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Health-related quality of life, mental and sleep disorders of patients with pulmonary arterial hypertension participating in the Polish PH treatment program

Kathie Sarzyńska[1,2*], Beata Jankowska-Polańska[3,4]

1 Department of Nursing and Midwifery, Wroclaw Medical University, 5 Bartla Street, 51-618 Wroclaw, Poland
2 Provincial Specialist Hospital in Wroclaw, Research and Development Center, 73a H. Kamieńskiego St., 51-124 Wroclaw
3 Faculty of Medicine, Technical University of Wroclaw, 50-370 Wroclaw, Poland
4 Research and Innovation Center, 4th Military Teaching Hospital, 50-981 Wroclaw, Poland
*Correspondence: kathie@vp.pl
E-mail addresses
kathie@vp.pl (K. Sarzyńska)
bpolanska@4wsk.pl (B. Jankowska-Polańska)
ORCID:
Kathie Sarzyńska 0000-0003-0470-354X
Beata Jankowska-Polańska 0000-0003-1120-3535
Address for correspondence
Kathie Sarzyńska
Provincial Specialist Hospital in Wroclaw, Research and Development Center, 73a H. Kamieńskiego St., 51-124 Wroclaw
phone number: +48 883 652 438

ABSTRACT

Pulmonary arterial hypertension (PAH) is a rare, chronic disease that leads to the development of a range of heart failure-related symptoms that directly affect an individual's functioning. The aim of the study was to assess health-related quality of life (HRQoL) of the PAH patients. Participants completed several specific survey questionnaires. The mean scores obtained in each domain of the PAH-SYMPACT HRQoL questionnaire were: cardiopulmonary symptoms 1.02±0.57, cardiovascular symptoms 0.54±0.5, physical effects 1.24±0.9, mental effects 0.99±SD=0.82. The average scores on the anxiety and depression scales were 5.33±3.31, 4.4±3.78, respectively, which indicates their low severity. The mean score obtained on the Pittsburgh Sleep Quality Scale (PSQI) was 8.7±4.64 and represented the poor sleep quality affecting the study group. Sleep disorders were present in the majority of the subjects and had the most significant impact on reducing the HRQoL in the study group. Anxiety and depressive disorders were present in a small percentage of the subjects. The results showed that the best predictor of HRQoL evaluation is sleep quality assessment.

Keywords: Health-related quality of life (HRQoL); pulmonary arterial hypertension (PAH)

1. INTRODUCTION

Pulmonary arterial hypertension (PAH), is the first WHO group of pulmonary hypertension (PH). It is a rare, chronic, debilitating and progressive disorder characterized by obstructive remodeling of distal pulmonary vessels, result of right heart failure and death if not properly treated [1-5]. The current definition of pre-capillary PH is updated as mPAP (normal mean pulmonary artery pressure) > 20 mmHg, PCWP (pulmonary capillary wedge pressure) ≤ 15 mmHg, and PVR (pulmonary vascular resistance) >2WU (Wood units) in 2022 ESC/ERS PH guideline [6]. The new lower mPAP value reflects a greater appreciation for normal values (14±3 mmHg) and their prognostic relevance [6]. Similarly, the pulmonary vascular resistance

(PVR) has been reduced and a lower value of >2 WU and now defines a pre-capillary pattern of PH [7].

The prevalence of PAH is estimated at 5.8 people per million [8] with the incidence of 15-50 cases per million annually [9]. Since the widespread use of the various new treatment options, the long-term prognosis of PAH has significantly improved [10]. The median survival rates, presented in the REVEAL registry, are 86 and 61% at one and five years after diagnosis respectively, despite the use of targeted therapy [11]. Prolonged survival in PAH has drawn attention to the disease burden that PAH imposes on patients and health care systems [12]. Many researchers have shown disturbances in health-related quality of life (HRQoL) in this group of patients [13-17]. Abnormalities were observed in the overall assessment of HRQoLand in specific domains, depending on the type of questionnaire used. The most common HRQoL assessment instruments used by researchers for patients with PAH are generic or non-specific for the disease, and therefore may not accurately reflect the disease burden of PAH [13]. In the past, the measure of optimal treatment of PAH patients, was the assessment of functional, exercise capacity and hemodynamic parameters performed during right heart catheterization. However, improvements in physiological measures, such as resting pulmonary hemodynamic variables, are often not perceived by patients as providing recognizable benefits in daily life. It is suggested that HRQoL is a more relevant patient-reported measure in determining the effectiveness of therapeutic interventions and may be the starting point for possible treatment modification [18]. The factors that help determine the HRQoL of patients with PAH are related to the severity of cardiovascular and cardiopulmonary symptoms specific to this disease, as well as Cognitive/Emotional and Physical Impacts [19]. Due to the importance of determining the HRQoL of patients with PAH, it is necessary to use a research tool dedicated to patients with this particular condition, that takes into account the specifics of this disease entity, i.e. the type of symptoms and their impact on the patient's life [18, 19]. Considering the above, researchers developed the PAH-specific PRO instrument Pulmonary Arterial Hypertension-Symptoms and Impact (PAH-SYMPACT; Actelion Pharmaceuticals Ltd, Allschwil, Switzerland) questionnaire [20].

The use of questionnaires of HRQoL assesment, in daily clinical or outpatient practice in Poland is currently not widespread. Perhaps more frequent use such tools would be a good way to draw attention to symptoms and problems that significantly reduce a given patient's HRQoL. Therefore, the purpose of this study was to determine the HRQoL, anxiety, depression and sleep disorders among patients with PAH, and being treated for such condition

in a national hypertension therapy program. In addition, the study analyzed the impact of selected sociodemographic, and clinical variables on HRQoL for such patients.

2. MATERIALS AND METHODS

2.1. Study description

The study was conducted in two pulmonary arterial hypertension treatment centers. The basic criteria for inclusion in the study was: diagnosed PAH, confirmed by right heart catheterization and additional necessary mandatory tests, in accordance with current guidelines [6], age >18 years and chronic treatment (>6 months). Exclusion criteria was: diagnosed psychiatric/mental disorder or cognitive impairment that would prevent completion of the questionnaire, patients being treatment-naïve, mobility impairment due to neurological or skeletal-muscular conditions, pulmonary hypertension due to left heart disease, lung diseases and/or hypoxemia, chronic thromboembolic pulmonary hypertension (CTEPH), pulmonary hypertension with unclear and/or multifactorial mechanisms and lack of written consent to participate in the study. Before beginning the study, the design was reviewed and approved by the Bioethics Committee, in accordance with the Declaration of Helsinki (numbers: KB – 538/2019 and KB - 101/2023).

All participants were members of a national PAH drug therapy program, based on current ESC/ERS guidelines [6]. Each patient was under constant care of a multidisciplinary team, which included the following medical professions: a nurse, a physiotherapist and a doctor, according to the recommendations [6]. Consultations with the therapeutic team took place regularly, on average every 1-3 months, and each patient qualified to participate in the study had at least 3 series of meetings with the therapeutic team. Based on data from electronic medical records and paper questionnaires, the results of three standard noninvasive clinical prognostic measures: WHO-FC (World Health Organization Functional Classification), 6MWT (6 minutes walk test) and NT-proBNP (N-terminal pro b-type natriuretic peptide) were analyzed. Moreover, the study participants completed a self-authored survey and standardized questionnaires assessing: the quality of life (PAH-SYMPACT), the occurrence of anxiety and depression (HADS), sleep disorders (PSQI).

2.2. Research tools

1. Self-authored questionnaire - questions about gender, age, education and marital status and a rating by participants the intensity of selected PAH symptoms using the VAS scale.

2. Analysis of the patient's medical documentation - values of clinical parameters, i.e. NYHA/WHO functional class, TAPSE value, 6MWT result, NT-proBNP value and mean BMI score.

3. The Pulmonary Arterial Hypertension Symptoms and Impact Questionnaire (PAH-SYMPACT) is a PAH-specific patient-reported outcome scale that assesses patients' quality of life in four aspects: cardiorespiratory symptoms, cardiovascular symptoms, physical effects and cognitive/emotional impacts.

PAH-SYMPACT questionnaire has two constructs: PAH symptoms and PAH impact. The domains of PAH symptoms are cardiorespiratory symptoms and cardiovascular symptoms; the domains of PAH impacts are physical impacts and cognitive/emotional impacts. PAH-SYMPACT consists of 23 items (12 for the PAH symptom construct with a 24-hour recall period and 11 for the PAH effects construct with a one-week recall period). Scores for each item have five options ranging from 0 to 4, with higher scores indicating greater symptom severity or worse impact. There are no standards to say from how many points there are severe symptoms or major effects. However, since the subscale scores are averages of answers to single questions, they can be interpreted according to the key to a single question, in which 0 means no symptoms/effects, 1 - slight severity, 2 - medium severity, 3 - severe, and 4 - very severe. [21-22]. This questionnaire proved to have good internal consistency as indicated by Cronbach's alpha of above 0.7 for each subscale. Spearman's correlation coefficient in the test-retest study is above 0.9 for each subscale [23].

4. HADS – Hospital anxiety and depression scale, is an instrument to assess anxiety and depression. The HADS questionnaire is divided into two scales including seven items each for the screening of depression and anxiety. The total scores for depression and for anxiety range between 0 and 21 [24]. The HADS scores were interpreted according to the key to this scale, according to which:

- a score of 0 to 7 points indicates the absence of disorders,
- a score of 8 to 10 points indicates a borderline condition,
- a score of 11 to 21 points indicates a pronounced disorder.

Higher scores indicate greater depression and/or anxiety. We considered a score $\geqslant 8$ to represent symptoms of anxiety or depression. The HADS presented good internal consistency with Cronbach's α at 0.84 and 0.78 for depression and anxiety subscales, respectively, and 0.88 for the whole questionnaire [25, 26].

5. Pittsburgh Sleep Quality Index (PSQI) was used to assess participants' subjective sleep difficulties over the past 4 weeks. The PSQI consists of 18 items (excluding co-sleeping items), which are combined into seven subscales: sleep duration, sleep disturbances, sleep latency, daytime dysfunction, sleep efficiency, sleep quality and sleep medication. The subscales are scored on a 4-point scale. A global score with a possible range of 0 to 21 is obtained by adding the subscores; higher scores indicate worse overall sleep quality. The Pittsburgh Sleep Quality Index (PSQI) scale was used to assess sleep quality over the past month. The PSQI scale includes seven components that together make up the overall sleep quality score. The scale consists of 19 questions, which the patient answers on a scale of 0 to 3, where 0 means no problem and 3 means a serious problem in the category. A score above 5 suggests sleep quality problems and potential sleep disorders. This questionnaire proved to have high internal consistency as indicated by Cronbach's alpha of 0.83 and sufficient internal homogeneity [27].

2.3. Statistical analysis

Assessment of the consistency of empirical distributions of all continuous (e.g., age, BMI, etc.) and discrete (quality of life scores, etc.) quantitative variables with theoretical normal distributions was checked using the Kolmogorov-Smirnov test with Lilliefors correction and the Shapiro-Wilk test. For quantitative variables, mean values (M), standard deviations (SD), medians, lower (Q1) and upper (Q3) quartiles, and extreme values: smallest (Min) and largest (Max) were calculated. In the tables, variables with distributions close to normal were characterized by mean and standard deviation - M (SD), variables with distributions significantly different from normal were presented as medians and quartile range - Me [Q1; Q3]. For qualitative variables (nominal, e.g., gender, marital status, etc.) and ordinal variables (e.g., education level, WHO class, etc.), counts (n) and percentages (%) were calculated and collected in contingency (multivariate) tables. Correlations between quantitative variables were analyzed using Spearman's correlation coefficient. The significance level of 0.05 was adopted in the analysis. Therefore, all p values below 0.05 were interpreted as indicating significant relationships. Additionally, linear regression was used, a method based on linear combinations of variables and parameters that fit the model to the data. The analysis was performed in R, version 4.3.1 [28].

3. RESULTS

3.1. Study group

The patients' physical and pulmonary function profiles are presented in Table 1. The mean patient age was 57 ± 16,2 years. Most of the participants were women, n=66 (74.2%). Most patients (55%) were taking two medications for PAH and as many as 67.4% reported being in a relationship. Only 13.5% participants declared use of oxygen. The largest group were patients with WHO FC II (52%) and those with idiopatic type of PAH (60,7%). Among a range of PAH manifestations, symptoms such as dyspnea, fatigue and weakness were observed to be most severe (Table 1).

TABLE 1. Characteristics of the study group.

Characteristic	*Values (N = 89)*
Female:	*66 (74.2%)*
Age, mean (SD), years	*57.0 (16.2)*
Median [Q1; Q3]	*61 [43; 69]*
Min - Max	*19 - 85*
Marital status:	
In a relationship, n (%)	*60 (67.4%)*
Single, n (%)	*29 (32.6%)*
Education:	
Basic, n (%)	*16 (18.0)*
Vocational education, n (%)	*28 (31.5)*
Secondary, n (%)	*29 (32.6)*
Higher, n (%)	*16 (18.0)*
BMI (kg/m^2):	
Median [Q1; Q3]	*24.4 [20.7; 27.8]*
Min - Max	*13.3 – 43.0*
WHO FC:	
II	*46 (51.7%)*
III	*41 (46.1%)*
IV	*2 (2.2%)*
Oxygen use	
Yes	12 (13.5%)
No	*77 (86.5%)*

Drug therapy	
Monotherapy	20 (22.5%)
Duotherapy	49 (55%)
Tripple Therapy	20 (22.5%)
PAH etiology:	
Idiopathic	*54 (60.7%)*
Connective tissue disease	*16 (18.0)*
Congenital heart disease	*11 (12.3)*
Portal hypertension	*3 (3.4)*
After medication	*5 (5.6)*
Clinical parameters:	
6MWD, mean (SD), m	*399.3 (135.6)*
TAPSE, median [Q1; Q3], mm	*19 [17; 23]*
NT-proBNP, median [Q1; Q3], pg/ml	*496 [225; 1605]*
Number of hospitalizations due to worsening symptoms of pulmonary hypertension:	
Median [Q1; Q3]	*4 [3; 6]*
Min - Max	*0 - 20*
Pulmonary hypertension symptoms, VAS (1 – 10):	
Chest pain, Median [Q1; Q3]	*1 [1; 3]*
Dyspnea, Median [Q1; Q3]	*4 [1; 5]*
Cardiac arrhythmias, Median [Q1; Q3]	*1 [1; 4]*
Hemoptysis, Median [Q1; Q3]	*1 [1; 1]*
Fainting, Median [Q1; Q3]	*1 [1; 1]*
Weakness, Median [Q1; Q3]	*4 [1; 6]*
Fatigue, Median [Q1; Q3]	*6 [4; 7]*
Bleeding related to anticoagulant therapy	*1 [1; 2]*
Headaches, Median [Q1; Q3]	*3 [1; 4]*
Jaw pain, Median [Q1; Q3]	*1 [1; 3]*
Skin redness, Median [Q1; Q3]	*3 [1; 4.5]*
Diarrhea, Median [Q1; Q3]	*3 [1; 5]*

Note: Data expressed as n, %, mean (standard deviation) or median (25th percentile–75th percentile) as appropriate. Abbreviations: NTproBNP, N-terminal pro B-type natriuretic peptide; 6MWT- 6 minuts walk test; BMI, body mass index.

3.2 Health-related quality of life, anxiety, depression and sleep quality of the study group

The HRQoL determined by the PAH-SYMPACT questionnaire describes the results in 4 domains identified in Table 2 below. The mean scores obtained in each domain were: cardio-pulmonary symptoms 1.02 (SD=0.57), cardiovascular symptoms 0.54 (SD=0.5), physical effects 1.24 (SD=0.9), mental effects 0.99 (SD=0.82). Marked impairment on the anxiety scale, was exhibited by 8% of the subjects, while on the depression scale it was 9% of the subjects. However, as many as 70% of participants had poor sleep quality (Table 2).

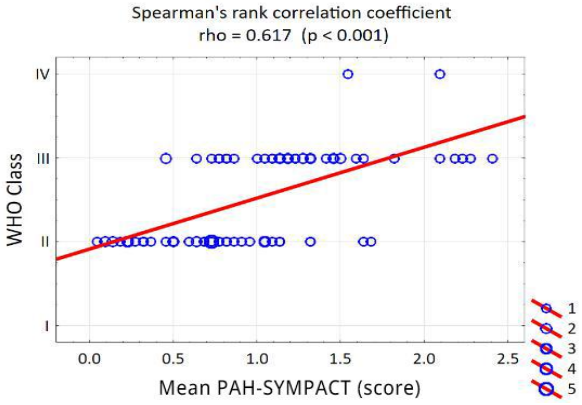
TABLE 2. The results of the PAH-SYMPACT, HADS and PSQI questionnaire and % of patients with impairment HRQoL.

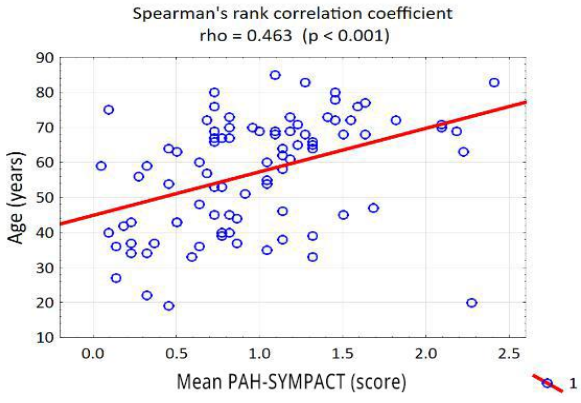
Survey	**Mean (SD)**	**Min-Max**	**% of patients**
PAH-SYMPACT			
Cardiopulmonary	1.02 (0.57)	0-2.5	
symptoms	0.54 (0.5)	0-2.2	61% reduced quality of life in at least one
Cardiovascular	1.24 (0.9)	0-3.86	domain (mean score > 1)
symptoms	0.99 (0.82)	0-3.75	
Physical effects			
Mental effects			
HADS			
Anxiety	5.33 (3.31)	0-15	8% anxiety (score ⩾8)
Depression	4.4 (4.64)	0-16	9% depression (score ⩾8).
PSQI	8.7 (4.64)	0-19	70% poor sleep quality (score >5); Disturbances at the seven PSQI components: • 40.4% poor subjective sleep quality (score >1) • 36 % increased sleep latency, (score >1) • 24.7% short sleep duration, ≤5 h of sleep per night

	• 37.1% reduced sleep efficiency (<85%) • 39.3% sleep disturbances, (score >1) • 12.4% sleep medications use, (score >1) • 36% daytime dysfunction, (score >1)

3.3. Correlation of the selected factors with the quality of life

The study found statistically significant correlations with the most common indicators, which are used to assess patient-reported treatment effects. Positive correlations ($r>0$) indicate that the higher the value of a factor, the lower the quality of life. In contrast, negative correlations ($r<0$) indicate that the lower the value of a factor, the lower the quality of life. The age, BMI, WHO FC, NT-proBNP result, correlates significantly ($p<0.05$) and positively ($r>0$) with PAH-SYMPACT mean score but negative correlation ($r<0$) was observed for the 6MWT score (Figure 1).





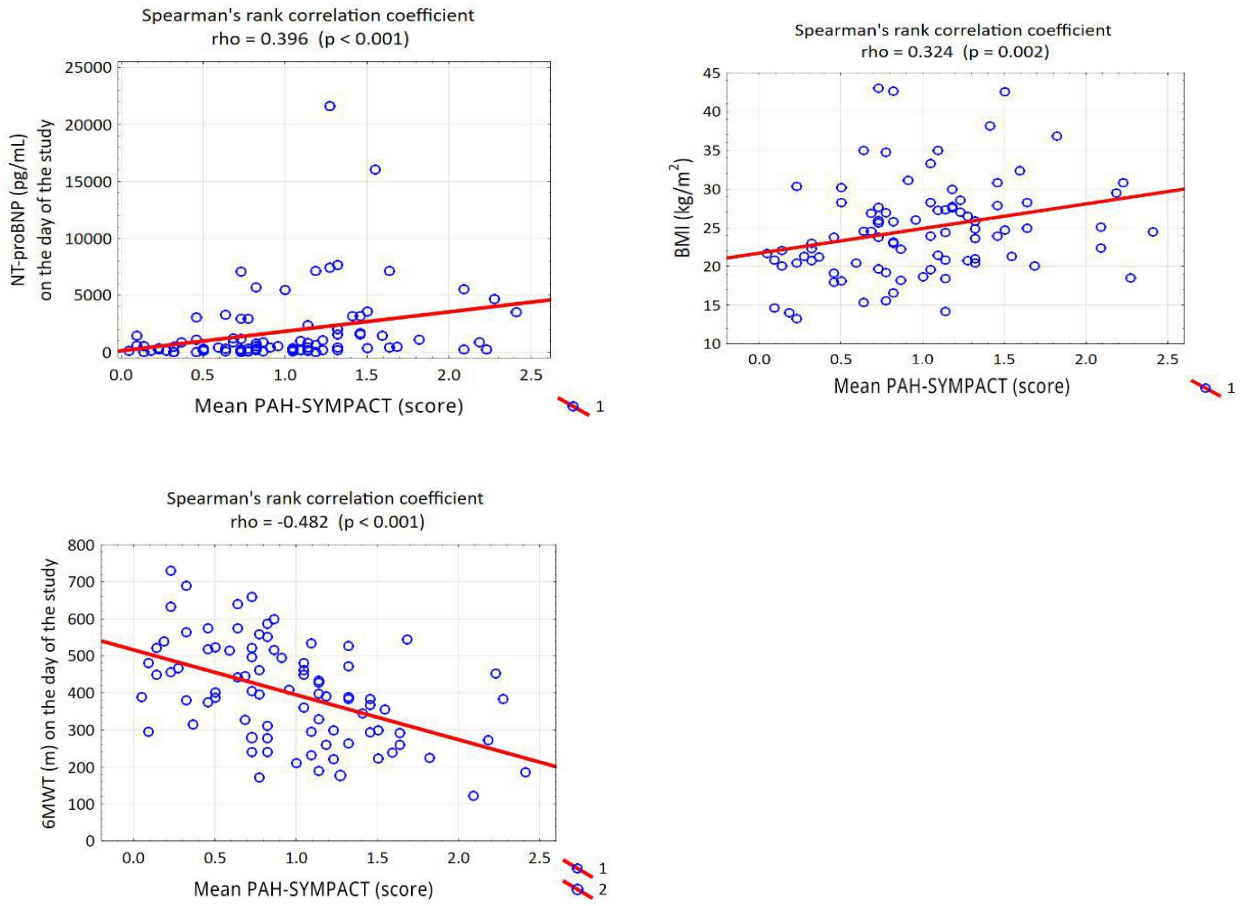


FIGURE 1. Spearman correlation of the selected factors (NT-proBNP, WHO FC, BMI, age, 6MWT) with the PAH-SYMPACT questionnaire.

Correlations of the HADS and PSQI questionnaires with individual domains of the PAH-SYMPACT questionnaire were performed. Anxiety, depression and sleep quality correlate significantly ($p<0.05$) and positively ($r>0$) with cardiopulmonary symptoms, cardiovascular symptoms, physical effects and mental effects (Table 3).

TABLE 3. Correlations of the PAH-SYMPACT questionnaire with individual questionnaires

PAH-SYMPACT	**HADS-A**	**HADS-D**	**PSQI**
	Spearman's correlation coefficient		

for assessing anxiety/depression (HADS) and sleep quality (PSQI).

Cardiopulmonary symptoms	r=0.456*	r=0.401*	r=0.41*
Cardiovascular symptoms	r=0.301*	r=0.319*	r=0.323*
Physical effects	r=0.355*	r=0.49*	r=0.6*
Mental effects	r=0.521*	r=0.534*	r=0.39*

*statistically significant ($p<0.05$)
HADS-A: Hospital anxiety and depression scale-anxiety subscale
HADS-D: Hospital anxiety and depression scale-depression subscale

3.4. Sleep quality and mental health as predictors of HRQoL in PAH patients.

In order to determine the numerical impact of the intensity of anxiety, depression and poor sleep quality on the results of individual domains of the PAH-SYMPACT questionnaire, multivariate linear regression was performed. It showed that each point on the anxiety scale and on the PSQI scale (poor sleep quality) increases cardiopulmonary symptoms by an average of 0.44 points and 0.31 points, respectively. Moreover, each point on the PSQI scale (poor sleep quality) increases cardiovascular symptoms by an average of 0.26 points and physical effects by an average of 0.8 points. However, each point on the depression scale increases mental effects by an average of 0.69 points (Table 4).

TABLE 4. Multivariate linear regression model of the PAH-SYMPACT questionnaire with the HADS and PSQI questionnaires.

Dependent variable	Independent variable	Regression coefficient	95%CI		*p*
Cardiopulmonary symptoms	HADS: Anxiety	0.44	0.01	0.87	**0.046** *
	HADS: Depression	0.09	-0.29	0.47	0.656
	PSQI	0.31	0.05	0.58	**0.021** *
Cardiovascular symptoms	HADS: Anxiety	0.16	-0.24	0.56	0.43
	HADS: Depression	0.12	-0.23	0.48	0.493
	PSQI	0.26	0.02	0.5	**0.04** *
Physical effects	HADS: Anxiety	0.08	-0.56	0.72	0.799
	HADS: Depression	0.55	-0.02	0.11	0.06
	PSQI	0.8	0.41	0.18	**<0.001** *
Mental effects	HADS: Anxiety	0.58	-0.02	0.17	0.062
	HADS: Depression	0.69	0.16	0.21	**0.013** *
	PSQI	0.17	-0.19	0.54	0.346

p - multivariate linear regression
* relationship statistically significant ($p<0.05$)

Overall, poor sleep quality were identified as the strongest predictors of HRQoL impairment across domains of PAH-SYMPACT questionnaire.

4. Discussion

According to the authors' knowledge, this research is the first comprehensive assessment of the quality of life of Polish patients with PAH, which simultaneously assesses the quality of life, sleep disorders and the occurrence of anxiety/depression of patients treated chronically. The most important results are: the general HRQOL of patients with PAH is moderate. Patients showed a low prevalence of anxiety and depression. The majority of subjects showed poor sleep quality. The multivariate linear regression showed that the best indicator for determining the HRQOL of PAH patients is the assessment of their sleep quality.

Evaluation of the HRQOL of patients with PAH, helps determine the patient's health status and the progression of ongoing treatment, which is reflected in the patient's clinical condition [18]. HRQoL has been used as an end point in clinical trials in PAH and has been recommended to help determine whether patients might derive benefit from treatment [19]. The specific PAH-SYMPACT questionnaire is relatively new tool in HRQoL investigations. Of the multitude of tools available for PRO assessment, the only tool we could identify which met criteria for regulatory use (FDA-approved) as well as capturing disease impact was PAH-SYMPACT [20]. The mean scores obtained in each domain of the above questionnaire were similar to those previously published [20-23, 29]. In our own study, the mean PAH-SYMPACT score indicated a moderate reduction in patients' HRQoL, as evidenced by the low severity of symptoms and effects of the disease. The subscales of cardiovascular and cardiopulmonary symptoms indicate their mild severity. However, 61% of patients reported a mean score >1.0 in at least one domain, indicating more than mild worsening of PAH symptoms or effects. A similar result was obtained in a study by DuBrock et al (2022), where 69% reported a mean score >1.0 in at least one domain [29]. In both studies, patients showed a number of common features such as: similar: mean age, 6MWT result, values of hemodynamic parameters, majority of subjects were female and % of patients on monotherapy. However, the DuBrock el al. [29] study, also included a small group of patients who had not yet started treatment, in contrast to our own study.

In the study that used the PAH-SYMPACT questionnaire as a tool to assess quality of life, the highest average score (mean) was observed in the physical domain. This results are related to the progression of the disease and its impact on daily functioning. In contrast, patients showed the least severity of symptoms in the cardiovascular symptoms domain, which includes questions about the intensity of experiencing cardiac arrhythmias, chest pain, and lightheadedness [20-23, 29]. The results indicate that the severity of cardiovascular symptoms in patients with PAH, is not as severe as the severity of cardiopulmonary symptoms, where higher values were obtained.

Studies analyzing the prevalence of depression among patients with PAH have shown that it ranges from 9% to 40% [16; 30-33]. In our findings, the percentage of respondents with depression was only 9%. Similarly, we obtained a low rate of 8% for anxiety, while other studies have found the percentage of PAH patients with anxiety disorders ranged from 23-43% [16; 30-33]. The reasons for the small number of subjects with marked depressive and anxiety disorders, may be related to the individualized interdisciplinary medical care model used in the study, which could affect the psychological patient's well-being. Studies indicate that interdisciplinary team activities in the care of patients with chronic diseases improved their quality of life [34], and the study conducted by Bekelman et al. [35] demonstrated that a multidisciplinary team could reduce depressive symptoms in patients with heart failure. Moreover, since all participants were members of a free cost PAH drug program funded by the Polish National Health Fund and therefore, patients were not exposed to the financial burden, which could also have affected the results. The connection between the cost of treatment and its impact on the mental state of PAH patients has previously demonstrated [36], that, the higher the individual cost to the individual patient, the higher the risk of mental disorders such as anxiety or depression.

This study confirmed sleep disturbances are common in PAH patients (70% with poor sleep quality) and associated with psychological distress and HRQOL. Studies conducted by others researchers, also indicate that poor sleep quality was present in a significant percentage of PAH patients. Similar results were obtained by Matura el al. [37] and Batal et al. [38] where the percentage of patients classified as poor sleepers was 66% and 72.5% respectively. The problem of sleep disorders among patients with PAH is not fully understood [39]. They may be one of the effects of the progression of the disease (increasing dyspnea) and unoptimized treatment [6]. Current evidence demonstrates a higher prevalence of impaired sleep quality, sleep-disordered breathing, sleep-related hypoxia, and restless leg syndrome in patients with

PAH. Recent data suggest that sleep-related hypoxia is strongly linked to reduced right ventricular function and higher risk of transplantation or death [40, 41]. In various stages of PAH patients, experience shortness of breath (dyspnea), which is one of the symptoms of the disease [1, 4]. Exercise-related dyspnea during the day is different from dyspnea occurring during nighttime rest, which is most often caused by hypoxemia [39]. A study performed by Rafanan et al. [42], found that 77% of PAH patients had significant nocturnal hypoxemia. Other studies demonstrated that sleep disordered breathing was present in 71% of subjects [43]. In 2002, Schulz et al. [44] investigated the presence of periodic breathing (PB) in patients with PAH. It has been shown that PB was detected in almost one in three patients (30%) with advanced primary pulmonary hypertension and has been reversed by nocturnal nasal oxygen. In our research, only 13.5% of respondents reported using oxygen in the past 24 hours. The low percentage of patients using oxygen may have caused sleep disturbances. However, limited studies have investigated the role of oxygen therapy or positive airway pressure therapy improving symptoms or outcomes. In patients with PAH, untreated and undetected nocturnal hypoxemia can have deleterious effects and can worsen not only the patient's quality of life, but also the progression of the underlying disease [45]. In a recent published study by Benjamin et al. patients with PH underwent long-term O2 therapy (LTOT). This study highlights the importance of LTOT in terms of exercise capacity in patients with PAH and CTEPH. The LTOT was well tolerated and led to significant improvement of 6MWD [46]. In addition, studies indicate that in patients with heart failure, sleep disturbances worsen with advancing age [47], which was also demonstrated in our study. The multivariate linear regression model, used highlighted that the sleep quality assessment was the best predictor of HRQOL in our subjects, which was also shown by Nunes et al. (2009) [48]. Adequate diagnosis and management of sleep abnormalities could lead to improvement of patient’s HRQoL. To confirm the obtained results, further studies are necessary on a larger group of patients in several PAH treatment centers.

Limitations

A limitation of the study was that it was conducted in the same region, but in two different pulmonary hypertension treatment centers. Another limitation of the study was the lack of a control group. In addition, due to the relatively small number of subjects, however PAH is a disease with a rare occurrence in the general population. To avoid bias, a number of exclusion criteria were applied to the study.

5. CONCLUSIONS

Health-related quality of life is reduced in the majority of PAH patients on chronic treatment in at least one domain. The reduction of quality of life in the domain of physical effects is most significant. Anxiety and depression are not present in most PAH patients, in contrast to sleep quality disorders.

Disclosures

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