TURKISH JOURNAL OF CARDIOVASCULAR **NURSING**



The Relationship Between Symptom Status and Health-Related Quality of Life in Patients with Pulmonary Arterial Hypertension

Pulmoner Arteriyel Hipertansiyonlu Hastaların Semptom Durumu ve Sağlıkla İliskili Yasam Kalitesi Arasındaki İliski

ABSTRACT

Objective: This study was conducted to investigate the relationship between symptom status and health-related quality of life in patients with pulmonary arterial hypertension.

Methods: This was a descriptive study, and 31 outpatient pulmonary arterial hypertension patients were enrolled. The "Data Gathering Form," the "Symptom Status Questionnaire-Heart Failure," and the "Short-Form Health Survey" were used to collect research data.

Results: The mean age of the patients was 47.4 ± 15.3 years and 51.6% of them were female. The majority of the patients were in World Health Organization functional class II and not working (80.6%). The most common symptoms experienced by the patients were "fatigue or lack of energy" (96.8%) and "shortness of breath during the daytime" (80.6%). The mean total Symptom Status Questionnaire-Heart Failure score was significantly lower in the working group compared to the non-working group (P < .001). Except for the "general health" and "social functioning" of the SF-36 sub-dimensions, there were significant differences between the high and low Symptom Status Questionnaire-Heart Failure groups in all other sub-dimensions (P < .05). There was a weak negative correlation between the "general health" sub-dimension of Short-Form Health Survey and Symptom Status Questionnaire-Heart Failure. All other sub-dimensions of Short-Form Health Survey had moderate negative correlations with Symptom Status Questionnaire-Heart Failure.

Conclusion: It was found that there was a negative correlation between the symptom status of patients with pulmonary arterial hypertension and health-related quality of life regular evaluation of patients' symptom status, and health-related quality of life is recommended to provide comprehensive care and improve long-term quality of life.

Keywords: Pulmonary arterial hypertension, symptom status, quality of life

Amaç: Bu çalışma pulmoner arteriyel hipertansiyonlu (PAH) hastaların semptom durumu ve sağlıkla ilişkili yaşam kalitesi (SİYK) arasındaki ilişkinin belirlenmesi amacıyla yapıldı.

<mark>Yöntem:</mark> Çalışma tanımlayıcı olarak tasarlandı ve çalışmaya poliklinikte takip edilen 31 PAH hastası dahil edildi. Arastırmanın verileri "Veri Toplama Formu," "Kalp Yetersizliği Semptom Durum Ölçeği (KYSDÖ)" ve "Kısa Form Yaşam Anketi (SF-36) ile toplanmıştır.

Bulgular: Hastaların yaş ortalaması 47,4 ± 15,3 yıl olup, %51,6'sı kadındı. Hastaların çoğunluğu Dünya Sağlık Örgütü fonksiyonel sınıf li'deydi ve çalışmıyordu (%80,6). Hastaların en sık yaşadıkları semptomlar "yorgunluk veya enerjisizlik" (%96,8) ve "gündüz nefes darlığı" (%80,6) idi. Çalışma grubunda çalışmayan gruba göre ortalama toplam KYSDÖ puanı anlamlı olarak daha düşüktü (P < ,001). SF-36 alt boyutlarının "genel sağlık" ve "sosyal işlevsellik" dışındaki diğer tüm alt boyutlarında yüksek ve düşük KYSDÖ grupları arasında anlamlı farklılıklar bulunmuştur (P < 0.05). SF-36'nın "genel sağlık" alt boyutu ile KYSDÖ arasında zayıf bir negatif korelasyon vardı. SF-36'nın diğer tüm alt boyutları, KYSDÖ ile orta düzeyde negatif korelasyona sahipti.

Sonuç: PAH'lı hastaların semptom durumu ve SİYK arasında negatif korelasyon olduğu saptandı.

Anahtar Kelimeler: Pulmoner arteriyel hipertansiyon, semptom durumu, yaşam kalitesi

ORIGINAL ARTICLE

Sevda Türen¹ Kadriye Memiç Sancar² Selahattin Türen²

- ¹ Department of Nursing, Faculty of Health Sciences, İstanbul Kültür University, İstanbul, Turkey
- ² Department of Cardiology, Health Sciences University Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training Research Hospital, İstanbul, Turkey

Corresponding author:

Sevda Türen

oxtimes sevdamercanhm@hotmail.com

Received: September 16, 2022 Accepted: November 14, 2022 Publication Date: December 22, 2022

Cite this article as: Türen S, Memiç Sancar K, Türen S. The relationship between symptom status and health-related quality of life in patients with pulmonary arterial hypertension. Turk J Cardiovasc Nurs 2022;13(32):173-179.

DOI: 10.5543/khd.2022.2225123



Copyright@Author(s) - Available online at khd.tkd.org.tr.

Content of this journal is licensed under a Creative Commons Attribution-NonCommercial 4.0 International License.

Introduction

Pulmonary arterial hypertension (PAH) is a chronic progressive disease characterized by elevated pulmonary artery pressure and ultimately right ventricular dysfunction.¹ According to the European Society of Cardiology (ESC)/European Respiratory Society (ERS) 2015 guidelines for the diagnosis and treatment of pulmonary hypertension, the prevalence of PAH in adults is estimated to be 15 cases per million people.² In studies conducted in different countries, the prevalence is shown as approximately 15–50 million-¹.¹.¹.³.⁴ According to the epidemiological study conducted on the Turkish population, the annual prevalence is 9.6/million.⁵ Therefore, PAH is included in the rare diseases group.

Since PAH has no specific signs and symptoms, it is very difficult to diagnose. The mean time from the onset of symptoms to diagnosis is 2 years. Patients are asymptomatic in the early period, and as time progresses, signs and symptoms of heart failure (HF) appear. In addition to symptoms of exertional dyspnea, fatigue, and weakness, individuals may also experience chest pain and syncope. The presence and severity of symptoms seriously affect the quality of life of patients.

Although there have been developments in the treatment of PAH in recent years, the difficulty of diagnosis causes a delay in treatment. The goal of PAH treatment is to increase the quality of life and control or reduce symptoms, keeping the patient in the low-risk group and reducing mortality. If PAH is not treated and controlled effectively, it can lead to death.² In addition to the treatment of PAH, nurses play an important role in the management of care. Close follow-up and evaluation of symptoms are very important in the care of these patients.⁷ Therefore, international guidelines emphasize the vital importance of a multidisciplinary approach.²

Health-related quality of life (HRQoL) is an individual's personal perception of their general and daily quality of life.8 In patients with PAH, HRQoL is closely related to PAH symptoms.9 Symptoms of HF increase as PAH progresses. A decrease in the daily living activities and functional capacity of the individual, repeated hospitalizations, and therefore a decrease in HRQoL occur with the increase in HF symptoms.^{2,6,10} In their study (n=55), Halim et al¹¹ also reported that PAH patients had low HRQoL. In a study by Matura et al⁶ (n=191), the most interfering symptoms reported were fatigue, shortness of breath (SOB) with exertion, and difficulty sleeping. The physical components of HRQoL were worse in PAH while the mental health components were actually better than the U.S. normative data from the SF-36.6 Knowing the symptoms experienced by patients and the severity of these symptoms is one of the issues that should be focused on in the care of patients. To the best of our knowledge, there is little information in the literature about the symptom status and HRQoL of patients with PAH. Therefore, this study was conducted to investigate the relationship between symptom status and HRQoL in patients with PAH.

Research Questions

 What are the socio-demographic characteristics of patients with PAH?

- 2. What is the symptom status of patients with PAH?
- 3. What is the HRQoL of patients with PAH?
- 4. What is the relationship between symptom status and HRQoL in patients with PAH?

Methods

Study Population and Procedure

The study conforms to the ethical principles outlined in the Declaration of Helsinki. Appropriate permissions were obtained from the institution where the study was conducted. The study was approved by the University Institutional Review Board (IRB date and number: 23.09.2021/2021.65). Both verbal and written (informed consent) approval were obtained from patients who met the study criteria. The participants were assured that their responses would remain anonymous and confidential.

Patient Selection

Patients who were 18 years of age or older, had no communication problems, agreed to consent, and were diagnosed with PAH by a cardiologist in accordance with the ESC 2015 criteria² were included in the study. Patients who had communication problems, declined to consent, or had recently been hospitalized were excluded from the study.

Thirty-seven consecutive patients with PAH were screened for eligibility. Three patients declined to participate in the study, 2 patients had communication problems, and 1 patient had recent hospitalization. After the exclusion of these patients, the final study population consisted of 31 patients.

Data Gathering Form

The data gathering form was prepared by the researchers in light of the literature review. This form consisted of 17 questions regarding socio-demographic characteristics (age, gender, marital status, education level, comorbidities, etc.) and information about PAH (duration of illness, knowledge, hospitalization within the last year, regular physician follow-up, medications, etc.).

Symptom Status Questionnaire-Heart Failure

The questionnaire was developed in 2015 by Heo et al. 12 and it was adapted into the Turkish language by Gök Metin and Gülbahar.¹⁰ The questionnaire consists of 7 questions used to assess the presence, frequency, severity, and distress of common physical HF symptoms, including daytime dyspnea, dyspnea when lying down, fatigue, chest pain, edema, difficulty sleeping, and dizziness or loss of balance. Patients are asked to indicate the presence of each symptom during the past 4 weeks. If no symptoms exist, the score is 0. If a patient has experienced a symptom, the patient is asked about the frequency, severity, and distress of the symptom. Responses on frequency range from 1 (less than once per week) to 4 (nearly daily), severity responses range from 1 (slight) to 4 (very much), and distress responses range from 0 (not at all) to 4 (very much). The total score on the questionnaire is calculated by summing the total scores of all the symptoms. Possible total scores range from 0 to 84, with higher scores indicating more severe symptoms. As per the reliability and validity study performed in Turkish, Cronbach's alpha was found to be 0.86 for the SSQ-HF. In this study, it was found to be 0.94.

Short-Form Health Survey

The Short-Form Health Survey (SF-36) is one of the most widely used questionnaires to assess HRQoL in both general populations and PAH patients. It was developed by Ware et al¹³, and it was adapted into the Turkish language by Koçyiğit et al.¹⁴ Short-Form Health Survey consists of 2 main components (physical component and mental component) and 8 subdomains. Instead of giving a single score on the questionnaire, each sub-domain is evaluated within itself, and scores range between 0 and 100. A high score obtained from the SF-36 refers to a better HRQoL. As per the reliability and validity study performed in Turkish, Cronbach's alpha was found for each subgroup between 0.73 and 0.76. In this study, it was found to be between 0.74 and 0.78.

Statistical Analysis

Continuous variables were expressed as means $(\bar{x}) \pm SD$, and categorical variables were expressed as numbers and percentages. The normality of the continuous variables (SF-36 and SSQ-HF) was tested using the Shapiro-Wilk test. Patients were divided into 2 groups based on their SSQ-HF mean score as patients with a low SSQ-HF score (≤31) and a high SSQ-HF score (>31). We used the mean because the data showed a normal distribution. The SSQ-HF scores of the patients in the 2 groups were compared across socio-demographic and PAHrelated characteristics of the patients using the Student t-test or one-way analysis of variance test for continuous variables and the Chi-square test or Fisher's exact test for categorical variables. Post-hoc analyses were performed where appropriate using Bonferroni correction. Relationships between variables are analyzed by using Pearson correlation analyses. Two-sided P values of .05 were considered significant for all tests. Statistical analysis was performed using the Statistical Package for the Social Sciences version 20.0 for Windows (IBM SPSS Corp.; Armonk, NY, USA).

Results

Socio-Demographic and Clinical Characteristics of the Patients

The mean age of the patients included in the study was 47.4 ± 15.3 (min. 24 and max. 75) years, and the duration of the PAH was 61.6 ± 65.3 (min. 2 and max. 283) months. The majority of the patients were female (51.6%), married (71.0%), had an education level less than high school (80.6%), not working (80.6%), and living with a family member (96.8%). While 67.7% of the patients were in World Health Organization (WHO) functional class II, 25.8% had comorbidities, and 48.4% had been hospitalized in the previous year. In addition, 96.8% of the patients reported that they had knowledge about PAH, and 93.5% of them were compliant with regular physician follow-up. The baseline characteristics of patients are shown in Table 1.

Symptom Status of Patients

The total SSQ-HF score of the patients included in the study was 31.0 \pm 18.0 (min. 5 and max. 66). When the symptoms experienced by the patients were examined, the most common symptoms were "fatigue or lack of energy" (96.8%) and "shortness of breath during the daytime" (80.6%) (Table 2). The mean total SSQ-HF score was significantly lower in the

working group compared to the non-working group (P=.001). Furthermore, the working ratio was significantly lower in the group with a high total SSQ-HF score than in the group with a low SSQ-HF score (P=.02) (Table 1).

Patients' Health-Related Quality of Life

Considering the patients' SF-36 scores, "bodily pain" had the highest score (64.68 \pm 24.73), and "role physical" had the lowest score (29.84 \pm 41.54). The SF-36 scores of the patients in the high and low SSQ-HF groups were compared and all subdimension scores of SF-36 were lower in patients with high SSQ-HF. Except for the "general health" and "social functioning" of the SF-36 sub-dimensions, there were significant differences between the 2 groups (high and low SSQ-HF) in all other sub-dimensions (P < .05). In addition, the mean physical component summary (PCS) and mental component summary (MCS) scores, which are the main components of the SF-36, were significantly higher in the group with a low total SSQ-HF score (P=.002, P=.005, respectively) (Table 3).

The Relationship Between Patients' Symptom Status and Health-Related Quality of Life

While there was a weak negative correlation (r=-0.364, P=.04) between the "general health" sub-dimension of SF-36 and SSQ-HF, all other sub-dimensions of SF-36 had moderate negative correlations with SSQ-HF (r=-0.512, P=.003; r=-0.401, P=.02; r=-0.566, P=.01; r=-0.507, P=.004; r=-0.411, P=.02; r=-0.415, P=.02; r=-0.488, P=.005) (Table 4).

Discussion

Pulmonary arterial hypertension is a rare and progressive disease that has a substantial negative impact on HRQoL. Therefore, the assessment of symptom status and HRQoL of patients with PAH is important in terms of PAH treatment research and care management. This study was conducted to investigate the relationship between symptom status and HRQoL in patients with PAH.

Studies in the literature indicate that PAH is more common between the ages of 30 and 60 and in women.^{1,15,16} In the ESC/ERC guideline, it is reported that the age at diagnosis is 50-65 years, and although it is variable, the female gender is dominant.2 According to studies conducted in Turkey, Kaymaz et al¹⁷ reported (a multicentric study encompassing 1501 patients from 20 adult cardiology centers) that the majority of the patients (69.0%) were women and the mean age was 44.8 \pm 5.45. In their epidemiological study, Pektas et al¹⁸ reported that the majority of the patients were over the age of 45 and of female gender (female-male ratio of 2.2:1). In this study, the mean age of the patients was 47.4 ± 15.3 years, and the majority of the patients (51.6%) were women. The mean age was found to be consistent with studies when compared to the literature. However, although the female gender was higher in number, its ratio was lower than in the literature.

Patients with PAH face many symptoms that affect HRQoL, and this limits the individual's daily life. Evaluation of the symptoms experienced by patients is vital in determining treatment and care goals.^{2,7,18,19} As reported in the literature, patients with PAH frequently experience dyspnea and fatigue.^{2,18} In parallel

Table 1. Patients' Socio-Demographic and Clinical Characteristics and Symptom Status Comparisons Across These Characteristics Total Score of SSQ-HF $SSQ-HF \leq 31$ SSQ-HF > 31n (%) Mean ± SD t, F Ρ N (%) N (%) \mathbf{X}^2 P Age (years) 24-45 years 16 (51.6) 28.62 ± 16.87 -0.900*.37 11 (68.8) 5 (31.2) 1.551 .21 ≥46 years 15 (48.4) 34.53 ± 19.65 7 (46.7) 8 (53.3) Gender Female 1.198* .24 8 (50.0) 0.883 16 (51.6) 35.25 ± 17.63 8 (50.0) .34 Male 15 (48.4) 27.47 ± 18.55 10 (66.7) 5 (33.3) Marital status Married 22 (71.0) 30.59 ± 19.91 -0.421*.67 13 (59.1) 9 (49.9) 0.003 .85 Single 9 (29.0) 33.67 ± 13.99 5 (55.6) 4 (44.4) **Education level** <High school 25 (80.6) 31.88 ± 18.20 0.243* .80 14 (56.0) 11 (44.0) 0.226 .63 >High school 6 (19.4) 29.83 ± 19.87 4 (66.7) 2 (33.3) Working status <.001 .02 Working 6 (19.4) 14.83 ± 7.65 2.754* 6 (100.0) 0 (00.0) 5.373 Not working 25 (80.6) 35.48 ± 17.79 12 (48.0) 13 (52.0) **Economic status** .12 4.922 Income more than expenses 4 (12.9) 34.00 ± 16.99 2.242** 3 (75.0) 1 (25.0) .08 Income partially covers 15 (48.4) 24.80 ± 15.88 11 (73.3) 4 (26.7) expenses Income less than expenses 12 (38.7) 39.00 ± 19.49 4 (33.3) 8 (66.7) Health insurance 30.03 ± 17.87 Yes 28 (90.3) 1.372* .18 17 (60.2) 11 (39.3) 0.834 .36 No 3 (9.7) 45.00 ± 19.00 1 (33.3) 2 (66.7) Home status Living with a family member 30 (96.8) 0.191* .85 17 (56.7) 13 (43.3) 0.746 31.60 ± 18.51 .38 Living alone 1 (3.2) 28.00 ± 0.00 1 (100.0) 0(0.0)Duration of illness (months) 2-36 months 16 (51.6) 28.81 ± 17.17 -0.839*.40 10 (62.5) 0.267 6 (37.5) .60 ≥37 months 15 (48.4) 34.33 ± 19.44 8 (53.3) 7 (46.7) Presence of comorbidities 0.971* .34 1,873 Yes 8 (25.8) 36.87 ± 19.50 3 (37.5) 5 (62.5) .17 No 23 (74.2) 15 (65.2) 8 (34.8) 29.61 ± 17.80 **Knowledge about PAH** Yes 30 (96.8) 32.07 ± 18.23 0.975* .33 17 (56.7) 13 (43.3) 0.746 .38 No 1 (3.2) 14.00 ± 0.00 1 (100.0) (0.0)Sufficient knowledge Sufficient 27 (90.0) 1.009* .32 .19 33.18 ± 18.78 14 (51.9) 13 (48.1) 3.317 Insuffcient 3 (10.0) 22.00 ± 7.94 3 (100.0) 0(0.0)Hospitalization within the last year Yes 15 (48.4) 35.47 ± 19.44 1.187* .24 8 (53.3) 7 (46.7) 0.267 .60 No 16 (51.6) 27.75 ± 16.73 10 (62.5) 6 (37.5)

(Continued)

Table 1. Patients' Socio-Demographic and Clinical Characteristics and Symptom Status Comparisons Across These Characteristics (Continued)

		Total Score of SSQ-HF SSQ-HF		$SSQ\text{-HF} \leq 31$	SSQ-HF > 31			
	n (%)	Mean ± SD	t, F	P	N (%)	N (%)	X ²	P
Regular physician follow-up								
Yes	29 (93.5)	31.52 ± 18.23	0.038*	.97	17 (58.6)	12 (41.4)	0.057	.81
No	2 (6.5)	31.00 ± 25.45			1 (50.0)	1 (50.0)		
WHO functional class								
Class II	21 (67.7)	28.43 ± 17.78	-1.373*	.18	14 (66.7)	7 (33.3)	1.978	.16
Class III	10 (32.3)	37.90 ± 18.31			4 (40.0)	6 (60.0)		
Medications ^µ								
Endothelin receptor antagonis	st							
Yes	26 (83.9)	32.23 ± 18.90	-0.514*	.61	15 (57.7)	11 (47.3)	1.000	.65
No	5 (16.1)	27.60 ± 15.24			3 (60.0)	2 (40.0)		
Spironolactone								
Yes	22 (71.0)	34.00 ± 18.42	-1.212*	.23	12 (54.5)	10 (45.5)	0.696	.41
No	9 (29.0)	25.33 ± 17.12			6 (66.7)	3 (33.3)		
Furosemide								
Yes	20 (64.5)	30.80 ± 17.53	.278*	0.78	12 (60.0)	8 (40.0)	1.000	.53
No	11 (35.5)	32.73 ± 20.21			6 (54.5)	5 (45.5)		
Phosphodiesterase type 5 inh	ibitors							
Yes	19 (61.3)	36.26 ± 18.22	-1.919*	.06	9 (47.4)	10 (52.6)	0.158	.12
No	12 (38.7)	23.92 ± 16.10			9 (75.0)	3 (25.0)		
Prostacyclin analog								
Yes	3 (9.7)	21.67 ± 8.50	1.795*	.13	3 (100.0)	0 (0.0)	0.245	.18
No	28 (92.3)	32.53 ± 18.74			15 (53.6)	13 (46.4)		

CAD, coronary artery disease; COPD, chronic obstructive pulmonary disease; DM, diabetes mellitus; HL, hyperlipidemia; HT, hypertension; PAH, pulmonary arterial hypertension; SSQ_HF, Symptom Status Questionnaire-Heart Failure; WHO, World Health Organization.

Significant difference at P < .05; value in bold: significant; *Students t-test (t), **ANOVA (F), **More than one option was selected.

Table 2. Comparison of Patients' Symptom	Status and Quality of Life

		SSQ-HF ≤31	SSQ-HF >31	Statistics	
SF-36	$\bar{x} \pm SD$ (minmax.)	$f x \pm SD$	$f x \pm SD$	t	P
Physical Functioning	45.97 ± 23.64 (5-90)	56.11 ± 22.26	31.92 ± 18.09	3.22	.003
Role Physical	29.84 ± 41.54 (0-100)	41.67 ± 46.18	13.46 ± 28.16	2.10	.04
Bodily Pain	64.68 ± 24.73 (30-100)	72.78 ± 25.29	53.46 ± 19.70	2.29	.02
Vitality	45.00 ± 21.37 (10-90)	51.67 ± 24.37	35.77 ± 11.87	2.40	.02
General Health	43.06 ± 20.32 (10-85)	46.39 ± 21.81	38.46 ± 17.84	1.07	.29
Social Functioning	54.43 ± 23.84 (0-100)	60.42 ± 24.72	46.15 ± 20.66	1.69	.10
Role Emotional	38.71 ± 41.36 (0-100)	51.85 ± 44.61	20.51 ± 28.99	2.36	.02
Mental Health	53.93 ± 17.76 (12-92)	59.33 ± 19.22	46.46 ± 12.70	2.10	.04
Physical Component summary	45.89 ± 20.07 (17-85)	54.24 ± 21.09	32.33 ± 11.31	3.38	.002
Mental Component summary	48.02 ± 18.91 (17-84)	55.82 ± 19.05	37.22 ± 12.74	3.05	.005

Significant difference at P < .05; value in bold: significant; Student's t-test (between SSQ-HF ≤31 and SSQ-HF >31).

Table 3. Items of Failure	the Symptom Status Q	uestionnaire-Heart
SSQ-HF	n (%)	$\bar{x} \pm SD$ (minmax.)
Shortness of bro	eath during daytime	
Yes	25 (80.6)	5.4 ± 3.2 (0-10)
No	6 (19.4)	
Shortness of brea	ath when lying down	
Yes	18 (58.1)	4.6 ± 4.4 (0-11)
No	13 (41.9)	
Fatigue or lack of	energy	
Yes	30 (96.8)	7.1 ± 2.6 (0-11)
No	1 (3.2)	
Chest pain		
Yes	16 (51.6)	3.1 ± 3.4 (0-10)
No	15 (48.4)	
Leg or ankle swel	ling	
Yes	16 (51.6)	4.2 ± 4.3 (0-11)
No	15 (48.4)	
Difficulty sleepin	g at night	
Yes	14 (45.2)	$3.4 \pm 4.2 (0-12)$
No	17 (54.8)	
Dizziness or loss	of balance	
Yes	18 (58.1)	3.6 ± 3.7 (0-12)
No	13 (41.9)	
Total	31 (100.0)	$31.0 \pm 18.0 (5-66)$

with the literature, the patients in this study reported that they most frequently experienced "fatigue or lack of energy" (96.8%) and "shortness of breath during daytime" (80.6%). In addition, the HRQoL of patients with a high SSQ-HF score was found to be significantly lower (P < .05, Table 2).

In the ESC/ERC 2015 guideline, it is emphasized that patients with PAH should be evaluated for HRQoL.² Studies have shown that patients' HRQoL is affected by the further progression of PAH disease. 20,21 In a study (n=108) by Von Visger et al. 9 the HRQoL of patients with PAH was found to be moderate. The HRQoL of the patients was found to be low by Halim et al 11 (n=55). In a systematic review evaluating the PAH patients' HRQoL (20 studies, n=3392), SF-36 PCS was found to be in

the range of 25.0-80.1 and MCS in the range of 33.2-76.0. 22 In a meta-analysis, which included 11 studies, the mean of SF-36 PCS and MCS were determined as 37.2 and 46.4, respectively. 20 In this study, PCS and MCS were 45.89 \pm 20.07 and 48.02 \pm 18.91, respectively (Table 3). The HRQoL of the patients included in this study was found to be higher when compared to the literature. We believe that this could be due to the patients included in the study consisting of only outpatients with WHO functional classes II and III.

Except for "general health" and "social functioning," patients with high SSQ-HF scores had significantly lower scores for all sub-dimensions of SF-36 (P < .05). In addition, significant differences were found in the main components of SF-36, which are PCS and MCS, between the high and low SSQ-HF score groups (P = .002, P = .005, respectively). The total score of SSQ-HF was significantly higher in those who were not working (P < .001). Also, patients with a high SSQ-HF score had a significantly lower working ratio (P = .02). Because patients with a high SSQ-HF score have a lower working ratio, the severity of symptoms is even more important in these young patients who are in the active working period. This is indicated by a low "role physical" score as well, which is a sub-dimension of the SF-36 that evaluates both working and daily life activities.

All sub-dimensions of the patients' SF-36 were found to have a negative correlation with the mean SSQ-HF total score (Table 4). It was revealed that an increase in the patients' PAH-related symptoms was associated with a decrease in their HRQoL.

Limitations

The main limitation of this study was the inclusion of a limited number of patients in a single center. Furthermore, because only outpatients and WHO functional classes II and III patients were included in the study and WHO functional class IV patients and inpatients were excluded, the findings of this study cannot be generalized to these groups.

Conclusion

It was found that "fatigue or loss of energy" was the most common symptom experienced by the patients in this study. Also, an increase in symptoms negatively affected the working status of the patients. Furthermore, a negative correlation was found between the symptom status of the patients and HRQoL. Regular evaluation of patients' symptom status and HRQoL is recommended to provide comprehensive care and improve long-term quality of life.

Table 4. Relationship Between the Patients' Health-Related Quality of Life and Symptom Status

		SF-36							
		Physical Function	Physical Role	Bodily Pain	Vitality	General Health	Social Function	Emotional Role	Mental Health
SSQ-HF	r	-0.512	-0.401	-0.566	-0.507	-0.364	-0.411	-0.415	-0.488
	P	.003	.02	.01	.004	.04	.02	.02	.005

r, correlation coefficient; using Pearson's correlation analyses

Ethics Committee Approval: Ethics committee approval was received for this study from the ethics committee of Istanbul Kültür University (Date: September 23, 2021, Decision No: 2021.65).

Informed Consent: Written informed consent was obtained from patients who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept - S.T., K.M.S., S.T.; Design - S.T., K.M.S., S.T.; Supervision - S.T.; Resources - S.T., K.M.S., S.T.; Materials - S.T., S.T.; Data Collection and/or Processing - K.M.S., S.T.; Analysis and/or Interpretation - S.T., K.M.S., S.T.; Literature Search - S.T., S.T.; Writing Manuscript - S.T., K.M.S., S.T.; Critical Review - S.T., K.M.S., S.T.

Declaration of Interests: The authors declare that they have no conflicts of interest.

Funding: The authors declared that this study has received no financial support.

References

- Prins KW, Thenappan T. World Health Organization Group I Pulmonary hypertension: epidemiology and pathophysiology. Cardiol Clin. 2016;34(3):363-374. [CrossRef]
- Galiè N, Humbert M, Vachiery JL, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: the Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). Eur Heart J 2016;37(1):67-119. [CrossRef]
- Hoeper MM, Humbert M, Souza R, et al. A global view of pulmonary hypertension. Lancet Respir Med. 2016;4(4):306-322. [CrossRef]
- National Organization for Rare Disorders (NORD). Rare disease database; 2021. Available at: https://rarediseases.org/rare-disea ses/pulmonary-arterial-hypertension/. Accessed June 4, 2022.
- Pektas MB, Pektas A, Emren SV, et al. An epidemiological study of pulmonary hypertension in Turkish adult population. *Mustafa Kemal Üniv Tip Derg*. 2015;6(24):24-31.
- Matura LA, McDonough A, Carroll DL. Health-related quality of life and psychological states in patients with pulmonary arterial hypertension. J Cardiovasc Nurs. 2014;29(2):178-184. [CrossRef]
- 7. Türen S. Pulmoner Hipertansiyon. In: N Enç, ed. *Kardiyovasküler Hemşirelik*. Istanbul: Nobel Tıp Kitabevleri; 2019:235-241.
- Heo S, Lennie TA, Moser DK, Dunbar SB, Pressler SJ, Kim J. Testing of a health-related quality of life model in patients with heart

- failure: a cross-sectional, correlational study. *Geriatr Nurs*. 2022;44: 105-111. [CrossRef]
- Von Visger TT, Kuntz KK, Phillips GS, Yildiz VO, Sood N. Quality of life and psychological symptoms in patients with pulmonary hypertension. *Heart Lung*. 2018;47(2):115-121. [CrossRef]
- Gök Metin Z, Gülbahar M. Kalp yetersizliği semptom durumu ölçeğinin Türkçe geçerlik ve güvenirlik çalışması. HUHEMFAD-JOHUFON. 2020;7(2):95-103.
- Halimi L, Suehs CM, Marin G, et al. Health-related quality of life and disease progression in pulmonary arterial hypertension patients: a 3-year study. ERJ Open Res. 2021;7(3):00617-2020. [CrossRef]
- Heo S, Moser DK, Pressler SJ, Dunbar SB, Mudd-Martin G, Lennie TA. Psychometric properties of the Symptom Status Questionnaire-Heart Failure. J Cardiovasc Nurs. 2015;30(2):136-144.
 [CrossRef]
- 13. Ware JE, Snow KK, Kosinski M, et al. *SF-36 Health Survey Manual and Interpretation Guide*. Boston: New England Medical Center, the Health Institute; 1993.
- 14. Koçyiğit H, Aydemir Ö, Fişek G, Ölmez N, Memiş AK. Kısa Form-36 (KF-36)'nın Türkçe versiyonunun güvenilirliği ve geçerliliği. *İlaç Tedavi Derg*. 1999;12(2):102-106.
- 15. Levine DJ. Pulmonary arterial hypertension: updates in epidemiology and evaluation of patients. *Am J Manag Care*. 2021;27(3) (suppl):S35-S41. [CrossRef]
- 16. Badesch DB, Raskob GE, Elliott CG, et al. Pulmonary arterial hypertension: baseline characteristics from the REVEAL Registry. *Chest*. 2010;137(2):376-387. [CrossRef]
- Kaymaz C, Mutlu B, Küçükoğlu MS, et al. Preliminary results from a nationwide adult cardiology perspective for pulmonary hypertension: Registry on clinical outcoMe and sUrvival in pulmonaRy hypertension Groups (SIMURG). Anatol J Cardiol. 2017;18(4):242-250. [CrossRef]
- 18. Wapner J, Matura LA. An update on pulmonary arterial hypertension. *J Nurse Pract*. 2015;11(5):551-559. [CrossRef]
- 19. Ünsar S, Özdemir Ö, Yenici Bulut E. Pulmonary Hypertension and Nursing care. *Turk J CardioVasc Nurs*. 2019;10(21):17-24.
- Sarzyńska K, Świątoniowska-Lonc N, Dudek K, et al. Quality of life of patients with pulmonary arterial hypertension: a meta-analysis. Eur Rev Med Pharmacol Sci. 2021;25(15):4983-4998. [CrossRef]
- Yorke J, Deaton C, Campbell M, et al. Symptom severity and its effect on health-related quality of life over time in patients with pulmonary hypertension: a multisite longitudinal cohort study. BMJ Open Respir Res. 2018;5(1):e000263. [CrossRef]
- 22. Gu S, Hu H, Dong H. Systematic review of health-related quality of life in patients with pulmonary arterial hypertension. *Pharmacoeconomics*. 2016;34(8):751-770. [CrossRef]