

EmPHasis-10 Health-Related Quality of Life and Exercise Capacity in Chronic Thromboembolic Pulmonary Hypertension After Balloon Angioplasty: A Cross-Sectional Study

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Abstract

Background

Whether pulmonary haemodynamic parameters and functional capacity are associated with quality of life (QoL) in patients with chronic thromboembolic pulmonary hypertension (CTEPH) remains unknown. This study aimed to evaluate disease-specific QoL with the emPHasis-10 questionnaire and assess its determinants in CTEPH patients with normalised pulmonary haemodynamics.

Methods

This cross-sectional study included 143 patients with CTEPH (median age, 68 [58–75] years; men/women, 51/136; use of home oxygen therapy [HOT], 51 patients [27%]) after balloon pulmonary angioplasty (BPA) with normalised mean pulmonary artery pressure (PAP) <25 mmHg at rest. Right heart catheterisation was performed, followed by the 6-minute walk test (6MWD) and the emPHasis-10 questionnaire.

Results

The median PAP and pulmonary vascular resistance were 18 (15–21) mmHg and 2.2 (1.7–2.9) Wood units, respectively. The median emPHasis-10 score was 14 (range, 8–24), whereas the median 6MWD was 447 (385–517) m. Univariate linear regression analysis showed that the emPHasis-10 score was associated with 6MWD ($\beta = -0.476$ [95% CI -0.604, -0.348], $p < 0.001$) and HOT ($\beta = 0.214$ [95% CI 0.072, 0.356], $p = 0.003$), but not with haemodynamic parameters. Multiple regression analysis revealed that a higher emPHasis-10 score was associated with lower 6MWD ($\beta = -0.475$ [95% CI -0.631, -0.319], $p < 0.001$).

Conclusion

Health-related QoL was associated with exercise capacity and the use of HOT, but not haemodynamic parameters, in patients with CTEPH after BPA with normalised haemodynamics. Improvements in exercise capacity may lead to further improvements in QoL.

Introduction

Chronic thromboembolic pulmonary hypertension (CTEPH) is an important cause of pulmonary hypertension (PH) and is associated with significant morbidity and mortality. Its main clinical manifestations include exertional dyspnoea, accompanied by a marked reduction in exercise capacity and quality of life (QoL).^{1–4} While pulmonary endarterectomy (PEA) has traditionally served as the primary treatment for patients with CTEPH, therapeutic options have expanded with the development of

balloon pulmonary angioplasty (BPA), leading to an improvement in prognosis.⁵⁻⁷ Recently, therapeutic decision-making for patients with CTEPH have considered not only disease-specific outcomes (e.g., improvement in prognosis and relief of PH) but also patient-centred outcomes (e.g., improvement in QoL and alleviation of symptom burden).⁸ In the 6th World Symposium on PH,⁹ disease-specific measures of health-related QoL were highlighted as relevant and important endpoints in clinical trials. These measures should also be integrated into daily clinical practice for the treatment of CTEPH, as has been performed for other cardiovascular diseases such as heart failure.¹⁰

The emPHasis-10 questionnaire is a unidimensional disease-specific patient-reported outcome assessment of QoL in patients with PH,¹¹ which can be used in both clinical trials and routine care. It consists of 10 questions on the important components of PH, such as breathlessness, fatigue, lack of energy, social restrictions, and concerns regarding the effect of the disease on the patient's family and friends. EmPHasis-10 scores have been correlated with exercise capacity and haemodynamics in the PH patient population, including those with CTEPH.^{12, 13} A part of its utility in clinical care stems from its strong association with long-term prognosis. Despite the recent proposal to redefine PH by lowering the mean pulmonary artery pressure (PAP) threshold from 25 mmHg to 20 mmHg, it remains unknown whether pulmonary haemodynamic parameters and/or functional capacity are associated with QoL among CTEPH patients with a mean PAP of less than 25 mmHg. This represents an important knowledge gap, as these modifiable determinants may be potential targets for interventions aiming to improve QoL among patients with CTEPH treated with BPA.

We recently reported that impaired functional capacity was common in CTEPH patients with improved haemodynamics after BPA.¹⁴ Furthermore, we found that the 6-minute walk distance (6MWD), a widely used parameter of functional capacity in daily life, was mainly defined by peripheral factors, but not pulmonary haemodynamics at rest.¹⁵ In addition, some patients still struggled to wean from home oxygen (O₂) therapy (HOT), even after haemodynamic normalisation; this may have adversely affected QoL.

Methods

Study design and patient recruitment

This study aimed to evaluate disease-specific QoL with emPHasis-10 and assess its determinants in CTEPH patients with normalised pulmonary haemodynamics after BPA.

This retrospective study included patients with CTEPH at Kyorin University Hospital who underwent BPA at >6 months previously. The inclusion criteria were as follows: admission for routine follow-up at our hospital between June 2018 and August 2020 and PAP <25 mmHg by right heart catheterisation (RHC). The 6-minute walk test (6MWT), RHC, and emPHasis-10 QoL assessment were performed at least 6 months after the patients' BPA session. This study was approved by the Committee for Clinical Studies and Ethics of Kyorin University School of Medicine.

Rhc

RHC was performed using a 6-F double-lumen balloon-tipped flow-directed Swan-Ganz catheter (Harmac Medical Products, Inc., Buffalo, NY, USA) via the transjugular approach. Baseline haemodynamic data were recorded; the zero-reference level (mid-chest) was adjusted at the start of pressure measurement, and the pulmonary artery wedge pressure (PAWP) was obtained as the mean value of the arterial trace during occlusion. Measurements were obtained at the end of a normal expiration, with the patients in the supine position in the resting state, to assess the right chamber, right atrial pressure, PAP (mean PAP, systolic PAP, and diastolic PAP), and PAWP. O₂ saturation in arterial blood (SaO₂) in the radial or femoral artery and O₂ saturation in the pulmonary artery (SvO₂) were measured. Cardiac output (CO) was determined by the Fick method using the following formula: $CO \text{ (L/min)} = VO_2 / 1.34 \text{ haemoglobin} \times (SaO_2 - SvO_2)$. Pulmonary vascular resistance (PVR) (Wood units) was determined via $(\text{mean PAP} - \text{PAWP}) / \text{CO}$.

6mwt

The 6MWT was performed according to the guidelines published by the American Thoracic Society and the European Respiratory Society.¹⁶ Participants were instructed to walk as fast and as far as possible for 6 min and to take as many breaks as possible for 6 min without O₂ supplementation. Each participant went through the 6MWT in a quiet hospital corridor with a 20-m marked track; chairs were used instead of cones to support turning if needed. The total distance walked was recorded as the 6MWD (to the nearest m). To ensure adequate risk management, participants were constantly monitored with an electrocardiogram and peripheral O₂ saturation (SpO₂) was assessed by a finger pulse oximeter.

Assessment of QoL

The self-administered emPHasis-10 questionnaire was distributed to the patients after the 6MWT. Each patient completed the questionnaire after any queries they had were addressed.

The emPHasis-10 is a PH-specific health-related QoL questionnaire developed through joint research by the University of Manchester and the PH Association UK.¹¹ EmPHasis-10 consists of 10 items, each on a scale of 0–5; the total maximum score is 50, with a higher score indicating a poorer health status and QoL. Translation into several languages is currently underway, which will facilitate international comparisons of this QoL metric. The Japanese version was generated via translation, followed by back translation, and validated in a cohort that included patients with CTEPH. The final version was approved by the developers of the original English version and published in 2018.¹²

Statistical analysis

Data are presented as median (range) for continuous variables, while categorical variables are presented as absolute counts and percentages (%). The correlations between the emPHasis-10 score and functional

and haemodynamic parameters were determined using Spearman's rank correlation. Univariable linear regression analysis was used to explore the linear correlation between the emPHasis-10 score and functional variables (6MWD), sociodemographic variables (age, sex, and body mass index [BMI]), use of HOT, and haemodynamic parameters (mean PAP, PAWP, CO, and PVR). A multiple regression analysis was then performed to determine the standardised regression coefficients. A sub-analysis was conducted, in which patients with and without HOT were compared with the Mann–Whitney U test or chi-square test, as appropriate. Statistical significance was set at $p < 0.05$. Data were analysed using Easy R version 1.41.

Results

Patient characteristics

Patient demographics are presented in Table 1. A total of 187 health status assessments were performed in 143 patients with CTEPH. The median age of the patients was 68 (range, 58–75) years, and 73% were women. HOT was used in 51 (27%) patients, and the time interval since the last BPA was 26 (11–48) months. Blood data and haemodynamic parameters were as follows: plasma brain natriuretic peptide, 17 (9–34) pg/dL; mean PAP, 18 (15–21) mmHg; PAWP, 8 (6–9) mmHg; CO, 4.4 (3.5–5.4) L/min; and PVR, 2.2 (1.7–2.9) Wood units. The 6MWD was 447 (385–517) m. Soluble guanylate-cyclase stimulator and oral prostacyclin analogue were present in 43 (23%) and 23 (12%) patients, respectively. All patients were taking anticoagulants. The emPHasis-10 total score was 14 (8–24) points.

Table 1
Baseline characteristics of the study patients

	(<i>n</i> = 187)
Age, years	68 (58–75)
Sex (male/female), <i>n</i>	51/136
BMI, kg/m ²	24.1 (21.5–26.7)
BNP level, pg/dL	17.0 (9.0–34.0)
Hb level, g/dL	13.2 (12.5–14.1)
HOT, <i>n</i> , %	51 (27%)
Time interval from final BPA, months	26 (11–48)
Medical treatment	
SGCSs, <i>n</i> , %	43 (23%)
Prostacyclin analogue, <i>n</i> , %	23 (12%)
ERA, <i>n</i> , %	19 (10%)
PDE5i, <i>n</i> , %	16 (9%)
Haemodynamics	
Mean RAP, mmHg	3 (3–5)
Systolic PAP, mmHg	31 (26–35)
Diastolic PAP, mmHg	8 (6–11)
Mean PAP, mmHg	18 (15–21)
PAWP, mmHg	8 (6–9)
SaO ₂ , %	94 (93–96)
SvO ₂ , %	71 (68–73)
CO, L/min	4.4 (3.5–5.4)

Values are presented as median (interquartile range).

6 MWD, 6-minute walk distance; BMI, body mass index; BNP, B-type natriuretic peptide; BPA, balloon pulmonary angioplasty; CO, cardiac output; ERA, endothelin receptor antagonist; Hb, haemoglobin; HOT, home oxygen therapy; PAP, pulmonary artery pressure; PAWP, pulmonary artery wedge pressure; PDE5i, phosphodiesterase type V inhibitor; PVR, pulmonary vascular resistance; RAP, right atrial pressure; SaO₂, arterial oxygen saturation; SGCS, soluble guanylate cyclase stimulator; SvO₂, mixed venous oxygen saturation.

	(n=187)
PVR, Wood unit	2.2 (1.7–2.9)
6MWD, m	447 (385–517)
emPHasis-10, score	14 (8–24)
Values are presented as median (interquartile range).	
6 MWD, 6-minute walk distance; BMI, body mass index; BNP, B-type natriuretic peptide; BPA, balloon pulmonary angioplasty; CO, cardiac output; ERA, endothelin receptor antagonist; Hb, haemoglobin; HOT, home oxygen therapy; PAP, pulmonary artery pressure; PAWP, pulmonary artery wedge pressure; PDE5i, phosphodiesterase type V inhibitor; PVR, pulmonary vascular resistance; RAP, right atrial pressure; SaO ₂ , arterial oxygen saturation; SGCS, soluble guanylate cyclase stimulator; SvO ₂ , mixed venous oxygen saturation.	

The distribution of the emPHasis-10 sub-items is shown in Figure 1. The score for each item was as follows: item 1 (I am not/very frustrated by my breathlessness), 1 (0–3) point; item 2 (Being breathless never/always interrupts my conversations), 1 (0–2) point; item 3 (I do not/always need to rest during the day), 1 (0–2) point; item 4 (I do not/always feel exhausted), 2 (1–3) point; item 5 (I have no/lots energy at all), 2 (1–3) point; item 6 (When I walk up one flight of stairs I am not/very breathless), 2 (0–3) point; item 7 (I am/not confident out in public places/ crowds despite my PH), 1 (0–2) point; item 8 (PH does not/completely control my life), 2 (1–3) point; item 9 (I am independent/completely dependent), 1 (0–2) point; and item 10 (I never/always feel like a burden), 1 (0–3) point.

Relationship between emPHasis-10 total score and functional/haemodynamic parameters

A significant moderate correlation was found between the emPHasis-10 total score and the 6MWD ($\rho = -0.454$, $p < 0.001$). However, no correlations were observed between the emPHasis-10 total score and mean PAP ($\rho = 0.007$, $p = 0.929$) or PVR ($\rho = 0.080$, $p = 0.275$) (Figure 2).

Factors influencing the emPHasis-10 total score

The items that showed a significant association with the emPHasis-10 total score in the univariate linear regression analysis were age ($\beta = 0.165$ [95% confidence interval (CI) 0.022, 0.308], $p = 0.024$), use of HOT ($\beta = 0.214$ [95% CI 0.072, 0.356], $p = 0.003$), and 6MWD ($\beta = -0.476$ [95% CI -0.604, -0.348], $p < 0.001$). Multiple linear regression analysis showed a significant association between the emPHasis-10 total score and 6MWD, with a standardised partial regression coefficient of $\beta = -0.475$ (95% CI -0.631, -0.319, $p < 0.001$) (Table 2).

Table 2
Association of haemodynamic variables and physical function with emPHasis-10

	Univariate		Multivariate ($R^2 = 0.21; p < 0.001$)	
	Coefficient (95% CI)	p-value	Coefficient (95% CI)	p-value
Age	0.165 (0.022, 0.308)	0.024	0.010 (-0.141, 0.162)	0.892
Female	0.098 (-0.046, 0.243)	0.181	-0.019 (-0.161, 0.124)	0.796
BMI	0.08 (-0.064, 0.226)	0.271	-0.004 (-0.182, 0.174)	0.962
Use of HOT	0.214 (0.072, 0.356)	0.003	0.088 (-0.056, 0.231)	0.230
Mean PAP	0.024 (-0.121, 0.169)	0.745	-0.118 (-0.281, 0.046)	0.156
PAWP	-0.044 (-0.189, 0.101)	0.548	0.037 (-0.125, 0.198)	0.653
CO	0.054 (-0.091, 0.199)	0.460	0.062 (-0.117, 0.241)	0.494
6MWD	-0.476 (-0.604, -0.348)	<0.001	-0.475 (-0.631, -0.319)	<0.001

6MWD, 6-minute walk distance; BMI, body mass index; CI, confidence interval; CO, cardiac output; HOT, home oxygen therapy; PAP, pulmonary artery pressure; PAWP, pulmonary artery wedge pressure.

Comparison between patients with and without HOT

The baseline characteristics of patients with HOT ($n = 51$) and without HOT ($n = 136$) are shown in Table 3. Patients with HOT had a significantly higher mean PAP and PVR than those without HOT. EmPHasis-10 total scores were significantly higher in patients with HOT than in those without HOT (18 [9–28] vs. 12 [7–21], $p = 0.005$). The following sub-items were significantly higher in patients with HOT: item 1, 2 (1–3) vs. 1 (0–3) ($p = 0.007$); item 5, 3 (2–3) vs. 2 (1–3) ($p = 0.012$); item 6, 3 (1–4) vs. 1 (0–3) ($p < 0.001$); item 8, 3 (2–3) vs. 2 (1–3) ($p = 0.012$); item 9, 1 (0–3) vs. 1 (0–2) ($p = 0.049$); and item 10, 2 (0–3) vs. 1 (0–2) ($p = 0.041$) (Figure 3). Among the patients with HOT, 6MWD was also significantly associated with emPHasis-10 scores ($\rho = -0.362, p = 0.020$).

Table 3
Comparison between patients with and without home oxygen therapy

	Without HOT (n = 136)	With HOT (n = 51)	p-value
Age, years	67 (55–75)	70 (65–77)	<0.001
Sex (male/female), n	40/96	11/40	0.357
BMI, kg/m ²	24.1 (21.7–26.5)	24.4 (21.5–27.9)	0.761
BNP level, pg/dL	16.0 (8.0–26.0)	27.0 (11.3–51.3)	0.004
Time interval from final BPA, months	27 (12–48)	24 (11–46)	0.563
Haemodynamics			
Mean PAP, mmHg	17 (14–20)	20 (17–22)	<0.001
PAWP, mmHg	7 (6–9)	8 (6–9)	0.434
SaO ₂ , %	95 (93–96)	94 (92–95)	0.066
SvO ₂ , %	71 (68–74)	70 (68–73)	0.109
CO, L/min	4.4 (3.5–5.3)	4.4 (3.5–5.4)	0.936
PVR, Wood unit	2.2 (1.6–2.8)	2.5 (2.0–3.5)	0.007
6MWD, m	467 (396–545)	400 (340–445)	<0.001
emPHasis-10, score	12 (7–21)	18 (9–28)	0.005
Values are presented as median (interquartile range).			
6MWD, 6-minute walk distance; BMI, body mass index; BNP, B-type natriuretic peptide; BPA, balloon pulmonary angioplasty; CO, cardiac output; HOT, home oxygen therapy; PAP, pulmonary artery pressure; PAWP, pulmonary artery wedge pressure; PVR, pulmonary vascular resistance; SaO ₂ , arterial oxygen saturation; SvO ₂ , mixed venous oxygen saturation.			

Discussion

This study showed that QoL, as measured with the disease-specific scale emPHasis-10, remained impaired in CTEPH patients with normalised haemodynamics after BPA. While the QoL score was associated with 6MWD, it was not associated with haemodynamic parameters. The administration of HOT was also associated with impaired QoL. Among patients with HOT, the QoL score was associated with 6MWD.

EmPHasis-10 in patients with PH

A previous validation study of emPHasis-10, which included patients with CTEPH, was

conducted by Takeyasu et al.¹² in Japan. The emPHasis-10 total score was 19.4 ± 10.6 points; scores were increased with higher World Health Organisation (WHO) functional classes (I, 5.0 ; II, 17.0 ± 1.4 ; III, 24.1 ± 2.2 ; and IV, 39.5 ± 2.5). These results were consistent with those of the original emPHasis-10 report by Yorke et al.,¹¹ which was based on patients in the United Kingdom and Ireland. The median emPHasis-10 total score was 14 (8–24) points, which corresponded to the midpoint between WHO functional classes I and II.^{11,12} This implied that QoL was not completely equivalent to WHO functional class I, despite haemodynamic improvement.

Recently, Lewis et al. reported that emPHasis-10 was an independent prognostic marker in patients with either idiopathic pulmonary artery hypertension or connective tissue disease-associated pulmonary artery hypertension, thus emphasising the importance of emPHasis-10 assessment.¹⁷ There are limited reports on emPHasis-10 in patients with CTEPH.¹⁸ While the majority can be treated by surgical and/or catheter interventions that do not apply to other patients with PH, the clinical progression and prognosis of these patients vary. Thus, further studies are needed to clarify the prognostic utility of emPHasis-10 in patients with CTEPH.

Determinants of QoL in patients with CTEPH

Studies using generic instruments have reported impaired QoL in CTEPH patients with a mean PAP >25 mmHg after treatment.⁴ Halank et al.¹⁹ used the SF-36 questionnaire to assess QoL in patients with severe CTEPH and concluded that mental disorders, exercise capacity, long-term HQT, right heart failure, and age played more important roles in the impairment of QoL than haemodynamic parameters at rest. Kamenskaya et al.²⁰ also used the SF-36 questionnaire and reported impaired QoL even after 1 year of PEA. Urushibara et al.²¹ found that QoL was also impaired after PEA and medical treatment; physical function items were especially associated with PVR and 6MWD. Using the EuroQol-5 dimensions questionnaire, Minatsuki et al.²² observed impaired QoL after BPA and medical treatment; they also reported a significant correlation with mean PAP and 6MWD.

Generic QoL assessment scales may be unable to assess specific conditions inherent to patients with PH. Therefore, it is necessary to use disease-specific QoL scales that assess factors directly affected by disease pathology and symptoms.²³ Several observational cohort studies have demonstrated that haemodynamics and exercise capacity are related to disease-specific QoL in patients with moderate to severe PH. In the original report, which included patients with varying types and severity of PH, emPHasis-10 was shown to have a moderate correlation with 6MWD.¹¹ Among patients with severe PH, including those with CTEPH, the emPHasis-10 score was moderately correlated with both 6MWD and mean PAP.¹³ Lewis et al.¹⁷ reported that the emPHasis-10 score was modestly correlated with 6MWD and weakly correlated with pulmonary haemodynamics in a large cohort with severe PAH of various aetiologies. To our knowledge, the present study is the first to evaluate disease-specific QoL and its determinants among CTEPH patients with a mean PAP <25 mmHg. We demonstrated that the emPHasis-10 score was related

to the 6MWD, but not to haemodynamic parameters. These findings suggest that exercise capacity plays a more important role in QoL among CTEPH patients with normalised haemodynamics. Furthermore, emPHasis-10 may be a convenient alternative to the 6MWD as an assessment of exercise capacity in this patient group.

Improvement in QoL

Exercise training has been reported to improve exercise capacity and QoL in patients with PH. Randomised controlled trials^{24–26} evaluating the efficacy of individually tailored interventions, including exercise therapy and respiratory training, in patients with PH (who were on stable medications) have reported improved exercise capacity, health-related QoL, and cardiopulmonary parameters. A study that prospectively recruited 35 consecutive patients with confirmed invasive and inoperable CTEPH or residual CTEPH²⁷ found that exercise training was associated with improvements in exercise capacity and QoL. Notably, in a recent prospective cohort study of patients with CTEPH after BPA,²⁸ exercise training improved QoL and exercise capacity. Based on our data, it is plausible that exercise training can contribute to an improvement in QoL in CTEPH patients with more normalised haemodynamics; this needs to be evaluated in further studies.

QoL in patients with HOT

The use of HOT was associated with impaired QoL. The following emPHasis-10 sub-items were found to be significantly lower among patients treated with HOT: frustration due to shortness of breath, vitality, shortness of breath on stairs, lack of control over life events due to PH, independent living, and burden on family and friends. This was unexpected, as HOT prevents the progression of PH and improves hypoxemia.^{29,30} However, it is consistent with previous studies reporting that HOT decreases QoL.^{19,31} Possible factors related to impaired QoL may have included the patients' perceived restrictions on their daily activities, embarrassment about using O₂ with a nasal cannula in public, distress at losing their independence, and worry due to the possibility that their O₂ supply would be completely depleted if they were to leave their house.³² We need to be cautious about the unnecessary use of HOT in CTEPH patients with haemodynamic improvement. The use of HOT was associated with impaired haemodynamics, even among patients with a mean PAP <25 mmHg. This highlights the need for further study on whether additional treatment with BPA and/or medication adjustments could facilitate the discontinuation of HOT and lead to further improvement in QoL and physical function.

Study limitations

This study has some limitations. Our study adopted a cross-sectional observational design without a control group. While QoL was assessed, we were unable to verify the patients' psychological status, housing circumstances, family support, or other social aspects of their daily lives. The potential influence of these factors should be considered in future studies.

Conclusions

In patients with CTEPH who underwent BPA with haemodynamic normalisation, disease-specific QoL was suboptimal and related to exercise capacity and the use of HOT, but not pulmonary haemodynamics. Further studies are needed to determine whether improvements in exercise capacity could lead to further improvements in QoL.

Abbreviations

6MWD: 6-minute walk test

6MWT: 6-minute walk test

BPA: balloon pulmonary angioplasty

BMI: body mass index

CO: cardiac output

CTEPH: chronic thromboembolic pulmonary hypertension

HOT: home oxygen therapy

SpO₂: peripheral O₂ saturation

PAP: pulmonary artery pressure

PAWP: pulmonary artery wedge pressure

PEA: pulmonary endarterectomy

PH: pulmonary hypertension

PVR: pulmonary vascular resistance

QoL: quality of life

SaO₂: O₂ saturation in arterial blood

SvO₂: O₂ saturation in the pulmonary artery

Declarations

Ethics approval and consent to participate:

This study was approved by the Committee for Clinical Studies and Ethics of Kyorin University School of Medicine. Due to the observational nature of the study and the provision of usual care, written informed consent was waived.

Consent for publication:

Not applicable.

Availability of data and materials:

The datasets generated during 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 conflicts of interest.

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There was no specific funding received for this study.

Authors' contributions:

Dr. Goda, PT Sakamoto, and PT Tobita designed the study, analysed the data, and wrote the manuscript. Dr. Kohno, Dr. Yamada, Dr. Satoh, and Dr. Soejima reviewed the data and edited the manuscript. Dr. Takeuchi, Dr. Kikuchi, Dr. Goda, and Dr. Inami performed the RHC.

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Figures

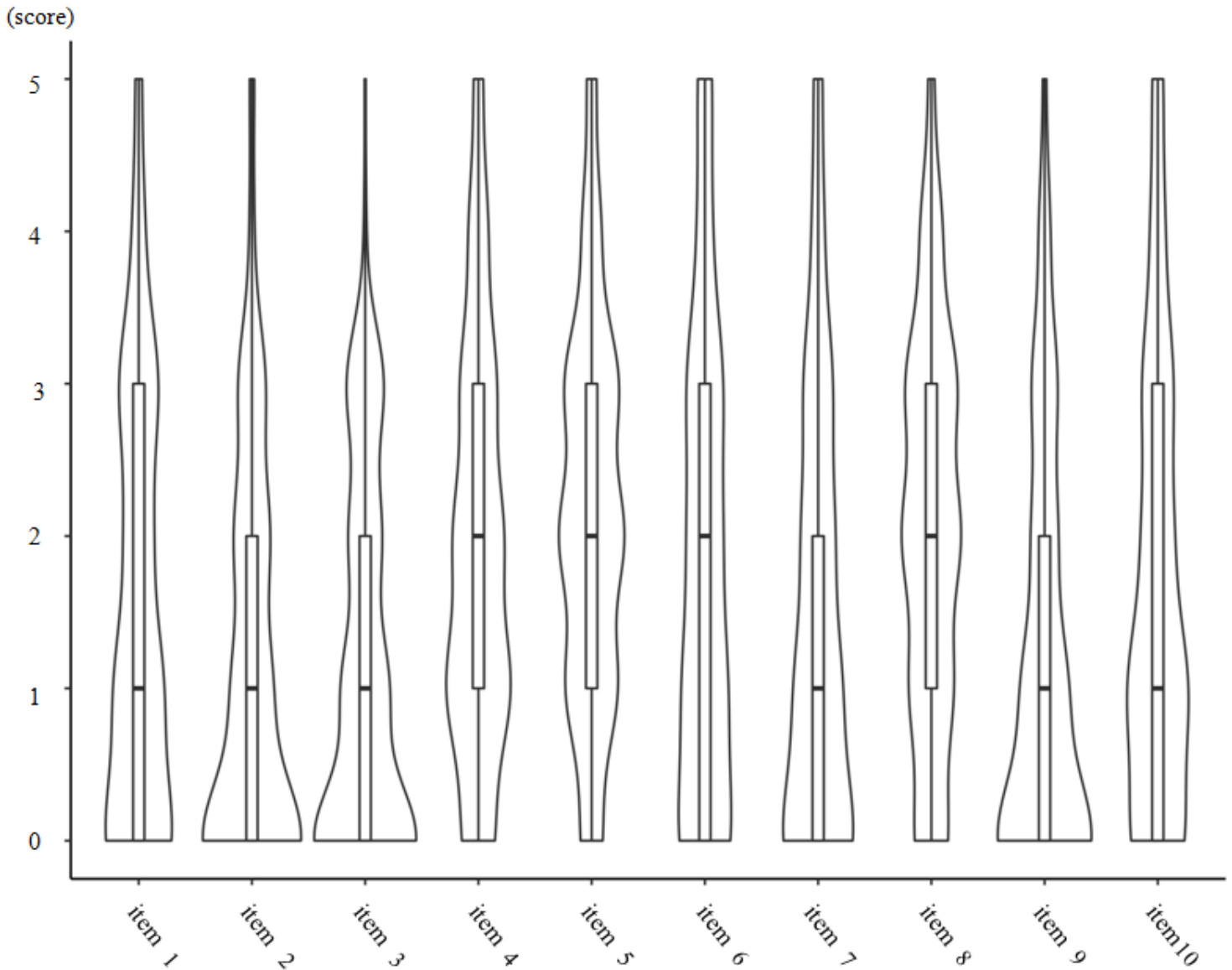


Figure 1

Distributions of sub-items in emPHasis-10. Item 1 (I am not/very frustrated by my breathlessness); item 2 (Being breathless never/always interrupts my conversations); item 3 (I do not/always need to rest during the day); item 4 (I do not/always feel exhausted); item 5 (I have no/lots energy at all); item 6 (When I walk up one flight of stairs I am not/very breathless); item 7 (I am/not confident out in public places/ crowds

despite my pulmonary hypertension); item 8 (Pulmonary hypertension does not/completely control my life); item 9 (I am independent/completely dependent); item 10 (I never/always feel like a burden)

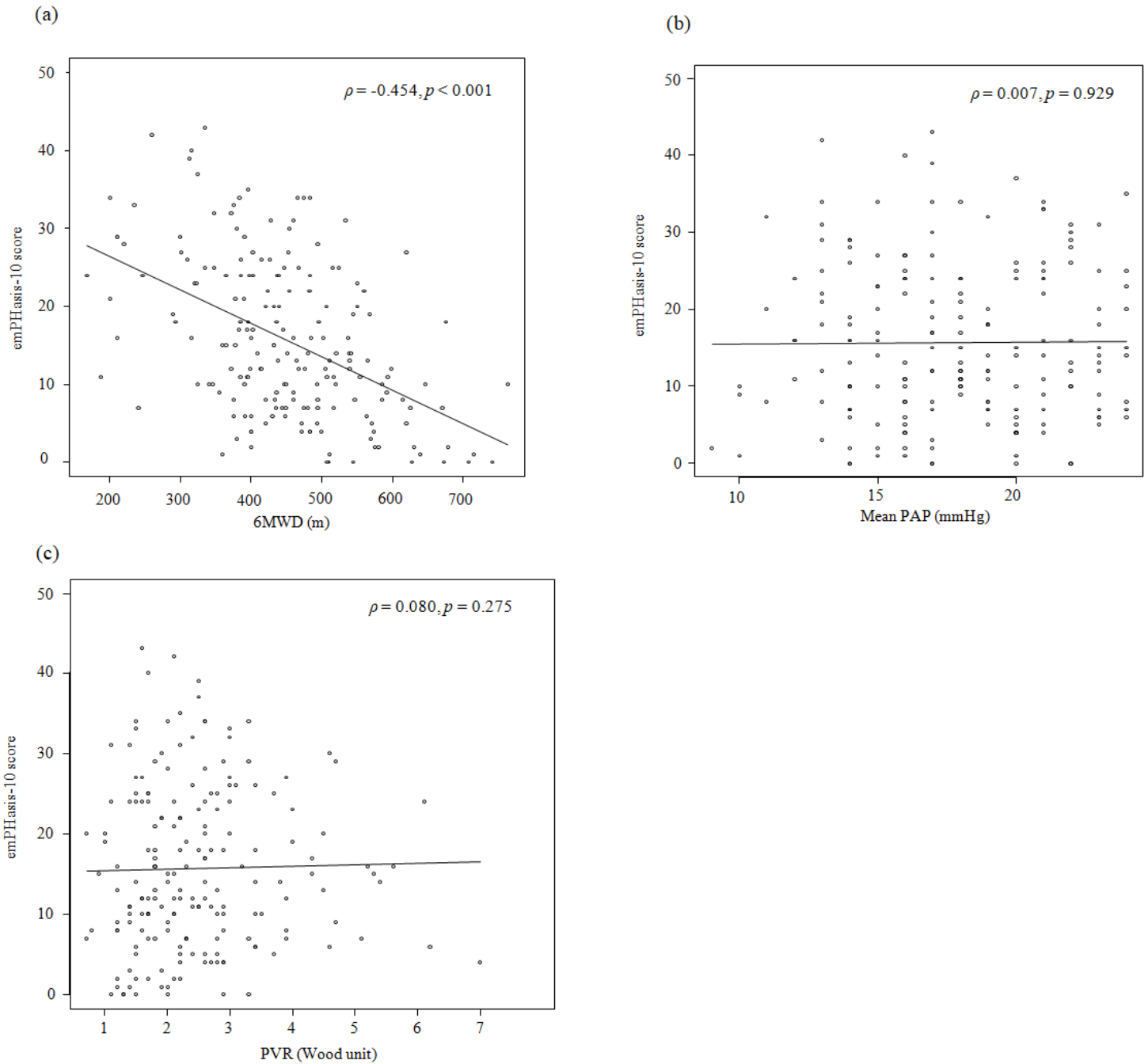


Figure 2

Correlations of the emPHasis-10 score with 6MWD (a), PAP (b), and PVR (c). PAP, pulmonary artery pressure; PVR, pulmonary vascular resistance; 6MWD, 6-minute walk distance

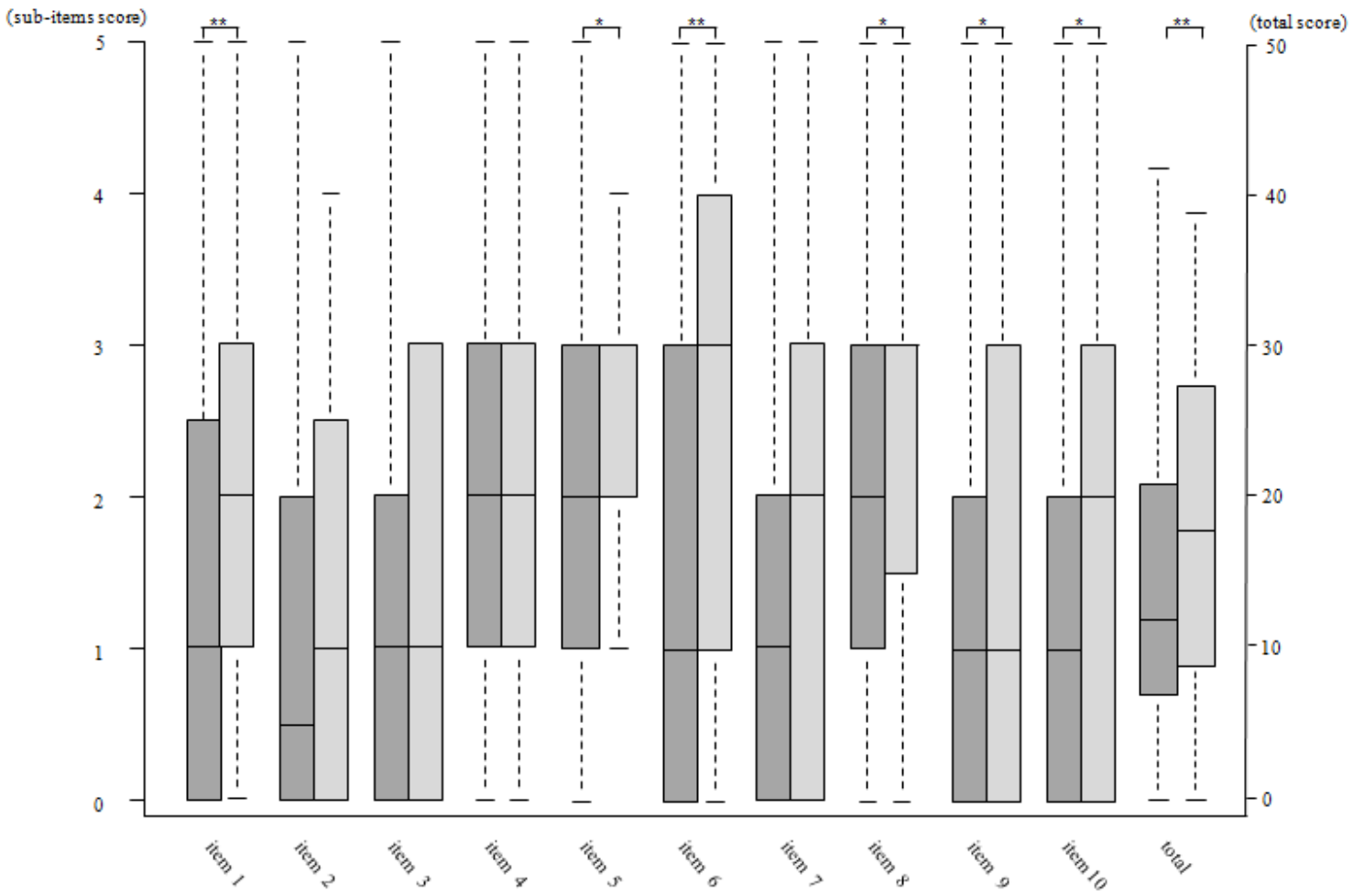


Figure 3

Comparisons between patients with and without home oxygen therapy in the sub-items and total score of the emPHasis-10. Home oxygen therapy is indicated by light grey bars. The absence of home oxygen therapy is indicated by dark grey bars. * $p < 0.05$ and ** $p < 0.01$.