ORIGINAL ARTICLES

Validity and Reliability of the St George's Respiratory Questionnaire in Adults With Cystic Fibrosis

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OBJECTIVE: To study self-perceived quality of life in adults with cystic fibrosis (CF), and to assess the validity of the St George's Respiratory Questionnaire (SGRQ) for use in these patients.

PATIENTS AND METHODS: We studied 37 adults with CF who were in stable condition as indicated by their respiratory and nutritional status. Disease severity was assessed by spirometry used in conjunction with a modified National Institutes of Health (NIH) scoring system and the Bhalla scale. Nutritional status was evaluated by measuring height and weight, calculating body mass index, analyzing bioelectric impedance, and performing various laboratory tests. The patients' quality of life was assessed using the SGRQ.

RESULTS: SGRQ scores were higher (indicating poorer quality of life) among patients with CF than in the general population or among patients with chronic obstructive pulmonary disease. Internal consistency coefficients indicated the SGRQ had good reliability (Cronbach's alpha, 0.864). Women with CF tended to score higher than men, although this difference was not statistically significantly. A statistically significant relationship was observed between SGRQ score and severity of pulmonary impairment (the more severe the impairment, the worse the patient's quality of life). Statistically significant positive relationships were found between SGRQ domains and age, body mass index, and body fat percentage. Significant negative relationships were found between these domains and the modified NIH score, the Bhalla score, forced expiratory volume in 1 second (expressed as a percentage of predicted), and somatomedin C and zinc levels.

CONCLUSIONS: Self-perceived quality of life is worse among adults with CF than in the general population or among patients with chronic obstructive pulmonary disease. The SGRQ is a valid instrument for analyzing health-related quality of life in adults with CF as it discriminates very well between different degrees of severity of pulmonary impairment and has acceptable internal consistency.

Key words: *Cystic fibrosis. Quality of life. St George's Respiratory Questionnaire.*

Validez y fiabilidad del Cuestionario Respiratorio de St. George en población adulta con fibrosis quística

OBJETIVO: Estudiar la percepción de la calidad de vida (CV) en una población adulta con fibrosis quística (FQ) y valorar la validez del Cuestionario Respiratorio St. George (SGRQ) en estos pacientes.

PACIENTES Y MÉTODOS: Hemos estudiado a 37 personas adultas con FQ, estables desde el punto de vista respiratorio y nutricional. Se realizó una valoración de la gravedad de la enfermedad mediante espirometría y los sistemas NIH (National Institutes of Health) modificado y Bhalla, así como una valoración nutricional mediante la medición del peso, la talla, el índice de masa corporal, impedanciometría bioeléctrica y parámetros analíticos. Se estimó la percepción de la CV mediante el SGRQ.

RESULTADOS: Las personas con FQ tienen mayores puntuaciones (peor CV) que la población general y que los afectados de enfermedad pulmonar obstructiva crónica (EPOC). El coeficiente alfa de Cronbach de la puntuación total fue de 0,864. Las mujeres con FQ presentan puntuaciones más altas que los varones, aunque sin alcanzar la significación estadística. Se observa una gradación de las puntuaciones (de forma estadísticamente significativa) en función de la gravedad de la afectación pulmonar (a mayor gravedad, peor CV). Se han encontrado relaciones positivas, estadísticamente significativas, entre las dimensiones del SGRQ y la edad, el índice de masa corporal y el porcentaje de masa grasa, y negativas con el NIH modificado, el Bhalla, el volumen espiratorio forzado en el primer segundo (expresado en porcentaje) y las concentraciones de cinc y somatomedina C.

CONCLUSIONES: Los adultos con FQ tienen peor percepción de la CV que la población general y que los afectados de EPOC. El SGRQ es válido para analizar la CV relacionada con la salud de las personas adultas con FQ, ya que discrimina muy bien entre los distintos grados de gravedad de la función pulmonar y presenta una adecuada consistencia interna.

Palabras clave: *Fibrosis quística. Calidad de vida. Cuestionario St. George (SGRQ).*

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Introduction

Cystic fibrosis is an autosomal recessive hereditary disorder. The culprit gene, which was identified in 1989, is located on the long arm of chromosome 7 and codes for the cystic fibrosis transmembrane conductance regulator (CFTR) protein.[1] The *CFTR* gene functions as a chloride channel and mutations give rise to defective chloride transport in epithelial cells affecting primarily the respiratory, hepatobiliary, gastrointestinal, and reproductive systems, the pancreas, and the sweat glands.[2] This defect gives rise to dysfunctions in various exocrine glands, the most important manifestations of which are an increase in electrolytes in sweat, pancreatic insufficiency, and inflammation and infection of the airways.[3] Respiratory involvement is the principal cause of morbidity and mortality in patients with cystic fibrosis and is, together with malabsorption, the most common mode of presentation.[4]

The life expectancy of these patients has increased considerably as a result of advances in care,[5] and it is consequently of paramount importance that this increase in "quantity" of life should also be accompanied by an improvement in quality of life. However, both the progression of the disease itself and the treatments used to manage it often have a negative impact on the patient's perceived quality of life.

Assessment of health-related quality of life (HRQL) in these patients provides additional information on the impact of the disease that cannot be obtained by means of other purely physical tests such as lung function or nutritional status. It is also a useful tool for describing the situation in a comprehensible way for health professionals, patients, and relatives. Certain clinical parameters, such as forced expiratory volume in 1 second (FEV_1) and body mass index (BMI), have prognostic value with respect to morbidity and mortality but are poor predictors of the patients' perceived lack of well-being. A valid and reliable measure is, therefore, needed to quantify perception of quality of life.[6]

HRQL is measured by way of questionnaires. Generic questionnaires, such as the 36-item general health short form questionnaire (SF-36), have been used in adults with cystic fibrosis but do not appear to discriminate well between degrees of disease severity or detect changes over time in HRQL caused by disease progression. Disease-specific questionnaires have also been designed, but none of these have been adequately validated in the Spanish population.[7] The St George's Respiratory Questionnaire (SGRQ) has been validated in the general population in Spain and for several respiratory diseases including asthma, chronic obstructive pulmonary disease (COPD),[8] and clinically stable noncystic fibrosis bronchiectasis.[9]

In light of the above, our objective was to study HRQL in adults with cystic fibrosis using the SGRQ, and to analyze both the validity and internal consistency of this measure in such patients.

Patients and Methods

Selection of Patients

We studied a group of adults with cystic fibrosis who were attending the specialized clinic in the Hospital Carlos Haya in Malaga on a regular basis and who fulfilled the following inclusion criteria: *a)* met the diagnostic criteria for cystic fibrosis[10]; *b)* had completed pubertal development (Tanner stage 4) and had a bone age of 15 years or older; and *c)* had been clinically stable for 2 months before entering the study (no hospital admissions, respiratory exacerbations, or weight gain or loss greater than 3% of their usual weight).

Assessment of Pulmonary Involvement

A detailed clinical history recording the evolution of disease from the time of diagnosis until enrollment in the study was available for each patient. The history also provided information on the degree of involvement of respiratory and digestive systems, as well as details of the other organs and systems affected and a record of the treatment the patient had received for the disease.

These patients attended the cystic fibrosis clinic for check-ups every 2 or 3 months. Clinical variables were recorded routinely at each visit, and treatment was reviewed and adjusted if necessary. A sputum sample was taken for microbiological study including culture on general media, culture on selective media for the pathogens found commonly in patients with cystic fibrosis, and bacterial counts. We analyzed initial colonization with germs commonly found in these patients, considering the first appearance of the germ in sputum (at least 3 positive sputum samples) regardless of its persistence at the time of the study.

Lung function was studied in all patients by way of spirometry with plethysmography. Disease severity was assessed using the modified National Institutes of Health (NIH) scoring system, which is based on clinical, radiographic, and spirometric findings.[11] We also used the Bhalla[12] scale, a scoring system that measures lung involvement on the basis of chest computed tomography. On both of these scales, the lower the final score obtained, the worse the clinical, radiographic, and spirometric (NIH) or radiographic (Bhalla) findings.

Assessment of Nutritional Status

Nutritional status was assessed in all patients using the following examinations and tests:

1. Anthropometric assessment. Height and weight were measured and the BMI calculated. Participants were classified as malnourished when their BMI was under 18.5 kg/m^2. The data for anthropometric parameters published by Alastrué et al[13] for each sex and age were used to estimate ideal weight. Percentage of ideal body weight was calculated using the following formula: [actual weight (kg) × 100]/ideal or desirable weight (kg).

2. Hand dynamometry (Collin Dynamometer, AS Medizintechnik, Tuttlingen, Germany). The mean of 3 consecutive measurements of the dominant limb was recorded. The results were expressed in kilograms.

3. Bioelectrical impedance. Measurements were made with a multifrequency bioimpedance meter (Bioscan Multifrequencia, Tecnología Médica SL, Barcelona, Spain). Three consecutive measurements were taken under baseline conditions (fasting, with an empty bladder, and resting), and the mean value was calculated. Body fat and lean body mass were calculated using the formulas recommended by Pencharz and Azcue[14] and Segal et al.[15]

4. Laboratory tests. The following laboratory tests were carried out: complete blood count and biochemistry including zinc, iron, albumin, and somatomedin C levels (this last was measured by radioimmunoassay).

Other Clinical Aspects

Various clinical characteristics were taken into consideration:

1. Genotype. We classified the *CFTR* genotypes according to the phenotypic effects of the mutations as published in the medical literature and the primary mechanism of the defective *CFTR*.[16] The genotypes were then classified as either severe or mild. The following were including in the group of severe mutations: DF508, N1303K, G542X, 17/17-8 G → A, Q890X, P2055, 1811+1,6kbA → G, 2184insA, 2869insG, 621+1G → T, G551D, 712-1G → T, P2055. A genotype with 2 severe mutations was classified as severe, whereas those with combinations of 2 mild mutations or 1 severe and 1 mild mutation were classified as mild.

2. Abnormal carbohydrate metabolism. After a challenge with an oral glucose load administered in accordance with the recommendations of the American Diabetes Association Report published in 1997, the patients were classified into 3 categories according to whether they were diabetics or had abnormal fasting glucose levels, or carbohydrate intolerance.[17]

Quality of Life

The participants completed the SGRQ, which was designed specifically to measure quality of life in respiratory diseases and has been validated in the Spanish population. The object was to assess their perception of their health and relate this to nutritional status.[8] The self-administered questionnaire was subsequently reviewed in the patient's presence to ensure that all the questions had been answered.

The results, expressed as means (SD), were compared with those of a representative sample of the general population comprising both healthy persons and individuals with COPD.[18]

Statistical Analysis

The statistical analysis was carried out using the SPSS software, version 11.0 for Windows. The results of the questionnaires were entered into a database for analysis. Quantitative variables were expressed as means (SD) and qualitative variables as percentages. For comparisons between groups, the nonparametric Mann–Whitney test was used for quantitative variables and the χ^2 test for qualitative variables. The Kruskal–Wallis test was used to compare quantitative variables between the 3 groups, and the Spearman test to analyze correlations between 2 sets of values. After the possible associations between variables had been identified, a forward multiple regression analysis was applied. The dependent variable was the total score for each of the questionnaire domains and the explanatory variables were those identified in the simple regression analysis. The internal consistency of the SGRQ was analyzed using Cronbach's α. Statistical significance was set at a value of P less than .05.

TABLE 1
Clinical Characteristics and Analytical Data*

	Total (n=37)	Men (n=15)	Women (n=22)
Age, y	24.5 (12)	22.7 (10.3)	25.7 (13.4)
Age at diagnosis, y	11.9 (16.4)	12.5 (14.3)	11.5 (17.9)
Patients diagnosed as adults†	10 (27%)	5 (33%)	5 (22.7%)
Sweat test (conductivity)	106.5 (20.2)	110.3 (14.4)	103.9 (23.4)
Modified NIH score	75.6 (16.4)	82.5 (14.6)	70.8 (16.2)‡
Bhalla scale	15.4 (3.7)	16.9 (3.5)	14.3 (3.5)‡
FEV_1, mL	2258.5 (1187.1)	3109.3 (1154)	1678.5 (81)§
FEV_1, %	63.6 (27.4)	76 (26.6)	55.2 (25.1)‡
FVC, mL	3060.3 (1285.1)	4050 (1214.7)	2385.5 (819)‖
FVC, %	71.1 (21.5)	80.7 (21.2)	64.6 (19.6)‡
FEV_1/FVC	70.3 (14.2)	74.5 (11.7)	67.4 (15.3)
Weight, kg	57.5 (12)	64.2 (12.7)	52.8 (8.7)¶
Percentage ideal weight	91.9 (14.2)	88.4 (14.9)	93.5 (13.5)
Height, cm	162 (8)	167.7 (6.9)	158.2 (6.8)
BMI, kg/m2	21.8 (4)	22.7 (3.9)	21.2 (3.8)
Percentage ideal BMI	101.5 (17.5)	103.4 (17.9)	100.8 (18.2)
BMI <18.5 kg/m^2†	7 (19%)	1 (7.1%)	6 (28.6%)
Lean body mass, kg	42.7 (8.3)	49.7 (7.6)	37.5 (4.1)§
Lean body mass, %	74.8 (9.5)	78.8 (7.4)	72.1 (10.2)
Body fat, kg	14.8 (7.1)	13.7 (5.8)	15.5 (8.2)
Body fat, %	25.2 (9.5)	21.2 (7.3)	27.9 (10.2)
Dynamometry, kg	0.35 (0.12)	0.45 (0.1)	0.28 (0.07)‖
Hemoglobin, g/dL	13.5 (1.4)	14.6 (0.8)	12.8 (1.3)‖
Glucose, mg/dL	102.9 (32.1)	99.3 (30.9)	104.8 (33.8)
Cholesterol, mg/dL	129.3 (36.9)	123.4 (28.3)	134.1 (41.9)
HDL-C, mg/dL	46.7 (10.8)	41.9 (6.4)	50.4 (11.7)¶,‖
LDL-C, mg/dL	64.6 (32.5)	60.9 (27.1)	67.4 (36.9)
Triglycerides, mg/dL	86.9 (36.3)	95.1 (38.3)	77.9 (31.9)
Zinc, µg/dl	73.2 (13.8)	75.4 (12.7)	70.9 (14.4)
Albumin, g/dL	3.9 (0.4)	4.1 (0.4)	3.8 (0.5)‡

*Data are expressed as means (SD) unless otherwise indicated. HDL-C indicates high density lipoprotein cholesterol; LDL-C, low density lipoprotein cholesterol; FEV_1, forced expiratory volume in 1 second; FVC, forced vital capacity; BMI, body mass index; and NIH, National Institutes of Health.
†Data are numbers (percentage). ‡P<.05 between men and women. §P<.005 between men and women. ‖P<.001 between men and women. ¶P<.01 between men and women. (Mann–Whitney test for all comparisons.)

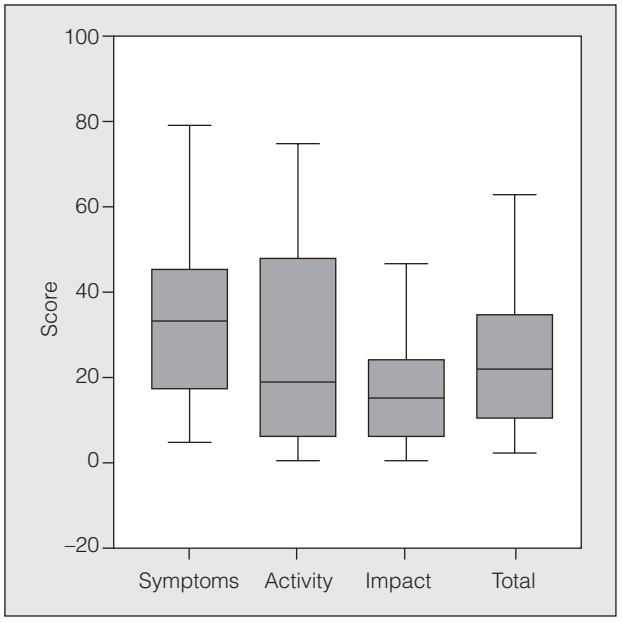

Figure 1. Distribution of St George Respiratory Questionnaire Scores.

Results

Clinical Characteristics

A total of 37 patients (15 men and 22 women) were included in the study. The 2 principal areas of clinical involvement on diagnosis were the respiratory (n=18, 48.6%) and digestive (n=17, 45.9%) systems. The other 2 patients (5%) had been diagnosed in the course of a family study. Overall, 27% of patients had been diagnosed as adults (over 16 years of age). FEV_1 (expressed as a percentage of predicted) was more than 80% in 35% of the participants; between 40% and 80% in 40%; and below 40% in 24%. At some time in their lives, 59% of the patients had been colonized by *Haemophilus influenzae*, 75% by *Pseudomonas aeruginosa* or *Burkholderia cepacia* (previously *Pseudomonas cepacia*), and 81% by *Staphylococcus aureus*. Twenty-three patients (62.2%) had had at least one respiratory exacerbation during the 12-month period before the study. Pancreatic insufficiency was evident in 70%. Seven patients (19%) had diabetes mellitus, although only 3 required pharmacological treatment, and 4 others (11%) had some other carbohydrate metabolism abnormality. Somatomedin C levels were normal in all the participants. Eleven patients (30%) were taking therapeutic dietary supplements, and these represented 18.9% of their total calorie intake (628 [279] kcal/d). Table 1 shows the main clinical and anthropometric characteristics and analytical data for all the participants.

Quality of Life

Table 2 shows the results of the analysis of SGRQ scores with respect to both general description and internal consistency. The figure provides a clearer overall picture of the SGRQ scores obtained.

The correlation (Spearman coefficient) between each item and the scale or domain to which it belongs was analyzed to evaluate the internal consistency and reliability of the questionnaire (defined as the degree of homogeneity of the responses to the set of items within a single domain). Cronbach's α was calculated for the aggregate score of the questionnaire and for each of the 3 domains. The Spearman correlation coefficient for the questionnaire items varied for the different domains. In the symptoms domain it fluctuated between 0.37 and 0.77 and was always significant. In the activity domain, the correlations ranged between 0.06 and 0.82, and most were significant. In the impact domain they ranged between 0.08 and 0.67 and were significant for the most part. Cronbach's α was 0.864 for the aggregate score, 0.61 for the symptoms subscale, 0.87 for the activity subscale, and 0.49 for the impact subscale. The response rate for questions 6 and 8 (symptoms domain) and questions 10 and 14 (impact domain) was under 37% because the response to each of these items was conditioned by the answer to another question. These questions were, therefore, excluded from the calculation of both Cronbach's α coefficient and the correlation coefficients between the 3 domains and their component items.

Table 3 shows the SGRQ results as mean absolute values for the whole group and by sex. No statistically significant differences were observed between the 2 populations, although women tended to score higher.

In order to analyze the effect of lung function impairment on HRQL, we divided the sample into 3 categories: FEV_1 >80%, FEV_1 40%-80%, and FEV_1 <40% (Table 4). A correlation was found between HRQL scores and lung function, with lower SGRQ scores among patients with FEV_1 over 80% and higher SGRQ scores among patients with FEV_1 less than 40%; these differences were significant for all 3 domains.

TABLE 2
General Description and Internal Consistency of the Results of the St George's Respiratory Questionnaire*

Variables	Symptoms	Activity	Impact	Total
Mean (SD) score	35.29 (19.3)	28.9 (25.2)	18.6 (14.6)	24.5 (16.8)
Unanswered items, %†	0.5	0	0.7	0.4
Range	4.42-79.3	0-74.7	0-54.3	1.93-62.4
Ceiling effect	0	0	0	0
Floor effect	0	8	1	0
Item-domain correlation†	0.37-0.77	0.06-0.82	0.08-0.67	
Cronbach's α†	0.61	0.87	0.494	0.864

*Floor effect refers to the number of patients with the minimum score (0 points); ceiling effect, the number of patients with the maximum score (100 points).
†Excluding questions that depend on a response to another item or the patient's situation (numbers 6, 8, 10, and 14).

TABLE 3
Quality of Life (St George's Respiratory Questionnaire)*

	Total (n=37)	Men (n=15)	Women (n=22)
Domains			
Symptoms	35.29 (19.3)	28.69 (16.7)	39.68 (19.96)
Activity	28.9 (25.2)	21.68 (25.2)	33.7 (24.6)
Impact	18.6 (14.6)	17.3 (13.2)	19.5 (15.7)
Total score	24.5 (16.8)	20.5 (16)	27.2 (17.1)

*The data are expressed as means (SD). P=NS, Mann–Whitney test.

TABLE 4
Quality-of-Life Scores by Severity of Lung Impairment*

	FEV_1 >80% (n=12)	FEV_1 = 40%-80% (n=14)	FEV_1 <40% (n=9)	*P*†
Domains				
Symptoms	23.7 (13.4)	35.6 (17.7)	50.3 (19)	.0043
Activity	18.8 (20.8)	24.3 (22.2)	49.6 (25.2)	.01
Impact	12.4 (11)	15.3 (10.1)	32.1 (14.6)	.0025
Total score	16.2 (12.4)	21.4 (13.9)	40.4 (16.3)	.0013

*Data are expressed as means (SD). FEV_1 indicates forced expiratory volume in 1 second.
†*P* between the 3 groups (Kruskal–Wallis).

Analysis of the correlation between HRQL and a history of respiratory exacerbations revealed that patients who had experienced at least 1 exacerbation during the year prior to the study had higher HRQL scores than those who had had no exacerbation; this difference was significant for both the impact domain and the total score, and almost significant for the symptoms domain. The results are shown in Table 5. However, we found no significant differences in quality of life scores between the patients who had a history of colonization by pathogens *(P aeruginosa, B cepacia, S aureus*, and/or *H influenzae)* and those who did not. Nor were any differences found when the results were analyzed by genotype, or by the presence of pancreatic insufficiency, abnormalities in carbohydrate metabolism, or malnutrition (Table 5).

The Spearman correlation coefficient between the scores for each of the 3 questionnaire domains and the total score ranged from 0.63 to 0.94 ($P<.0001$). Analysis of the relationships between quality-of-life scores and various clinical and anthropometric parameters revealed positive correlations between the scores obtained on the questionnaire (higher scores indicating poorer quality of life) and age, BMI, and percent body fat (Table 6). Conversely, negative correlations were found with the Bhalla scale, the NIH modified scoring system, FEV_1, percent lean body mass, and zinc and somatomedin C levels (Table 6).

In the multiple linear regression analysis, the variables age, BMI, and FEV_1 (expressed as a percentage of predicted value) explained 54% (r^2=0.54) of the variance in the overall quality-of-life score, 37% in the symptoms domain, 44% in the activity domain, and 52% in the impact domain.

Discussion

To date we are not aware of any studies in the literature that assess HRQL in adults with cystic fibrosis using the SGRQ. Although this test was not designed specifically for use in patients with cystic fibrosis, we decided to use it because it had been validated for the Spanish population and because earlier studies had shown that deterioration of lung function is the primary cause of decline in quality of life among individuals with this disease.[6,19] Our study demonstrates that the SGRQ has strong internal consistency when it is used in adult patients with cystic fibrosis. Cronbach's α was 0.86, a score high enough to make the questionnaire useful for comparing groups of patients

TABLE 5
Quality-of-Life by Various Clinical Parameters*

		Domains			
	No.	Symptoms	Activity	Impact	Total Score
Exacerbations					
Yes	23	43.1 (19.6)	35 (25.6)	21.7 (14.5)	29.3 (16.6)
No	14	25.8 (15.1)	21 (19.9)	12 (10)†	17 (12)†
Genotype					
Severe	18	34.5 (22)	21.6 (25)	13.9 (14)	19.7 (17)
Mild	19	36.1 (16)	34.3 (23)	21.5 (12)	27.7 (15)
Malnutrition					
No	29	36.1 (19)	29.3 (26)	19.5 (14)	25.2 (17)
Yes	8	32 (23)	21.9 (21)	10.7 (8)	17.7 (10)
Carbohydrate abnormalities					
No	26	39.4 (21)	29.4 (24)	18.4 (13)	24.2 (16)
Yes	11	33.6 (19)	23.9 (27)	15.8 (15)	22.2 (17)
Exocrine pancreatic insufficiency					
No	11	35.4 (16)	38.4 (21)	22.4 (11)	27.9 (11)
Yes	26	35.3 (21)	23.8 (25)	16.1 (14)	22.2 (17)
Albumin					
>3.5 g/dL	31	32.5 (19)	24.7 (23)	15.6 (12)	21.2 (15)
<3.5 g/dL	6	49.9 (14)	44.6 (25)	28.5 (18)	37 (18.6)

*The data are expressed as means (SD).
†*P* <.05, Mann–Whitney test.

TABLE 6
Correlation Coefficients Between the St George's Respiratory Questionnaire Quality-of-Life Domains and Clinical, Anthropometric, and Analytic Parameters*

	Symptoms		Activity		Impact		Total	
	r	*P*	*r*	*P*	*r*	*P*	*r*	*P*
Age	0.49	.003	0.53	.001	0.51	.002	0.57	.000
Modified NIH	–0.61	.000	–0.47	.004	–0.48	.004	–0.55	.001
Bhalla scale	–0.4	.017	–0.29	.097	–0.34	.049	–0.36	.033
FEV_1,%	–0.46	.006	–0.4	.015	–0.4	.018	–0.46	.006
BMI	0.15	.4	0.38	.024	0.48	.003	0.42	.011
Lean body mass,%	–0.61	.000	–0.61	.000	–0.59	.001	–0.65	.000
Body Fat,%	0.61	.000	0.61	.000	0.59	.001	0.64	.000
Zinc	–0.4	.02	–0.38	.026	–0.39	.024	–0.43	.011
Somatomedin C	–0.53	.002	–0.53	.002	–0.57	.001	–0.6	.000
Albumin	–0.23	.19	–0.16	.36	–0.26	.13	–0.24	.18
Glucose	0.35	.037	0.23	.18	0.21	.24	0.27	.12
Cholesterol	0.12	.49	0.28	.1	0.13	.46	0.21	.23
Dynamometry	–0.36	.038	–0.24	.16	–0.1	.57	–0.23	.2

* NIH indicates National Institutes of Health; FEV_1, forced expiratory volume in 1 second; BMI, body mass index.

(according to Nunnally,[20] an α value above 0.7 is adequate for intergroup comparisons).

The SGRQ has 3 component domains: symptoms, activity, and impact. The first of these measures the frequency and presence of symptoms such as dyspnea, cough, and expectoration. The second evaluates how the patients' activities are limited by dyspnea (for example, walking more slowly than others, having to stop and rest when climbing stairs, or being unable to ride a bicycle). The third domain assesses more psychosocial aspects, such as the problems that may be associated with taking medication.

In our study, patients with cystic fibrosis scored higher on the SGRQ than a reference population,[18] and these higher scores represent a poorer quality of life despite the fact that the study group was younger. They also perceived their health as being worse than did an older reference population of patients with COPD.[18]

Scores in the reference population were significantly higher among the older cohorts, an unsurprising finding since many individuals in such older cohorts have respiratory diseases and other chronic conditions. Similarly, a strong positive correlation was also found in our sample between all 3 SGRQ domains and age (the older the patient was, the poorer their quality of life), a finding possibly related to progressive deterioration of lung function and other factors difficult to evaluate in this study. These findings are similar to those reported in earlier studies using questionnaires designed specifically for patients with cystic fibrosis.[21,22]

When we analyzed the results by sex, we found that the women tended to have higher scores than men (although this difference was not significant). However, in the reference population of COPD patients, men scored higher than women.[18] This difference may be due to the fact that the women in our study were in worse clinical condition than the men; their lung function impairment was greater and they tended to score lower on both the modified NIH system and the Bhalla scale (with statistically significant differences in all cases). These results were similar to the findings of earlier studies using disease-specific quality-of-life questionnaires in patients with cystic fibrosis.[7,22,23] However, these differences between men and women were not reported in other studies using generic questionnaires in these patients.[19]

When the results were analyzed according to FEV_1 (>80%, 40%-80%, <40%), we found a statistically significant correlation in all 3 domains between severity of lung function impairment and SGRQ scores. This finding supports the validity of the questionnaire, confirming that it discriminates adequately between groups of patients according to the severity of their lung function impairment. Many studies confirm this correlation between lung function and generic or disease-specific quality-of-life questionnaires in patients with cystic fibrosis.[7,21,22,24] These findings are also consistent with the results of studies using the SGRQ in patients with noncystic fibrosis bronchiectasis.[9]

As expected, we found correlations between the SGRQ domains and the modified NIH, a scoring system that measures clinical and radiographic parameters and includes certain aspects related to quality of life. We also found correlations with the Bhalla scale, although they were less marked than the correlations with the modified NIH. The correlation with the activity domain was not statistically significant probably because the Bhalla system is based solely on radiographic parameters.

Patients with a history of at least 1 exacerbation of their respiratory disease during the 12 months prior to the study scored higher, and this difference was statistically significant for both the impact domain and the aggregate score. This difference may possibly be related to the psychosocial problems caused by the impact of frequent respiratory exacerbations on leisure activities, work situation, psychological well-being, and so on.

Unlike the authors who used measures designed specifically for cystic fibrosis, we found no significant changes in SGRQ scores when the results were analyzed by genotype, BMI, or the presence of pancreatic insufficiency or abnormalities in the metabolism of carbohydrates.[7,22] Perhaps this was because the SGRQ is

specific for respiratory diseases but does not discriminate well between different types of associated systemic involvement.

Unlike other authors, we found no significant differences between the SGRQ scores of malnourished (BMI <18.5 kg/m^2) and adequately nourished patients. In our study, BMI and percent body fat correlated with the total SGRQ score and the activity and impact domains, indicating that more obese patients have a poorer perception of their quality of life. This finding contradicts the results of earlier studies of disease-specific questionnaires for cystic fibrosis.[7,21,25,26] Possibly the results of our study were affected by the presence in our sample of several older obese and overweight patients who had been diagnosed as adults. While these patients did not present digestive involvement, they had high scores due to the severity of their respiratory impairment. In fact, in the multiple regression analysis the combination of age, BMI, and FEV_1 explained a great part of the variance in all the SGRQ domains (between 37% for the symptoms domain and 54% for the overall score). Moreover, we found a negative correlation between SGRQ scores and certain parameters indicative of muscular function and lean body mass including dynamometry and somatomedin C levels; the lower the results were for these parameters, the worse the patient's quality of life. This finding may indicate that quality of life scores are, to some degree, dependent on the patient's functional nutritional state. Zinc levels were also lower in patients with poorer quality of life. Zinc is related to nutritional status, and zinc levels decrease through malabsorption in inflammatory states.[27] No statistically significant relationship was found between the SGRQ scores and other nutritional parameters related to morbidity and mortality that are reduced in inflammatory states (albumin for example).[28] However, patients with low albumin levels (<3.5 g/dL) scored higher on the SGRQ (Tables 5 and 6).

In summary, patients with cystic fibrosis present a poorer quality of life than the general population and adults with COPD.[18] The SGRQ is a valid instrument for assessing HRQL in such patients because it has acceptable internal consistency, discriminates well between differing degrees of pulmonary impairment, and correlates significantly with measures of severity such as the NIH score and the Bhalla scale.

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