Perception of disease severity in adult patients with cystic fibrosis

Percepção da gravidade da doença em pacientes adultos com fibrose cística

Paulo de Tarso Roth Dalcin, Greice Rampon, Lílian Rech Pasin, Sinara Corrêa Becker, Gretchem Mesquita Ramon, Viviane Ziebell de Oliveira

Abstract

Objective: To evaluate the perception of disease severity in patients with cystic fibrosis (CF), investigating its relationship with clinical score, radiographic score, respiratory function tests, adherence to treatment and perception of self-care practices. Methods: Prospective, cross-sectional study involving CF patients treated in a program for adults with CF. The perception of disease severity, adherence to treatment and reported self-care practices were evaluated by means of questionnaires. Clinical data, Shwachman-Kulczycki clinical score, Brasfield radiographic score and spirometry were obtained for all of the patients. Results: Of the 38 patients studied, 3 (7.9%) patients rated their perception of health status as well below average; 5 (13.2%), as below average; 15 (39.5%), as average; 10 (26.3%), as above average; and 5 (13.2%), as well above average. The perception of disease severity correlated significantly with clinical score ($r = 0.43$, $p = 0.007$), FVC ($r = 0.34$, $p = 0.034$), FEV1 ($r = 0.38$, $p = 0.019$) and self-care practices ($r = 0.33$, $p = 0.044$), but not with degree of adherence ($r = -0.03$, $p = 0.842$) and radiographic score ($r = 0.33$, $p = 0.51$). Conclusions: The perception of disease severity correlated with objective measurements of disease severity (clinical score and respiratory function tests) and with reported self-care practices, but not with adherence to treatment.

Keywords: Severity of illness index; Cystic fibrosis; Patient compliance; Respiratory function tests.

Resumo

Objetivo: Avaliar a percepção da gravidade da doença em pacientes com fibrose cística (FC), investigando sua relação com escore clínico, escore radiológico, testes de função pulmonar, adesão ao tratamento e percepção de autocuidado. Métodos: Estudo transversal, prospectivo, envolvendo pacientes com FC atendidos em um programa para adultos com FC. A percepção da gravidade da doença, a adesão ao tratamento e o relato de autocuidado foram avaliados por questionários. Foram obtidos de todos os pacientes dados clínicos, escore clínico de Shwachman-Kulczycki, escore radiológico de Brasfield e espirometria. Resultados: De 38 pacientes estudados, 3 (7,9%) relataram percepção de sua saúde como muito abaixo da média; 5 (13,2%), como abaixo da média; 15 (39,5%), como na média; 10 (26,3%), como acima da média; e 5 (13,2%), como muito acima da média. A percepção da gravidade da doença correlacionou-se significativamente com o escore clínico ($r = 0,43$, $p = 0,007$), CVF ($r = 0,34$, $p = 0,034$), VEF1 ($r = 0,38$, $p = 0,019$) e com relato de autocuidado ($r = 0,33$, $p = 0,044$), mas não com o grau de adesão ($r = -0,03$, $p = 0,842$) e escore radiológico ($r = 0,33$, $p = 0,51$). Conclusões: A percepção da gravidade da doença se relacionou com medidas objetivas de gravidade da doença (escoros clínico e testes de função pulmonar) e com relato de autocuidado, mas não com a adesão ao tratamento.

Descritores: Índice de gravidade de doença; Fibrose cística; Cooperação do paciente; Testes de função pulmonar.

Introduction

Cystic fibrosis (CF) is a genetic disease of autosomal recessive inheritance.$^{1,2}$ In general, CF presents as multisystemic impairment, characterized by progressive lung disease, exocrine pancreatic insufficiency, liver disease, intestinal motility disorder, male infertility and high concentrations of sweat electrolytes.$^{3,4}$ It is an irreversible disease, and, until recently, its evolution did not allow patients to survive until adolescence. However, in recent decades,
advances in the knowledge and treatment of CF have increased the survival of these patients.\(^5\)

The increased longevity of CF patients has resulted in a greater proportion of age-related medical problems and complications related to disease progression, requiring an increasingly complex treatment program.\(^6\) This treatment requires lifelong patient adherence to an extensive self-care program.\(^7\-9\)

One of the aspects that can determine how involved individuals are in their treatment is the perception that these individuals have of the severity of their disease.\(^10\) Strategies to improve patient health need to take into consideration patient attitudes toward and perceptions of the disease.\(^11\)

The objective of the present study was to evaluate the perception of disease severity in patients with CF, investigating its relationship with clinical score, radiographic score, respiratory function tests, adherence to treatment and perception of self-care practices.

**Methods**

This was a prospective, cross-sectional study, carried out at a single center. The study was approved by the Ethics in Research Committee of the Hospital de Clínicas de Porto Alegre (HCPA, Porto Alegre Hospital de Clínicas). All patients aged 18 or older, or legal guardians in the case of patients under 18 years, gave written informed consent.

The study population comprised patients monitored via the HCPA Program for Adults with CF.

We included patients aged 16 or older and diagnosed with CF in accordance with the consensus criteria.\(^3\-4\,12\) In addition, patients were required to have been clinically stable for the preceding 30 days. Clinical stability was defined as no clinical evidence of exacerbation, no modifications in the therapeutic regimen and no hospitalizations.

Patients who failed to complete any of the questionnaires used in the study were excluded.

The clinical characteristics of the disease were obtained using a specific form. The inclusion in the study and subsequent clinical evaluation were performed in an outpatient visit.

The Shwachman-Kulczycki clinical evaluation score was used.\(^13\) The score was always determined by the same member of the team of physicians. This score comprises four criteria: general activity; physical examination; nutritional aspects; and chest X-ray findings. Each of the criteria is scored on a scale ranging from 5 to 25 points (better performance equals higher score). The maximum score is 100 points and represents a patient in excellent clinical conditions.

Spirometry and conventional chest X-rays were performed within 7 days of the study evaluation.

Spirometry was performed using a spirometer (v. 4.31a; Jaeger, Würzburg, Germany), in accordance with the guidelines currently available.\(^14\) We determined the postbronchodilator values of FVC, FEV\(_1\), and the FEV\(_1\)/FVC ratio, which are expressed in percentage of predicted for gender, age and height.\(^15\) A pulse oximeter (NPB-40; Nellcor Puritan Bennett, Pleasanton, CA, USA) was used to measure SpO\(_2\) at rest and in room air.

All individuals underwent anterior and lateral chest X-rays. The radiographic score was determined by a senior pulmonologist using the Brasfield et al. radiographic scoring system.\(^16\) The pulmonologist was blinded to patient clinical status, patient identity and the study outcome measures. In this scoring system, the following characteristics are scored in ascending order by degree of severity: air trapping (0 to 4); linear markings (0 to 4); nodular cystic lesions (0 to 4); extensive lesions in the air space (0, 3 or 5); and overall severity (0 to 5). The total score is obtained using the following formula:

\[
25 - \text{the total score obtained in the 5 characteristics examined}
\]

The maximum score is 25 points, and corresponds to a chest X-ray with no alterations, whereas the minimum score is 3 points, and corresponds to a chest X-ray presenting considerable alteration.

Adherence was evaluated using a questionnaire adapted from another study.\(^17\) The questionnaire addressed the following therapeutic aspects: question 1) respiratory therapy; question 2) physical activity; question 3) hypercaloric diet; question 4) pancreatic enzymes; question 5) vitamins A, D, E and K; question 6) antibiotic nebulization; and question 7) DNase nebulization. For each question, the patient was instructed to indicate the weekly frequency of use of the therapeutic modality considered: a) every
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day or almost every day; b) approximately 3-5 days per week; or c) less than 3 days per week or never. Questions 3 to 7 also included a fourth option (option d): “not indicated”. Each question was scored as follows: option a = 2 points; option b = 1 point; and option c = 0 points. An adherence score was calculated based on the quotient between the total score obtained and the number of points possible. A score of 1 would correspond to full adherence, and a score of 0 would correspond to no adherence. For questions 3 through 7, the responses were excluded from the calculation of the overall score if option d was checked.

Patient perception of disease severity and self-care practices was evaluated using questions adapted from another study.(18) The questions were formulated and presented as follows:

“This question refers to how you see the severity of your disease. We would like you to read it carefully and check the option that best describes your perception. In comparison with that of other patients with CF, how would you rate your health status: a) well below average; b) below average; c) average; d) above average; or e) well above average?” Responses to this question were scored on a scale of 1-5 (a = 1 and e = 5).

“This question refers to the degree to which you are personally involved in managing your disease. We would like you to read it carefully and check the option that best describes your perception. Overall, how would you rate your self-care practices: a) very poor; b) poor; c) fair; d) good; e) very good; or f) excellent?” Responses to this question was scored on a scale of 1-6 (a = 1 and f = 6).

The questions were administered outside the ambulatory care environment by a member of the research team who was not involved in the treatment of the patient.

Statistical analysis

Data were entered into a Microsoft® Excel 2000 database, after which they were processed and analyzed using the Statistical Package for the Social Sciences program, version 13.0 (SPSS Inc., Chicago, IL, USA), and the Number Cruncher

Figure 1 - Correlations of the perception of disease severity with clinical score, FEV₁, radiographic score and perception of self-care practices. Perception of disease severity: 1 = well below average; 2 = below average; 3 = average; 4 = above average; and 5 = well above average.
A descriptive analysis of the variables under study was performed. Quantitative data were expressed as mean ± standard deviation or as median and interquartile range (IQR). Qualitative data are expressed as n and % of all cases.

The perception of disease severity was considered an outcome measure, whereas clinical score, pulmonary function tests, radiographic score and perception of self-care practices were considered independent variables.

Spearman’s linear correlation test was used to determine the correlation between the perception of disease severity and the quantitative variables of the study. This correlation analysis took into consideration the perception of disease severity in five groups of patients, which were formed based on the five categories of perception of disease severity previously described.

In order to compare the groups, the classification of the perception of disease severity was summarized into three categories—well below or below average, average and above or well above average—and the five groups were thus merged into three. Continuous variables were compared using one-way ANOVA or the Kruskal-Wallis test. Tukey’s post hoc test was used for variables with normal distribution. The Kruskal-Wallis Z test was used for variables without normal distribution. Qualitative data were analyzed using the chi-square test.

The level of statistical significance was set at p < 0.05. All statistical tests used were two-tailed.

Results

Between August of 2005 and February of 2006, 41 patients treated in a program for adults with CF were eligible for inclusion in the study. Of those 41 patients, 2 declined to participate in the study, and there was one case in which the patient did not submit to clinical evaluation during the study period. Therefore, the final sample consisted of 38 patients.

Of the 38 patients included, 20 were female and 18 were male. The mean age was

<table>
<thead>
<tr>
<th>Variable</th>
<th>Perception</th>
<th>Well below or below average (n = 8)</th>
<th>Average (n = 15)</th>
<th>Above or well above average (n = 15)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, years</td>
<td>Well below or below average</td>
<td>23.0 ± 4.0</td>
<td>24.7 ± 8.3</td>
<td>23.3 ± 5.9</td>
<td>0.788</td>
</tr>
<tr>
<td>Gender, male/female</td>
<td>Average</td>
<td>3/5</td>
<td>7/8</td>
<td>8/7</td>
<td>0.767</td>
</tr>
<tr>
<td>Age at diagnosis, years</td>
<td>Above or well above average</td>
<td>11.0 (15.3)</td>
<td>9.0 (19.0)</td>
<td>11 (14.5)</td>
<td>0.904</td>
</tr>
<tr>
<td>Marital status, n (%)</td>
<td>Single</td>
<td>8 (21.1)</td>
<td>12 (31.6)</td>
<td>11 (28.9)</td>
<td>0.276</td>
</tr>
<tr>
<td></td>
<td>Married</td>
<td>0</td>
<td>3 (7.9)</td>
<td>2 (5.3)</td>
<td>0.956</td>
</tr>
<tr>
<td></td>
<td>Separated or divorced</td>
<td>0</td>
<td>0</td>
<td>2 (5.3)</td>
<td>0.067</td>
</tr>
<tr>
<td>Level of education, n (%)</td>
<td>Junior high</td>
<td>1 (2.6)</td>
<td>1 (2.6)</td>
<td>1 (2.6)</td>
<td>0.612</td>
</tr>
<tr>
<td></td>
<td>High school</td>
<td>2 (5.3)</td>
<td>8 (21.1)</td>
<td>9 (23.7)</td>
<td>0.145</td>
</tr>
<tr>
<td></td>
<td>University</td>
<td>5 (13.2)</td>
<td>6 (15.8)</td>
<td>5 (13.2)</td>
<td>0.067</td>
</tr>
<tr>
<td></td>
<td>Student, n (%)</td>
<td>4 (50.0)</td>
<td>4 (26.7%)</td>
<td>6 (40.0)</td>
<td>0.153</td>
</tr>
<tr>
<td>Worker, n (%)</td>
<td>No</td>
<td>0</td>
<td>9 (23.7)</td>
<td>8 (21.1)</td>
<td>0.067</td>
</tr>
<tr>
<td></td>
<td>Part-time</td>
<td>2 (5.3)</td>
<td>1 (2.6)</td>
<td>1 (2.6)</td>
<td>0.956</td>
</tr>
<tr>
<td></td>
<td>Full-time</td>
<td>6 (15.8)</td>
<td>5 (13.2)</td>
<td>6 (15.8)</td>
<td>0.153</td>
</tr>
<tr>
<td>Self-care practices, points</td>
<td>1-3 times the MW</td>
<td>1 (2.6)</td>
<td>3 (7.9)</td>
<td>3 (7.9)</td>
<td>0.145</td>
</tr>
<tr>
<td></td>
<td>3-10 times the MW</td>
<td>5 (13.2)</td>
<td>9 (23.7)</td>
<td>10 (63.2)</td>
<td>0.067</td>
</tr>
<tr>
<td></td>
<td>&gt; 10 times the MW</td>
<td>2 (5.3)</td>
<td>3 (7.9)</td>
<td>2 (5.3)</td>
<td>0.956</td>
</tr>
<tr>
<td>Adherence score, points</td>
<td>0.73 ± 0.16</td>
<td>0.84 ± 0.11</td>
<td>0.76 ± 0.17</td>
<td>0.153</td>
<td></td>
</tr>
</tbody>
</table>

MW: (national) minimum wage. aMean ± SD; and bmedian (interquartile range). Chi-square test for categorical variables; and one-way ANOVA or Kruskal-Wallis test for quantitative variables.
Table 2 - Objective measurements of severity in each group of perception of disease severity.

<table>
<thead>
<tr>
<th>Variable</th>
<th>Well below or below average (n = 8)</th>
<th>Average (n = 15)</th>
<th>Above or well above average (n = 15)</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>BMI*</td>
<td>20.4 ± 2.5</td>
<td>20.7 ± 3.4</td>
<td>21.2 ± 2.4</td>
<td>0.772</td>
</tr>
<tr>
<td>S-K clinical score#, points</td>
<td>70 (15)</td>
<td>65 (25)</td>
<td>85 (18)</td>
<td>0.018</td>
</tr>
<tr>
<td>FVC%, % of predicted</td>
<td>52.0 ± 20.4</td>
<td>63.8 ± 21.8</td>
<td>74.1 ± 24.0</td>
<td>0.091</td>
</tr>
<tr>
<td>FEV1, % of predicted</td>
<td>39.0 ± 17.2</td>
<td>50.6 ± 26.2</td>
<td>63.5 ± 29.3</td>
<td>0.102</td>
</tr>
<tr>
<td>FEV1/FVC, % of predicted</td>
<td>76.4 ± 17.4</td>
<td>79.0 ± 16.4</td>
<td>81.1 ± 16.6</td>
<td>0.824</td>
</tr>
<tr>
<td>SpO2, %</td>
<td>96.6 ± 1.3</td>
<td>96.4 ± 1.5</td>
<td>96.2 ± 2.8</td>
<td>0.895</td>
</tr>
<tr>
<td>Radiographic score#, points</td>
<td>15 (4)</td>
<td>16 (6)</td>
<td>18 (6)</td>
<td>0.164</td>
</tr>
</tbody>
</table>

BMI: body mass index; and S-K: Shwachman-Kulczycki. *Mean ± SD; and #median (interquartile range). One-way ANOVA or Kruskal-Wallis test for quantitative variables. Kruskal-Wallis post-hoc Z test: medians significantly different if letters are different.

23.8 ± 6.5 years. All but one of the patients were Caucasian. The mean clinical score was 75 points (IQR, 24), the mean FEV1 was 54.5% ± 27.2%, and the median radiographic score was 16.5 points (IQR, 6.0). Of the 38 patients included, 27 presented the classic form of the disease, with chronic lung disease and pancreatic insufficiency, and 11 presented the non-classic form, with pancreatic insufficiency only.

In comparison with that of other individuals with CF, 3 (7.9%) patients rated their health status as well below average; 5 (13.2%) rated it as below average; 15 (39.5%) rated it as average; 10 (26.3%) rated it as above average; and 5 (13.2%) rated it as well above average.

Two (5.3%) patients rated their self-care practices as poor, 9 (23.7%) rated them as fair, 16 (42.1%) rated them as good, 10 (26.3%) rated them as very good, and 1 (2.6%) rated them as excellent.

We can observe (Figure 1) that the perception of disease severity correlated significantly with clinical score (r = 0.43, p = 0.007), FVC in % of predicted (r = 0.34, p = 0.034), FEV1 in % of predicted (r = 0.38, p = 0.019) and reported self-care practices (r = 0.33, p = 0.044). The perception of disease severity did not correlate significantly with age (r = 0.02; p = 0.992), age at diagnosis (r = −0.008; p = 0.962), SpO2 (r = 0.12; p = 0.465), radiographic score (r = 0.33; p = 0.51) or adherence score (r = 0.033; p = 0.842). Reported self-care practices correlated significantly with adherence score (r = 0.57; p < 0.0001).

The comparison of the three groups of perception of disease severity (Table 1) revealed that there was no statistically significant association for age (p = 0.788), gender (p = 0.767), age at diagnosis (p = 0.904), marital status (p = 0.276), level of education (p = 0.612), being a student (p = 0.515), being a worker (p = 0.067), self-care practices (p = 0.145), family income (p = 0.956) or adherence score (p = 0.153). There was a significant association among the three groups for clinical score (p = 0.018), and the group in which perception of health status was rated as above or well above average had a significantly higher score than did the other two groups. There was no significant association among the groups for the following variables (Table 2): body mass index (p = 0.772); FVC in % of predicted (p = 0.091); FEV1 in % of predicted (p = 0.102); FEV1/FVC ratio in % of predicted (p = 0.824); SpO2 (p = 0.895); and radiographic score (p = 0.164).

Discussion

This cross-sectional study addressed how adult patients with CF perceive the severity of their disease and what are the implications of this subjective perception. In our sample, 20% of the patients rated their health status, in comparison with that of other individuals with CF, as below or well below average, whereas 39.5% rated it as average and 39.5% rated it as well above average. This subjective perception correlated moderately with objective measures of disease severity, such as the Shwachman-Kulczycki clinical score and pulmonary function tests. In addition, the patients who perceived greater disease severity reported fewer self-care practices. However, an extremely important finding was that the perception of disease severity did not correlate with self-reported degree of adherence to the conventional treatment.

When the patients were evaluated by group of perception of disease severity, the clinical
score was found to be higher in the group in which the subjective perception of health status was classified as above or well above average. However, no differences were found in terms of clinical score between the group in which perception was classified as average and the group in which perception was classified as below or well below average. One hypothesis generated from this finding is that the subjective perception of the patients discriminates the severity of their disease when it is mild. As the disease progresses, the subjective perception ceases to discriminate the progressive health impairment, especially when the disease progresses from a moderate to a severe stage. However, this hypothesis needs to be confirmed in prospective cohort studies.

Another important aspect identified in the present study is the fact that the patients who perceived greater disease severity reported fewer self-care practices. This can be explained by the fact that, with the progression of the disease, the complexity of the therapeutic regimens increases, demanding more time and dedication from the patient in order to administer the treatment appropriately.

Our study can be compared with two studies mentioned below.

One group of authors studied 60 patients with CF treated in a program for adults in order to evaluate their perception of disease severity.[18] The mean FEV$_1$ in the patients studied was 61% of predicted, and 83% of the patients rated their health status as above or well above average. The perception of self-care practices did not correlate with the perception of disease severity. In contrast, our study included patients with more severe disease. The mean FEV$_1$ was 54.5% of predicted, and only 39.5% of the patients rated their health status as above or well above average. Nevertheless, we found a weak-to-moderate correlation between the perception of self-care practices and the perception of disease severity. In addition, the perception of self-care practices was found to correlate significantly with the adherence score.

Disease severity in CF patients is related to its variability in phenotypic expression. The evaluation of this severity requires the use of objective parameters such as scores, which can be clinical, radiographic, tomographic, echographic or scintigraphic,[19] as well as pulmonary function tests.[20,21] The disease differs from patient to patient, especially regarding the degree of pulmonary and gastrointestinal impairment. Objective measurements of severity are used to evaluate the extent of the organic injury, compare patient clinical severity, evaluate the effects of the therapeutic interventions and determine the prognosis of the disease. Each form of evaluation contributes to the evaluation of one dimension of the disease and can be useful in a specific clinical situation.[19]

In the present study, three objective measurements were used to evaluate the severity of CF: clinical score, pulmonary function tests and radiographic score. Whereas pulmonary function tests and the radiographic score evaluate only the degree of lung disease, the Shwachman-Kulczycki clinical score[13] evaluates four different patient dimensions: general activities of daily living; clinical test findings; nutritional aspects and radiographic aspects of the lung. The emphasis on the evaluation of lung disease severity is due to the fact that, although CF is a multisystemic disease, pulmonary involvement is the principal cause of morbidity and mortality, thereby constituting the principal determinant of disease severity and prognosis.[22]

Disease perceptions represent organized cognitive constructs or patient beliefs about their own disease. It has been demonstrated that these perceptions are important behavioral determinants and are associated with relevant outcomes in clinical practice, such as adherence to treatment, functional recovery and quality of life.[23,24] Disease perceptions were initially evaluated through semi-structured interviews with patients. However, this method lacks psychometric validity and does not show sufficient reproducibility. More recently, disease perceptions have been evaluated through questionnaires, visual analog scales and representation through drawings.[23] In the present study, the instrument used was a questionnaire that emphasized how patients rated their health status in comparison with that of other patients with CF. The questio-
naire is simple and self-explanatory, as well as being easily and rapidly administered.

The present study had certain limitations. One difficulty in studying the patient perception of disease severity is the lack of instruments validated for that purpose. We opted for the strategy of adapting a questionnaire presented in another study.\(^\text{(18)}\) We believe that the objectivity and simplicity of the options proposed as a reference for disease severity, as well as the fact that the questionnaire was easily translated into Portuguese, reduced the possible interference of this limiting factor. In addition, the cross-sectional design, used in the present study, does not provide sufficient evidence to define the temporal sequence of the associations among perception of disease severity, perception of self-care practices, degree of adherence to treatment and objective disease progression. Furthermore, the fact that 2 patients refused to participate in the study might have contributed to the overestimation of the relationship between the perception of disease severity and objective measurements of severity, on the hypothesis that this occurred precisely because these patients had an impaired perception of their health status.

Although recent studies have demonstrated that disease perception is associated with significant outcomes in chronic diseases, this remains an emerging area of clinical research.\(^\text{(23)}\) The present study, which evaluated patients treated in a program for adults with CF, provides additional information on patient perception of disease severity and self-care practices, showing that such perception is associated with the degree of adherence to treatment and objective disease severity. This information can contribute to a multidisciplinary therapeutic approach to improving the health of patients with CF. However, since this was a cross-sectional study, this population of patients should be monitored prospectively so that we can obtain more accurate estimates of the alterations in the perception of disease severity over time. In addition, intervention studies should be designed to determine the implications of such alterations on clinical outcomes, pulmonary function and quality of life.

In conclusion, the present study showed that the perception of disease severity in patients with CF correlates with objective measurements of disease severity, such as the Shwachman-Kulczycki clinical score and pulmonary function tests. However, the perception of disease severity does not correlate with the degree of self-reported adherence to the conventional treatment.

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**References**


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