

Original Article

Health-Related Quality of Life in Patients With Alpha-1 Antitrypsin Deficiency: A Cross Sectional Study[☆]



Margarida Torres Redondo,^{a,*} Elsa Campoa,^b Luis Ruano,^{c,d} Maria Sucena^a

^a Pulmonology Department, Centro Hospitalar de São João, Oporto, Portugal

^b Oncology Department, Centro Hospitalar do Algarve, Faro, Portugal

^c Department of Clinical Epidemiology, Predictive Medicine and Public Health, University of Porto Medical School, Portugal

^d EPIUnit, Institute of Public Health, University of Porto, Portugal

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ABSTRACT

Background: Measures of health related quality of life (HRQoL) in patients with α1-antitrypsin deficiency (AATD) can help to determine the impact of the disease and provide an important insight into the intervention outcomes. There is few data regarding this issue in the literature. The aim of this study is to assess the relationship between HRQoL and gender, functional parameters and history of hospitalizations in patients with AATD.

Methods: This is a cross-sectional study of 26 patients with severe AATD recruited in the pulmonology outpatient clinic at a tertiary care medical center. Social-demographic, clinical and functional parameters were recorded and HRQoL was assessed with the Portuguese version of the medical outcome study short form-36 (SF-36) self-administered questionnaire.

Results: Older patients, females and patients with at least one hospitalization in the previous year due to respiratory disease had statistical lower scores in some dimensions of the SF-36 questionnaire. Superior FEV1 and higher distance mark in the 6-min walking test distance influenced positively several dimensions of the questionnaire. Higher scores in the mMRC scale influenced negatively the HRQoL.

Conclusions: These data suggests that older and female patients with AATD have worse HRQoL. Hospitalizations and functional markers of respiratory disease progression influenced negatively the HRQoL, suggesting that the SF-36 questionnaire could be useful as an outcome for AATD patients with lung involvement.

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Calidad de vida relacionada con la salud en pacientes con déficit de alfa-1 antitripsina: estudio transversal

RESUMEN

Introducción: Las medidas de calidad de vida relacionada con la salud (CVRS) pueden ayudar a determinar los efectos de la enfermedad de los pacientes con déficit de α1-antitripsina (DAAT) y proporcionar una perspectiva valiosa de los resultados de las intervenciones. Este tema se ha abordado poco en la literatura, y el objetivo de este estudio fue evaluar si existe alguna relación entre la CVRS y el sexo, los parámetros funcionales y los antecedentes de hospitalización de los pacientes con DAAT.

Métodos: Para este estudio transversal se reclutaron 26 pacientes con DAAT grave que eran atendidos en las consultas externas de neumología de un hospital terciario. Se registraron parámetros sociodemográficos, clínicos y funcionales, y se evaluó la CVRS mediante la versión portuguesa del cuestionario de salud SF-36.

Resultados: Los pacientes de mayor edad, de sexo femenino y los que habían sido hospitalizados por enfermedad respiratoria al menos una vez durante el año anterior mostraron puntuaciones más bajas en

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* Corresponding author.

E-mail address: margarida.tredondo@gmail.com (M. Torres Redondo).

algunas dimensiones del cuestionario SF-36. Los valores más altos de FEV₁ y distancias recorridas más largas en la prueba de la marcha de 6 min tuvieron una influencia positiva sobre varias dimensiones del cuestionario, mientras que las puntuaciones más altas en la escala MRCm influyeron negativamente en la CVRS.

Conclusiones: Los resultados muestran que la CVRS de los pacientes de mayor edad y las mujeres con DAAT es peor. Las hospitalizaciones y los marcadores funcionales de progresión de la enfermedad respiratoria tuvieron una influencia negativa sobre la CVRS, lo que indica que el cuestionario SF-36 podría ser de utilidad como medida de resultados de los pacientes con DAAT y afectación pulmonar.

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Introduction

Alpha 1-Antitrypsin deficiency (AATD), though often underdiagnosed, is one of the most common genetic respiratory disorders worldwide, affecting from 1 in 2000 to 1 in 5000 individuals.¹ AATD predisposes to lung and liver disease and some other conditions.^{2,3}

Alpha 1-Antitrypsin is the prototype member of the serine protease inhibitor (serpin) superfamily of proteins, and is caused by mutations in the *SERPINA1* gene located in the long arm of chromosome 14.^{1,4} This genetic defect results in decreased serum levels of α 1-antitrypsin, leading to low alveolar concentrations and protease excess, causing emphysema.^{1,4}

In chronic respiratory diseases, such as chronic obstructive pulmonary disease (COPD), measures of Health Related Quality of Life (HRQoL) are frequently used as descriptive instruments or as outcome measures.⁵ Since no cure has yet been found for most chronic respiratory diseases, a major goal of care is to improve HRQoL.

Given the lack of studies assessing HRQoL in patients with AATD and the increasing importance of HRQoL measures, the aim of this study was to assess the relationship between HRQoL and gender, functional parameters and history of hospitalizations (previous year) in patients with AATD.

Methods

Study Design and Setting

This is a cross-sectional study of patients with AATD recruited from January to June 2013 (6 months) in the pulmonology outpatient clinic at the University Hospital São João, a tertiary hospital. The research protocol was submitted to and approved by the local research ethics committee.

Patients

The sample included 26 consecutive patients attending the pulmonology outpatient clinic, with an AATD diagnosis confirmed by genetic analysis (genotypes ZZ and SZ). Written informed consent was obtained from each patient. Illiterate patients and those with physical and mental disabilities prior to the diagnosis of AATD were excluded.

Clinical and Pulmonary Function Assessment

Social-demographic data were recorded, namely age, gender, educational level, social and employment status. Patients also underwent clinical evaluation that included dyspnea severity measured on the modified Medical Research Council (mMRC) scale,⁶ assessment of smoking status, presence of significant comorbidities and current medication. The results from the genetic analysis were also retrieved. The number of hospitalizations due to respiratory disease in the previous year was retrospectively analyzed using clinical records. Each patient was tested for spirometry and lung volumes (MasterScreen™ Body; Jaeger, Würzburg,

Germany) according to international guidelines.^{7,8} The 6-min walking test (6MWT) was performed using the methodology described by the American Thoracic Society and the European Respiratory Society.⁹

Quality of Life

HRQoL was assessed using the Portuguese version of the short form-36 (SF-36) health survey questionnaire.^{10,11} The SF-36 questionnaire was used as it is considered a well-researched and reliable instrument that has been culturally adapted and validated for the Portuguese population.¹¹ Moreover, it has been used as an outcome measure in different studies in chronic respiratory diseases, such as COPD.

The questionnaire was administered during the patient's first visit to the pulmonology outpatient clinic in the study period.

Statistical Analysis

Continuous variables are described with their mean and standard deviation. Categorical variables are described by their relative frequencies (in percentage).

The independent samples *t*-test was used to compare the SF-36 scores between genders, genotypes, patients untreated vs patients treated with augmentation therapy, and patients without vs patients with hospitalizations due to respiratory disease in the previous year. Linear regression was used to analyze the association between the SF-36 scores and age, forced expiratory volume in 1 second (FEV₁), forced vital capacity (FVC) and 6-min walking test distance (6MWT).

P-values <0.05 were considered statistically significant.

Results

Socio-demographic characteristics of AATD patients are presented in Table 1. The mean age of the sample population was 48.2 years, 73.1% (Standard Error, SE=0.09) of the patients were married and 19.2% (SE=0.08) single. Regarding the education level, 46.2% (SE=0.10) had more than basic education.

The main baseline clinical characteristics of patients are described in Table 2. The AAT phenotype was ZZ for 15 patients (57.7%, SE=0.10) and SZ for 11 (42.3%, SE=0.10). All patients had lung involvement with emphysema or bronchiectasis on chest tomography. The majority of patients (80.8%, SE=0.08) had respiratory symptoms: dyspnea in 18 (69.2%, SE=0.09), chronic bronchitis in 16 (61.5%, SE=0.10) and wheezing in 10 (38.5%, SE=0.10). Liver disease was found in 50% (SE=0.10) of patients.

Most patients had a ventilatory obstructive pattern, with a mean FEV₁ of 64.9% predicted, and 38.5% (SE=0.10) of patients with a FEV₁ of less than 50% predicted. The mean 6MWT was 387.2 meters, and the mean desaturation was 8.2%.

During the study period, 23 patients (88.5%, SE=0.06) were treated with long-acting bronchodilators, whereas 19 (73.1%,

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