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Evaluation of quality of life predictors in adolescents and young adults with cystic fibrosis

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ABSTRACT

Background: Cystic fibrosis (CF) is a chronic disease that has an impact on Health-Related Quality of Life (HRQoL).
Objectives: To identify demographic and clinical factors associated with HRQoL in adolescents and young adults with CF.
Methods: The sample comprised adolescent and young adult patients with CF. They completed the Cystic Fibrosis Quality of Life (CFQoL) questionnaire, which includes *Physical, Social, Treatment, Chest Symptoms, Emotional Functioning, Future Concerns, Relationships, Body Image*, and *Career* dimensions. We examined the relationships between gender, age, body weight, FEV1, pain, sleep, anxiety, depression and HRQoL.
Results: The sample comprised 95 patients (aged 14–25 years; female/male: 43.1/56.8%). The lowest CFQoL score was observed in *Future Concerns*. FEV_1 and body weight were positively associated with *Physical Functioning* (ß = 0.21; $P < 0.01$) and *Body Image* (ß = 0.30; $P < 0.01$), respectively. Females perceived themselves more negatively in *Future Concerns* (ß = −0.26; $P < 0.01$), *Relationships* (ß = −0.17; $P < 0.01$) and *Career Concerns* (ß = −0.20; $P < 0.01$) than males. Pain intensity (ß = −0.37), anxiety (ß = −0.39) and poor sleep quality (ß = −0.21) were negatively associated with global CFQoL ($P < 0.001$).
Conclusions: Pain intensity, anxiety and quality of sleep have the broadest impact on HRQoL. Regular assessment of psycho-emotional functioning, quality of sleep and pain intensity may improve a patient's well-being.

Introduction

Cystic fibrosis (CF) is an incurable, genetically determined disease that affects many systems of the human body. Clinically, it is characterized by chronic obstructive bronchial lesions, respiratory tract infections and digestive disorders (1). Despite the implementation of new methods of treatment (2–4), CF still leads to premature death.

According to available cystic fibrosis registers there are around 70,000 CF patients worldwide (5). Currently, the median age of survival in CF patients living in highly developed countries is over 40 years (1,6), and adults account for more than half of the patients (5).

Currently, in Poland about 2000 people suffer from CF, and the percentage of sufferers over 18 years of age does not exceed 30% (7). In 2014 the median age at the time of death of CF patients was 24.2 years. This results from the insufficient access of CF patients to multidisciplinary medical care that would comply with European standards (e.g. dietetics, physiotherapists and psychologists) (7,8).

Fluctuations in the course of CF with its gradual progression, worsening of physical fitness, multiple pain symptoms (9,10) and sleep disorders (11,12) lead to the gradual loss of independence, deterioration of social functioning and may induce anxiety and depression (13,14), a lowering of the quality of life in many areas (15) and a reduction in the motivation to undertake proactive health-oriented actions.

Identifying factors that determine quality of life in individuals with CF allows for planning comprehensive medical care, including

Abbreviation list: CF, cystic fibrosis; HRQoL, Health Related Quality of Life; CFQoL, Cystic Fibrosis Quality of Life; FEV_1, Forced Expiratory Volume in 1 second; NRS, Numeric Rating Scale; HADS sub-scales, Anxiety (HADS-A) and Depression (HADS-D), Hospital Anxiety and Depression Scale; AIS, Athens Insomnia Scale; BMI, Body Mass Index

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early detection of disorders (e.g. deficiency of body weight, pain, insomnia, anxiety, depression) and the implementation of treatment that is adequate to the individual needs of a young CF patient. Providing high quality medical care is particularly important for Polish CF patients due to their shorter survival time compared to their peers from highly developed countries (5). The aim of the study was to identify demographic and clinical factors associated with HRQoL among Polish patients with CF aged 14–25 years.

Material and methods

Design

The study employed a cross-sectional questionnaire design. This research project was approved by the Bioethics Commission (no. 122.6120.275.2015) and was implemented in accordance with the ethical principles of the Declaration of Helsinki.

Sample and setting

The study was conducted at the Department of Pneumonia and Cystic Fibrosis of the Institute of Tuberculosis and Lung Diseases in Rabka Zdroj, Poland.

The criteria for inclusion in the study were as follows: aged 14–25 years; of both sexes; consent to participate in the study obtained from the patients or their legal guardians (if the subjects were under the age of 18).

We excluded people who had diagnosed mental illness, alcohol abuse disorders, intellectual disability, deterioration of the clinical condition within the last month (e.g. increased dyspnoea, cough or sputum according to the patient's self-report), or traumatic events within the last 6 months (e.g. death, breakdown of a relationship, job loss), organ transplantation, non-invasive assisted breathing, or terminal health.

Variables and measures

Health-Related Quality of Life

The Cystic Fibrosis Quality of Life (CFQoL) questionnaire, created by Gee et al. (16) and adapted to Polish conditions by Dębska et al. (17), was used to assess HRQoL in CF patients. The CFQoL questionnaire contains 52 questions and consists of the following domains: *Physical Functioning, Social Functioning, Treatment Issues, Chest Symptoms, Emotional Functioning, Future Concerns, Relationships, Body Image, Career Concerns.*

Scores are calculated for each domain (range: 0–100 points) and then transformed into the global result (global CFQoL, range: 0–100 points). Higher scores represent better HRQoL.

Demographic and clinical variables

Demographic and clinical factors were recorded (15). Demographic factors included gender and age. Clinical factors included: Forced Expiratory Volume in 1 second (FEV_1), body weight, body height, Body Mass Index (BMI), pain (NRS scores), anxiety (HADS-A scores), depression (HADS-D scores) and sleep quality (AIS scores). Demographic date, FEV_1, body weight, body height, and BMI were obtained from the subjects' medical records. Intensity of pain, emotional state (anxiety, depression), and sleep quality were assessed using standardized research tools.

Pulmonary function was determined based on the results of spirometry of the last 4 weeks, resulting in FEV_1 as a percentage of the predicted value. Determination of the degree of disease progression was based on the following criteria: $FEV_1 > 70\%$ – early stage of the disease, $FEV_1 < 70\%$ and $>40\%$ – intermediate stage of the disease, and $FEV_1 < 40\%$ – advanced (severe) condition (5,18).

Pain intensity was assessed using the Numeric Rating Scale (NRS), where 0 meant no pain and 10 maximum pain. Numeric scores of 1–3, 4–6, and 7–10 corresponded to mild, moderate and severe pain (19,20).

Anxiety and depression were evaluated using the Hospital Anxiety and Depression Scale – HADS (21,22), which was validated for Polish conditions. This scale can be used in both adults and young people. It consists of 7 items grouped in sub-scales: anxiety (HADS-A) and depression (HADS-D). The answers describe the patient's well-being over the last week and are coded on a scale of 0–3 points. The total score amounts to 0–21 points (0–7 points = norm, 8–10 = borderline, ≥11 points = clinical levels of symptoms).

The Polish version of the Athens Insomnia Scale – AIS (23) was used to evaluate the quality of sleep. The scale allows for the quantitative measurement of the symptoms of insomnia, including trouble sleeping at night and worsening of daytime functioning. The subject marked a given symptom on a scale of 0–3 points (0 = no symptom, 3 = significant severity of the symptom), provided that it occurred at least three times a week during the last month. The overall score is in the range of 0–24 points. The higher the score, the worse the quality of sleep. A score of 8 points or more indicates insomnia symptoms.

Statistical methods

Qualitative variables are expressed as numbers representing counts of particular events occurring and adequate percentages. Quantitative variables are expressed as means and standard deviations (for normal distributions) or medians with upper and lower quartiles (for non-normal distributions). The Shapiro–Wilk test was used to assess the normality of qualitative variable distributions.

The comparison of global CFQoL and its domains between male and female groups was evaluated using the Mann–Whitney U-test.

Correlations between all variables were verified using the Pearson correlation coefficient (for normally distributed variables); otherwise, the Spearman correlation coefficient was used. We applied the interpretation of correlation coefficients according to Ratner (24): a correlation coefficient below 0.3 represents a week correlation, a correlation coefficient between 0.3 and 0.7 means a moderate correlation, while a correlation coefficient higher than 0.7 corresponds to a high correlation.

Multivariable linear regression models were calculated to find the relation between the global CFQoL (dependent variable) and both demographic and clinical parameters (independent variables). The backward step-wise approach was used to find the best multivariable model with independent variables selected based on the simple linear regression models. Only independent variables highly and moderately correlated with global CFQoL were included in the backward step-wise regression. The same methodology was used for independent variable selection in multivariable models with each global CFQoL domain as a dependent variable. Due to the high and moderate correlations between independent variables, the variance inflation factor (VIF) was used to detect multicollinearity in all final regression models (VIF < 5 was assumed as acceptable) (25). The results of all multivariable regression models were presented as standardized regression coefficients (ß) and their 95% confidence intervals (CI). *P*-values lower than 0.05 were considered statistically significant. All calculations were performed using *STATISTICA* version 12 (StatSoft®, Poland).

The sample size analysis was done using the G*Power program v. 3.0.10 (26). The study required at least 78 patients for detecting a 10% difference in global CFQoL using a significance level of 0.05 with powered to have 90% chance. The necessary values of global CFQoL are based on our previous study (27).

Results

The data from 95 patients were subjected to the final analysis. The studied data were complete, without any missing information. Our cohort accounted for 34.9% of the total of CF patients who were admitted to our hospital between February 10, 2016 and February 28, 2017, and 90.7% of the total CF patients were between the age of 14–25 years. Fig. 1 presents a flow diagram of the recruitment process.

Patient characteristics

The median age of subjects participating in the study was 19 years old, and the majority of patients were male (n = 54, 56.8%). The mean BMI was 19.8 kg/m^2 among all patients, and the prevalence of underweight patients (BMI < 18.5) was 30.5% (n = 29) – only 2 patients (2.1%) were overweight (BMI ≥ 25). An early stage bronchopulmonary disease was diagnosed in 43 patients (45.3%), 40 subjects (42.1%) were at the medium stage, and 12 patients (12.6%) met the criteria for the advanced stage. Clinical levels of depressive symptoms were reported in 6 patients (6.3%) and "borderline" levels of HADS-D scores in 12 subjects (12.6%), whereas 16 patients (16.8%) reported anxiety symptoms (in the clinical symptom range) and "borderline" levels of HADS-A scores in 17 subjects (17.9%). About 40.0% (n = 36) of the subjects had insomnia symptoms. Over the prior 2 weeks, mild, moderate and severe pain was reported by 8 (20%), 18 (45%) and 14 (35%) patients, respectively. About 90% of patients (n = 35) indicated one source of pain. The most common was abdominal pain (n = 20, 55.5%), followed by headache (n = 19, 52.7%) and chest pain (n = 15, 41.7%). The demographic and clinical characteristics of the sample are shown in Table 1.

Health-Related Quality of life

The highest median results of CFQoL calculated for the whole dataset were obtained in the domain: *Physical Functioning*, while the lowest results were seen in the domain: *Future Concerns* The comparison of global CFQoL and its domains between females and males showed that differences of *Social Functioning, Treatment* and *Body Image* domains are not statistically significant (P> 0.05), whereas the global *CFQoL* and the rest of its domains are significantly different

Table 1
Demographic and clinical characteristics of the sample.

Variables			
	Gender	Female	41 (43.1%)
		Male	54 (56.8%)
	Age (years)		19 (16–22)
Clinical	Body height (cm)		166.9 ± 9.6
	Body weight (kg)		55.5 ± 11.1
	BMI (kg/m^2)		19.8 ± 2.8
	FEV1 (% of pred.)		68.2 ± 22.7
	HADS-D scores		3 (1–6)
	HADS-A scores		6 (3–9)
	AIS-scores		6 (3–9)
	NRS-scores		0 (0–5)

Results presented as numbers (percentages) or mean (standard deviation) or median (upper and lower quartile); BMI – body-mass index; FEV_1 – forced expiratory volume in 1 s; HADS – Hospital Anxiety and Depression Scale, D - depression (range: 0–21 pts), A – anxiety (range: 0–21 pts); AIS – Athens Insomnia Scale (range: 0–24 pts); NRS – Numeric Rating Scale (range: 0–10 pts).

Table 2
Characteristics of Health-Related Quality of Life in adolescent and young adults with cystic fibrosis.

Quality of life	Total n = 95	Gender	
		Female	Male
Global CFQoL	66 (52–79)	60 (48–73)*	69 (59–81) *
CFQoL domain:			
Physical functioning	84 (70–92)	74 (64–92) *	86 (72–94) *
Social functioning	80 (65–95)	75 (65–85)	87 (65–95)
Treatment issues	66 (47–80)	66 (40–80)	67 (47–80)
Chest symptoms	70 (50–90)	60 (35–75) *	77 (55–90)*
Emotional functioning	77 (55–92)	67 (45–85)*	84 (67–95)*
Future concerns	40 (30–63)	33 (13–46)*	53 (37–67)*
Relationships	64 (44–74)	52 (42–70)*	67 (54–80)*
Body image	60 (40–80)	60 (47–80)	57 (33–80)
Career concerns	60 (40–75)	50 (35–60)*	65 (50–80)*

CFQoL – Cystic Fibrosis Quality of Life Questionnaire (range: 0–100 pts); results presented as median, upper and lower quartile; *P< 0.05.

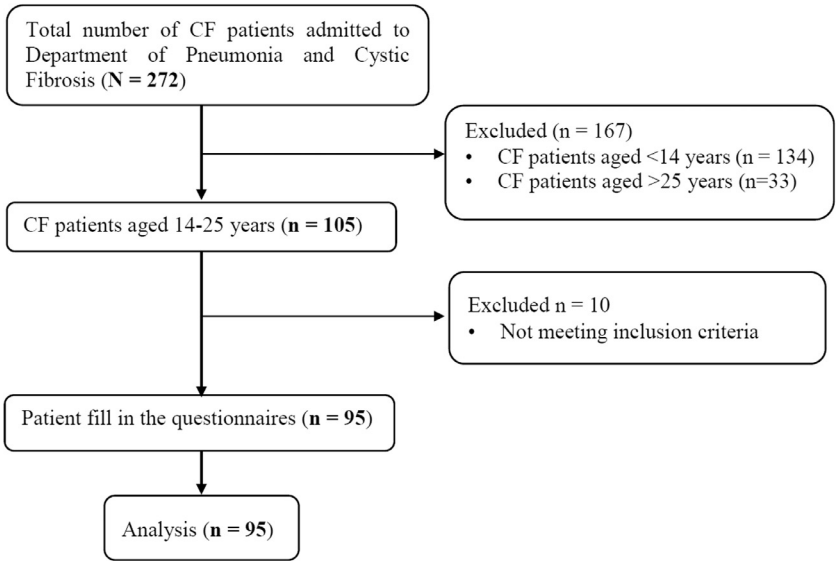

Fig. 1. Flow diagram of the recruitment process.

between females and males ($P< 0.05$). Table 2 contains the medians and quartiles (lower and upper) for the global CFQoL and its domains calculated for both the whole dataset and gender.

Assessment of selected, potential predictors of Health-Related Quality of Life

Table 3 shows the linear regression models for *Global CFQoL* and its domains in adolescents and young adults with CF. All presented models are statistically significant ($P< 0.05$). In the case of global *CFQoL* only AIS scores, NRS scores and HADS-A scores were statistically significant in the regression model. All parameters in this model are negative, which means that the global *CFQoL* decreases with the increase of one of these predictors by one unit, whereas the rest of the variables are stable. The model is quite well fitted ($R^2 = 0.54$). The contribution to the *Physical Functioning* domain comes from HADS-A scores, FEV_1 (% pred), NRS scores, and HADS-D scores. Only for FEV_1 (% pred) does one unit increase cause the increase of the *Physical Functioning* mean value by about 0.21. The remaining parameters are negatively associated with *Physical Functioning*. The fitting of the model is comparable to the previous one ($R^2 = 0.53$). AIS scores and NRS scores have an influence on *Social Functioning*. Both coefficients are negative, so one unit increase of AIS scores (or NRS scores) is associated with *Social Functioning* decreasing. Both *Treatment Issues* and *Chest Symptoms* depend on HADS-A scores and NRS scores only. NRS scores, HADS-D scores, and HADS-A scores have a negative impact on the mean of *Emotional Functioning* (all regression coefficients are below zero). This model is very well fitted, and the coefficient of determination is close to 60%. *Future Concerns* is linearly related with NRS scores, HADS-A scores, and gender. Females see their future worse than males (negative regression coefficient for females). The domain *Relationships* depends on gender, NRS scores and HADS-D scores. Female gender was associated with lower scores in *Relationships* in comparison to males. AIS scores and body weight influence the *Body Image* domain. The higher the body weight, the better the perception of one's own body. Only gender and HADS-D scores contribute to the *Career Concerns* domain. The higher HADS-D scores were correlated with lower scores in the *Career Concerns* domain. In relation to gender, females perceive their career prospects (e.g. maintaining a suitable job or college course) worse than males.

In all obtained regression models VIF ranged between 1.1 and 2.4, indicating that multicollinearity did not influence the linear regression results.

Discussion

The study results indicated that patients with CF have the lowest of HRQoL in the domain: *Future Concerns*. Pain intensity, anxiety and depression, quality of sleep, gender, FEV_1 and body weight were important predictors of HRQoL in adolescents and young adults with CF.

In our study, the lowest score of HRQoL was observed in the domain: *Future Concerns*, similarly to English studies (28,29). On the other hand, in studies conducted among Greek patients with CF, Stofa et al. (30) found poor HRQoL in as many as five out of nine domains: *Symptoms, Future Concerns, Relationships, Body Image* and *Career Concerns*. Differences in HRQoL in patients with CF from Poland and Greece are probably the result of the age of the study patients. Patients from Greece were older (47% of the population were over 28 years of age) than those from Poland.

Both our studies and those conducted by others (28–31) found the highest scores in the following domains: *Physical Functioning, Social Functioning* and *Emotional Functioning*. The good results obtained in the aforementioned domains may indicate the effectiveness of the adaptation to this chronic disease (32).

Our study has shown that moderate or severe pain was quite a common symptom of CF. We found that 42% of our cohort (40% of adolescents and 46% of adults) had at least one episode of pain over the prior 2 weeks. Lechtzin et al. (10) reported a prevalence of 58% in adolescents who had pain one or more times per month. Sermet-Gaudelus et al. (33) demonstrated that 89% of adults with CF had at least one episode of pain in the previous month. Similar to Lechtzin (10), we found an association between pain intensity and HRQoL. Pain had a negative influence on as many as seven out of nine domains of HRQoL. Lechtzin et al. (10) reported that pain intensity reduced HRQoL in four out of eight domains (*Physical, Emotional, Body Image* and *Digestion*).

The analysis showed that anxiety symptoms were more frequent than depressive symptoms. It was also demonstrated that the level of depression and anxiety had an effect on most HRQoL domains, with the level of depression related more to physical activity, and the level of anxiety more to emotional state. The level of anxiety was also a factor lowering HRQoL in the domains: *Chest Symptoms* and *Treatment Issues*. It is likely that the patients experienced greater anxiety and thus a greater burden of treatment as health problems resulting from the intensified progression of the disease. An analysis of the literature indicated that people with CF do not experience greater psychological problems than healthy people, and compared to some other chronic diseases, their problems are the same or even smaller (34–36).

The study also demonstrated that quality of sleep is one of the predictors of HRQoL in CF patients. Worse quality of sleep was associated with lower HRQoL in the *Physical Functioning* and *Body Image* domains. Bouka et al. (37) also noted the negative impact of sleep deprivation on the domains representing mental health (i.e. *Vitality, Emotional Functioning* and *Social Functioning*).

The meta-analysis conducted by Habib et al. (15) revealed that gender, which is the second most frequently studied HRQoL-related factor, was an important predictor of HRQoL in CF patients. Females achieved lower HRQoL in the following domains: *Physical Functioning, Emotional Functioning, Social Functioning* and *Chest Symptoms*. In our study females reported poorer HRQoL for global CFQoL and domains: *Physical Functioning, Emotional Functioning, Chest Symptoms, Future Concerns, Relationships and Career Concerns* than males. However, our study demonstrated a significant effect for gender as a predictor of HRQoL for *Relationships, Future Concerns,* and *Career Concerns* domains. Our findings are in line with the study by Gee et al. (38), who also found that females reported worse HRQoL for concerns for the future and career issues than males. This was despite having similar health status as indicated by objective clinical measures. The authors suggested that the difference in HRQoL between males and females was based on their perceptions of health rather than their actual health status.

There are plenty of data indicating that a decrease in FEV_1 is a significant predictor of the progression of changes in the respiratory system (5,39). Our study confirmed that the progression of bronchopulmonary disease had a significant impact on HRQoL in the domain *Physical Functioning*. Gee et al. (16,28,38) demonstrated that FEV_1 positively correlated with all nine CFQoL domains.

Scientific reports indicate a relationship between HRQoL and the severity of the disease and nutritional status (40,41). Apart from FEV_1, BMI is one of the most frequently studied HRQoL-related factors; *Body Image* is also connected with nutritional status (15). This theory has been confirmed by our results, showing that the higher the body weight, the better the HRQoL in the *Body Image* domain.

Although age was not associated with HRQoL, there is evidence of this relationship. The studies conducted by Hochwälder et al. (42) demonstrated that young adults (18–25 years) achieved higher HRQoL values than older age groups in the following domains: *Physical Functioning, Social Functioning, Chest Symptoms* and *Body Image*. With age, patients with CF have a worse mental

Table 3
Regression analyses for the Health-Related Quality of Life in adolescents and young adults with cystic fibrosis.

Quality of life	Predictors	Simple standardized regression coefficient (95% CI)	Multivariable standardized regression coefficient (95% CI)	Model
Global CFQoL	NRS scores	−0.51 (−0.69 to −0.34)*	−0.21 (−0.39 to −0.04)*	R^2 = 0.54; P< 0.001
	HADS-A scores	-0.62 (-0.78 to -0.45)*	-0.37 (-0.52 to -0.23)*	
	AIS scores	-0.54 (-0.71 to -0.37)*	-0.39 (-0.57 to -0.22)*	
	Gender ^{women}	-0.29 (-0.49 to -0.10)*		
	Height	0.36 (0.17 to 0.56)*		
	Body weight	0.28 (0.08 to 0.48)*		
	FEV_1 (% pred)	0.11 (-0.09 to 0.32)		
	HADS-D scores	-0.61 (-0.78 to -0.45)*		
CFQoL domains				
Physical Functioning	FEV_1 (% pred)	0.19 (-0.005 to 0.39)	0.21 (0.07 to 0.35)*	R^2 = 0.53; P < 0.01
	HADS-A scores	-0.56 (-0.73 to -0.40)*	-0.21 (-0.43 to -0.004)*	
	NRS scores	-0.49 (-0.67 to -0.37)*	-0.34 (-0.49 to -0.19)*	
	HADS-D scores	-0.61 (-0.77 to -0.45)*	-0.35 (-0.57 to -0.14)*	
	Gender ^{women}	-0.20 (-0.40 to -0.002)**		
	Height	0.30 (0.10 to 0.49)*		
	Body weight	0.28 (0.08 to 0.47)*		
	AIS scores	-0.44 (-0.62 to -0.25)*		
Social Functioning	AIS scores	-0.47 (-0.65 to -0.29)*	-0.37 (-0.54 to -0.21)*	R^2 = 0.37; P< 0.01
	NRS scores	-0.50 (-0.67 to -0.32)*	-0.41 (-0.58 to -0.24)*	
	Gender^{women}	-0.19 (-0.39 to 0.009)		
	Height	0.33 (0.13 to 0.52)*		
	Body weight	0.23 (0.02 to 0.49)**		
	FEV_1 (% pred)	0.12 (-0.08 to 0.32)		
	HADS-D scores	-0.47 (-0.65 to -0.29)*		
	HADS-A scores	-0.44 (-0.62 to -0.25)*		
Treatment issues	HADS-A scores	-0.33 (-0.52 to -0.14)*	-0.23 (-0.42 to -0.05)*	R^2 = 0.25; P< 0.01
	NRS scores	-0.46 (-0.64 to -0.28)*	-0.40 (-0.59 to -0.22)*	
	Gender ^{women}	-0.02 (-0.22 to 0.19)		
	Height	0.16 (-0.04 to 0.36)		
	Body weight	0.15 (-0.05 to 0.35)		
	FEV_1 (% pred)	0.16 (-0.04 to 0.36)		
	HADS-D scores	-0.27 (-0.47 to -0.07)*		
	AIS scores	-0.25 (-0.45 to -0.05)**		
Chest symptoms	NRS scores	-0.38 (-0.57 to -0.19)*	-0.28 (-0.46 to -0.11)*	R^2 = 0.29; P < 0.01
	HADS-A scores	-0.47 (-0.67 to -0.28)*	-0.40 (-0.58 to -0.22)*	
	Gender ^{women}	-0.26 (-0.46 to -0.07)*		
	Height	0.35 (0.16 to 0.54)*		
	Body weight	0.27 (0.08 to 0.47)*		
	FEV_1 (% pred)	0.13 (-0.07 to 0.33)		
	HADS-D scores	-0.37 (-0.56 to -0.18)*		
	AIS scores	-0.38 (-0.57 to -0.19)*		
Emotional functioning	HADS-D scores	-0.69 (-0.83 to -0.53)*	-0.26 (-0.46 to -0.06)*	R^2 = 0.59; P < 0.01
	NRS scores	-0.38 (-0.57 to -0.19)*	-0.19 (-0.32 to -0.04)*	
	HADS-A scores	-0.72 (-0.87 to -0.59)*	-0.49 (-0.68 to -0.29)*	
	Gender ^{women}	-0.27 (-0.47 to -0.08)*		
	Height	0.32 (0.12 to 0.51)*		
	Body weight	0.17 (-0.03 to 0.37)		
	FEV_1 (% pred)	0.02 (-0.18 to 0.23)		
	AIS scores	-0.58 (-0.74 to -0.41)*		
Future concerns	NRS scores	-0.33 (-0.52 to -0.13)*	-0.22 (-0.40 to -0.04)*	R^2 = 0.27; P < 0.01
	Gender women	-0.39 (-0.58 to -0.20)*	-0.26 (-0.45 to -0.08)*	
	HADS-A scores	-0.41 (-0.60 to -0.22)*	-0.27 (-0.46 to -0.08)*	
	Height	0.10 (-0.10 to 0.31)		
	Body weight	0.13 (-0.07 to 0.33)		
	FEV_1 (% pred)	0.14 (-0.07 to 0.34)		
	HADS-D scores	-0.30 (-0.50 to -0.11)*		
	AIS scores	-0.33 (-0.53 to -0.14)*		
Relationships	Gender ^{women}	-0.27 (-0.47 to -0.08)*	-0.18 (-0.35 to -0.003)*	R^2 = 0.30; P < 0.01
	NRS scores	-0.43 (-0.61 to -0.24)*	-0.30 (-0.48 to -0.12)*	
	HADS-D scores	-0.45 (-0.63 to -0.26)*	-0.32 (-0.51 to -0.14)*	
	Height	0.34 (0.15 to 0.53)*		
	Body weight	0.21 (0.01 to 0.41)**		
	FEV_1 (% pred)	0.02 (-0.18 to 0.23)		
	HADS-A scores	-0.39 (-0.58 to -0.20)*		
	AIS scores	-0.39 (-0.58 to -0.20)*		
Body image	Body weight	0.21 (0.01 to 0.41)*	0.30 (0.12 to 0.48)*	R^2 = 0.24; P<0.01
	AIS scores	-0.39 (-0.58 to 0.20)*	-0.36 (-0.54 to -0.18)*	
	Gender ^{women}	-0.27 (-0.47 to -0.07)*		
	Height	0.34 (0.15 to 0.53)*		
	FEV_1 (% pred)	0.02 (-0.18 to 0.23)		
	HADS-D-scores	-0.45 (-0.63 to -0.26)*		

(*continued*)

	HADS-A scores	-0.39 (-0.58 to -0.20)**		
	NRS scores	-0.43 (-0.62 to -0.24)**		
Career concerns	Gender [women]	-0.26 (-0.46 to -0.07)*	-0.20 (-0.38 to -0.02)*	$R^2 = 0.22$; $P < 0.01$
	HADS-D scores	-0.39 (-0.59 to -0.21)*	-0.41 (-0.60 to -0.23)*	
	Height	0.22 (0.02 to 0.42)**		
	Body weight	0.21 (0.005 to 0.40)**		
	FEV_1 (% pred)	0.12 (-0.08 to 0.32)		
	HADS-A scores	-0.43 (-0.61 to -0.24)*		
	NRS scores	-0.32 (-0.51 to -0.12)*		
	AIS scores	-0.38 (-0.57 to -0.18)*		

Only independent variables highly and moderately correlated with global CFQoL were included in the backward step-wise regression (BMI and age were not correlated with global CFQoL); *$P < 0.01$; **$P < 0.05$.

CFQoL – Cystic Fibrosis Quality of Life Questionnaire; HADS – Hospital Anxiety and Depression Scale, D – depression, A – anxiety; AIS – Athens Insomnia Scale; NRS – Numeric Rating Scale; R^2 – adjusted determination coefficient; Cl – confidence interval.

state and health condition. The older the patient, the worse the HRQoL in most domains (30).

A discussion about the effects of clinical variables on HRQoL in patients with CF indicated that HRQoL should be treated as a trait rather than a condition. This was also confirmed by authors who sought the cause of the lack of changes in HRQoL over time, despite the progression of the disease (27,43,44). In spite of the observed relationships, it is difficult to identity factors that unambiguously determine HRQoL; therefore, this issue requires further research. Knowledge of the factors determining HRQoL is important for monitoring treatment and planning nursing care.

Implications for clinical practice

Regular assessment of psycho-emotional functioning, quality of sleep, pain intensity and the efficacy of pain management may improve a patient's well-being and reduce the social and material costs associated with treatment of patients with CF. Self-assessment scales (e.g. HADS or AIS) should be routinely used for screening for symptoms of anxiety and depression or sleep disturbance in CF patients.

Limitations of the study

Our study contains some possible limitations: the assessment of anxiety and depression using only screening tools, the evaluation of sleep without objective tests (e.g. polysomnography), the lack of a control group and information on how often patients reported pain during the day, small sample.

Conflicts of interest

None.

Conclusions

Pain intensity, the presence of anxiety and depression, poor quality of sleep, body weight, FEV_1 and gender are predictors of HRQoL in adolescents and young adults with CF.

Authors' contribution

Lucyna Tomaszek was responsible for study design, statistical analysis, data interpretation and writing the draft of the article.

Grażyna Dębska was responsible for study design, analyzing, and writing the draft of the article.

Grażyna Cepuch was responsible for study design supervised, literature search, and completed the draft of the article.

Marlena Kulpa and Lidia Pawlik were responsible for collecting data, literature search.

Elżbieta Broniatowska was responsible for statistical analysis and data interpretation.

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