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Quality of life of children and adolescents with cystic fibrosis: the importance of body image and the impact of nutritional status, age and race/skin color on patients' and caregivers' perceptions

Qualidade de vida de crianças e adolescentes com fibrose cística: importância da imagem corporal e impacto do estado nutrional, idade e raça/cor na percepção dos pacientes e responsáveis

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Abstract

Objective: To identify factors related to the quality of life of children and adolescents with cystic fibrosis. *Methods:* Cross-sectional study of patients with cystic fibrosis, aged 7-18 years, from a Pediatric Referral Center in Brazil. The Cystic Fibrosis Questionnaire was used to evaluate the quality of life. The Mann-Whitney Test and the Spearman Correlation Test were used for the statistical analysis. The significance level adopted was 5%. *Results:* The domains with the lowest averages were body image (64.3±29.6) and weight (47.6±42.4), in the patient's perception. In the parents' perception, weight also presented the lowest average (52.8±43.7). The treatment domain had greater impact on the quality of life of patients aged 14 years ($p=0.012$) and older, and eating had greater impact among children younger than 14 years ($p=0.04$). The white individuals had better scores than the black/brown individuals in the body image domain ($p=0.049$). A moderate positive correlation was found between pBMI/A and the respiratory domain and a strong positive correlation between pBMI/A and body image. *Conclusions:* The correlation between pBMI/A and body image suggests that low weight negatively affects body image, which deserves attention, since body image and weight were the domains with the lowest scores in the patient's perception, and weight also had the lowest score in the perception of those responsible for the patients. In addition, low weight increased the impact of respiratory symptoms on the quality of life. The impact of treatments was greater in older patients, while the impact of eating was greater in younger patients. Race/skin color had an impact on the body image domain.

Keywords: Cystic fibrosis. Body image. Nutrition assessment. Body mass index. Quality of life. Malnutrition.

Resumo

Objetivo: Identificar os principais fatores relacionados à qualidade de vida de crianças e adolescentes com fibrose cística. *Métodos:* Estudo transversal, com indivíduos com fibrose cística, entre 7 e 18 anos, de um Centro de Referência Estadual. A qualidade de vida foi avaliada com o *Cystic Fibrosis Questionnaire*. Testes usados na análise estatística: Mann-Whitney e Correlação de Spearman. Nível de significância adotado: 5%. *Resultados:* Na percepção dos pacientes, os domínios da qualidade de vida com piores médias foram imagem corporal (64,3±29,6) e peso (47,6±42,4). Na percepção dos responsáveis, peso também apresentou a pior média (52,8±43,7). O impacto dos tratamentos na qualidade de vida foi maiora partir dos 14 anos (p=0,012), e o impacto da alimentação foi maior entre menores de 14 anos (p=0,04). Indivíduos brancos apresentaram melhores escores do que negros/pardos no domínio imagem corporal (p=0,049). Houve correlação positiva moderada entre pIMC/I e o domínio respiratório e correlação positiva forte entre pIMC/I e imagem corporal. *Conclusões:* A correlação entre pIMC/I e imagem corporal sugere que o baixo-peso afeta negativamente a imagem corporal, o que merece atenção, pois imagem corporal e peso foram os domínios com piores pontuações na percepção dos pacientes, sendo que o peso também teve a pior pontuação na percepção dos responsáveis. Além disso, o baixo-peso aumentou o impacto dos sintomas respiratórios na qualidade de vida. O impacto dos tratamentos aumentou com a idade, enquanto o da alimentação diminuiu com a idade. Também se observou impacto da raça/cor no domínio imagem corporal da qualidade de vida.

Palavras-chave: Fibrose cística. Imagem corporal. Avaliação nutricional. Índice de massa corporal. Qualidade de vida. Desnutrição.

Introduction

Cystic fibrosis (CF) is a genetic, autosomal recessive, chronic and progressive disease caused by mutations in a gene located in the long arm of chromosome 7.[1,2] Such mutations interfere with the regulation of ion transport, having as consequences dehydration of mucus secretions and its increased viscosity, favoring the obstruction of the exocrine gland ducts, followed by inflammatory reactions and, therefore, it is a multisystemic disease.[3] The phenotypic characteristics, the systems affected and the severity of the disease vary according to the type of mutation.[4]

Although the respiratory system affected by CF is the main cause of morbidity and death from this disease,[3,4] gastrointestinal dysfunctions are also very important due to the obstruction of the pancreatic and biliary ducts.[4] In the most severe cases, the obstruction of the pancreatic ducts by mucus begins in the intrauterine life, and the patients show pancreatic insufficiency at birth or shortly after birth.[4] Individuals with less severe mutations may or may not develop chronic obstructive pancreatitis and pancreatic insufficiency during life.[4] Obstruction of pancreatic ducts hinders the release of enzymes into the duodenum, causing digestive disturbances and malabsorption of fats, proteins and carbohydrates, and chronic diarrhea with bulky, fatty stools and protein-calorie undernutrition.[3]

Advances in the knowledge of CF, its early diagnosis and the evolution of treatment, with new drugs and emphasis on multidisciplinary team care, contributed to increase this population's lifespan.[4] US data revealed for the first time that there are more adults than children with this disease.[5]

It is vital that improvements in the quality of life (QoL) accompany the life span increase.[6] For this reason, specific instruments to assess the QoL of individuals with CF have been developed and improved.[7,8]

QoL evaluation questionnaires provide a standardized, valid and reliable perspective of patients and family members about the benefits and limitations of an intervention, information that cannot be obtained in the analyses of other clinical outcomes.[9] In addition, regular evaluations of QoL and longitudinal records allow to identify and monitor the difficulties experienced by patients over time,[10] contributing to technical analyses and the development of specific health policies.[11]

Factors that affect the QoL adversely have been studied,[10] but more knowledge is necessary so that public policies with an impact on this outcome can be planned accordingly. Thus, this study aimed to identify the major QoL-related factors in children and adolescents with CF.

Methods

Cross-sectional study approved by the Research Ethics Committee of the Federal University of Espírito Santo (UFES) (Process no. 352/572, CAAE: 16947313.1.0000.5060), conducted with patients with cystic fibrosis undergoing treatment from November 2015 to August 2016 at the *Hospital Estadual Infantil Nossa Senhora da Glória (HINSG)*, the single child referral hospital for CF treatment in the state of Espírito Santo (ES), Brazil.

The criteria for inclusion were patients with a diagnosis of CF, aged between 7 and 18 years, registered and undergoing treatment at the HINSG hospital. Criteria for exclusion were patients and/or parents who refused to participate; patients who did not have a cognitive condition that allowed interaction with the researcher and were absent from the assessment sessions during the data collection period.

The total of patients registered and on treatment at HINSG were 49 during the study period, and 30 were excluded due to the following criteria: 27 did not attend for the scheduled assessment appointments, 2 (autism), 1 (neuropathy). The convenience sample comprised 19 patients.

The assessment began after signature of the Free, Informed Consent Form and the Agreement Statement, when necessary. Information on personal data, age, colonization bacteria, bacterial colonization time, passive or active smoking, were questions asked to the patients/caregivers and confirmed through medical records.

To classify the patients regarding the disease severity, we used the Shwachman Score (< 40 = severe; 41-55 = poor; 56-70 = average; 71-85 = good; and 86-100 = excellent).[12]

In the nutritional assessment, the body mass index (BMI) was calculated to determine the percentile of body mass index for age (pBMI/A), using the *WHO Anthro Plus, 2007software, version 1.0.4*. The nutritional status was determined as recommended by the World Health Organization.[13]

The *Cystic Fibrosis Questionnaire* (CFQ), translated into Portuguese language and validated in 2006,[14] was administered. The four Portuguese versions of the CFQ were used: CFQ for groups of patients aged 6-11 years; CFQ for groups aged 12-13 years; CFQ for children aged 14 years and older; CFQ for the parents/caregivers of children aged 6-13 years. There are 25 questions divided into eight domains (physical, emotional, social, body image, eating disturbances, treatment burden, respiratory and digestive symptoms) for the two groups of 6-11 years and 12-13 years; 50 questions divided into 12 domains (social role, vitality, weight, health and the domains previously cited) for children aged 14 years and older, and 44 questions divided into 12 domains (physical, emotional, social, social role, eating, treatment, respiratory, digestive, vitality, health, body image and weight) for the parents/caregivers. The scale of each domain ranges from 0 to 100. Higher values express better QoL.

For the 7-11 years group, the questionnaire was administered by an interviewer with the aid of answer cards. The other questionnaires were filled out by the patients themselves and parents, with special care to prevent influence on the answers between both.

For the statistical analysis, the *Statistical Package for Social Sciences* (IBM SPSS Statistics 22, IBM, Armonk, NY, US) was used. The Mann-Whitney test was used to assess the differences between the groups, due to the normality rejection hypothesis. To analyze correlations between the QoL domains the Shwachman Score and BMI values, the Spearman's correlation was used. The correlation strength was determined by the correlation coefficient value (p), according to the following criteria: if $0 < p < 0.3$, there is a weak correlation; if $0.3 < p < 0.6$, there is a moderate correlation; if $0.6 < p < 0.9$, there is a strong correlation; if $0.9 < p < 1$, there is a very strong correlation.[15] The significance level adopted was 5%.

Results

The patients' mean age was 12.21 ± 3.78 years. The demographic and clinical characteristics that predominate in the sample were: male individuals (63.2%), Caucasian individuals (68.4%), nonpassive smokers (73.74%), colonized by bacteria (73.7%), with score "good" in the Shwachman Score (57.9%) and normal weight (89.5%) (Table 1).

Out of 14 colonized patients, eight were colonized by *Staphylococcus aureus* (57.14%) and six by *Pseudomonas aeruginosa* (42.85%), with colonization time varying from 1 to 6 years (mean time of 3.14 ± 1.9 years).

The Shwachman Score mean value was 78.16 ± 10.30.

The characterization of the sample with regard to pBMI/A is described on Table 2.

All participants attended school and received care from an interdisciplinary team, including physiotherapist and nutritionist.

In the group of children aged between 7 and 13 years, the highest and lowest means in the QoL domains were, respectively, 87.05± 12.38 for the "treatment" domain, and 59.26± 30.47 in the "body image" domain. In the group comprised of children aged 14 years and over, the best mean found was for the "eating" domain (95.24± 12.59), whereas the lowest mean was for the "weight" domain (47.61± 42.42). In general, the domain that had the best mean value was "eating" (83.64± 19.01) and the domains with the lowest means were "weight" (47.61± 42.42) and "body image" (64.33± 29.55) (Table 3).

Table 1. Demographic and clinical characteristics of CF patients according to age. Vitoria, ES, 2015/2016.

Variables	7-13 years	>14 years	Total
	n (%)	n (%)	n (%)
Sex			
Male	8 (66.7)	4 (57.1)	12 (63.2)
Female	4 (33.3)	3 (42.9)	7 (36.8)
Race / Skin color			
White	9 (75.0)	4 (57.1)	13 (68.4)
Black / Brown	3 (25.0)	3 (42.9)	6 (31.6)
Passive smoker?			
Yes	3 (25.0)	2 (28.6)	5 (26.3)
No	9 (75.0)	5 (71.4)	14 (73.7)
Colonized?			
Yes	9 (75.0)	5 (71.4)	14 (73.7)
No	3 (25.0)	2 (28.6)	5 (26.3)
Shwachman Score			
Excellent (86-100)	2 (16.7)	1 (14.3)	3 (15.8)
Good (71-85)	7 (58.3)	4 (57.1)	11(57.9)
Average (56-70)	2 (16.7)	2 (28.6)	4 (21.0)
Poor (41-55)	1 (8.3)	-	1 (5.3)
Severe (< 40)	-	-	-
Nutritional status (BMI/A)			
Normal	10 (83.3)	7 (100)	17 (89.5)
Thin	1 (8.3)	-	1 (5.25)
Very thin	1 (8.3)	-	1 (5.25)
Total	**12 (100)**	**7 (100)**	**19 (100)**

CF= cystic fibrosis; BMI/A: body mass index for age.

Table 2. Characterization of CF patients according to pBMI/A. Vitória, ES, 2015/2016.

Age (years)	Variables	N	Median	Mean	Standard Deviation
7-13	pBMI/A	12	18.45	34.37	32.38
>14	pBMI/A	7	30.80	30.31	24.51
Total	pBMI/A	19	21.80	32.88	29.07

CF: cystic fibrosis; pBMI/A: percentile of the body mass index for age.

Table 3. Scores for the quality of life domains of the *Cystic Fibrosis Questionnaire* according to the perception of children/adolescents and their parents. Vitória-ES, 2015/2016.

Groups	Domains	N	<value	>value	Mean ±SD
7-13 years	Physical	12	33.3	100.0	67.6 ±18.8
	Emotional	12	41.7	91.7	73.4 ±15.0
	Social	12	38.1	100.0	70.2 ±16.6
	Eating	12	44.4	100.0	76.9 ±19.2
	Treatment	12	66.7	100.0	87.1 ±12.4
	Respiratory	12	41.7	100.0	61.1 ±18.2
	Digestive	12	0.0	100.0	63.9 ±33.2
	Body image	12	0.0	100.0	59.3 ±30.5
>14 years	Physical	7	54.2	95.8	79.2 ±14.4
	Emotional	7	60.0	100.0	80.9 ±16.5
	Social	7	55.6	94.4	73.1 ±16.1
	Eating	7	66.7	100.0	95.2 ±12.6
	Treatment	7	0.0	77.8	61.9 ±28.6
	Respiratory	7	50.0	88.9	71.4 ±12.2
	Digestive	7	66.7	100.0	85.7 ±13.9
	Vitality	7	25.0	91.7	66.7 ±24.5
	Health	7	22.2	88.9	77.8 ±24.9
	Body image	7	33.3	100.0	73.0 ±27.9
	Social role	7	41.7	100.0	81.0 ±20.8
	Weight	7	0.0	100.0	47.6 ±42.4

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Total (children and adolescents)	Physical	19	33.3	100.0	71.9 ±17.8
	Emotional	19	41.7	100.0	76.2 ±15.6
	Social	19	38.1	100.0	71.3 ±16.0
	Eating	19	44.4	100.0	83.6 ±19.0
	Treatment	19	0.0	100.0	77.8 ±22.8
	Respiratory	19	41.7	100.0	64.9 ±16.7
	Digestive	19	0.0	100.0	72.0 ±29.3
	Vitality	7	25.0	91.7	66.7 ±24.5
	Health	7	22.2	88.9	77.8 ±24.9
	Body image	19	0.0	100.0	64.3 ±29.6
	Social role	7	41.7	100.0	81.0 ±20.8
	Weight	7	0.0	100.0	47.6 ±42.4
Parents (of 7-13 yr children)	Physical	12	20.8	95.8	66.6 ±20.5
	Emotional	12	53.3	100.0	85.0 ±14.5
	Social	12	8.3	100.0	68.8 ±26.2
	Eating	12	0.0	100.0	66.7 ±35.5
	Treatment	12	0.0	88.9	60.2 ±26.6
	Respiratory	12	7.0	100.0	59.4 ±27.8
	Digestive	12	22.2	100.0	73.2 ±22.5
	Vitality	12	40.0	93.3	68.3 ±18.2
	Health	12	0.0	100.0	61.6 ±33.3
	Body Image	12	44.4	100.0	81.5 ±19.7
	Weight	12	0.0	100.0	52.8 ±43.7

From the parents' perspective, the domain that exhibited the highest mean was "emotional" (84.98± 14.54), and the domain with the lowest mean was "weight" (52.78± 43.71) (Table 4).

In the statistical comparison of the QoL scores between the different age groups, the 14 to18-year group showed higher values in the "eating" domain (p=0.04) than the group with ages between 7-13 years. On the other hand, in the "treatment" domain, the children aged 7-13 years had higher scores (p=0.012) (Table 4).

When comparing the parents' perception of the QoL of their children with the children's own perception of their QoL, there was a significant difference in the "treatment" domain only, where the children had higher scores than their parents (p=0.003) (Table 4).

Table 4. Scores of the QoL domains and comparisons between different age groups and 7-13 year children with CF and their parents. Vitória-ES, 2015/2016.

Domains	Comparison between age groups			Comparison: children/parents		
	Age* (n)	Mean+SD	p	Groups (n)	Mean ±SD	p
Physical	7-13 (12)	67.59 ±18.77	0.203	Children (12)	67.59±18.77	0.799
	≥ 14 (7)	79.17 ±14.41		Parents (12)	66.62±20.49	
Emotional	7-13 (12)	73.42 ±14.99	0.611	Children (12)	73.42±14.99	0.060
	≥ 14 (7)	80.94 ±16.52		Parents (12)	84.98±14.54	
Social	7-13 (12)	70.24 ±16.63	0.966	Children (12)	70.24±16.63	0.887
	≥14 (7)	73.09 ±16.09		Parents (12)	68.75±26.15	
Eating	7-13 (12)	76.87 ±19.22	0.042[1]	Children (12)	76.87±19.22	0.932
	≥14 (7)	95.24 ±12.59		Parents (12)	66.67±35.52	
Treatment	7-13 (12)	87.05 ±12.37	0.012[1]	Children (12)	87.05±12.37	0.0031
	≥14 (7)	61.93 ±28.59		Parents (12)	60.20±26.59	
Respiratory	7-13 (12)	61.12 ±18.22	0.148	Children (12)	61.12±18.22	0.977
	≥14 (7)	71.36 ±12.23		Parents (12)	59.39±27.82	
Digestive	7-13 (12)	63.91±33.21	0.124	Children (12)	63.91±33.21	0.478
	≥14 (7)	85.73±13.91		Parents (12)	73.16±22.45	
Body Image	7-13 (12)	59.26±30.47	0.302	Children (12)	59.26±30.47	0.052
	≥14 (7)	73.01±27.87		Parents (12)	81.49±19.72	
Vitality	7-13 (0)					
	≥14 (7)	66.67±24.54		Parents (12)	68.33±18.22	
Health	7-13 (0)					
	≥14 (7)	77.79±24.86		Parents (12)	61.59±33.29	
Weight	7-13 (0)					
	≥14(7)	47.61±42.42		Parents (12)	52.78±43.72	
Social role	7-13 (0)					
	≥14 (7)	80.96±20.80				

[1] Statistically significant difference for the Mann-Whitney test ($p < 0.05$); QoL: quality of life; CF: cystic fibrosis; SD: standard deviation

Regarding race/skin color, the white individuals (n=13; mean = 72.65 ±31.64) had higher scores than the black/brown individuals (n = 6; mean =46.28 ±13.02) in the "body image" domain (p=0.049). No differences were found in the other domains between these groups.

It was not observed a relationship between the QoL domains and passive smoking or bacterial colonization. There was a positive correlation with moderate strength between the BMI/A and the "respiratory" domain, and a strong positive correlation between the BMI/A and the "body image" domain (Table 5). So, the higher the individuals' BMI/A value, the higher the scores found in the "respiratory" and "body image" domains.

There was no correlation between the Shwachman Score with any of the QoL domains (Table 6).

Table 5. Correlation between the QoL domains and the BMI/A scales of children and adolescents with CF. Vitória-ES, 2015/2016.

Quality of life	pBMI/A	
	p	p-value
Physical	0.280	0.245
Emotional	0.069	0.780
Social	0.078	0.752
Eating	0.270	0.263
Treatment	-0.134	0.584
Respiratory	0.493	0.032
Digestive	0.068	0.783
Vitality	0.288	0.531
Health	-0.178	0.702
Body image	0.683[1]	0.001
Social role	0.200	0.667
Weight	0.055	0.907

[1] Statistically significant for Spearman correlation; QoL: quality of life; CF: cystic fibrosis; BMI/A: body mass index for age; p: coefficient of correlation; pBMI/A: percentile of the body mass index for age.

Table 6. Correlation between the QoL domains and the Shwachman Score. Vitória-ES. 2015/2016.

Quality of life	Shwachman Score	
	p	p-value
Physical	0.095	0.699
Emotional	-0.424	0.071
Social	-0.022	0.927
Eating	0.129	0.598
Treatment	0.065	0.790
Respiratory	0.388	0.101
Digestive	-0.172	0.481
Vitality	-0.312	0.496
Health	0.327	0.474
Body image	0.314	0.191
Social role	-0.191	0.682
Weight	-0.452	0.309

Spearman correlation; QoL: quality of life; p: Coefficient of Correlation

Discussion

The overall mean score in the CFQ was 71.25, similar to the mean score found in other study conducted with clinically stable patients.[16] However, reference data are lacking in the literature. In Holland, the CFQ (except for the “treatment” domain) was administered to 478 healthy individuals aged 6 to 20 years, showing that even healthy children and adolescents may not have reached maximal scores, since only 34.8% of the individuals reached the maximal score on the assessed domains.[17]

The domains that had the lowest overall mean value were “weight” and “body image”, which means that these were the two domains with the highest negative impact on the QoL of these individuals. The low scores on the "body image" domain was also observed in other study with patients with CF.[16] In turn, healthy children and adolescents had higher scores on this domain in all ages.[15] CF may affect the physical appearance due to delayed growth, difficulty in gaining weight and delayed puberty development,[18] secondaries to calorie-protein malnutrition, which contributes to the adverse impact of the disease on the "weight" and "body image" domains.

Age had an impact on two QoL domains, "eating" and "treatment". Adolescents aged 14 years and over had a higher score on the "eating" domain, when compared to the 7 to 13-year group. In addition, "eating" (95.2 ±12.6) was the domain that had the highest mean score in children aged 14 years and older; and the "weight" domain had the lowest mean value (47.6 ± 42,4). These findings corroborate other Brazilian study conducted in the state of São Paulo, where higher and lower scores on this domain were found at this age group.[19] Evidences have shown that adolescents have a preference for processed foods, with high calorie content,[20,21] considered more pleasurable than healthier foods.[21] In contrast, it is also observed among adolescents a demand imposed by themselves for self-control in eating, associated with a frequent attempt to limit the intake of these foods that give them pleasure in order to fit body aesthetic standards.[21] It is possible that the high scores for CF adolescents in the "eating" domain is related to the perception of their freedom to eat caloric foods, which give them pleasure, without the restrictions imposed by the risk of gaining weight that other adolescents of their relationship have. On the other hand, depending on the severity of the digestive system impairment, adolescents with CF will need to receive digestive enzymes replacement[4] at every meal, which contributes to the negative impact on the "treatment" domain in their perception of QoL. The 14-18 years old group exhibited a significantly lower score on the "treatment" domain, when compared to the 7-13 years old group, showing that the treatment has a more adverse impact on the perception of QoL by adolescents than children, which was also observed in other studies.[19-22] The treatment strict routine interferes with the adolescents' activities and social relations, which is also and often the main reason for their peers to become aware of the colleague's disease.[18] Added to this is the growing complexity of the treatment due to comorbidities, aggravation of the disease over time,[22] and the fact that, during adolescence, the individual begins to take responsibility, at least partially, for his treatment,[18] which increases his perception of its impact.

Corroborating previous studies,[23] in the parents' perception, the "treatment" domain had a significantly higher impact on the QoL of their children than on the perception of the children younger than 14 years, possibly because at this age parents still take responsibility for the treatment,[18] which contributes to a reduction of its impact on their child's life.

A recent study showed that the mother's perception of her child's body weight tends to be distorted, especially when the child is overweight or obese, in which cases mothers tend to underestimate their children's weight. However, such distortion is present, although less frequently, in mothers of low-weight children, who may overestimate their children's weight.[24] This study suggests that the parents of children with CF are aware of the difficulty that their children have in gaining weight due to the disease. This may be the reason why they overestimate the weight of their children's, since "weight" was the domain that received the lowest score according to the parents' perception, despite most of the children had normal weight during the study. This result reveals the parents' concerns with the nutritional deficiencies caused by the disease and with the difficulty of their children to reach the ideal weight for their age.[25-28] The fact that most of the patients had normal weight at the time of assessment is possibly due to the interdisciplinary team work, including nutritionist and physiotherapist, who participate in the monitoring of all patients registered in the program, as set out by current guidelines for CF treatment.[25] In another Brazilian study carried out in the state of São Paulo, the "weight" domain also received the lowest score in the parents' perception.[19]

Guilt, impotence and shame are frequent feelings associated with the children's thinness by mothers of malnourished children.[29] However, no studies were found about the feelings of mothers of patients with CF with respect to their children's low weight, so this is a field for future investigations, considering that the present study showed that, according to the parents' perception, weight has a major negative impact on the children's QoL.

The patients who declared themselves black or brown presented scores significantly lower for the "body image" domain when compared with individuals who declared themselves white. A US study showed that Afro-American and Latin-American children and adolescents with CF had lower scores on the "physical", "body image", "health" and "social role" domains.[30] No other studies were found that assessed individuals with CF about the impact of the race/skin color on the QoL domain. However, a study conducted with college black students with eating disorders showed that the body image for these women is negatively influenced by their experiences with racial discrimination and external pressure to fit an aesthetic standard that was conceived based on the physical traits of white women.[31] The results found in the present study with regard to race/skin color are likely to be related to similar factors due to the stereotypes of beauty imposed by society, based on physical characteristics of white individuals.[32]

A positive correlation with moderate strength was found between the "respiratory" domain and pBMI/A, indicating that a lower pBMI/A is associated with a higher impact of respiratory symptoms on the QoL perception of these individuals. Although such relation was not observed in other studies conducted with adolescents and adults,[22] it is well established in the literature that the nutritional status is associated with pulmonary function impairments in patients with CF.[26-28,33]

Corroborating other studies,[22,34] a strong positive correlation was observed between the "body image" domain and the pBMI/A, indicating that the lower the weight, the poorer the body image is. This result deserves attention, if we take into account that "body image" and "weight" were the two domains with the lowest score, as discussed earlier in this study, which means that they are the domains that had a more negative impact on the QoL of children and adolescents, according to their own perception, as well as their parents' perception, in the case of the "weight" domain.

There is a consensus in the literature that an early colonization with *P. aeruginosa* has a significant impact on the prognosis of patients with CF,[35] being an important predictor of morbidity and death.[36] However, although the majority of the participants of the study is colonized with bacteria, no differences were found between the patients colonized or not regarding the QoL domains. It is possible that this is due to the clinically stable condition of the patients assessed, because this result corroborates other study, which assessed the impact of bacterial colonization with *P. aeruginosa,* in the "respiratory" domain of the CFQ, on 100 clinically stable patients with CF, and did not show a significant difference between the colonized or not colonized patients.[37]

No correlation was found between the Shwachman Score and QoL, which differs from the study conducted by Cohen et al.[19] It is worth emphasizing that in the present study 14 individuals (73.9%) were classified as "excellent" or "good" according to this scoring, which possibly can explain the absence of correlation, due to the reduced number of individuals in the category "average" (n=4) and "poor" (n=1), and absence of individuals in the category "severe".

Five patients in this study reported being passive smokers. However, there was no difference in the QoL of these individuals when compared with the group whose parents/caregivers were nonsmokers. These data corroborate a recent study conducted in Hungary, which found that passive smoking did not have an impact on the QoL of children with CF.[38] It was not found any other study addressing the impact of passive smoking on the QoL of children or adults with CF, but it is worth noting that the negative impact of passive smoking on the pulmonary function of these individuals has been well documented.[39]

The two main limitations of the present study were the impossibility of assessing longitudinal variations because it is a cross-sectional study, and the reduced number of the sample, which did not allow to assess other categorizations. However, we should take into account that, according to the Brazilian Cystic Fibrosis Registry,[40] the state of Espirito Santo has 135 patients registered and undergoing treatment for CF, including adults and children, and the study was carried out at the single pediatric referral center for CF treatment in ES, which had 49 patients in the age groups studied registered at the time of assessment. Thus, although small, the sample corresponds to approximately 39% of the individuals with CF in this age group.

Conclusions

The strong positive correlation between pBMI/A and the “body image” domain suggests that low weight has a negative impact on the body image of children and adolescents with CF. This result deserves attention, taking into account that “body image” and “weight” were the domains that most affected adversely the QoL according to the patients’ perception, and in the parents’ perception of “weight”.

The pBMI/A had a moderate positive correlation in the perception of QoL regarding the “respiratory” domain, showing that patients with lower weight have a sharper perception of the adverse impact of respiratory symptoms on the QoL.

Race/skin color also had an impact on the “body image” domain. In addition, it was found that the impact of the treatment on the QoL increases with age at the age groups studied; that eating disturbances have more impact on children than adolescents, although it is the domain with the highest overall scores; and that in the parents’ perception, the treatment burden has a greater impact on the QoL than in the children’s perception.

These results emphasize the need for optimized, prioritized strategies, and included in the SUS network (the Brazilian HealthCare System), for the monitoring and correction of nutritional deficiencies and muscle mass gain in individuals with CF, with participation of a multi-professional team, including nutritionists, physiotherapists and physical educators that work in these areas.

It is important that further studies on this subject be carried out in different regions of the country, with larger samples and longitudinal assessment, so that the improvement of the QoL of this population can be increasingly effective, considering that Brazil has major cultural, social, educational and ethnic differences that can interfere with the perception of the body image and QoL.

Collaborators

da Silva LA participated in the project design, data collection, data analysis and in writing the manuscript; Lima ACP participated in the project design, data collection, data analysis and in writing the manuscript; Wittmer VL contributed to the project design, data analysis and revision of the manuscript; FMG Liberato FMG contributed to the project writing, supervised data collection

and contributed to the writing and final revision of the manuscript; Arpini LSB contributed to the data analysis, writing and final revision of the manuscript; Paro FM participated in the project design, supervised data collection and analysis and contributed to the writing and final revision of the manuscript.

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