Original Article

Symptom Interference Severity and Health-Related Quality of Life in Pulmonary Arterial Hypertension

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Abstract

Context. While assessing symptom severity is an important component of evaluating symptoms, understanding those symptoms that interfere with patients' lives is also key. Pulmonary arterial hypertension (PAH) is a chronic disease resulting in right heart failure and increased mortality. Patients with PAH experience multiple symptoms but we do not know which symptoms and to what extent their symptoms interfere with daily life.

Objectives. To: 1) describe the prevalence of those symptoms that interfere with life; 2) describe the severity of symptom interference; and 3) determine those sociodemographic and clinical characteristics and interfering symptoms associated with health-related quality of life (HRQOL) in patients with PAH.

Methods. A convenience sample of 191 patients with PAH completed a sociodemographic form, the Pulmonary Arterial Hypertension Symptom Interference Scale (PAHSIS) and the Medical Outcomes Survey Short Form-36 to measure HRQOL. Hierarchical multiple linear regression was used to analyze demographic and medical characteristics along with symptom interference from the PAHSIS as predictors of HRQOL from the composite mental and physical health summary scores of the Short Form-36.

Results. The most interfering symptoms reported were fatigue, shortness of breath with exertion, and difficulty sleeping. Age, gender, functional class, oxygen use, fatigue, dizziness, and Raynaud phenomenon were associated with the HRQOL physical health summary scores. The symptoms fatigue and SOB while lying down were associated with the HRQOL mental health summary scores.

Conclusion. Patients with PAH are experiencing multiple symptoms that are interfering with their HRQOL and ability to function. J Pain Symptom Manage 2016;51:25–32.

Key Words
Symptoms, interference, quality of life, pulmonary arterial hypertension

Introduction

The importance of the symptom experience as well as the severity of symptoms can impact a person's activities of daily living, and this is important in overall quality of life. Pulmonary arterial hypertension (PAH) is a life-limiting disease that affects primarily young to middle-aged women.[1] The etiology for PAH is varied, with more than half of these patients diagnosed with idiopathic PAH. There are additional associated disorders that include connective tissue disease (e.g., systemic sclerosis); portopulmonary hypertension, congenital heart disease, schistosomiasis, and side effects of certain drugs (e.g., fenfluramine/phentermine).[1] PAH is characterized by elevated mean pulmonary artery pressures of 25 mm Hg or greater and a mean capillary wedge pressure less

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than 15 mm Hg; these increased pressures result in remodeling of the right heart and ultimately right heart failure and eventual death.[1] Initial symptoms include fatigue and shortness of breath (SOB) on exertion. These symptoms may be attributed to other cardiopulmonary disorders resulting in a delay in diagnosis. Diagnosis is often one of exclusion and may require multiple health care encounters to rule out other disorders.[2]

Patients are commonly treated with diuretic therapy, anticoagulation, and PAH-approved drugs that promote vasodilation. Common classes of PAH-approved medications include calcium channel blockers, endothelin receptor antagonists, phosphodiesterase type 5 inhibitors, and prostanoid analogues. Pharmacological treatments can be complex especially the prostanoid analogues, which are commonly administered intravenously, subcutaneously, or inhaled six to nine times per day.[3] Although pharmacological therapies have reduced mortality, the mortality rate still remains high, with a median survival of seven years.[4] Recent Registry to Evaluate Early and Long-term Pulmonary Arterial Hypertension Disease Management data show five-year survival for previously diagnosed patients with PAH at 65.4% compared with 61.2% for newly diagnosed patients.[5]

Patients with PAH have multiple symptoms that can be severe and impair their health-related quality of life (HRQOL).[6,7] Some of the most commonly reported symptoms that can be severe are fatigue, SOB on exertion, and difficulty sleeping. Although patients have limits in their physical functioning, they also report depressive symptoms[8] and anxiety.[9] Patients with PAH describe how they must readjust their lives and tailor their activities because of such symptoms as SOB and fatigue.[10]

While assessing the severity of symptoms, it is important to gauge how much symptoms interfere with the patient's life to understand how patients manage and cope with their symptoms while living with a chronic illness. Even though a symptom may not be reported as severe, it may dramatically interfere with a patient's ability to perform activities of daily living and care for themselves and/or their families. Patients with PAH have impairment in the ability to function that adversely affects their HRQOL.[11,12] Others have found decreased HRQOL in PAH with increased symptom burden and a lack of awareness of the availability of palliative care.[13] Symptoms and physical functioning are important to assess in PAH[14,15] along with how symptoms interfere with patients' lives.

The University of San Francisco Symptom Management Model was the theoretical framework used for this study.[16] There are three dimensions, which include the symptom experience, symptom management strategies, and symptom outcomes. Understanding which PAH symptoms interfere the most and the extent of their interference is important to understand to develop and implement symptom management strategies. Much of the research investigating symptom interference has been conducted in oncology patients.[17–23] Commonly reported symptoms that are severe and interfere with patients' lives include fatigue,[23] pain, and sleep disturbance.[24] We aimed to extend the knowledge of the symptom experience in patients with PAH. By understanding this phenomenon, we can determine those symptoms that most interfere with patients' lives, so that targeted interventions can be developed and tested. The objectives of this study were to 1) describe the prevalence of those symptoms that interfere with life; 2) describe the severity of symptom interference; and 3) determine those sociodemographic and clinical characteristics and interfering symptoms associated with HRQOL in PAH.

Methods

Sample and Setting

This cross-sectional study included self-identified patients with World Health Organization Group I PAH, which includes the PAH etiologies: idiopathic, familial, connective tissue disease, congenital heart disease, HIV, portopulmonary hypertension, and drugs associated with causing PAH such as anorexigens. The inclusion criteria for this convenience sample included adults 18 years and older who were able to read and write English. Two hundred sixty-two people with PAH were recruited. A total of 191 patients completed the study. The response rate was 69%.

Before recruitment began, the study was approved by both the university and hospital institutional review boards. A PAH clinic, local support groups, and international conferences, along with the Pulmonary Hypertension Association Web site, served as recruitment sites. At the PAH clinic, the nurse practitioner approached the patients to gauge their interest. If they were interested, the principal investigator (PI) then approached the patient to answer questions regarding the study. At the PAH support groups and the conferences, the facilitator introduced the PI who would briefly discuss the study, and interested patients approached the PI after the meeting. The administrator for the Pulmonary Hypertension Association Web site posted an advertisement describing the study. Those people with PAH contacted the PI. Questionnaires were either completed and returned to the PI on site or participants mailed back the questionnaires to the PI using a self-addressed stamped envelope provided in each packet. The study packet