# Cultural adaptation of the Polish version of the PAH-SYMPACT questionnaire (PL)

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**Research Article**

**Keywords:** Pulmonary arterial hypertension, PAH, PAH-SYMPACT, validation

**Posted Date:** April 30th, 2021

**DOI:** https://doi.org/10.21203/rs.3.rs-453005/v1

# Abstract

**Background:** Pulmonary arterial hypertension (PAH) is a rare chronic disease that significantly affects patients' ability to perform daily activities, decreasing physical and mental well-being and social functioning. Symptoms of the disease increase as it progresses, leading to disability and death. In addition, the quality of life of patients deteriorates as the disease progresses and may reflect the effectiveness of the therapy used. Most questionnaires used in PAH clinical trials are generic quality-of-life (QoL) measures that may do not adequately reflect the clinical status and symptoms, changes in health status, or prognosis of patients with PAH. The purpose of this study was to translate and culturally adapt the Polish version of the PAH-SYMPACT questionnaire to reflect the severity of symptoms and their effects on patients' daily functioning.

**Material and methods**: The study included 55 patients with PAH treated, as part of a pulmonary hypertension treatment program in Poland. Validation of this questionnaire met the guidelines for translation and validation of questionnaires, which are set out in the document on the process of translation and adaptation of World Health Organization tools. Reliability was tested using a measure of internal consistency (Cronbach's α) and test-retest reliability. Correctness was confirmed using known group validity.

**Results**: Moderate internal consistency (Cronbach's) was found for all questionnaire domains (cardiopulmonary symptoms α=0.706; cardiovascular symptoms α=0.806, physical effects α=0.904, psychological effects α=0.817) and test-retest reliability was satisfactory. The PAH-SYMPACT questionnaire (PL) met all validation criteria and can be used to assess quality of life in patients with PAH.

**Conclusion:** Psychometric evaluation of the Polish version of the PAH-SYMPACT (PL) indicates that it is a reliable and valid measure tool to detect quality of life patients with PAH.


# Introduction

PAH is a disease whose course leads to the development of right ventricular failure and death despite optimal treatment [1]. A study published by Kopeć et al. in 2020 shows that the estimated prevalence of patients with PAH in Poland is 30.8/million adults. [2, 3 Recent advances in therapy have improved symptom severity, functional capacity and survival in PAH to varying degrees, but PAH remains an incurable disease. A median survival was estimated of 7 years [4]. Prediction of survival in PAH is thus an important consideration for the management of patients with this disease and informs individual therapeutic decisions [5].

The main symptom of PAH is dyspnoea. However, the experience of this as well as other symptoms is subjective, so tools are needed that are appropriately sensitive to patient needs and patient outcomes. PAH study endpoint recommendations now emphasize the importance of measuring patient-reported outcome as a secondary endpoint in clinical trials. [7]

In clinical practice, various questionnaires have been used to assess quality of life in patients with PAH, one of the first specific tools used for this purpose is the Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR). This questionnaire has been widely used in several countries among patients with PH over the last decade [8–10]. Another specific tool is the PAH-SYMPACT questionnaire, which does not have a validated Polish version, unlike the CAMPHOR questionnaire. The CAMPHOR questionnaire differs from the PAH-SYMPACT in being developed on the theoretical basis of the needs-based model of quality of life, rather than a measure of symptoms and impacts [7].

In the few studies carried out so far in Poland among patients with PAH only generic tools have been used, i.e. general tools for quality of life assessment regardless of the type of disease entity [11], which may not reflect the symptoms and effects of PAH and their impact on QoL. Due to the need for research in this group of patients, we have undertaken to translate and validate a tool that can be used to assess quality of life and relate it to data presented by other centers in Europe and worldwide. Further work is thus mandatory to validate PAH-specific questionnaires that are responsive to clinical changes as well as to determine their interpretability.

# Material And Methods

### Questionnaires used in psychometric validation process

- The Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR) questionnaire is a specific, standardized tool for assessing quality of life in patients with pulmonary hypertension. The CAMPHOR questionnaire was developed in the United Kingdom and subsequently validated for use in the United States as an instrument that measures health related quality of life (HQoL). CAMPHOR consists of three parts, which form scales about how the subject feels on the day the questionnaire is completed. The symptom scale contains a total of 25 items (maximum score = 25), the functional scale contains 15 items (maximum score = 30), and the quality of life section contains 25 items (maximum score = 25). The higher the score, the more severe the impairment for each part of the questionnaire [12].
- WHO-QOL BREF - a standardized, abbreviated version of the generic questionnaire - in the Polish language version, assessing the quality of life of a patient with any disease entity. It contains 26 questions analysing four domains of life and, separately, perception of quality of life and self-assessment of health status. Larger numbers mean better quality of life. The perception of quality of life and health consist of one question each from the questionnaire (questions number 1 and 2, respectively), so the results can be interpreted according to the content of the answers to these questions. The other domains of quality of life are made up of multiple questions, and there are no standards to determine which scores represent good or poor quality of life. However, all domains are expressed on the same scale, so quality of life can be compared across domains [13].
- PAH-SYMPACT (Pulmonary Arterial Hypertension-Symptoms and Impact) is a specific questionnaire to assess the quality of life of patients with pulmonary arterial hypertension. The questionnaire assesses the severity of cardiopulmonary and cardiovascular symptoms of PAH as well as the

physical and psychological impact of the disease. Each subscale of the questionnaire takes values from 0 to 4 and a higher score indicates more severe symptoms/more effects of the disease. There are no norms allowing to say from how many points we speak about strong symptoms or big effects. However, since the subscale scores are averages of the answers to individual 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 [7].

## Patients

The study was conducted between March 1, 2019 and January 30, 2020 at the Wroclaw Hospital, which is one of the three centers in Lower Silesia that provide treatment for patients with pulmonary hypertension under the Pulmonary Arterial Hypertension Treatment Program of the National Health Fund. The research team, consisting of two physicians and two nurses, defined the qualification criteria of the patients in the study protocol. The inclusion criteria were: age > 18 years, diagnosis of pulmonary arterial hypertension, entry into the national pulmonary hypertension database, written informed consent of the patient for participation in the study, health condition allowing participation in the study and completion of questionnaires. Patients were excluded from the study if they met at least one of the following exclusion criteria: age < 18 years, pulmonary hypertension other than PAH, coexistence of severe chronic diseases (left ventricular heart failure, ischemic heart disease, neoplastic diseases, acute respiratory diseases), cognitive impairment (Down syndrome), diagnosis of mental disease, which impeded conscious participation in the study both at the stage of qualification and later follow-up. Among the subjects treated for pulmonary hypertension at the center during the study period, 36 subjects were disqualified because they did not meet the study inclusion criteria with the main reasons being non-arterial pulmonary hypertension, genetic disease, mental illness, and refusal to participate in the study. Therefore, 55 patients attending routine follow-up appointments scheduled at the hospital outpatient clinic were eligible for the study. Patients at the visit were asked to complete a paper version of the questionnaire survey with questionnaires: PAH-SYMPACT. The questionnaire was administered twice approximately 2–4 weeks apart. Patients also completed the WHO-QoL BREF at the first visit. Demographics (sex, age, marital status, occupation) and disease characteristics (time since diagnosis, and disease severity) was also collected. Fifty-two patients participated in the second study; three patients dropped out of the re-survey without giving a reason.

## The process of cultural adaptation of the Polish version of the PAH-SYMPACT questionnaire

PAH-SYMPACT is a specific tool to measure the symptoms of pulmonary arterial hypertension and assess their effects (Fig. 1).

Identification of impairments in the symptoms domain is conducted in two subscales: cardiopulmonary symptoms and cardiovascular symptoms. In contrast, assessment of the severity of disease effects is conducted in two domains: physical effects and psychological effects. PAH-SYMPACT is a new tool, validated in the English language version for the US population [7], which does not have a validated Polish version.

Permission to translate and validate the PAH-SYMPACT questionnaire was obtained from MAPI Research Gate. MAPI Research Gate provided an English version of the questionnaire along with instructions for the translation and adaptation procedures from English to the selected language. We followed the standard translation protocol, which has a number of steps that include a forward translation, a panel of experts, a back translation, pretesting and creation of the final version [14]. Three steps were used in the adaptation of the Polish version of PAH-SYMPACT.

### Step 1

Translation process (Questionnaire forward translation, Questionnaire back translation)

The original PAH-SYMPACT was independently translated to Polish by 2 native speakers of Polish who are proficient in English: a medical doctor, nurse and a psychologist all with academic and clinical experience. To support accuracy and unambiguity of the final translated version, the translation was compared with the original English version and discussed. The expert panel verified the wording and meaning of all questions, as well as the clarity and correctness of the instructions with regard to the possible application of the questionnaire in Polish patients. After the expert panel discussed the suggested changes, the final version of the questionnaire was sent to a professional translation agency, with a request to revise the resulting translated Polish version of PAH-SYMPACT. After minor grammatical corrections, the final Polish version of PAH-SYMPACT(PL) was obtained. A British native speaker who is a professional English language editor at the Wroclaw Medical University, who lives in Poland and speaks Polish well, was asked to do a back-translation of the PAH-SYMPACT (Polish to English). The translator did not participate in the expert panel and was blinded to the content of the original PAH-SYMPACT. After completing the back-translation, the translator was shown the original PAH-SYMPACT and was asked to compare it with a back-translated version of PAH-SYMPACT(PL) in order to report the potential discrepancies of the meaning. The resulting version was verified by a professional translation agency. The purpose of this was to check whether the translated Polish version of the questionnaire corresponds to the original English version in terms of grammar and whether the translation captures the same context of understanding of each question due to cultural differences.

### Step 2

Cognitive debriefing interviews

The final Polish version, approved by the author of the original questionnaire, was verified in a pilot study. For this purpose 15 people were then interviewed. All were non-medically educated, patients with PAH, no apparent cognitive impairment. Their level of education ranged from primary to higher. The interview was performed in a semi-structured way according to general guidelines [15] and a protocol for questionnaire pretesting [16]. The aim of the interview was to assess the degree of understanding of the introduction, each statement, and the rating scale labels of the translated questionnaire. The feedback received was used to construct the final Polish version of the PAH-SYMPACT(PL) and to complement further quantitative analysis. Written informed consent was received from all participants before taking part. The

pilot study showed that patients had a good understanding of the questions included in the PAH-SYMPACT(PL) questionnaire, did not need additional explanations and were able to complete the questionnaire on their own. A final report was prepared from the whole translation process and the results were submitted to MAPI Research Gate, which accepted all translation steps undertaken.

### Step 3

Psychometric validation

To determine the construct validity of the translated Polish version of the PAH-SYMPACT (PL) questionnaire, we calculated the reliability coefficient (Cronbach's alpha) for each subscale and determined the internal consistency of the translated version using the test-retest method.

### Ethical Considerations

The study was approved by the Bioethics Committee of the Wroclaw Medical University, Poland (approval number 538/2019). All patients provided informed consent, and were informed that they could withdraw from the study at any time. The study protocol was carried out in accordance with the tenets of the Declaration of Helsinki and Good Clinical Practice guidelines.

### Description of statistical methods

The analysis of quantitative variables (i.e., expressed by number) was conducted by calculating the mean, standard deviation, median, quartiles, minimum, and maximum. The analysis of qualitative variables (i.e., not expressed by number) was performed by calculating the number and percentage of occurrences of each value. Comparison of the values of quantitative variables in the two groups was performed using the Mann-Whitney test. The internal consistency of PAH-SYMPACT was assessed using Cronbach's alpha. Convergent validity was assessed by correlating PAH-SYMPACT and CAMPHOR and WHOQoL-BREF scores. Correlations between test and retest scores were analyzed using Spearman's correlation coefficient (as scores did not have a normal distribution). A significance level of 0.05 was assumed in the analysis. Thus, all p values below 0.05 were interpreted as indicating significant correlations. The analysis was performed in the program R, version 3.6.3 (*R Core Team (2019). R: A language and environment for statistical computing. R Foundation for Statistical Computing, Vienna, Austria. URL* https://www.R-project.org/*.)*

# Results

### Study group

More than half (52.73%) of the respondents were over 60 years of age (mean 56.22 ± 17.25). Most of the respondents were female (63.6%) and 69% of the respondents were in relationship. Most of the respondents had vocational education (38.1%) and the smallest group of respondents had higher education (10.91%) (Table 1).

Table 1
Selected socio-clinical characteristics of the study group patients.

| Feature | | n | % |
|---|---|---|---|
| **Age** | 18– 20 | 2 | 3,64% |
| | 21–30 | 3 | 5,45% |
| | 31–40 | 8 | 14,55% |
| | 41–50 | 6 | 10,91% |
| | 51–60 | 7 | 12,73% |
| | 61–70 | 18 | 32,73% |
| | 71–80 | 11 | 20,00% |
| **Sex** | Women | 35 | 63,64% |
| | Men | 17 | 30,91% |
| | No answer | 3 | 5,45% |
| **Education** | Basic | 9 | 16,36% |
| | Vocational | 21 | 38,18% |
| | Secondary | 19 | 34,55% |
| | Higher | 6 | 10,91% |
| **WHO Functional Class** | I | 1 | 1,82% |
| | II | 25 | 45,45% |
| | III | 27 | 49,09% |
| | IV | 2 | 3,64% |
| **Relationship Status** | In relationship | 38 | 69,09% |
| | Single | 17 | 30,91% |
| **Year of PAH treatment initiation** | 2016 | 28 | 50,91% |
| | 2017 | 13 | 23,64% |
| | 2018 | 9 | 16,36% |
| | 2019 | 5 | 9,09% |

*6MWT- 6 minuts walk test, WHO- World Health Organization, TAPSE- Tricuspid annular plane systolic excursion, NT-proBNP- Ventricular natriuretic peptide, PDE5- A phosphodiesterase type 5.

| Feature | | n | % |
|---|---|---|---|
| **Wartość TAPSE*** | 5–10 | 1 | 1,82% |
| | 11–15 | 526 | 9,09% |
| | 16–20 | 10 | 47,27% |
| | 21–25 | 6 | 18,18% |
| | 26–30 | 8 | 10,91% |
| | 31–35 | 1 | 14,55% |
| **6MWT*** | 0–100 | 2 | 3,64% |
| | 101–200 | 12 | 21,82% |
| | 201–300 | 10 | 18,18% |
| | 301–400 | 12 | 21,82% |
| | 401–500 | 9 | 16,36% |
| | 501–600 | 2 | 3,64% |
| | 601–700 | 8 | 14,55% |
| **NT-proBNP* value** | 0–2000 | 34 | 61,82% |
| | 2001–4000 | 8 | 14,55% |
| | 4001–6000 | 3 | 5,45% |
| | 6001–8000 | 10 | 19,18% |
| **Medication** | PDE5* inhibitors + Prostanoids inhaled + Endothelin receptor antagonists | 5 | 9,10% |
| | PDE5* inhibitors + Endothelin receptor antagonists + Prostanoids intravenous | 7 | 12,72% |
| | Prostanoids intravenous + PDE5* inhibitors | 8 | 14,55% |
| | PDE5* inhibitors+ Endothelin receptor antagonists | 16 | 29,1% |
| | Endothelin receptor antagonists + Prostanoids intravenous | 2 | 3,60% |
| | Prostanoids inhaled + PDE5* inhibitors | 3 | 5,5% |
| | PDE5* inhibitors | 7 | 12,73% |
| | Prostanoids intravenous | 1 | 1,8% |

*6MWT- 6 minuts walk test, WHO- World Health Organization, TAPSE- Tricuspid annular plane systolic excursion, NT-proBNP- Ventricular natriuretic peptide, PDE5- A phosphodiesterase type 5.

| Feature | | n | % |
|---|---|---|---|
| | Endothelin receptor antagonists | 4 | 7,3% |
| | Prostanoids inhaled | 1 | 1,8% |
| | Riocyguat | 1 | 1,8% |
| | Monotherapy | 14 | 25,50% |
| | Dual therapy | 29 | 52,73% |
| | Tripletherapy | 12 | 21,8% |

*6MWT- 6 minuts walk test, WHO- World Health Organization, TAPSE- Tricuspid annular plane systolic excursion, NT-proBNP- Ventricular natriuretic peptide, PDE5- A phosphodiesterase type 5.

### Reliability of the PAH-SYMPACT subscales

The reliability for each subscale of the translated Polish version of the PAH-SYMPACT questionnaire was determined using Cronbach's alpha. In all 4 domains, all subscales were shown to be reliable as Cronbach's alpha coefficient for each subscale is above 0.7 (Table 2), an alpha above 0.7 is assumed to be a reliable scale [17]. Removing any of the items does not improve the alpha coefficient. Considering this, it can be concluded that the created scale is well constructed and does not need to be changed.

Table 2
Cronbach's alpha coefficient for each subscale of the PAH-SYMPACT questionnaire.

| Subscale | Cronbach's alpha |
|---|---|
| Cardiopulmonary Symptoms | 0,706 |
| Cardiovascular Symptoms | 0,806 |
| Physical effects | 0,904 |
| Psychological effects | 0,817 |

### PAH-SYMPACT consistency assessment

Consistency over time was examined by correlating the results of the first and second tests. The result indicated a very high consistency, between the repeated tests performed. Indeed, the correlation coefficient was always above 0.9 and was statistically significant ($p < 0.05$) for each subscale (Table 3). There were no cultural/linguistic differences between Poland and the creators' home country in terms of what daily life is like for PAH patients.

Table 3
Spearman correlation coefficient for each subscale of the PAH-SYMPACT questionnaire in the test-retest study.

| Subscale | Spearman correlation coefficient | p |
|---|---|---|
| Cardiopulmonary Symptoms | 0,945 | p < 0,001 |
| Cardiovascular Symptoms | 0,911 | p < 0,001 |
| Physical effects | 0,993 | p < 0,001 |
| Psychological effects | 0,935 | p < 0,001 |

### Quality of life determined by the PAH-SYMPACT questionnaire (PL)

The mean score on the cardiopulmonary symptoms subscale was 1.03 (or 1 when rounded), which can be interpreted as low severity. On another subscale of cardiovascular symptoms, the mean score was 0.56 (or 1 when rounded), which can be interpreted as low severity. On the other hand, the mean score on the physical effects subscale was 1.32 points (i.e., rounded 1), which can be interpreted as low severity. On the other hand, the mean score on the psychological effects subscale was 0.89 (or 1 when rounded), which can be interpreted as low severity (Table 4).

Table 4
Scores for each subscale of the PAH-SYMPACT questionnaire.

| PAH-SYMPACT | N | Mean | SD | Median | Min | Max | Q1 | Q3 |
|---|---|---|---|---|---|---|---|---|
| Cardiopulmonary Symptoms | 55 | 1,03 | 0,56 | 1,00 | 0,00 | 2,50 | 0,67 | 1,33 |
| Cardiovascular Symptoms | 55 | 0,56 | 0,56 | 0,40 | 0,00 | 2,20 | 0,10 | 0,90 |
| Physical effects | 55 | 1,32 | 0,94 | 1,14 | 0,14 | 3,86 | 0,50 | 2,00 |
| Psychological effects | 55 | 0,89 | 0,74 | 0,75 | 0,00 | 2,75 | 0,50 | 1,25 |

### Correlation coefficient of individual subscales of PAH-SYMPACT with subscales of CAMPHOR and WHO-QOL BREF questionnaires

Convergent validity was performed by correlating the results of individual subscales of the PAH-SYMPACT questionnaire with the results of individual subscales of the specific questionnaire for patients with pulmonary hypertension CAMPHOR, which has a Polish language version and cultural adaptation [18]. The second questionnaire with which the degree of correlation in individual domains of the PAH-SYMPACT(PL) questionnaire was compared was the WHOQoL BREF generic questionnaire, used to assess the quality of life of ill and healthy patients and applied, among others, in studies on the quality of life of patients with lung diseases [19].

Correlation results showed that each subscale of the specific CAMPHOR questionnaire correlated with all domains of the PAH-SYMPACT questionnaire in a statistically significant manner ($p < 0.05$). However, in the correlation between WHOQoL- BREF and PAH-SYMPACT, the only domain that correlated with all domains of the PAH-SYMPACT questionnaire was the physical domain.

In terms of the other domains of the WHO-QoL BREF questionnaire, the domains of self-perception of health, psychological domain, and social and environmental domains correlated with all domains of the PAH-SYMPACT except for the domain of cardiovascular symptoms. This may indicate that the use of a generic questionnaire does not adequately reflect the quality of life level of patients with PAH (Table 5).

Table 5
correlation coefficient of individual subscales of PAH-SYMPACT with subscales of CAMPHOR and WHO-QOL BREF questionnaires.

| **Variable** | **PAH-SYMPACT correlation coefficient** | | | |
|---|---|---|---|---|
| | **Cardiopulmonary symptoms** | **Cardiovascular symptoms** | **Physical effects** | **Psychological effects** |
| **CAMPHOR**: Symptoms | r = 0,564, p < 0,001 | r = 0,368, p = 0,006 | r = 0,773, p < 0,001 | r = 0,466, p < 0,001 |
| Physical fitness | r = 0,307, p = 0,023 | r = 0,295, p = 0,029 | r = 0,61, p < 0,001 | r = 0,266, p = 0,05 |
| Perception of quality of life | r = 0,552, p < 0,001 | r = 0,311, p = 0,021 | r = 0,709, p < 0,001 | r = 0,418, p = 0,001 |
| **WHO-QOL BREF**: Perceptions of self-reported health | r=-0,516, p < 0,001 | r=-0,077, p = 0,578 | r = 0,554, p < 0,001 | r = 0,399, p = 0,003 |
| Physical domain | r=-0,55, p < 0,001 | r=-0,354, p = 0,008 | r = 0,777, p < 0,001 | r = 0,638, p < 0,001 |
| Psychological domain | r=-0,426, p = 0,001 | r=-0,148, p = 0,282 | r=-0,616, p < 0,001 | r=-0,554, p < 0,001 |
| Social domain | r=-0,443, p = 0,001 | r=-0,178, p = 0,193 | r=-0,638, p < 0,001 | r=-0,413, p = 0,002 |
| Environmental domain | r=-0,347, p = 0,009 | r = 0,013, p = 0,927 | r=-0,402, p = 0,002 | r=-0,312, p = 0,02 |

# Discussion

This study shows that the Polish adaptation of PAH-SYMPACT was successful. The new language version meets the expectations of good internal consistency, test-retest reliability, and convergent and

known group validity. The resulting version reliably examines symptoms (resulting from the disease) as well as outcomes (consequences of symptoms) in patients with PAH.

Numerous studies confirm low quality of life in patients with PAH [9, 10, 20], which is lower the longer the disease persists despite optimal treatment [21]. In our study, the quality of life of patients with PAH was assessed according to the specific questionnaire as satisfactory and according to the generic one as inadequate for the group of patients in the terminal stage, which is confirmed by other studies [22]. Carefully developed QoL scales provide a holistic picture of the impact of disease and its treatment on the patient. In the case of chronic or terminal illness where no effective cure is available, emphasis should be placed on improving QoL as the goal of treatment [18].

To accurately identify the problems of patients with PAH, it is necessary to construct and use specific tools to help identify them. Recognition of abnormalities in quality of life assessment will help to improve the construction of therapeutic goals and will allow for the optimization of treatment methods. Currently, there is a deficit in the availability of Polish-language specific questionnaires for assessing the quality of life of patients with PAH, which is evident in studies conducted in Poland on this group of patients. In order to gain a better understanding of the problems of Polish patients with PAH, there is a need for specific tools which will help to identify and eliminate them [7].

Similar findings have been reported for previous adaptations of the questionnaire [6]. Cronbach's alpha coefficient for individual subscales of the PAH-SYMPACT questionnaire was always > 0.7, and Spearman's correlation coefficient in the test-retest study was > 0.9. Compared with the original American version of the questionnaire, where the adaptation process yielded coefficients > 0.8 and > 0.7, respectively, which indicates that the Polish version of the PAH-SYMPACT questionnaire has a very good design and is a reliable version of the questionnaire for assessing quality of life. Among the specific tools that can also examine quality of life in patients with PAH is the CAMPHOR questionnaire. In our study, each domain of the PAH-SYMPACT, correlated with all domains of the CAMPHOR questionnaire, which indicates that both tools can be interchangeably or concurrently used successfully in the assessment of quality of life of patients with PAH, although it should be emphasized that only the PAH-SYMPACT questionnaire is a questionnaire dedicated to a narrow group of PAH patients. An American cultural adaptation of the PAH-SYMPACT questionnaire [7] yielded similar results to our own study. For each domain: cardiopulmonary symptoms, cardiovascular symptoms, physical effects, psychological effects: 1.0 (SD = 0.5); 0.4 (SD = 0.5); 1.3 (SD = 0.9); 0.9 (SD = 0.8) compared to our own results, respectively: 1.03 (SD = 0.56); 0.56 (SD = 0.56); 1.32 (SD = 0.94); 0.89 (SD = 0.74). In both studies, the results should be interpreted as low severity of symptoms and effects. Only in the study by Chen et al [7] in the subscale of cardiovascular symptoms the result can be interpreted as the absence of symptoms. However, it should be emphasized that despite the similarities in both studies such as female predominance in the study group > 60%, mean age > 55, our study included patients chronically treated for PAH. In contrast, in the study by Chen et al [7], less than half of the subjects were treated for PAH in the past (46%) of which 42% were treated by monotherapy while in our study most of the subjects (52%) received dual therapy. The

low intensity of symptoms obtained in our study may indicate the effectiveness (optimality) of the therapy provided.

It is significant, that PAH-SYMPACT questionnaire is a relatively young tool, because of this, there are not many works in the literature using it to assess quality of life, nor many works presenting correlations of this tool with other questionnaires measuring quality of life. In our own work, we used the WHOQoL-BREF general questionnaire, which showed a worse correlation with the PAH-SYMPACT compared to the correlation with the specific CAMPHOR questionnaire. The PAH-SYMPACT domain with which no WHOQoL-BREF subscale correlated, except for the physical domain, was the cardiovascular symptoms domain. These results suggest that the WHOQoL-BREF questionnaire may not be an adequately sensitive tool for measuring quality of life in this specific group of PAH patients. However, a study by Chin et al (2018) [7] showed that another general quality of life assessment questionnaire, the SF-36, correlated with all domains of the PAH-SYMPACT questionnaire ($p < 0.05$), which may be evidence that only some general quality of life assessment questionnaires are not applicable in the assessment of the quality of life of patients with PAH, or another explanation for this situation should be sought. In the literature, there are also studies on the relationship between the specific CAMPHOR questionnaire and the SF-36 general quality of life assessment questionnaire. Both questionnaires in the study by Gardenton et al. correlated with each other in all domains in a statistically significant manner ($p < 0.05$) [7, 23]. However, Ganderton et al [8] noted that in clinical trials, the SF-36 has been shown to respond only moderately to changes following the application of some intervention in patients with PAH. In view of the above, it is believed that the best questionnaires for assessing quality of life or symptom severity are specific questionnaires dedicated to a particular group of patients, even if generic questionnaires are used, both tools should be used in the study. An important finding of this study is that the use of the PAH-SYMPACT specific questionnaire can support clinicians in the management and monitoring of patients undergoing treatment for pulmonary hypertension.

## Study Limitation

The study had limitations. One limitation was the difficulty in comparing Spearman correlation coefficients of the Polish version of the PAH-SYMPACT questionnaire with other validated versions of this tool in other languages, due to a lack of publications in this area, except for the English-language adaptation of the PAH-SYMPACT questionnaire. A one more limitation of this study is the sample size and single center study. However, it was designed to establish the suitability of the Polish PAH-SYMPACT rather than to describe in detail the impact of PH on patients.

## Practical Implications

The PAH-SYMPACT(PL) is a simple research tool that can be used in clinical practice or in research to evaluate the quality of life of the PAH population. The Polish version of the PAH-SYMPACT(PL) questionnaire can be used to assess quality of life, symptom severity and outcome, and to evaluate the effectiveness of treatment, thus providing valuable information on patient needs and clinical interventions. Use of the PAH-SYMPACT questionnaire is recommended for use in clinical practice and a

single-point measurement in cross-sectional studies. It may be particularly helpful in the planning activities of multidisciplinary teams.

# Conclusion

The cultural adaptation of the Polish version of the PAH-SYMPACT questionnaire demonstrated that it provides a new accurate and reliable instrument for assessing both HRQL and QoL in Polish patients with PAH in clinical practice. The results obtained from the cultural adaptation process of the Polish version of the PAH-SYMPACT questionnaire demonstrated that it is a consistent and reliable tool and the obtained correlation coefficients are largely consistent with the English language validation of the PAH-SYMPACT questionnaire. The PAH-SYMPACT is a simple research tool which can be used in standardised daily clinical practice to assess the quality of life of patients with PAH. The evaluation of QoL will allow care to be optimised and will support tailored educational and treatment iterventions. Moreover, using this instrument in everyday practice may improve patients' quality of life.

# Abbreviations

PH: Pulmonary Hypertension

PAH: Pulmonary Arterial Hypertension

PAH-SYMPACT: Pulmonary Arterial Hypertension-Symptoms and Impact

6MWT: 6 minuts walk test

CAMPHOR: The Cambridge Pulmonary Hypertension Outcome Review

WHO: World Health Organization

TAPSE: Tricuspid annular plane systolic excursion

NT-proBNP: Ventricular natriuretic peptide

PDE5: Phosphodiesterase type 5

QoL: quality of life

HRQoL: Health related quality of life

# Declarations

**Ethics approval and consent to participate:** The study was approved by the Bioethics Committee of the Wroclaw Medical University, Poland (approval number 538/2019). All study participants signed an

informed consent to participate in the study and were informed about the course, purpose and duration of the study.

**Consent for publication**: Not applicable.

**Availability of data and materials:** The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

**Competing interests:** The authors declare that they have no competing interests.

**Funding**: No special funding.

**Authors' contributions:** KS and BJP wrote the main manuscript text. JP prepared Tables and Figure. EM developed methods and materials section. GK described the validation process. All authors reviewed the manuscript. All authors have read and approved the manuscript.

**Acknowledgements:** Not applicable.

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# Figures

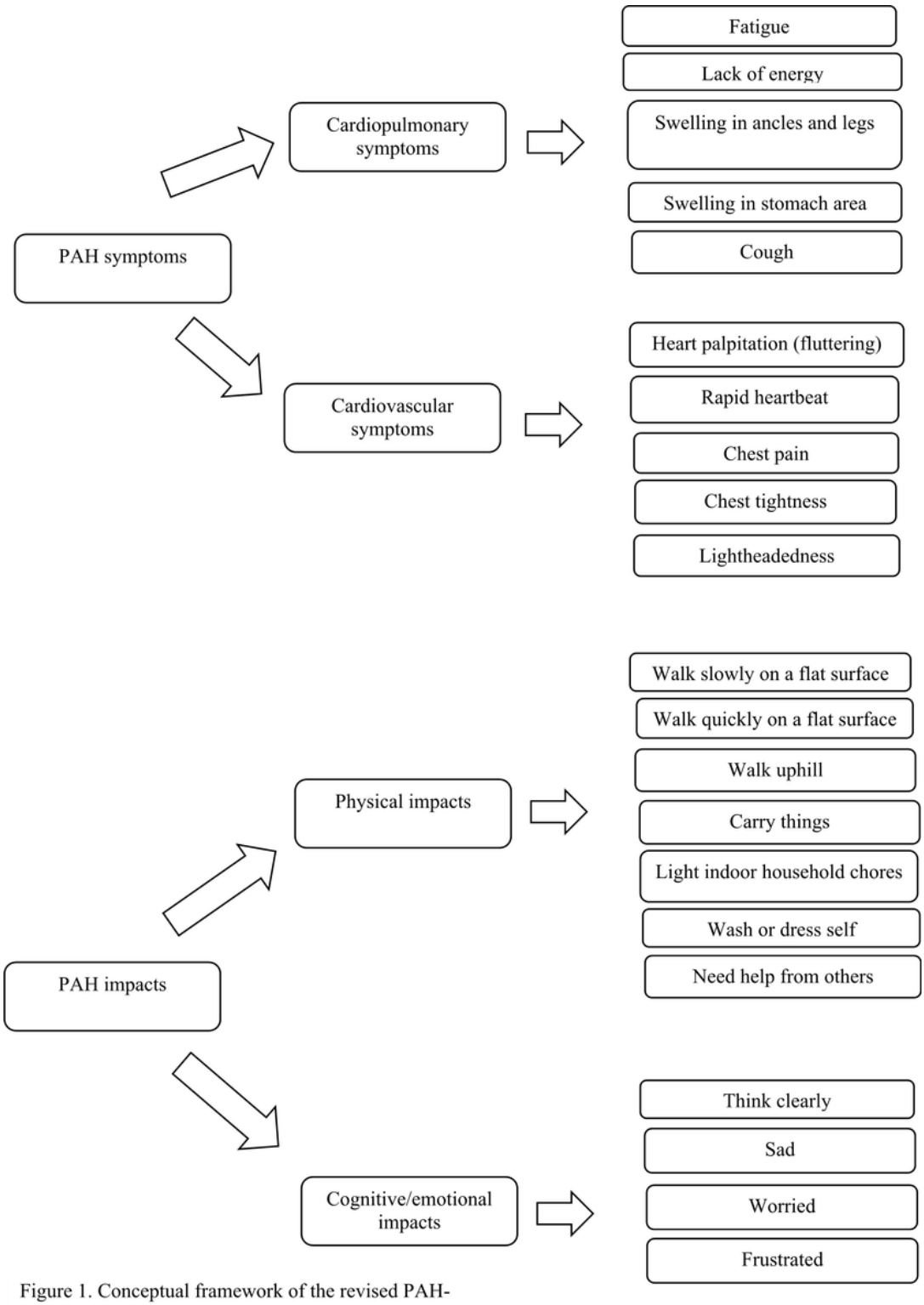


Figure 1. Conceptual framework of the revised PAH-

## Figure 1

Conceptual framework of the revised PAH-SYMPACT [7].