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DETERMINANTS OF HEALTH-RELATED QUALITY OF LIFE IN POLISH PATIENTS WITH CF – ADOLESCENTS' AND PARENTS' PERSPECTIVES

CZYNNIKI WPŁYWAJĄCE NA JAKOŚĆ ŻYCIA ZWIĄZANĄ ZE ZDROWIEM WŚRÓD POLSKICH PACJENTÓW Z MUKOWISCYDOZĄ – PERSPEKTYWA NASTOLATKÓW I RODZICÓW

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Abstract

Aim: 1. Evaluation of health-related quality of life (HRQOL) in adolescents with cystic fibrosis (CF). 2. Evaluation of HRQOL in children with CF from the parents' perspective. 3. Evaluation of the relationship between HRQOL and both medical and psychosocial factors.

Material and methods: Health-related quality of life was measured with the Cystic Fibrosis Questionnaire – Revised. Seventy patients with cystic fibrosis, aged 14-18 years completed the version for adolescents and adults (CFQ-R 14+) and 70 parents of children aged 6-13 years filled out the version for parents (CFQ-R 6-13). Scores ranged from 0 to 100, with higher scores indicating a better quality of life. Disease severity was assessed by lung function test, nutritional status, chronic *Pseudomonas aeruginosa* infection and type of CFTR gene mutation. Social indices i.e. the patient's school attendance and the parent's work status were collected.

Results: In the adolescents' opinion, Eating problems and Digestive functioning got the highest rate, whereas Vitality, Treatment burden, Health perceptions and Weight got the lowest. Boys estimated their Physical functioning significantly higher than girls.

When evaluating their children's quality of life, parents granted the highest score to Physical, Respiratory and Digestive functioning and the lowest results were attributed to Treatment burden. Nutritional status and lung function impairment turned out to be predictors of some other domains but not psychosocial ones. The chronic *Pseudomonas aeruginosa* infection had an influence on several quality of life areas from the parents' perspective. School attendance had a significant impact on many aspects of the adolescents' functioning.

Conclusions: 1. The study revealed that the health-related quality of life of CF children and adolescents is moderately good. 2. Digestive functioning was one of the highest scored domains, while Treatment burden was one of the lowest, according to both the adolescents' and the parents' perception. 3. The potential impact of disease severity was clearly marked in the group of younger children, whose health-related quality of life was assessed by the parents. Self-evaluation conducted by adolescents was more subjective. 4. School attendance was an important factor of the quality of life. Further research is required in order to find other psychosocial indices.

Key words: Cystic Fibrosis, Cystic Fibrosis Questionnaire, Health Related Quality of Life, Adolescent

Streszczenie

Cel: 1. Ocena jakości życia związanej ze zdrowiem wśród nastolatków z mukowiscydozą. 2. Ocena jakości życia związanej ze zdrowiem dzieci z mukowiscydozą z perspektywy rodzica. 3. Ocena związku pomiędzy jakością życia związaną ze zdrowiem i medycznymi oraz społecznymi czynnikami.

Materiał i metody: Jakość życia związana ze zdrowiem była mierzona Kwestionariuszem Jakości Życia w Mukowiscydozie. Siedemdziesięciu pacjentów z mukowiscydozą, w wieku 14-18 lat wypełniło

wersję przeznaczoną dla adolescentów i dorosłych (CFQ-R 14+) oraz 70 rodziców dzieci w wieku 6-13 lat wypełniło wersję dla rodziców (CFQ-R 6-13). Zakres możliwych punktów wahał się od 0 do 100, przy czym wyższy wynik świadczy o lepszej jakości życia. Stopień zaawansowania choroby określony był za pomocą testu wydolności płuc, stanu odżywienia, przewlekłego zakażenia *Pseudomonas aeruginosa* oraz typu mutacji genu CFTR. Zebrano również informacje dotyczące formy nauczania, z jakiej korzysta dziecko (uczęszcza do szkoły vs nauczanie indywidualne w warunkach domowych) oraz statusu zawodowego rodzica.

Wyniki: W ocenie nastolatków, Zachowania związane z jedzeniem i Funkcjonowanie układu oddechowego uzyskały najwyższe wyniki, podczas gdy Witalność, Ograniczenia związane z leczeniem oraz Masa ciała uzyskały najniższe. Chłopcy istotnie wyżej niż dziewczynki ocenili swoje Funkcjonowanie fizyczne. Z perspektywy rodzica, jakość życia ich dzieci jest najwyższa w obszarach: Funkcjonowanie fizyczne, Funkcjonowanie układu oddechowego i pokarmowego, a najniższe dotyczy Ograniczeń związanych z leczeniem. Stan odżywienia oraz funkcja płuc ujawniły się, jako predyktory części mierzonych obszarów, ale nie psychospołecznych aspektów. Przewlekłe zakażenia *Pseudomonas aeruginosa* wykazało istotny wpływ na większość obszarów jakości życia z perspektywy rodzica. Uczęszczanie do szkoły miało istotny wpływ na wiele aspektów funkcjonowania adolescentów.

Wnioski: 1. Badanie wykazało umiarkowanie dobrą jakość życia związaną ze zdrowiem dzieci i młodzieży z mukowiscydozą. 2. Funkcjonowanie układu pokarmowego było najwyżej ocenianym obszarem, podczas gdy Ograniczenia związane z leczeniem oceniono najniżej zarówno w grupie nastolatków, jak i rodziców. 3. Potencjalny wpływ stopnia zaawansowania choroby wyraźniej zaznaczył się w grupie młodszych dzieci, których jakość życia związana ze zdrowiem oceniana była przez rodziców. Samoocena dokonana przez nastolatków była bardziej subiektywna. 4. Zaobserwowano, że uczęszczanie do szkoły było istotnym czynnikiem wpływającym na jakość życia. W celu ustalenia innych czynników psychospołecznych konieczne są dalsze badania.

Słowa kluczowe: Mukowiscydoza, Kwestionariusz dla Mukowiscydozy, Jakość Życia Związana ze Zdrowiem, Nastolatek

INTRODUCTION

Mucoviscidosis (ang. Cystic Fibrosis – CF) is a multi-organ disease, the most common genetic disorder in the Caucasian population, affecting mainly the respiratory and digestive systems. Treatment is multidisciplinary and predominantly includes prevention and treatment of the broncho-pulmonary disease, and a nutritional and pancreatic enzyme replacement therapy. The daily treatment routine, which includes chest physiotherapy and a high-energy diet, is time-consuming [1, 2]. Maintaining rigorous therapeutic regimes is difficult for many patients and may cause significant emotional stress for the patients and their families [3, 4].

Advances in diagnostic possibilities and treatment therapies in recent years contribute to improved survival. The increasing life expectancy of patients raises the question of its quality. Besides clinical outcomes, the patients' perception of the illness and their subjective opinions are more respected during CF treatment. More attention is focused on the individualization of medical interventions and adapting them to the patient's lifestyle in order to increase not only longevity but also the quality of life [5].

In the past few years there has been significant progress in defining and measuring the health-related quality of life (HRQOL). It is multidimensional and includes such factors as: severity of the disease symptoms, the physical

condition of patients, their mental state, emotional and social functioning. It is emphasized that the assessment of the health-related quality of life is carried out by the patients, and it is a subjective assessment of their own well-being and daily functioning [6, 7]. European standards of care for patients with cystic fibrosis recommend the evaluation of the HRQOL during annual psychological assessment [8].

Health-related quality of life measures are more often disease specific. Besides aspects of the general quality of life: i.e. physical, social and emotional functioning, they also include specific ranges for cystic fibrosis, such as: the burden of treatment and the symptoms of the respiratory and digestive systems. Information about the patients provides a wide-ranging perspective about their life and makes it possible to subjectively evaluate the methods of treatment (for example treatment of pulmonary exacerbations) [9, 10].

Studies have shown that the quality of life assessment made by the patients differs from their parents' report. For this purpose a CFQ-R questionnaire was developed which is specific for the parents. [11, 12].

Our study objectives included:

1. Evaluation of the HRQOL in adolescents with CF.
2. Evaluation of the HRQOL in children with CF from the parents' perspective.
3. An attempt to establish the correlation between the HRQOL and medical and psychosocial factors.

METHODS

Participants and procedure

Patients treated at the CF Center at the Institute of Mother and Child, Warsaw, Poland, with a confirmed diagnosis of CF according to national standards [1], aged 6-18 years old were included to the study. Children with acute respiratory infection, other chronic illnesses, mental retardation or reading difficulties were excluded.

The Bioethical Committee in the Institute of Mother and Child in Poland approved the study and written informed consent and assent was obtained from the patient and parent in accordance with the Committee's principles.

Measures

Health-related Quality of life (HRQOL) was measured using the Polish version [13] of the Cystic Fibrosis Questionnaire-Revised (CFQ-R). The CFQ-R is a widely used disease-specific instrument, translated into several languages [14-16]. Two versions of the CFQ-R were used: the Cystic Fibrosis Questionnaire – Revised version for Teen/Adult (CFQ-R14+) for patients aged 14-18 years old (self-report) and Cystic Fibrosis Questionnaire – Revised (CFQ-R 6-13) version for parents of children aged 6-13 years old (the parents'-proxy report). The instrument for adolescents CFQ-R 14+ includes 50 questions which are divided into 12 domains, while the questionnaire for parents covers 44 items divided into 11 domains. Results in individual domains are calculated, standardized and can range from 0 to 100 points. Response choices include ratings on a 4-point Likert scale. Items in the questionnaire are expressed as either "negative" or "positive".

The lung function, nutrition status and chronic *Pseudomonas aeruginosa* infection were collected during the routine ambulatory visits in order to assess the disease severity. Lung function was evaluated by measuring FEV1, expressed as the percentage of the predicted value based on gender, age and height. Nutritional status was assessed using the BMI Z-score. BMI standardisation was made due to the wide range of age, height and weight in the patient group. For calculations, the national reference groups were used [17] and the equation: BMI Z-score = (BMI of patients – BMI of the reference population)/Standard deviation of the reference population was applied.

Some information was recorded from the patients' file, such as infection with *Pseudomonas aeruginosa* (PA) during the last year and a type of mutation of the CFTR gene. Chronic PA infection was defined as one where 50% or more of the sputum samples were positive in the preceding 12 months according to the Leeds criteria [18].

Information about demographic data, the parents' employment status and the children's school attendance were part of the CFQ-R.

Statistical analyses

Statistical analyses were performed using the Statistical Package for the Social Sciences for Windows version

15.0 (SPSS 15.0). Data were expressed as means (M) + standard deviation (SD), confidence intervals (CI) or frequencies (%). An assessment of the normality of data was performed for skewness and kurtosis.

Student t-test analyses were conducted to compare means with the significance level adopted as $p \leq 0.05$.

A hierarchic regression analysis was performed with two factors: BMI Z-score and FEV1% as predictors of various HRQOL domains. The children's age and sex were controlled, and also the relation with the child (mothers vs fathers) in the parents' version.

The (UNIANOVA) variance analysis was conducted to assess the relationships between HRQOL domains and categorized factors: the type of genetic mutation, the *Pseudomonas aeruginosa* status, the children's school attendance and the parents' working status. The children's age and sex and also the relation with the child (mothers vs fathers) in the parents' version, were controlled.

RESULTS

Data from 70 adolescents aged 14-18 years old as well as from 70 parents of children aged 6-13 years old were collected. Table I presents both groups' characteristics and demographic data (Table I).

The internal consistency (Cronbach's alpha) for both CFQ-R versions was calculated. The data are presented in Table II. Two of the CFQ-R 14+ domains and four of the CFQ-R 6-13 domains obtained Cronbach's alpha < 0.60 . It was Treatment burden and Social functioning for CFQ-R14+ and Emotional functioning, Treatment burden, Health perception and School functioning for CFQ-R 6-13 (Table II).

In the adolescents' opinion, Eating problems and Digestive functioning got the highest rate ($F=39.07$; $p=0.001$) whereas Vitality, Treatment burden, Health perceptions and Weight got the lowest one ($F=57.01$; $p=0.001$) in comparison with other aspects of the quality of life. Figure 1. shows all the means for CFQ-R 14+.

While evaluating their children's quality of life, parents gave the highest scores in the following domains: Physical, Respiratory and Digestive functioning ($F=68.00$; $p=0.001$). The lowest results were attributed to Treatment burden ($F=76.21$; $p=0.001$) in comparison with other assessed aspects described in the CFQ-R 6-13 questionnaire. All the means were shown in Figure 2.

The comparison of data for boys and girls displayed one statistically significant difference. It concerned Physical functioning, $t=2.12$; $p<0.05$. Boys estimated their physical efficacy higher than girls. In turn, the analysis of data elicited from the parents revealed no gender-related difference in the quality of life in any of the domains considered.

Health-related quality of life determinants

The survey followed the influence of the Disease severity formulated by BMI for age Z-score, FEV1%, mutation type and chronic *Pseudomonas aeruginosa* infection on the quality of life in particular domains. In the adolescents' assessment, FEV1% turned out to be a significant predictor in the following domains: Physical functioning ($B=0.23$;

Table I. Demographic data and group characteristics for patients and parents.

Tabela I. Dane demograficzne i charakterystyka grup dla pacjentów i rodziców.

	Adolescents' report <i>Opis adolescentów</i>	Parents' report <i>Opis rodziców</i>
Number <i>Liczba</i>	70	70
Mean age±SD <i>Średni wiek±SD</i>	14.41±2.61	10.54±2.41
Gender, n (%) <i>Płeć, n (%)</i>	59 (41.8) male 82 (58.2) female	33 (47.1) 37 (52.9)
Relation, n (%)/ <i>Relacja, n(%)</i>	NA	62 (88.6) mothers 8 (11.4) fathers
FEV1%, M±S.D.	73.20 ±25.69	78.76 ± 22.32
BMI Z-score, M ±S. D.	-0.78 ± 0.94	-0.32 ± 1.02
Chronic Pseudomonas aeruginosa infection, n (%) <i>Chroniczne zakażenie Pseudomonas aeruginosa</i>	44 (62.9)	36 (51.4)
Delta F 508 homozygous, n (%) <i>Delta F 508 homozygota</i>	26 (37.1)	36 (51.4)
School attendance, n (%) <i>Uczęszczanie do szkoły</i>	56 (80.0)	NA
Parent's working status, n (%) <i>Sytuacja zawodowa rodzica</i>	NA	34 (48.6)

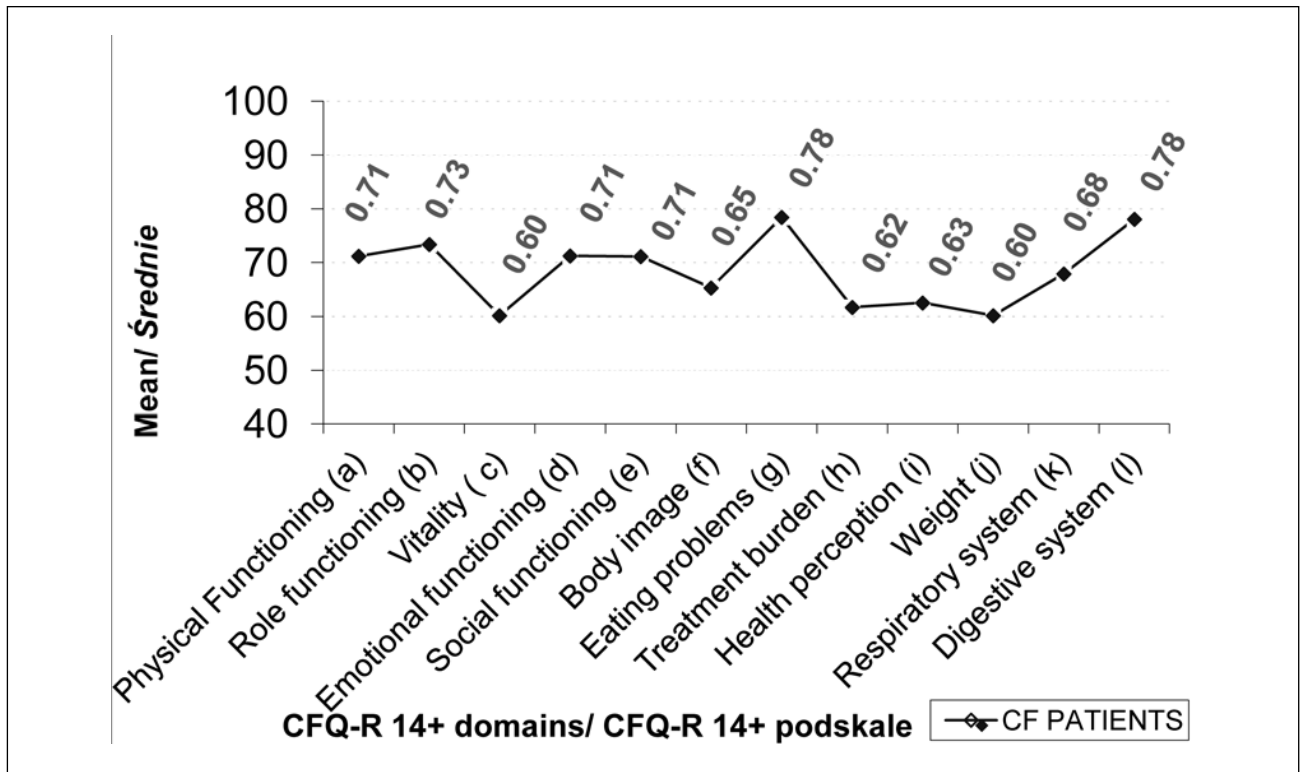
FEV1% predicted – forced expiratory volume in one second, BMI Z-score- Body Mass Index standardized with national charts/FEV1% predicted – *natężona objętość wydechowa pierwszosekundowa*, BMI Z-score – *Body Mass Index wystandaryzowany według norm polskich*.

Table II. Internal consistency coefficients for both CFQ-R versions.

Tabela II. Współczynniki estymacji wewnątrzgrupowej dla obu wersji kwestionariusza CFQ-R.

CFQ-R domains	CFQ-R 14+		CFQ-R 6-13	
	PL	Original	PL	Original
All	0.94	0.88	0.92	0.81
Physical functioning	0.91	0.94	0.91	0.91
Role functioning	0.77	0.83	NA	NA
Vitality	0.84	0.80	0.72	0.73
Emotional functioning	0.79	0.77	0.46	0.65
Social functioning	0.54	0.58	NA	NA
Body image	0.71	0.65	0.83	0.72
Eating problems	0.81	0.82	0.89	0.80
Treatment burden	0.49	0.51	0.56	0.59
Health perceptions	0.84	0.79	0.55	0.67
Weight	SIS	SIS	SIS	SIS
Respiratory symptoms	0.82	0.87	0.85	0.82
Digestive symptoms	0.72	0.68	0.75	0.63
School functioning	NA	NA	0.55	0.55

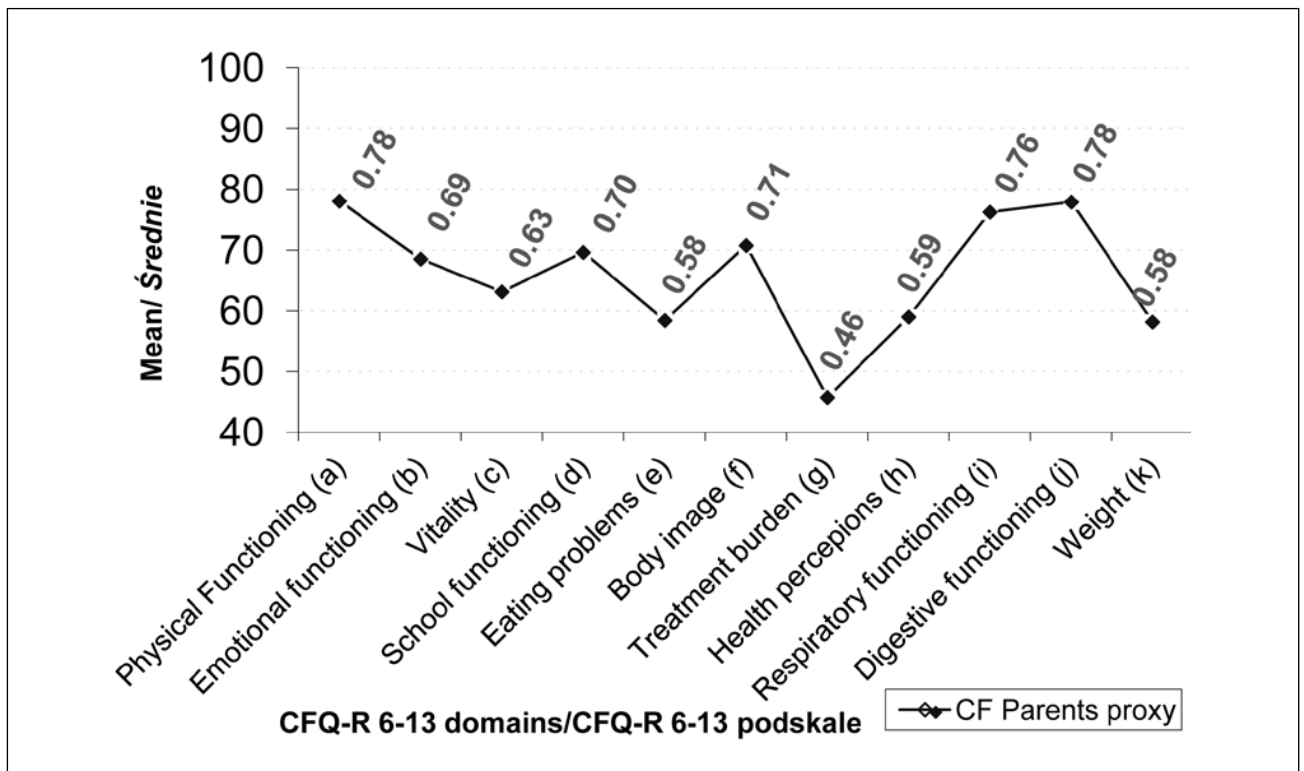
CFQ-R 14+ - Cystic Fibrosis Quality of Life – Revised questionnaire version for adolescents and adults/*Kwestionariusz Jakości Życia dla osób z mukowiscydozą w wieku 14 lat i powyżej*, CFQ-R 6-13 – Cystic Fibrosis Quality of Life – Revised questionnaire version for parents of children aged 6-13 years/*Kwestionariusz Jakości Życia dla rodziców dzieci z mukowiscydozą w wieku 6-13 lat*, PL – Polish results/*polskie wyniki*, Original – original Quittner's results [11]/*wyniki z badania Quittner*, NA – not applicable/*nie dotyczy*, SIS single-item scale (Cronbach's alpha cannot be calculated)/*podskala zawiera jedno pytanie (alfa Cronbacha nie może być policzone)*.



(a) – Fizyczne funkcjonowanie, (b) – Role społeczne, (c) – Witalność, (d) – Emocjonalne funkcjonowanie, (e) – Społeczne funkcjonowanie, (f) – Obraz ciała, (g) – Zachowania związane z jedzeniem, (h) – Ograniczenia związane z jedzeniem, (i) – Postrzeganie zdrowia, (j) – Masa ciała, (k) – Funkcjonowanie układu oddechowego, (l) – Funkcjonowanie układu pokarmowego.

Figure 1. Means in CFQ-R 14+ domains.

Wykres 1. Średnie w podskalach CFQ-R 14+.



(a) – Fizyczne funkcjonowanie, (b) – Emocjonalne funkcjonowanie, (c) – Witalność, (d) – Szkolne funkcjonowanie, (e) – Zachowania związane z jedzeniem, (f) – Obraz ciała, (g) – Ograniczenia związane z jedzeniem, (h) – Postrzeganie zdrowia, (i) – Funkcjonowanie układu oddechowego, (j) – Funkcjonowanie układu pokarmowego, (k) – Masa ciała

Figure 2. Means in CFQ-R 6-13 domains.

Wykres 2. Średnie w podskalach CFQ-R 6-13.

$p=0.03$) and Respiratory functioning ($B=0.23$; $p=0.04$). However, BMI Z-score proved to highly influence: Body image ($B=0.35$; $p=0.001$), Weight ($B=0.45$; $p=0.001$) and Eating problems ($B=0.23$; $p=0.04$).

Disease severity also considerably affected several quality of life domains from the parents' perspective. FEV1% was a significant predictor in the following domains: Physical functioning ($B=0.45$; $p=0.001$), Health perception ($B=0.42$; $p=0.001$), Weight ($B=0.43$; $p=0.01$), Respiratory functioning ($B=0.33$; $p=0.04$) and Vitality ($B=0.27$; $p=0.05$). The BMI Z-score of a child affected the evaluation of Body image ($B=0.27$; $p=0.04$) and Eating problems ($B=0.33$; $p=0.01$). Table III presents the results of both the CFQ-R 14+, as well as the CFQ-R 6-13 domains.

In order to observe the relation between the remaining determinants and the quality of life in particular domains,

the variance analysis (UNIANOVA) was applied. Quality of life domains were dependent variables, while delta F508 homozygous, chronic *Pseudomonas aeruginosa* infection and school attendance were covariant variables in the CFQ-R 14+ version, whereas in the CFQ-R 6-13 version, the quality of life domains were dependent variables and delta F508 homozygous, chronic *Pseudomonas aeruginosa* infection and parents' working status were covariant variables. The patients' age and sex were controlled, as well as the relation with the child (mother vs father) in the parents' version. In the group of adolescents, neither of two factors (type of mutation and chronic *Pseudomonas aeruginosa* infection) was found to exert a significant influence on any quality of life domain. In contrast, a social factor – school attendance – turned out to be an important factor influencing many quality of life domains described by the patients (Table IV).

Table III. Hierarchic regression analysis with two factors: BMI Z-score and FEV1% as predictors of CFQ-R domains. Children age, sex, as well as the relation with the child (mothers vs fathers) in parents' version, as a controlling factors.

Tabela III. Dwa czynniki: BMI Z-score i FEV1% jako predyktory w analizie regresji. W analizie kontrolowano wiek i płeć dziecka a także jego relację z rodzicem (matka vs ojciec) w wersji dla rodziców.

CFQ-R domains CFQ-R podskale	Adolescents report Opis adolescentów		Parents report Opis rodziców	
	BMI Z-score	FEV1%	BMI Z-score	FEV1%
	B (95% CI)	B (95% CI)	B (95% CI)	B (95% CI)
Physical functioning <i>Funkcjonowanie fizyczne</i>	0.02 (-4.52; 5.42)	0.23* (0.01; 0.42)	0.15 (-2.02; 8.98)	0.45** (0.23; 0.74)
Vitality <i>Witalność</i>	-0.07 (-6.20; 3.28)	0.20 (-0.02; 0.37)	0.06 (-3.38; 5.45)	0.27* (-0.00; 0.41)
Emotional functioning <i>Funkcjonowanie emocjonalne</i>	0.08 (-2.80; 5.62)	-0.06 (-0.21; 0.13)	0.11 (-2.85; 6.90)	0.21 (-0.05; 0.41)
Social functioning <i>Funkcjonowanie społeczne</i>	-0.06 (-4.70; 2.72)	0.08 (-0.10; 0.20)	NA	NA
Body image <i>Obraz ciała</i>	0.35** (4.28; 16.31)	0.09 (-0.13; 0.36)	0.27* (0.19; 14.08)	0.16 (-0.13; 0.52)
Eating problems <i>Zachowania związane z jedzeniem</i>	0.23* (0.22; 9.09)	0.10 (-0.10; 0.26)	0.33* (2.20; 20.83)	-0.05 (-0.52; 0.36)
Treatment burden <i>Ograniczenia związane z leczeniem</i>	0.15 (-1.72; 8.30)	-0.09 (-0.29; 0.12)	0.13 (-3.22; 9.06)	0.11 (-0.17; 0.41)
Health perception <i>Postrzeganie zdrowia</i>	0.21* (-0.16; 9.89)	0.09 (-0.12; 0.28)	0.19 (-1.23; 9.19)	0.42** (0.18; 0.67)
Weight <i>Masa ciała</i>	0.45** (9.01; 23.64)	-0.02 (-0.32; 0.27)	0.43** (6.03; 23.24)	0.06 (-0.30; 0.51)
Respiratory symptoms <i>Funkcjonowanie ukł. oddechowego</i>	-0.01 (-4.31; 4.13)	0.23* (0.01; 0.35)	0.06 (-3.46; 5.29)	0.33* (0.05; 0.46)
Digestive symptoms <i>Funkcjonowanie ukł. pokarmowego</i>	0.09 (-2.57; 6.18)	-0.04 (-0.21; 0.15)	0.12 (-2.82; 7.22)	0.16 (-0.10; 0.37)
School functioning <i>Funkcjonowanie szkolne</i>	NA	NA	0.17 (-2.32; 9.80)	0.22 (-0.05; 0.52)
Role functioning <i>Role społeczne</i>	0.12 (-1.90; 6.63)	0.03 (-0.15; 0.19)	NA	NA

CFQ-R – Cystic Fibrosis Questionnaire – Revised, FEV1% – forced expiratory volume in one second, BMI Z-score – Body Mass Index standardized with national charts, B – Beta, CI – confidence intervals, * $p<0.05$, ** $p<0.001$. / CFQ-R – Kwestionariusz Jakości Życia dla osób z mukowiscydozą, FEV1% predicted – natężona objętość wydechowa pierwszosekundowa, BMI Z-score – Body Mass Index wystandaryzowany według norm polskich, B-Beta, CI – przedziały ufności, * $p<0.05$, ** $p<0.001$.

Table IV. Results of the variance analysis (UNIANOVA), dependent variables: CFQ-R 14+ domains, covariant variables: chronic *Pseudomonas aeruginosa* infection, type of mutation and school attendance. The patient's age and sex were controlled.

Tabela IV. Wyniki analizy wariancji (UNIANOVA), zmienne zależne: podskale CFQ-R 14+, współzmiennie: chroniczne zakażenie *Pseudomonas aeruginosa*, rodzaj mutacji i uczęszczanie do szkoły. Wiek i płeć pacjenta były kontrolowane.

CFQ-R 14+ domains CFQ-R 14+podskale	Chronic PA infection Chroniczne zakażenie PA	Type of mutation Rodzaj mutacji	School attendance Uczęszczanie do szkoły
	F; p (η^2)	F; p (η^2)	F; p (η^2)
Physical functioning <i>Funkcjonowanie fizyczne</i>	0.37; 0.54 (0.007)	0.05; 0.02 (0.001)	7.39; 0.009 (0.12)
Emotional functioning <i>Funkcjonowanie emocjonalne</i>	0.21; 0.64 (0.004)	0.71; 0.40 (0.01)	3.69; 0.06 (0.06)
Body image <i>Obraz ciała</i>	0.25; 0.61 (0.005)	0.28; 0.59 (0.005)	3.94; 0.05 (0.06)
Eating problems <i>Zachowania związane z jedzeniem</i>	0.80; 0.37 (0.01)	2.41; 0.12 (0.04)	4.89; 0.03 (0.08)
Treatment burden <i>Ograniczenia związane z leczeniem</i>	0.15; 0.69 (0.003)	0.001; 0.97 (0.0001)	0.06; 0.79 (0.001)
Respiratory symptoms <i>Funkcjonowanie układu oddechowego</i>	0.52; 0.47 (0.01)	0.09; 0.76 (0.002)	0.83; 0.36 (0.01)
Digestive symptoms <i>Funkcjonowanie układu pokarmowego</i>	1.46; 0.23 (0.02)	0.55; 0.45 (0.01)	0.005; 0.94 (0.0001)
Vitality <i>Witalność</i>	0.95; 0.33 (0.01)	1.79; 0.18 (0.03)	1.07; 0.30 (0.02)
Role functioning <i>Role społeczne</i>	1.20; 0.27 (0.02)	0.06; 0.79 (0.001)	0.51; 0.47 (0.009)
Social functioning <i>Funkcjonowanie społeczne</i>	0.007; 0.93 (0.0001)	0.02; 0.87 (0.0001)	3.49; 0.06 (0.06)
Health perception <i>Postrzeganie zdrowia</i>	0.23; 0.62 (0.004)	0.06; 0.79 (0.0001)	9.15; 0.004 (0.14)
Weight <i>Masa ciała</i>	0.21; 0.64 (0.004)	0.34; 0.56 (0.006)	0.47; 0.49 (0.009)

CFQ-R 14+ - Cystic Fibrosis Quality of Life – Revised questionnaire version for adolescents and adults/ *Kwestionariusz Jakości Życia dla osób z mukowiscydozą w wieku 14 lat i powyżej.*

From the parents' perspective, delta F508 homozygous and parent's working status did not turn out to be significant factors, while chronic *Pseudomonas aeruginosa* infection proved to be an important factor related with many of the HRQoL domains (Table V).

DISCUSSION

Overall, the internal consistency of CFQ-R 14+ and CFQ-R 6-13 can be classified as comparable to Quittner's original results, but in the adolescents'/adults' versions there were two domains with unsatisfactory Cronbach's alpha results and in the parents' version there were four. The reason may be that in the US study there was a large sample of participants: 4679 adolescents/adults and 2145 parents, compared with our much smaller groups.

The most important result of the study was that in Poland, CF patients were generally a quite well-functioning group. Both the adolescents and the parents' perspectives were similar to those described in the literature [19-21]. Due to the different age groups of patients, the results of the adolescents' self-report and the parents' evaluation

of their children's quality of life could not be compared. It is probable that the parents' perception will change when the children grow and the illness progresses. It is assumed that the evaluation will be lower, but further research is needed.

Comparably to other studies, adolescents did not report problems concerning nutrition and their digestive system. The young people were well-educated and aware of the need of a high-calorie diet and pancreatic enzyme supplementation during meals. The adolescents attributed the lowest scores in their evaluation to Treatment burden, Health perception, Vitality and Weight. At the age of adolescence, Health perception in many Polish patients differed significantly from that of adolescents from countries with more advanced healthcare systems [22, 23]. Along with the course of the disease, the number of duties concerning the treatment rises, which was observable in the Treatment burden domain assessment. Confinement associated with time-consuming treatment was especially severe in the group of adolescents for whom the need for peer relations and social acceptance were extremely important [24]. Besides, adolescents were fully aware of

Table V. Results of variance analysis (UNIANOVA), dependent variables: CFQ-R 6-13 domains, covariant variables: chronic *Pseudomonas aeruginosa* infection, type of mutation and parent's working status. Patient age, sex and relation with parent (mother vs father) were controlled.

Tabela V. Wyniki analizy wariancji (UNIANOVA), zmienne zależne: podskale CFQ-R 6-13, współzmiennie: chroniczne zakażenie *Pseudomonas aeruginosa*, rodzaj mutacji i sytuacja zawodowa rodzica. Wiek i płeć pacjenta oraz jego relacja z rodzicem (matka vs ojciec) były kontrolowane.

CFQ-R 6-13 domains CFQ-R 6-13 podskale	Chronic PA infection Chroniczne zakażenie PA	Type of mutation Rodzaj mutacji	Parent's working status Sytuacja zawodowa rodzica
	F; p (η^2)	F; p (η^2)	F; p (η^2)
Physical functioning <i>Funkcjonowanie fizyczne</i>	9.48; 0.004 (0.19)	0.08; 0.77 (0.002)	0.02; 0.86 (0.001)
Emotional functioning <i>Funkcjonowanie emocjonalne</i>	1.67; 0.20 (0.04)	1.44; 0.23 (0.03)	0.15; 0.69 (0.004)
Body image <i>Obraz ciała</i>	3.69; 0.05 (0.09)	0.14; 0.70 (0.004)	0.54; 0.46 (0.01)
Eating problems <i>Zachowania związane z jedzeniem</i>	2.22; 0.14 (0.05)	0.01; 0.90 (0.0001)	0.59; 0.44 (0.01)
Treatment burden <i>Ograniczenia związane z leczeniem</i>	1.95; 0.17 (0.04)	0.008; 0.92 (0.0001)	0.42; 0.51 (0.01)
Respiratory symptoms <i>Funkcjonowanie układu oddechowego</i>	4.28; 0.04 (0.09)	0.31; 0.58 (0.008)	0.60; 0.44 (0.01)
Digestive symptoms <i>Funkcjonowanie układu pokarmowego</i>	3.82; 0.05 (0.08)	0.90; 0.34 (0.02)	0.95; 0.33 (0.02)
Vitality <i>Witalność</i>	3.83; 0.05 (0.08)	0.22; 0.63 (0.06)	1.54; 0.22 (0.03)
School functioning <i>Funkcjonowanie szkolne</i>	3.97; 0.05 (0.09)	0.01; 0.91 (0.0001)	0.86; 0.35 (0.02)
Health perception <i>Postrzeganie zdrowia</i>	13.79; 0.001 (0.26)	0.02; 0.86 (0.0001)	0.98; 0.32 (0.02)
Weight <i>Masa ciała</i>	2.70; 0.10 (0.06)	0.92; 0.34 (0.02)	1.10; 0.30 (0.02)

CFQ-R 6-13 – Cystic Fibrosis Quality of Life – Revised questionnaire version for parents of children aged 6-13 years/ *Kwestionariusz Jakości Życia dla rodziców dzieci z mukowiscydozą w wieku 6-13 lat.*

their disease and were able to assess their health status objectively, which was revealed in the Health perception domain evaluation. It was noticed that young people were not satisfied with their weight, while they highly evaluated the functioning of their digestive system and did not complain about eating problems. In a social context, low weight is the goal for the majority of girls in the period of pubescence [25]. The situation is opposite in the case of boys for whom the athletic body image is more attractive. Our studies revealed that Polish CF patients (both boys and girls) were not satisfied with their weight. This may be explained by the intensive education concerning the importance of nutrition in the process of the development of the disease carried out by parents and the medical CF team.

In the study we were trying to find determinants of HRQOL. It was confirmed that the disease severity was a significant predictor of some aspects of quality of life. Patients with greater pulmonary impairment assessed their Physical and Respiratory functioning as lower. However, we did not observe the influence of this determinant on Body image or Health perception, which was reported

in other studies. These domains were rather affected by the BMI Z-score which proved a significant predictor of nutrition concerning domains: Weight, Eating problems and Body image but also Health Perception. Type of mutation of the CFTR gene and chronic *Pseudomonas aeruginosa* infection did not reveal a relation to any quality of life domain in the adolescents' assessment [3, 10, 12, 26]. The aspect concerning Physical functioning in particular exemplified the fact that the adolescents' assessment was more subjective and less dependent on objective indices. In the group of CF patients of the Institute of Mother and Child, boys manifested more advanced disease represented by FEV1% and BMI Z-score than girls. In turn, boys assessed their Physical functioning as higher. It is additional proof showing how subjective a patient's perception of his/her quality of life can be. In order to find other factors influencing HRQOL, we chose social variables. It turned out that young people attending school classes thought their quality of life was higher than those who had individual home teaching. This concerned satisfaction with social relationships, a better emotional condition, but also physical functioning and

health-related aspects. This significant result could be an important argument while making the decision of individual home teaching.

The evaluation of the quality of life by parents was more dependent on objective indices of health status, such as FEV1%, BMI Z-score and chronic *Pseudomonas aeruginosa* infection [10]. This especially concerns children with chronic PA infection whose parents assessed their functioning as lower in many quality of life domains. What is interesting, this factor did not show any relation with any domain of HRQOL in adolescents. The parents' working status did not influence the child's functioning assessment.

Our study was not free of weaknesses. It was conducted in one clinical centre and the interpretation of results in reference to the whole population of CF children in Poland can be difficult. Every CF centre has different diagnostic and treatment possibilities and different access to doctors, physiotherapists, dieticians or psychologists, which may result in the differences of assessing some aspects of HRQOL. Also, there is no data concerning individual teaching at home in the case of children aged 6-13. This is a result of the construct of CFQ-R. The adolescent and adult version CFQ-R 14+ includes a question concerning individual teaching and the version for a parent CFQ-R 6-13 includes a question about the parents' working status. When the study was planned, this variable was not considered to have such a significant impact on HRQOL and we did not put enough effort into getting the data from young children.

CONCLUSIONS

The study revealed that the health-related quality of life of CF children is moderately good. This refers to both the adolescents who provided a self-description and to younger children who were assessed by their parents. The domains which caused many problems in the descriptions were Treatment burden and Weight. The evaluation of HRQOL determinants confirmed earlier reports that objective and medical aspects, such as the disease severity, were manifested more clearly in the parents' assessment. Self-evaluation made by adolescents is more subjective. Individual teaching proved to be the only significant social factor. Searching for other predictors of HRQOL in the assessment of CF patients may be the objective of future research.

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