group, including statistical differences in two subscales: changes in health and general assessment of health condition. A more severe course of CF was postulated as the reason for this finding. Constant monitoring of health-related QoL of patients with CF is necessary. As there are only a few publications concerning this issue in Poland, this study could constitute the basis for conducting further analyses.

Conclusions

In general, patients with CF perceive their quality of life as poor.

There are differences in the assessment of quality of life between women and men. Despite only minor statistically significant differences, the QoL assessed by women is worse in all subscales than the QoL assessed by men.

There is a correlation between patients' QoL and age. The older the patients, the worse their quality of life in most subscales.

There are crucial differences in the QoL assessment between patients with CF and the control group encompassing individuals without CF. Patients with CF find their quality of life poorer.

Conflict of interest

The authors declare no conflict of interest.

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