Quality of life assessment in patients with cystic fibrosis by means of the Cystic Fibrosis Questionnaire*

Avaliação da qualidade de vida de pacientes com fibrose cística por meio do *Cystic Fibrosis Questionnaire*

Milena Antonelli Cohen, Maria Ângela Gonçalves de Oliveira Ribeiro, Antonio Fernando Ribeiro, José Dirceu Ribeiro, André Moreno Morcillo

Abstract

Objective: To assess the quality of life (QoL) of patients with cystic fibrosis (CF) followed at a university referral center for CF. **Methods:** A cross-sectional study involving application of the Cystic Fibrosis Questionnaire (CFQ) and Shwachman score in CF patients between April of 2008 and June of 2009. **Results:** The sample consisted of 75 patients. The mean age was 12.5 ± 5.1 years (range, 6.1-26.4 years). The patients were divided into three groups by age in years: group I (< 12), II (12-14), and III (≥ 14). The highest and lowest CFQ scores were for the nutrition domain in group III (89.3 ± 16.2) and the social domain in group II (59.5 ± 22.3), respectively. Groups I and III differed significantly regarding the treatment domain (p = 0.001). Regarding Shwachman scores, there were significant differences between patients scoring ≤ 70 and those scoring > 70 in the social (group I; p = 0.045), respiratory (group II; p = 0.053), and digestive (p = 0.042) domains. In group III, severity did not correlate with QoL. In groups I and II, patients with an FEV_1 < 80% of predicted did not differ from other patients for any CFQ domain. However, in group III, values for the following domains were significantly lower in patients with an FEV_1 < 80%: physical (p = 0.012); body image (p = 0.031); respiratory (p = 0.023), emotional (p = 0.041); and social role (p = 0.024). **Conclusions:** It is important to assess QoL in CF patients, because it can improve treatment compliance.

Keywords: Cystic fibrosis; Quality of life; Questionnaires.

Resumo

Objetivo: Avaliar a qualidade de vida (QV) de pacientes com fibrose cística (FC) acompanhados em um centro universitário de referência em atendimento a FC. **Métodos:** Estudo transversal com a aplicação do *Cystic Fibrosis Questionnaire* (CFQ) e escore de Shwachman em pacientes com FC entre abril de 2008 e junho de 2009. **Resultados:** Participaram 75 pacientes, com média de idade de 12,5 ± 5,1 anos (variação: 6,1-26,4 anos). Os pacientes foram divididos em três grupos de acordo com a idade: grupo I (< 12 anos), II (12-14 anos) e III (≥ 14 anos). As pontuações mais altas e mais baixas no CFQ foram para o domínio alimentação do grupo III (89,3 ± 16,2) e para o domínio social no grupo II (59,5 ± 22,3), respectivamente. Houve uma diferença significativa no domínio tratamento entre os grupos III e I (p = 0,001). Pacientes com escore de Shwachman ≤ 70 apresentaram diferenças significativas em relação aos domínios social (grupo I; p = 0,045), respiratório (grupo II; p = 0,053) e digestivo (p = 0,042) quando comparados aqueles com escores > 70. No grupo III, não se observou associação entre gravidade e QV. Nos grupos I e II, não se observou diferenças entre os pacientes com VEF_1 < 80% do previsto e os demais pacientes em todos os domínios do CFQ. Entretanto, no grupo III, as médias dos pacientes com VEF_1 < 80% do previsto foram significativamente menores nos domínios físico (p = 0,012), imagem corporal (p = 0,031), respiratório (p = 0,023), emocional (p = 0,041) e papel social (p = 0,024). **Conclusões:** A avaliação da QV em pacientes com FC é importante, pois contribui para uma melhor aderência ao tratamento.

Descritores: Fibrose cística; Qualidade de vida; Questionários.

* Study carried out at the State University at Campinas School of Medical Sciences, Campinas, Brazil.
Correspondence to: André Moreno Morcillo. Rua Antonio Adami, 166, Santa Isabel, CEP 13084-240, Campinas, SP, Brasil.
Tel 55 19 9146-6379 or 55 19 3521-7353. E-mail: morcillo@fcm.unicamp.br or morcillo@hc.unicamp.br
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Introduction

Cystic fibrosis (CF) is a multisystemic autosomal recessive hereditary disease that is life-threatening and is characterized by chronic lung disease, pancreatic insufficiency (causing poor digestion), malnutrition, and high concentrations of sweat electrolytes. This is due to thick layers of viscous mucous secretions that obstruct the exocrine gland ducts.[1-3]

The incidence of CF varies according to ethnicity, ranging from 1:2,000 to 1:5,000 White live births in Europe, the United States, and Canada.[3] In Brazil, the incidence ranges from 1:1,850 to 1:9,600.[3]

The life expectancy of individuals with CF who were born in the year 2000 is estimated to be approximately 35-40 years.[4] This has become possible because of advances that have led to more appropriate treatments, such as aggressive antibiotic therapy, regular respiratory therapy, and nutritional counseling, all of which require a strong daily commitment, which negatively affects the quality of life (QoL) of patients and their families.[1,2]

According to the definition given by the World Health Organization in 1995, QoL represents "the perception that individuals have of their position in life, in the context of the culture and system of values in which they live and in relation to their objectives, expectations, standards, and concerns". Therefore, for individuals to have a good QoL, it is necessary that they establish a good balance among the social, psychological, and physical domains, adjusting their expectations to the context in which they live.

In the last two decades, the measurement of health-related QoL has been more widely used. More recently, there has been progress regarding its measurement, in healthy individuals and in those with chronic diseases.[5] There is a consensus among some specialists that instruments designed to quantify QoL should always include fundamental domains, such as disease severity, physical symptoms, psychological symptoms, emotional symptoms, functional status, and social status.[5-8]

The importance of measuring QoL in CF was first described in 1986 by a group of specialists who attended a workshop at the National Heart, Lung and Blood Institute. Since then, QoL has been assessed in this group of patients, using COPD as a model, given that the treatment of COPD is as complex as is that of CF.[6,8]

There are two basic types of instruments to measure QoL: specific and generic.[9] Although generic instruments can be applied to different populations and different diseases, thereby allowing the comparison of ill and healthy individuals, they have low sensitivity for detecting small changes related to the clinical aspects and treatment of a specific disease.[6,9-12] In contrast, specific questionnaires focus on the signs and symptoms of only one disease, such as CF, making it possible to assess the effectiveness of new therapeutic measures, thereby facilitating economic planning and potentially improving treatment compliance. In addition, specific questionnaires make it possible to determine how the disease affects activities of daily living.[7,12,13]

The first questionnaire designed to assess QoL in CF, originally known as the Cystic Fibrosis Questionnaire (CFQ), was developed in France in 1996.[5] In 2005, the CFQ was translated to English and validated by Quitter et al., who modified the English-language version to have additional advantages, such as the possibility of applying it in younger children (6-11 years of age).[8]

In 2006, the CFQ was translated to Portuguese and validated for use in Brazil.[14] The translation was based on the English-language version, which consists of four questionnaires developed for different groups: patients ≥ 6 and < 12 years of age (CFQ_{6-11}); patients ≥ 12 and < 14 years of age (CFQ_{12-13}); patients ≥ 14 years of age (CFQ_{14+}); and parents of patients ≥ 6 and < 14 years of age ($CFQ_{Parents6-13}$).[14]

The objective of the present study was to employ the CFQ as a means of assessing the QoL of patients with CF followed at a university hospital referral center for CF.

Methods

This was a cross-sectional study including 75 patients (age, 6.1-26.4 years) under treatment at the Cystic Fibrosis Outpatient Clinic of the State University at Campinas *Hospital de Clínicas*, located in the city of Campinas, Brazil. At this facility, 120 patients were being followed, and 94 of those patients were between 6 and 27 years of age. All patients who appeared for a routine visit during the period of data collection

(from April of 2008 to June of 2009) were invited to participate in the study.

We applied the CFQ validated for use in Brazil by Rozov et al.[14] The CFQ_{6-11} was applied by the principal author of the study, whereas the other three questionnaires (CFQ_{12-13}, CFQ_{14+}, and $CFQ_{Parents6-13}$) were completed by the patients themselves or by their parents. By means of these instruments, the following domains are assessed: physical; body image; digestive; respiratory; emotional; social; nutrition; treatment; vitality; health; social role; and weight. The scores for each domain range from 0 to 100, high scores indicating better QoL. In general, scores above 50 are considered to reflect good QoL.

The patients were divided into three groups by age: those ≥ 6 and < 12 years of age (CFQ_{6-11} group); those ≥ 12 and < 14 years of age (CFQ_{12-13} group); and those ≥ 14 years of age (CFQ_{14+} group).

From a genetic standpoint, the patients were grouped into ΔF508 homozygotes, ΔF508 heterozygotes, and those with other mutations.

Disease severity was assessed by Shwachman score: < 40 = severe; 41-55 = moderate; 56-70 = mild; 71-85 = very mild; and 86-100 = extremely mild.[15] For the purposes of analysis, the patients were divided into two groups: those with Shwachman scores of 71-100 and those with Shwachman scores ≤ 70.

The most recent spirometry results were evaluated; the cut-off point for FVC, FEV_1, and the FEV_1/FVC ratio was 80% of predicted, whereas the cut-off point for $FEF_{25-75\%}$ was 70% of predicted.[16]

The Statistical Package for the Social Sciences, version 16.0 (SPSS Inc., Chicago, IL, USA), was used for data processing. The scores are expressed as means and standard deviations or as medians and ranges. The Mann-Whitney test was used for comparing two groups, whereas the Kruskal-Wallis test was used for comparing more than two groups. The level of significance was set at 5%.[17]

This study was approved by the Research Ethics Committee of the State University at Campinas School of Medical Sciences (Ruling

Table 1 - Data distribution by gender, race, per capita income, genetics, Shwachman score, and physical therapy in cystic fibrosis patients, by group.[a]

Characteristic	Group		
	CFQ_{6-11}	CFQ_{12-13}	CFQ_{14+}
	(n = 39)	(n = 12)	(n = 24)
Gender			
Male/Female, n/n	16/23	4/8	14/10
Race			
White/non-White, n/n	33/6	11/1	22/2
Per capita income, number of times the national minimum wage			
< 0.5	11 (28.2)	2 (16.7)	3 (12.5)
0.5 |– 1	9 (23.1)	4 (33.3)	11 (45.8)
1 |– 2	8 (20.5)	2 (16.7)	6 (25.0)
≥ 2	11 (28.2)	4 (33.3)	4 (16.7)
Genetics			
DF508 homozygote	13 (33.3)	3 (25.0)	6 (25.0)
DF508 heterozygote	13 (33.3)	8 (66.7)	4 (16.7)
Other mutations	13 (33.3)	1 (8.3)	14 (58.3)
Shwachman score			
71-100 (very mild/extremely mild)	27 (69.2)	8 (66.7)	10 (41.7)
56-70 (mild)	7 (17.9)	3 (25.0)	8 (33.3)
≤ 55 (moderate/severe)	5 (12.8)	1 (8.3)	6 (25.0)
Physical therapy			
No	11 (28.2)	6 (50.0)	14 (58.3)
Yes (from a professional)	28 (71.8)	6 (50.0)	10 (41.7)

CFQ: Cystic Fibrosis Questionnaire; CFQ_{6-11}: patients ≥ 6 and < 12 years of age; CFQ_{12-13}: patients ≥ 12 and < 14 years of age; and CFQ_{14+}: patients ≥ 14 years of age. [a]Values expressed as n (%), except where otherwise indicated.

no. 027/2008). All subjects completed the questionnaire only after giving written informed consent to participate in the study.

Results

The mean age of the patients was 12.5 ± 5.1 years (range: 6.1-26.4 years). Table 1 shows the distribution of the data by gender, race, per capita family income, and Shwachman score.

Of the 39 patients in the CFQ_{6-11} group, 28 (71.8%) received physical therapy from a professional, as did 6 (50.0%) of the 12 patients in the CFQ_{12-13} group and 10 (41.7%) of the 24 patients in the CFQ_{14+} group (Table 1).

Table 2 shows the spirometry data. Of the 75 patients evaluated, 41 had an FEV_1 < 80% of predicted (mean = 57.2 ± 16.4%), 30 had an FVC < 80% (mean = 60.5 ± 13.3%), 41 had an $FEF_{25-75\%}$ < 70% (mean = 37.4 ± 17.5%), and 29 had an FEV_1/FVC ratio < 80% of predicted (mean = 60.9 ± 15.5%). In the CFQ_{6-11} group, 14 patients (42.4%) had an FEV_1 < 80% of predicted, as did 6 (50.0%) of those in the CFQ_{12-13} group and 21 (87.5%) of those in the CFQ_{14+} group.

In the CFQ_{6-11} group, the mean scores ranged from 71.4 in the physical domain to 83.5 in the treatment domain. In the CFQ_{12-13} group, the mean scores ranged from 59.5 in the social domain to 75.0 in the digestive and nutrition domains. In the CFQ_{14+} group, the mean scores ranged from 63.9 in the weight domain to 89.3 in the nutrition domain. When we compared the mean scores of the three groups, we observed a statistically significant difference only between the CFQ_{6-11} group and the CFQ_{14+} group and only in the treatment domain (p = 0.001; Table 3).

Table 4 shows the data regarding the questionnaires completed by the parents. We found a significant difference between the CFQ_{6-11} and CFQ_{12-13} groups only in the emotional domain (p = 0.043). We observed good agreement between the mean scores on the $CFQ_{Parents6-13}$ and the information obtained by means of the questionnaires applied to the patients.

The CFQ_{6-11} group patients with Shwachman scores of 71-100 scored higher for the social domain than did other patients (76.4 ± 15.2 vs. 65.5 ± 17.6; p = 0.045); whereas, in the CFQ_{12-13} group, the same was true for the respiratory domain (79.2 ± 12.6 vs. 50.0 ± 34.0; p = 0.042). In the CFQ_{14+} group, there were no such differences. Among the patients with Shwachman scores of 71-100, the $CFQ_{Parents6-13}$ scores for the physical domain were higher when given by parents of CFQ_{6-11} group patients than when given by parents of CFQ_{12-13} group patients (90.2 ± 14.1 vs. 74.1 ± 25.3; p = 0.032), whereas the inverse was true for the nutrition domain (74.1 ± 30.4 vs. 93.1 ± 19.4; p = 0.019). In the CFQ_{12-13} group, patients with Shwachman scores of 71-100 scored higher for the body image

Table 2 - Distribution of the spirometry values in cystic fibrosis patients, by group.[a]

Parameter	Mean ± SD	Group		
		CFQ_{6-11}	CFQ_{12-13}	CFQ_{14+}
		(n = 39)[b]	(n = 12)	(n = 24)
FVC				
> 80% of predicted	96.0 ± 12.9%	23 (69.7)	7 (58.3)	9 (37.5)
< 80% of predicted	60.5 ± 13.2%	10 (30.3)	5 (41.7)	15 (62.5)
FEV_1				
> 80% of predicted	95.9 ± 10.6%	19 (57.6)	6 (50.0)	3 (12.5)
< 80% of predicted	57.2 ± 16.4%	14 (42.4)	6 (50.0)	21 (87.5)
FEV_1/FVC				
> 80% of predicted	90.9 ± 13.3%	26 (78.8)	8 (66.7)	6 (25.0)
< 80% of predicted	60.9 ± 15.5%	7 (21.2)	4 (33.3)	18 (75.0)
$FEF_{25\%-75\%}$				
> 70% of predicted	93.9 ± 19.5%	19 (57.6)	5 (41.7)	4 (16.7)
< 70% of predicted	37.4 ± 17.5%	14 (42.4)	7 (58.3)	20 (83.3)

CFQ: Cystic Fibrosis Questionnaire; CFQ_{6-11}: patients ≥ 6 and < 12 years of age; CFQ_{12-13}: patients ≥ 12 and < 14 years of age; and CFQ_{14+}: patients ≥ 14 years of age. [a]Values expressed as n (%), except where otherwise indicated. [b]Six patients in the CFQ_{6-11} group did not undergo spirometry.

Table 3 - Quality of life scores from the questionnaires related to cystic fibrosis patients, by group.[a]

Domain	Group						p*
	CFQ_{6-11}		CFQ_{12-13}		CFQ_{14+}		
	(n = 39)		(n = 12)		(n = 24)		
Physical	71.4 ± 25.3	77.8 (16.7-100.0)	66.2 ± 28.1	72.2 (5.6-100.0)	72.4 ± 24.9	70.8 (20.8-100.0)	0.824
Body image	77.5 ± 25.9	88.9 (0.0-100.0)	63.0 ± 23.4	61.1 (22.2-100.0)	78.7 ± 20.7	77.8 (33.3-100.0)	0.082
Digestive	78.6 ± 22.3	66.7 (0.0-100.0)	75.0 ± 20.7	66.7 (33.3-100.0)	88.4 ± 14.5	100.0 (66.7-100.0)	0.097
Respiratory	74.4 ± 16.0	75.0 (33.3-100.0)	69.4 ± 25.0	75.0 (0.0-100.0)	68.7 ± 15.4	75.0 (33.3-83.3)	0.493
Emotional	79.4 ± 11.5	79.2 (58.3-100.0)	71.5 ± 19.0	75.0 (25.0-100.0)	82.8 ± 15.2	86.7 (46.7-100.0)	0.129
Social	73.0 ± 16.5	71.4 (42.9-100.0)	59.5 ± 22.3	64.3 (14.3-90.5)	68.1 ± 16.1	66.7 (33.3-94.4)	0.168
Nutrition	79.8 ± 22.1	88.9 (22.2-100.0)	75.0 ± 28.5	77.8 (11.1-100.0)	89.3 ± 16.2	100.0 (55.6-100.0)	0.129
Treatment	83.5 ± 14.4	88.9 (33.3-100.0)	70.4 ± 24.3	77.8 (22.2-100.0)	64.8 ± 19.0	66.7 (33.3-100.0)	0.001**
Vitality					76.7 ± 16.7	83.3 (50.0-100.0)	
Health					65.3 ± 26.1	66.7 (22.2-100.0)	
Social role					77.4 ± 20.5	79.2 (33.3-100.0)	
Weight					63.9 ± 38.0	66.7 (0.0-100.0)	

CFQ: Cystic Fibrosis Questionnaire; CFQ_{6-11}: patients ≥ 6 and < 12 years of age; CFQ_{12-13}: patients ≥ 12 and < 14 years of age; and CFQ_{14+}: patients ≥ 14 years of age. [a]Values expressed as mean ± SD and as median (range). *Kruskal-Wallis test. **CFQ_{6-11} group vs. CFQ_{14+} group.

Table 4 - Quality of life scores from the questionnaires completed by the parents of patients with cystic fibrosis.[a]

Domain	Group				p*
	CFQ_{6-11}		CFQ_{12-13}		
	(n = 39)		(n = 12)		
Physical	85.3 ± 19.5	92.6 (25.9-100.0)	79.3 ± 23.2	85.2 (14.8-100.0)	0.167
Body image	81.5 ± 26.7	88.9 (0.0-100.0)	74.1 ± 29.3	77.8 (0.0-100.0)	0.295
Digestive	77.2 ± 20.4	77.8 (22.2-100.0)	74.1 ± 18.6	77.8 (33.3-100.0)	0.511
Respiratory	78.5 ± 15.9	77.8 (38.9-100.0)	80.6 ± 13.9	83.3 (55.6-100.0)	0.737
Emotional	86.2 ± 14.3	93.3 (33.3-100.0)	73.9 ± 22.6	83.3 (20.0-100.0)	0.043
Social	83.4 ± 18.3	88.9 (44.4-100.0)	77.8 ± 28.8	88.9 (22.2-100.0)	0.750
Nutrition	79.9 ± 28.7	100.0 (0.0-100.0)	70.8 ± 28.5	66.7 (33.3-100.0)	0.303
Treatment	78.1 ± 19.8	77.8 (33.3-100.0)	82.4 ± 20.4	83.3 (33.3-100.0)	0.452
Vitality	74.4 ± 15.9	80.0 (33.3-100.0)	71.1 ± 20.4	80.0 (26.7-93.3)	0.805
Health	80.3 ± 22.4	88.9 (11.1-100.0)	73.1 ± 27.0	77.8 (0.0-100.0)	0.246
Weight	60.7 ± 37.4	66.7 (0.0-100.0)	50.0 ± 43.8	50.0 (0.0-100.0)	0.487

CFQ: Cystic Fibrosis Questionnaire; CFQ_{6-11}: patients ≥ 6 and < 12 years of age; and CFQ_{12-13}: patients ≥ 12 and < 14 years of age. [a]Values expressed as mean ± SD and as median (range). *Mann-Whitney test.

domain than did other patients (86.1 ± 19.5 vs. 50.0 ± 33.3; p = 0.023) and the respiratory domain (88.2 ± 8.6 vs. 65.3 ± 8.3; p = 0.005).

In the CFQ_{6-11} and CFQ_{12-13} groups, the comparison between patients with an FEV_1 < 80% of predicted and other patients revealed no statistically significant difference for any domain, whereas, in the CFQ_{14+} group, scores for the following domains were lower in patients with an FEV_1 < 80% of predicted: physical (68.4 ± 24.1 vs. 100.0 ± 0.0; p = 0.012); body image (75.7 ± 20.4 vs. 100.0 ± 0.0; p = 0.031); respiratory (66.7 ± 15.4 vs. 83.3 ± 0.0; p = 0.023); emotional (80.6 ± 15.0 vs. 97.8 ± 3.9; p = 0.041); and social role (74.2 ± 19.9 vs. 100.0 ± 0.0; p = 0.024). In the evaluation of the $CFQ_{Parents6-13}$ scores, no statistically significant difference was found for any domain in the CFQ_{6-11} group, whereas, in the CFQ_{12-13} group, scores for the digestive domain were higher for the patients with an FEV_1 < 80% of predicted (87.0 ± 8.4 vs. 61.1 ± 16.8; p = 0.008).

Discussion

Our data indicate that the patients with CF followed at the center under study have good QoL, evidence of impaired QoL being observed among those > 14 years of age with an FEV_1 < 80% of predicted.

At our facility, disease severity in patients with CF is determined by the Shwachman scoring system, whereas the impact of the disease on the respiratory tract is determined by spirometry. Other methods, such as the Cystic Fibrosis Clinical Score and the six-minute walk test, are also used. Respiratory tract infections are monitored by microbiological techniques. The objective of all of these methods is to determine disease severity and the potential risk of respiratory tract infection. However, these methods cannot translate the patient perception of what it is like to live with a fatal chronic illness and therefore tell us little about the daily routine of patients and how they live.

Our results show that the QoL of patients ≥ 6 and < 14 years of age is satisfactory in all domains (health, physical functioning, psychosocial functioning, and treatment burden), whereas adolescents and adults report dissatisfaction with the treatment. This is possibly due to the fact that children are less anxious, less depressed, and more optimistic about fighting the disease than are adults.[18-20] In addition, self-perceived QoL varies from individual to individual and as values, beliefs, cultural context, and social context change their views over the years.[13,19]

Symptoms of anxiety and depression in adolescents and adults correlate with low scores on instruments designed to quantify QoL and have been described as risk factors for poor treatment compliance, increased morbidity, and increased health care use in chronic diseases.[19,21]

Among the parents of the children with CF who participated in this study, lower scores were observed only for the weight domain, suggesting

parental dissatisfaction with the gastrointestinal functioning of their children.

Some studies have shown differences between parent and child reporting of QoL, especially regarding physical and emotional aspects.[19,20,22-24] This is probably due to the high level of emotional distress experienced by parents. The health problems of children with CF limit their participation in physical activities, as well as in school and family activities, causing caregivers anxiety and depression.[23,24] In addition, concerns about the life expectancy, together with the expensive and demanding treatment regimens, contribute to the onset of depressive symptoms in caregivers, directly affecting activities of daily living and treatment compliance.[24]

Our study revealed that children with low Shwachman scores (≤ 70) show dissatisfaction with their social functioning (CFQ_{6-11} group) and their respiratory function (CFQ_{12-13} group), when compared with those with Shwachman scores of 71-100. These results suggest that disease severity correlates with the QoL of children. However, there have been no studies capable of detecting these correlations, possibly due to the predominant use of generic questionnaires. In adolescents and adults, our results showed no correlation between self-reported QoL and disease severity, as has been shown in other studies.[6,19,25]

Pulmonary impairment has been described in the literature as an important factor for the onset of emotional stress in individuals with CF, since it correlates with higher morbidity and mortality, requiring treatments that are even more demanding.[21,25]

Our results show that children ≥ 6 and < 14 years of age have a satisfactory QoL, regardless of FEV_1. In contrast, adolescents and adults with greater pulmonary impairment had worse scores in the respiratory, social role, body image, and physical domains. Other studies have also described these correlations, especially in the domains related to physical functioning and to body and health perception, in adolescents and adults with altered FEV_1.[21,26,27]

In the literature, there are conflicting results regarding the correlation between pulmonary impairment and reported QoL in individuals with CF. This is due to the use of different types of questionnaires and to the age bracket of the patients. When specific instruments are used, this correlation becomes more consistent, especially among adolescents and adults.[12,13]

Adolescents and adults with CF who have already developed pulmonary impairment have been shown to report worse QoL on the Quality of Well-Being scale.[26] However, in individuals under 18 years of age, no such correlation was observed.[28]

In three studies conducted in the 2000s,[12,13,29] the correlation between QoL and FEV_1 in individuals with CF was assessed by the Child Health Questionnaire, with conflicting results. In two of the studies,[12,13] no such correlation was found. However, the authors of the third study observed that adolescents and adults with abnormal FEV_1 reported a poor perception of their physical health.[29]

In a recent study involving the use of the CFQ, adults with CF and with altered FEV_1 were found to have lower scores in the domains related to physical functioning and health, indicating that anxiety, depression, and pulmonary function correlate with self-reported QoL.[4,21]

Most studies suggest that self-perceived health is directly related to age and to respiratory system impairment, becoming more evident in adolescents and adults with greater lung injury,[4,21,26] as observed in the present study.

The assessment of QoL in individuals with CF is important because it reveals the patient perception of what it is like to live with a fatal chronic illness and can improve treatment compliance.[5,7,12] In addition, it provides information for economic planning and makes it possible to determine the impact of new treatments, as shown in a recent study.[30]

Patients between 6 and 14 years of age have a satisfactory QoL in all domains, whereas adolescents and adults report dissatisfaction with the treatment. We consider the use of QoL questionnaires to be a low-cost, user-friendly method. However, only after these questionnaires have been studied more extensively will they actually be incorporated into the routine of health care facilities. Such assessments, when performed periodically, will provide information for potentially expanding the role of the families who care for and of the professionals who treat these patients.

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About the authors

Milena Antonelli Cohen
Physical Therapist. State University at Campinas *Hospital de Clínicas*, Campinas, Brazil.

Maria Ângela Gonçalves de Oliveira Ribeiro
Physical Therapist. Department of Pediatrics, State University at Campinas School of Medical Sciences, Campinas, Brazil.

Antonio Fernando Ribeiro
Professor. Department of Pediatrics, State University at Campinas School of Medical Sciences, Campinas, Brazil.

José Dirceu Ribeiro
Associate Professor. Department of Pediatrics, State University at Campinas School of Medical Sciences, Campinas, Brazil.

André Moreno Morcillo
Associate Professor. Department of Pediatrics, State University at Campinas School of Medical Sciences, Campinas, Brazil.