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RESEARCH ARTICLE

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Change in health-related quality of life at early follow-up in patients with pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension

Bodil Ivarsson[1] | **Anders Johansson**[1] | **Barbro Kjellström**[2,3]

[1]Clinical Sciences, Lund University, Lund, Sweden

[2]Department of Medicine, Karolinska Institute, Stockholm, Sweden

[3]Department of Clinical Physiology, Clinical Sciences, Lund University and Skåne University Hospital, Lund, Sweden

Correspondence
Barbro Kjellström, Department of Clinical Physiology, Clinical Sciences, Lund University and Skåne University Hospital. Skåne University Hospital, Lund SE-222 42, Sweden.
Email: barbro.kjellstrom@med.lu.se

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Abstract

Symptoms associated with pulmonary arterial hypertension (PAH) or chronic thromboembolic pulmonary hypertension (CTEPH) impact patient's health-related quality of life (HRQoL). Studies on change and if a minimal clinically important difference (MCID) in HRQoL is reached within a year after diagnosis are lacking. The aim was to investigate the change in HRQoL as well as the proportion of patients that reached MCID at an early postdiagnosis visit. The study included adult patients from the Swedish PAH & CTEPH registry, diagnosed 2008–2021, with Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR) at time of diagnosis and a follow-up. Data were analyzed as total population and dichotomized for sex, age (<65 vs. ≥65 years), time of diagnosis (≤2015 vs. >2015) and pulmonary hypertension (PH) subgroups. Data reported as median, interquartile range (IQR), and proportions (%). There were 151 patients (PAH = 119, CTEPH = 32) with an available CAMPHOR score at diagnosis and follow-up. CAMPHOR total sum was 31 (IQR: 21–43) and 25 (14–36); ($p < 0.001$) at diagnosis and follow-up, respectively. At follow-up, 56% had reached MCID in total sum, while for domains activity, symptoms, and QoL 27%, 33%, and 39% reached MCID, respectively. These results were independent of PH subgroup, diagnosis before or after 2015 and sex. Age below 65 years was related to improvements in activity and worsening of symptoms. In conclusion on a group level, improvements in CAMPHOR total sum as well as all domains were seen in the first year after diagnosis, however, only slightly more than half of the patients reached MCID for CAMPHOR total sum.

KEYWORDS
chronic illness, disease management, newly diagnosed, patient-reported outcome measures

INTRODUCTION

In pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH) the small pulmonary arteries become narrow and nonelastic, leading to increased right ventricular afterload and right heart failure.[1] Symptoms such as dyspnea and fatigue are common at all stages of disease progress while syncope and chest pain are associated with more severe disease.[2] These symptoms affect social activities, family life, and the ability to work.[3–5] Diagnostic delays are common[6,7] and increased healthcare consumption has been shown to start up to 5 years before diagnosis for both PAH and CTEPH.[8,9]

Patients' well-being is central in all health care and the use of health-related quality of life (HRQoL) instruments in clinical practice is important. Asking the patient about health, symptoms, quality of life (QoL), and functional status opens the possibility to understand and improve their well-being.[10] Both generic and disease-specific HRQoL instruments are used by patients with PAH and CTEPH.[3] HRQoL instruments have a history of primarily being considered a research tool and used in clinical trials.[11] However, the utility in clinical practice is growing, and in demand, but still, follow-up studies are rare.[12–14] A commonly used HRQoL instrument in PAH and CTEPH is the Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR).[15] Recently, the relevant minimal clinically important difference (MCID) of CAMPHOR has been determined.[14,16]

Therefore, the aim of this study was to investigate the change in CAMPHOR, as well as the proportion patients that reached the MCID, between diagnosis and a postdiagnosis visit in a national cohort of patients with PAH and CTEPH.

METHODS

Study population

All adult patients (age ≥ 18 years) with a diagnosis of PAH or CTEPH registered in the Swedish PAH & CTEPH registry (SPAHR) 2008–2021 were considered for the analyses. To be included in the study, the patient should have completed a CAMPHOR questionnaire at time of diagnosis and at a postdiagnosis visit (follow-up) with a minimum of 3 months and a maximum of 15 months apart.

SPAHR constitutes an open continuous registry of patients diagnosed with PAH or CTEPH.[17] All Swedish PAH/CTEPH specialist clinics (PH center) participate in SPAHR and the national coverage of patients diagnosed with PAH or CTEPH in the registry is >90%. SPAHR is approved by the National Board of Health and Welfare and by the Swedish Data Protection Authority. All patients were informed about their participation in SPAHR and had the right to decline.

Data collection

The CAMPHOR consists of three domains that measure QoL (25 questions) in parallel with symptoms (25 questions) as well as levels of physical activity (15 questions). Symptoms and QoL items are both scored as "yes/true" = 1 and "no/not true" = 0, possible maximal score is 25 for each item, physical activity has three possible responses (score 0–2), giving a possible maximal score of 30.[15] The total sum as well as the sum for each domain was used in the study. A higher sum indicates worsening and subsequently a decrease is considered an improvement. CAMPHOR has been translated and validated for use in Sweden.[18] MCID for CAMPHOR used in the present study were total sum 4 points, QoL 3 points, symptoms 4 points, and activity 4 points.[16]

CAMPHOR scores and demographic variables were obtained from SPAHR.

Data management

Demographic variables and CAMPHOR scores were analyzed with the statistical software SPSS®version 27.0 (SPSS Inc.). Data are presented for the entire population as well as for sex, age, pulmonary hypertension (PH) subgroups, and diagnosis before and after 2015. The nature of collected variables is regarded as qualitative perceptions and experiences and were accordingly analyzed with nonparametric tests (Mann–Whitney *U* test). Patient age were analyzed using *t*-test. Where appropriate, variables are reported as absolute- and relative frequencies and analyzed with χ^2 test. Unless otherwise indicated, the results are reported as the median and interquartile range (IQR: 1–3) or the mean ± standard deviation (SD), where appropriate. A p-value < 0.05 was considered statistically significant.

RESULTS

This study included 151 patients (PAH, $n = 119$; CTEPH, $n = 32$) with a mean age of 65 ± 14 years at the time of diagnosis and 64% were women (Table 1). The time from diagnosis to follow-up was 186 ± 78 days, with no difference between sexes (Table 1). PAH was more common among women while CTEPH was evenly distributed between the sexes (Table 2). At time of

TABLE 1 Patient characteristics shown for all and by sex and age groups.

	Subjects *n* (%)	*p* Value	Age (years) mean ± SD	*p* Value
All	151		65 ± 14	
Men	54 (36)	0.001[a]	64 ± 14	0.608[b]
Women	97 (64)		66 ± 15	
<65 years	54 (36)	0.001[a]	50 ± 12	0.001[b]
≥65 years	97 (64)		74 ± 5	

Note: Values presented as absolute and relative (%) frequencies or mean ± SD, as indicated.

[a]*p* Value based on χ^2 test.

[b]*p* Value based on parametric *t*-tests.

TABLE 2 Pulmonary hypertension diagnosis shown for all and by sex.

	Total	Men	Women
CTEPH	32 (22)	16 (50)	16 (50)
PAH	119 (78)	38 (32)	81 (68)
PAH subgroups			
IPAH/HPAH	52 (44)	20 (38)	32 (62)
APAH-CTD	56 (47)	11 (20)	45 (80)
APAH-CHD	3 (2)	2 (66)	1 (34)
APAH-Other	8 (7)	5 (63)	3 (37)

Note: Values presented as absolute and relative (%) frequencies.

Abbreviations: APAH, associated pulmonary arterial hypertension; CHD, congenital heart disease; CTD, connective tissue disease; CTEPH, chronic thromboembolic pulmonary hypertension; HPAH, hereditary pulmonary arterial hypertension; IPAH, idiopathic pulmonary arterial hypertension; PAH, pulmonary arterial hypertension.

TABLE 3 CAMPHOR scores and PH-specific treatment at diagnosis and follow-up.

	Diagnosis	Follow-up	*p* Value
CAMPHOR score			
Total sum	31 (21–43)	25 (14–36)	0.001[a]
Activity	10 (6–14)	8 (5–13)	0.001[a]
Symptoms	12 (9–16)	9 (6–14)	0.001[a]
Quality of life	8 (4–13)	6 (2–10)	0.001[a]
PH-specific treatment			
Mono	101 (67)	77 (51)	0.001[b]
Combination	43 (28)	68 (45)	
No treatment	7 (5)	6 (4)	
Pulmonary endarterectomy surgery	0	2 (1)	

Note: CAMPHOR score values presented as median and (1st–3rd quartile), PH-specific treatment presented as absolute and relative (%) frequencies.

Abbreviations: CAMPHOR, Cambridge Pulmonary Hypertension Outcome Review; PH, pulmonary hypertension.

[a]*p* Value based on Wilcoxon Signed Ranks test (CAMPHOR).

[b]χ^2 test (Medical treatment).

diagnosis, 95% received PH-specific treatment, whereof 67% monotherapy and 28% combination therapy; while at follow-up, 51% received monotherapy and 45% combination therapy, 4% remained untreated (Table 3). Two patients with CTEPH underwent pulmonary endarterectomy surgery (PEA) between diagnosis and follow-up.

Median CAMPHOR total sum improved from 31 (IQR: 21–43) points at diagnosis to 25 (14–36) points at follow-up (Table 3). Significant improvement was seen in all three domains: activity, symptoms, and QoL (Table 3).

From diagnosis to follow-up, 56% had reached MCID in total CAMPHOR sum, while for the domain's activity, symptoms, and QoL 27%, 33%, and 39% reached MCID, respectively (Table 4). There was no difference in neither total sum nor domain scores between the sexes, diagnosis year ≤2015 versus >2015, subgroups PAH versus CTEPH or IPAH/HPAH versus PAH-CTD (Table 4, Figures 1 and 2). A higher proportion of patients <65 years improved MCID for activity and worsened for symptom dimensions compared to patients ≥65 years (Figure 2).

DISCUSSION

Improvements in CAMPHOR total sum as well as in all three domains were seen in the first year after diagnosis. However, less than half of the patients reached MCID in CAMPHOR total sum and even lower proportions reached MCID for the activity, symptoms and QoL domains. These results were independent of subgroup diagnosis, diagnosis before or after 2015 and sex. Age below 65 years was related to improvements in activity and worsening of symptoms.

Both PAH and CTEPH are associated with symptoms of dyspnea and fatigue that might present years before diagnosis, and thus, many patients experience a long and

TABLE 4 Change in CAMPHOR scores from diagnosis to the follow-up visit.

	Improved	No change	Worsened	*p* Value
Total sum				
All	84 (56)	42 (28)	25 (16)	<0.001
PAH	66 (55)	33 (28)	20 (17)	0.987
CTEPH	18 (56)	9 (28)	5 (16)	
Activity				
All	40 (27)	100 (66)	11 (7)	<0.001
PAH	32 (27)	78 (66)	9 (8)	0.935
CTEPH	8 (25)	22 (69)	2 (6)	
Symptoms				
All	50 (33)	83 (55)	18 (12)	<0.001
PAH	40 (34)	65 (55)	14 (12)	0.967
CTEPH	10 (31)	18 (56)	4 (13)	
Quality of life				
All	59 (39)	71 (47)	21 (14)	<0.001
PAH	48 (40)	53 (45)	18 (15)	0.457
CTEPH	11 (34)	18 (56)	3 (9)	

Note: MCID were total sum = 4 points, activity = 4 points, symptoms = 4 points, and quality of life = 3 points.[16] An MCID decrease in score was considered improvement and an MCID increase in score was considered worsened. Values presented as absolute and relative (%) frequencies, $p < 0.05$. *p* Value based on χ^2 test.

Abbreviations: CAMPHOR, Cambridge Pulmonary Hypertension Outcome Review; CTEPH, chronic thromboembolic pulmonary hypertension; PAH, pulmonary arterial hypertension.

stressful path before being diagnosed.[8,9,19,20] The burden of symptoms and the long path to diagnose, together with information that the disease have a dire outcome despite lifelong treatment, will affect HRQoL not only at time of diagnosis, but also during the early phase after diagnosis.[2,6,20] The impairment in HRQoL experienced by patients diagnosed with PAH or CTEPH has been compared to patients with COPD or prostate cancer.[21]

Using a HRQoL instrument in the care of patients with PAH and CTEPH is strongly recommended in the 2022 European guidelines for diagnosis and treatment of patients with PH.[2] The recent presentation of an MCID for changes in CAMPHOR total sum and domains is an important step forward for the usability of CAMPHOR in clinical practice as well as in clinical studies.[14,16] In the present study, slightly more than half of the population had an improvement in CAMPHOR total sum from diagnosis to follow-up that met the MCID criteria. While approximately a third of the patients improved their symptoms and QoL, only a fourth improved activity, using the MCID criteria in presented by Bunclark et al.[16] These results were independent of subgroup diagnosis, diagnosis before or after 2015 and sex, while, as can be expected, a higher proportion of patients younger than 65 years at time of diagnosis improved activity at follow-up compared to older patients. Interestingly, while the same proportion improved symptoms in both age groups, in the younger age group, a higher proportion worsened symptoms, while for patients 65 or older, a majority had no change in the symptom domain.

At the follow-up visit, only half of the patients were started on combination treatment of PH-specific drugs, which can be regarded as undertreated according to guidelines.[2] One can speculate that if a higher proportion of patients had received combination treatment up front, more patients might have improved their HRQoL and reached the MCID thresholds even at an early follow-up. A reason for the somewhat high proportion of monotherapy even at follow-up might be related to a cautious treatment approach due to age and cardiopulmonary comorbidities as two thirds of the study population was 65 years or older. Thus, treatment side effect related symptoms might at least partly explain why a higher proportion of younger patients reported worsened symptoms while for older patients symptoms more often remained unchanged. Most important, the results in the present study confirm that patients with PAH and CTEPH continue to suffer from shortness of breath, fatigue, fear, anxiety, and activity intolerance despite improvements in PH-specific treatment and treatment strategies.[22,23]

In the present study, activity was the domain with the highest proportion of patients that did not reach an MCID threshold. The MCID threshold itself might have played a role. In a recent Dutch study by Hendriks et al.,[14] a third of the population reached MCID in all CAMPHOR domains, including activity. The discrepancy from the present study regarding the activity domain may be explained by the use of a MCID threshold for activity of three in the Dutch study,[14] whereas a threshold of four was used in the present study.[16] Interestingly, in both the British and Dutch studies a third of the patients reached the MCID for activity, though they were using different MCID thresholds.

It is very likely that the short follow-up time will also have contributed to the lack of MCID in CAMPHOR activity in the present study. It might take some time to regain the lost exercise tolerance and to trust that symptoms are less pronounced, and thus, 6 months might not be long enough to reach the MCID threshold. This highlights the importance to motivate all patients, independent of age and physical status, to be physically active. Involving a physiotherapist already at time of diagnosis will support patients to find a level of exercise applicable to their capability and need.[2] Physical training has previously been considered too risky and healthcare professionals may have

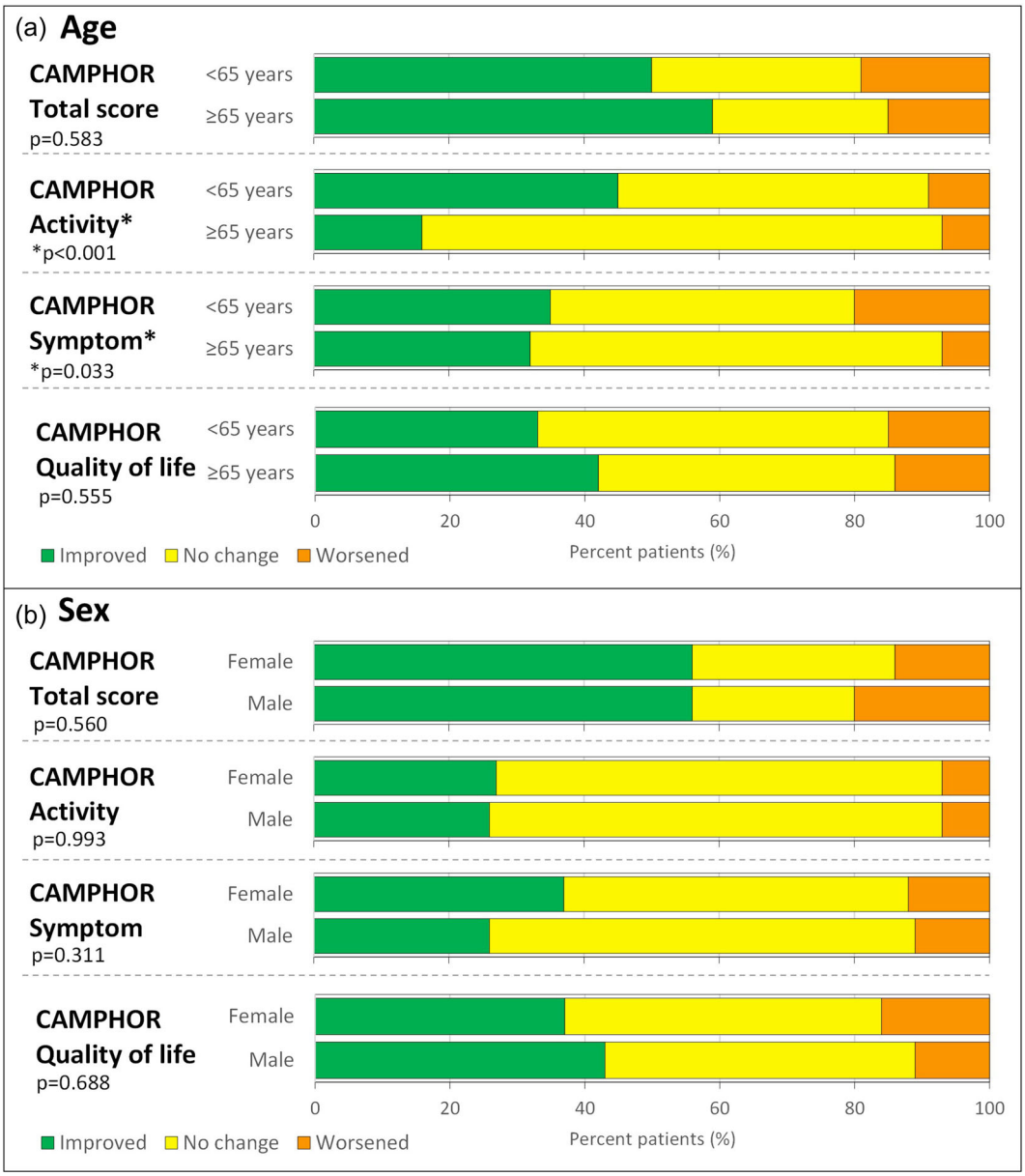

FIGURE 1 Relative change in Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR) scores from diagnosis to the follow-up visit shown by age (a) (<65 years = 54, ≥65 years = 97) and sex (b) (female = 97, male = 54). Minimal clinically important difference (MCID) were total sum = 4 points, activity = 4 points, symptoms = 4 points, and quality of life = 3 points.[16] An MCID decrease in score was considered improvement and an MCID increase in score was considered worsened.

focused more on medical treatment than physical activity. However, recent recommendations have shown that it is safe for patients with PAH and CTEPH, even those considered fragile, to participate in physical training.[24] In Sweden, most centers have a physiotherapist connected to the PH-team,[25] but the low proportion of patients that reached an MCID in the activity domain seen in the present study, and especially among older patients, indicate that more can be done to encourage patients to engage in physical activity. Contact with a local physiotherapist close to home with frequent contact and advise from the PAH center, might open possibilities to support patients being more active in their daily lives.

A previous study showed that patients with CTEPH that undergo PEA surgery improve their CAMPHOR scores and reached MCID criteria to a high degree.[26] These results could not be confirmed in the present study as only two patients underwent PEA, but the low number of patients with PEA might have affected the comparison between subgroups PAH and CTEPH.

The patients' view of their own health through self-assessment is a powerful tool in health care. A patient with

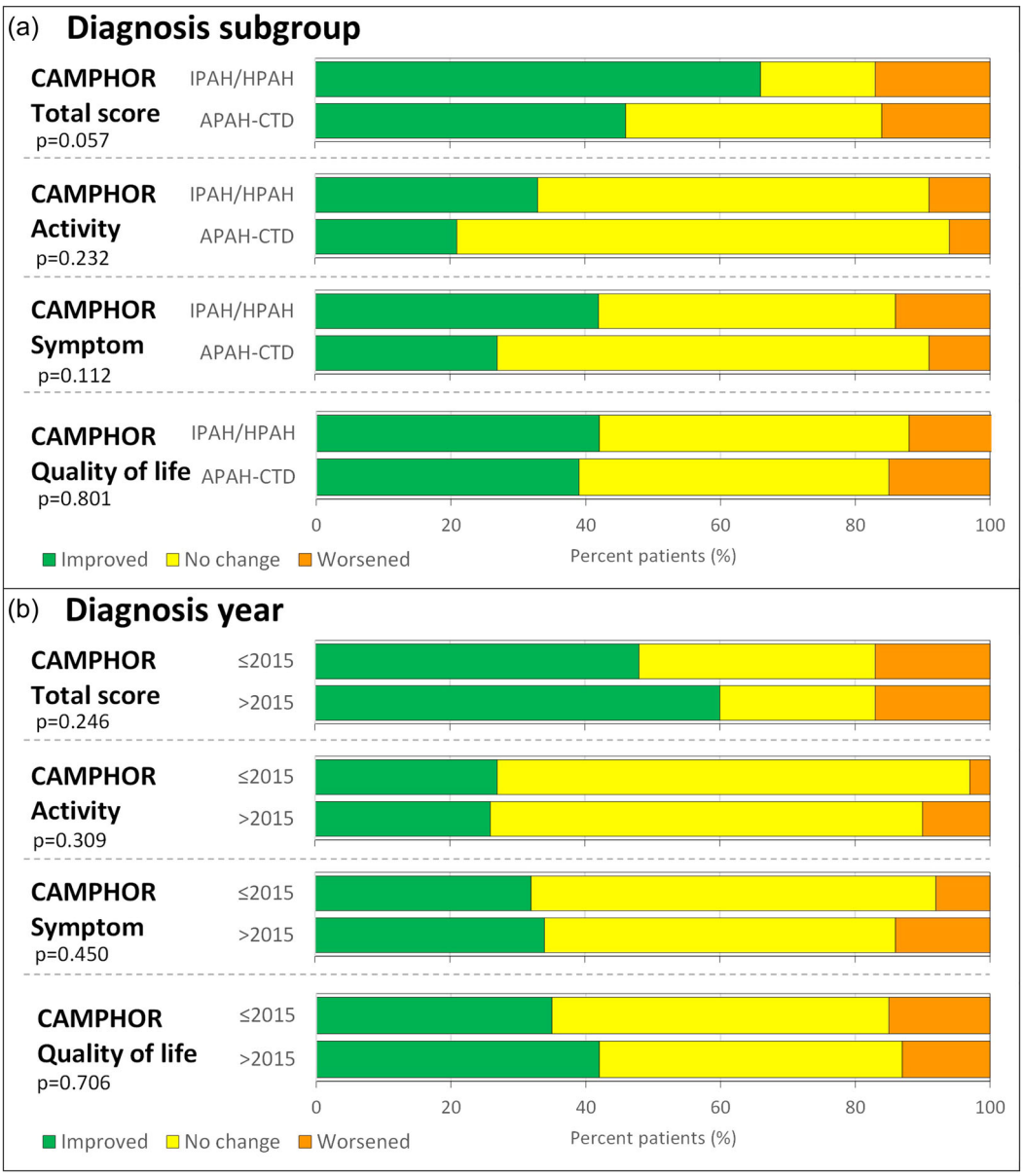

FIGURE 2 Relative change in Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR) scores from diagnosis to the follow-up visit shown by diagnosis subgroup (a) (idiopathic pulmonary arterial hypertension [IPAH], $n = 52$; associated pulmonary arterial hypertension[APAH]-connective tissue disease [CTD] = 56) and diagnosis year (b) (≤2015 = 60, >2015 = 91). Minimal clinically important difference (MCID) were total sum = 4 points, activity = 4 points, symptoms = 4 points, and quality of life = 3 points.[16] An MCID decrease in score was considered improvement and an MCID increase in score was considered worsened.

severely progressed disease may perceive the health status to be to satisfactory, while a patient with less progressed disease might rate the health less poorly.[27] Acceptance or nonacceptance of limitations in daily life, expectations on treatment that are fulfilled or not, and the fear of what the future might bring, will all affect HRQoL individually. However, when using a HRQoL like CAMPHOR with a 1-day recall, it might not reflect that some days are better, and some days are worse for the patient.[11]

Self-assessed HRQoL provide an important mean to facilitate the dialog between the patient and the healthcare professional to understand the patients burden of disease. It can help identify questions that should be asked to help maintain or increase the patient's well-being and functional capacity and to support patients to pay attention to important symptoms. It can be the start of a discussion about possible treatment side effects and help facilitate individualized treatment. HRQoL will support the healthcare professionals to provide a holistic care for the patient as well as increase the patients' own commitment to take responsibility for their health.

STRENGTHS AND LIMITATIONS

The major strength of the present study is that all PH centers in Sweden participate in SPAHR, which allows for the high national coverage of patients with PAH and CTEPH. CAMPHOR is used at all PH centers and is well accepted by the patients (The Swedish PH patient association survey, oral communication). Limitations include the possibility of selection bias, as only patients with CAMPHOR at baseline and a subsequent follow-up within a year were included. Reasons to why patients did not have CAMPHOR or a follow-up registered within the first year are not recorded in SPAHR. In addition, information as to why patients with CTEPH do not undergo PEA or the location of pulmonary lesions is not included in SPAHR. Finally, it has been determined that MCID values are important to evaluate at both the group and individual level.[28] This study only encompasses results of CAMPHOR MCID at the group level. Therefore, further studies are needed on CAMPHOR MCID at individual level.

CONCLUSION

Improvements in CAMPHOR total sum as well as all domains was seen in the first year after diagnosis, however, less than half of the patients reached an MCID. More focus should be put on how the patient experience the burden of living with the disease as well as to encourage participation in an exercise program.

AUTHOR CONTRIBUTIONS

Barbro Kjellström and Bodil Ivarsson contributed to the study concept and design. Material preparation, data collection, analyses, and writing were performed by all authors and all authors have read and approved the final manuscript. All authors meet the International Committee of Medical Journal Editors (ICMJE) criteria for authorship for this article, take responsibility for the integrity of the work, and have given their approval for this version to be published.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

ETHICS STATEMENT

This study complies with the Declaration of Helsinki and was approved by the Swedish Ethical Review Authority, Sweden (Dnr. 2018/373-321).

ORCID

Bodil Ivarsson http://orcid.org/0000-0002-5647-3929
Barbro Kjellström http://orcid.org/0000-0002-7936-1209

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