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ORIGINAL RESEARCH

Diagnostic Delays and Quality of Life in Japanese Patients with Pulmonary Hypertension: A Nationwide Survey

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ABSTRACT

***Introduction*:** Pulmonary hypertension (PH) is a rare and severe disorder that significantly affects patients' lives. However, a comprehensive picture of the diagnosis and treatment of this condition in Japan remains unclear. This study aimed to elucidate these aspects by conducting a nationwide survey targeting patients with PH and treating physicians.

***Methods*:** A cross-sectional survey was conducted among 160 patients with PH (119 with pulmonary arterial hypertension [PAH] and 41 with chronic thromboembolic pulmonary hypertension [CTEPH]), of whom 121 were female (75.6%), and 211 physicians across Japan. The questionnaires assessed patients' diagnostic journey, employment status, communication with physicians regarding treatment goals, health-related quality of life (HRQoL), and medication adherence.

***Results*:** Patients visited a mean of 2.3 medical facilities before receiving a PH diagnosis

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(PAH patients: 2.2 visits; CTEPH patients: 2.3 visits), with a mean time from symptom onset to diagnosis of 18.0 months (PAH: 20.2 months; CTEPH: 12.2 months). Employment and school attendance rates declined from 68.8% before diagnosis to 44.4% immediately after diagnosis, and further to 36.9% at the time of the survey. Discrepancies in communication about treatment goals were observed between patients and physicians, particularly in patients with CTEPH (82.9% of patients reported such discussions vs. 41.2% of treating physicians). Median HRQoL scores, as assessed by the emPHasis-10 questionnaire, indicated impairment (PAH: 21.5; CTEPH: 18.0), which worsened with increasing disease severity.

Conclusion: This nationwide study provides a comprehensive overview of the challenges faced by patients with PH in Japan. The findings suggest the essential need for earlier diagnosis, support for employment and education among patients, and improved patient-physician communication to reduce the burden of PH and enhance patient outcomes.

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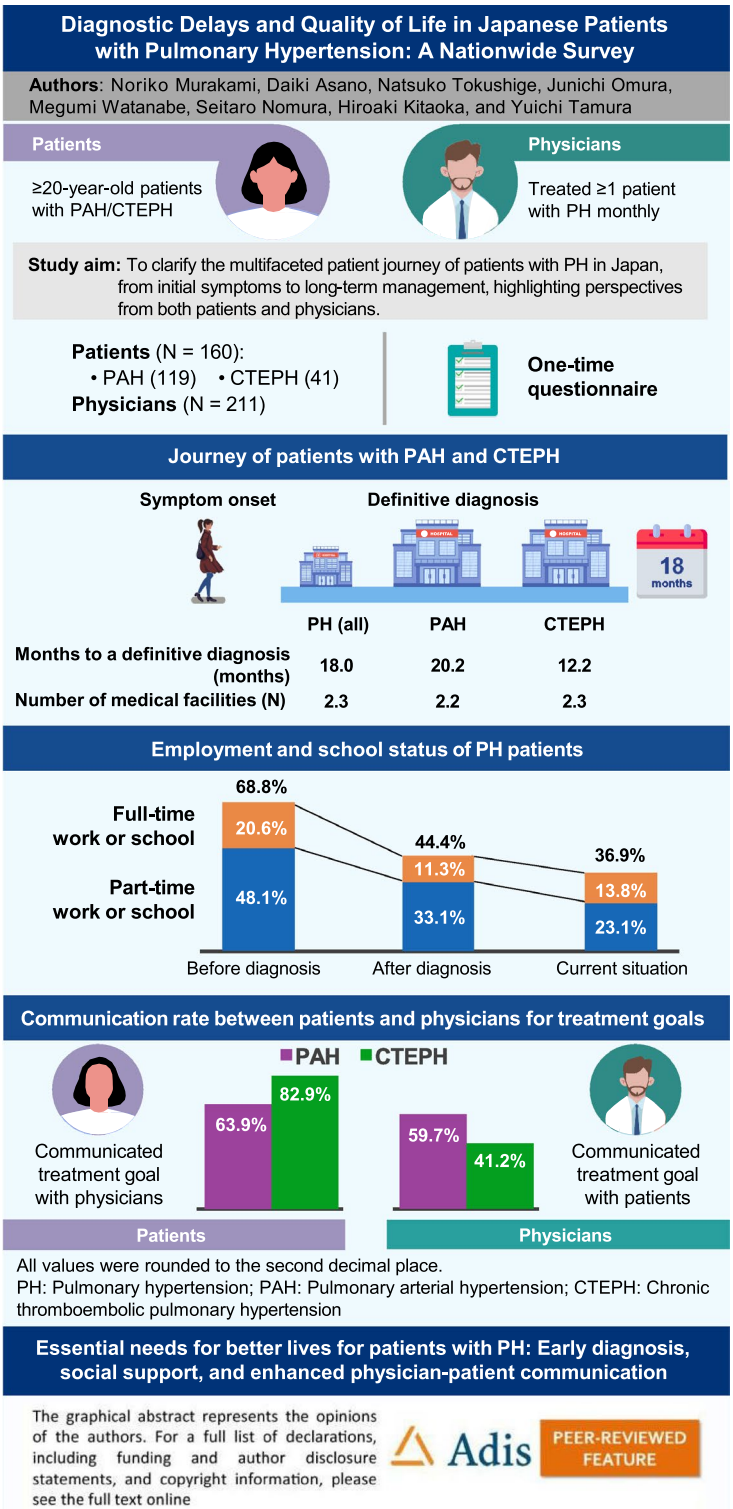

Keywords: Medication adherence; Patient journey; Patient-physician communication; Pulmonary hypertension; Quality of life

Key Summary Points

Why carry out this study?

Pulmonary hypertension (PH) places a significant burden on patients, yet comprehensive data on patient experiences and care pathways in Japan are limited, hindering optimization of clinical management strategies

This nationwide study aimed to clarify the multifaceted patient journey of patients with PH in Japan, from initial symptoms to long-term management, highlighting perspectives from both patients and physicians

What was learned from the study?

Our study revealed several critical insights, including the need to reduce the time from symptom onset to diagnosis, with a mean delay among the surveyed patients of 18.0 months

The study also exposed a substantial socioeconomic impact on these patients with PH, as employment and education rates dropped from 68.8% before diagnosis to 36.9% at the time of the survey, and identified a notable communication gap between patient and physician in terms of perceptions of treatment goals

We identified ongoing challenges in PH management in Japan, emphasizing the urgent need for earlier diagnosis, better patient-physician communication, and comprehensive support systems that address both the medical and socioeconomic aspects of PH care

DIGITAL FEATURES

This article is published with digital features, including a graphical abstract, to facilitate understanding of the article. To view digital features for this article, go to https://doi.org/10.6084/m9.figshare.28435892.

INTRODUCTION

Pulmonary hypertension (PH) is a pathophysiological disorder characterized by elevated blood pressure in the pulmonary arteries, which carry blood from the heart to the lungs. It is defined by a resting mean pulmonary artery pressure exceeding 20 mmHg, measured by right heart catheterization [1]. The two primary subtypes, pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH), are progressive vascular diseases that contribute to poor patient prognosis [2, 3]. Despite significant advancements in pharmacotherapy and invasive procedures, which have improved outcomes for patients with PAH and CTEPH in Japan, approximately 4500 patients with PAH and 5200 patients with CTEPH were estimated to be living with these conditions as of 2022 [4].

The PAH patient journey encompasses a series of experiences beginning with symptom onset and proceeding to hospital visits, diagnosis, and treatment, to daily life in the community. Understanding this journey has gained international recognition as a strategy to enhance healthcare quality [5]. Increasing duration of time from symptom onset to diagnosis (delay) correlates positively with risk stratification scores and mortality rates [6], making it critical to reduce delays in consultation, referral to specialized centers, and definitive diagnosis for both PAH and CTEPH. While the journey of patients with CTEPH has been reported in Japan, a comprehensive view of the journey of patients with PH, including those with PAH or CTEPH, remains unclear [7].

In recent years, health-related quality of life (HRQoL) has become a key consideration in treatment plans [8]. Studies have shown that a decline in HRQoL strongly predicts poor outcomes in patients with PAH and CTEPH [9, 10]. Addressing HRQoL deterioration is therefore urgent for improving PH prognosis, although this aspect of healthcare remains insufficiently

addressed. In clinical practice, patients often prioritize treatment impact on HRQoL, convenience, and daily life, while healthcare providers (HCPs) tend to focus on clinical disease progression [11–13]. These differences in communication priorities may hinder HRQoL improvements. Thus, patient-centered collaborative care, with effective communication, is increasingly important, although practical information in Japan is still limited.

Existing studies on HRQoL in Japan rarely incorporate both patient and physician perspectives within the same investigation. To address diagnostic delays, employment challenges, and shared decision-making (SDM) at a national scale, we designed a large-scale, cross-sectional survey covering patient-reported experiences and physician insights on treatment strategies, adherence, and communication. By recruiting participants across multiple regions, we aimed to capture the broader reality of PAH and CTEPH management in Japan and to highlight potential gaps that affect daily functioning and HRQoL.

Therefore, we conducted the first large-scale, questionnaire-based survey in Japan to clarify the patient journey, disease burden, medication adherence, and communication between patients with PAH or CTEPH and physicians.

METHODS

Design, Setting, and Participants

This study was a noninterventional, cross-sectional survey conducted using written or web-based questionnaires. The target participants were patients with PAH or CTEPH and the physicians treating them. Patients were eligible if they were adults aged ≥ 20 years, had been diagnosed with PAH or CTEPH, and had received treatment at any time. Physicians were included if they treated at least one patient with PH per month. Patients were recruited through hospitals or through PHA Japan, a Japanese PHA patient association (Kanagawa, Japan), and physicians were recruited via PHA Japan or the research company Ipsos Co., Ltd. (Tokyo, Japan). This study was performed in accordance with the Helsinki Declaration of 1964, and its later amendments and was approved by the Ethics Committee of Medical Corporation Heishinkai OPHAC Hospital ERC, Osaka, Japan (Approval No.: 014ERC). All participants reviewed detailed information about the study and provided informed consent either in writing or online. Those who did not consent were excluded from the study. The questionnaire was conducted with complete anonymity. Since all responses were completely anonymized, the total number of approached patients and physicians, as well as those who declined participation, remained unknown. Therefore, a formal response rate could not be determined.

Questionnaire

The questionnaire was specifically designed for this study and pre-reviewed by both physicians and patients with PAH/CTEPH, with the questions included in the analysis. The patient questionnaire comprised five sections. The first section collected patient information, including age, sex, residence, World Health Organization functional class (WHO-FC), and participation in the Japanese Pulmonary Hypertension Registry (JAPHR). As of 2023, JAPHR includes approximately 1300 patients with PAH from 55 centers across Japan [14], representing PAH care in specialized centers. The second section examined the patient journey, the third addressed treatment goals and social life, and the fourth focused on communication with HCPs. The final section assessed HRQoL and medication adherence using patient-reported outcome measures. HRQoL was evaluated using the emPHasis-10 questionnaire [15, 16], and medication adherence was measured using the 12-item Medication Adherence Scale [17]. This scale includes four subscales: medication adherence; collaboration with HCPs; willingness to access and use medication information; and how well medication fits the patient's lifestyle. Each item was rated on a 5-point Likert scale, ranging from 1 (never) to 5 (always), with total scores ranging from 12 to 60. Items 3 and 12 were reverse-scored, with higher total scores indicating better medication adherence. The physician

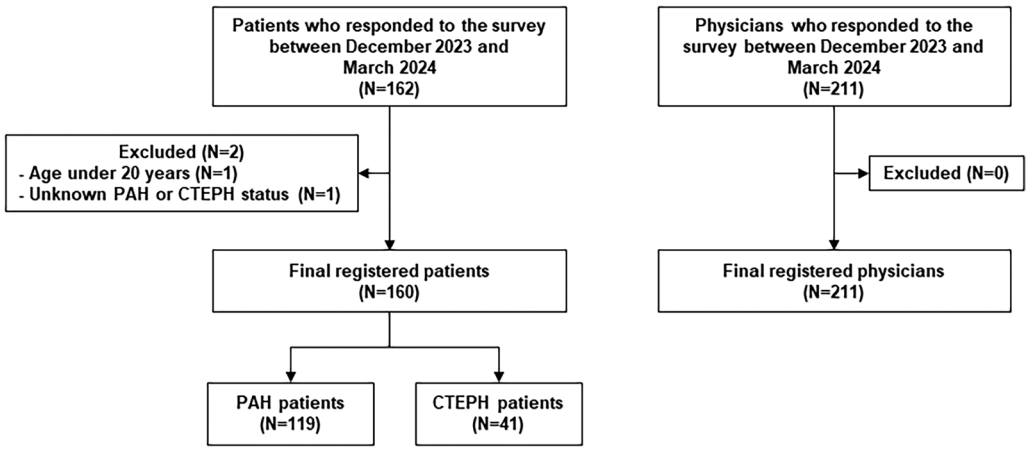

Fig. 1 Flow diagram showing the recruitment of patients and physicians. *CTEPH* Chronic thromboembolic pulmonary hypertension, *PAH* pulmonary arterial hypertension

questionnaire contained three sections. The first section collected physician information, including age, sex, residence, and medical specialty; the second section explored treatment practice patterns; and the third focused on communication with patients. The survey was conducted once per participant between December 2023 and March 2024.

Analysis

Statistical analyses were performed using SAS version 9.4 (SAS Institute Inc., Cary, NC, USA). The collected data were presented as the mean ± standard deviation (SD) if normally distributed or as the median with the minimum and maximum if skewed.

RESULTS

Clinical Characteristics of Patients with PH and Physicians

A total of 160 patients with PH (119 with PAH; 41 with CTEPH), among whom 121 (75.6%) were female, and 211 physicians were included in the study (Fig. 1). In the PAH group, the majority of patients (63 patients, 52.9%) were aged 41–60 years, and in the CTEPH group, most patients (20 patients, 48.8%) were aged 61–80 years. Idiopathic or heritable PAH was the most common subtype in the PAH group (67 patients, 56.3%). Stratification by WHO-FC among all patients with PAH and CTEPH showed that functional class (FC) II was most common type (80 patients, 50.0%), followed by FC I (37 patients, 23.1%), FC III (24 patients, 15.0%), and FC IV (3 patients, 1.9%). Patients were from various regions in Japan, with approximately 90% receiving care at PH centers (Table 1).

Among the physicians participating in the survey, most specialized in cardiology (34.6%), followed by respiratory medicine (25.1%) and rheumatology (14.2%). Over one half of participating physicians had > 5 years of experience treating PH, and approximately 30% of PAH physicians and 10% of CTEPH physicians had treated ≥ 6 patients in the previous year. About 50% of physicians were affiliated with PH centers (Table 2).

Patient Journey from Symptom Onset to a Definitive Diagnosis of PH

The patient journey is summarized in Table 3. mean, patients visited 2.3 (SD, 1.2) medical facilities before receiving a definitive diagnosis. The mean time to diagnosis was 20.2 (SD 49.6) months for patients with PAH and 12.2 (SD 17.5) months for those with CTEPH, with the results clearly showing a longer delay for a

Table 1 Characteristics of patients with pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension at the time of survey response

Patient characteristics	All patients (patients with PAH or CTEPH) (*N* = 160)	Patients with PAH (*N* = 119)	Patients with CTEPH (*N* = 41)
Age (years)			
20–40	29 (18.1)	25 (21.0)	4 (9.8)
41–60	73 (45.6)	63 (52.9)	10 (24.4)
61–80	44 (27.5)	24 (20.2)	20 (48.8)
≥ 81	10 (6.3)	4 (3.4)	6 (14.6)
Unknown	4 (2.5)	3 (2.5)	1 (2.4)
Sex			
Male	37 (23.1)	23 (19.3)	14 (34.2)
Female	121 (75.6)	94 (79.0)	27 (65.9)
Others	2 (1.3)	2 (1.7)	–
Classification of PAH			
Idiopathic/heritable	–	67 (56.3)	–
Associated with CTD	–	32 (26.9)	–
Associated with CHD	–	11 (9.2)	–
Others	–	9 (7.6)	
WHO functional class			
I	37 (23.1)	26 (21.9)	11 (26.8)
II	80 (50.0)	58 (48.7)	22 (53.7)
III	24 (15.0)	21 (17.7)	3 (7.3)
IV	3 (1.9)	1 (0.8)	2 (4.9)
Unknown	16 (10.0)	13 (10.9)	3 (7.3)
Residential area			
Northern Japan	35 (21.9)	24 (20.2)	11 (26.8)
Central Japan	96 (60.0)	73 (61.3)	23 (56.1)
Western Japan	27 (16.9)	20 (16.8)	7 (17.1)
No answer	2 (1.3)	2 (1.7)	–
Receiving treatment at a PH center			
Yes	140 (87.5)	104 (87.4)	36 (87.8)

Values in table are the number of patients (*N*), with the percentage (%) of the total number of patients in each group given in parenthesis

CHD Congenital heart disease, *CTD* connective tissue disease, *CTEPH* chronic thromboembolic pulmonary hypertension, *PAH* pulmonary arterial hypertension, *PH* pulmonary hypertension, *WHO* World Health Organization

diagnosis of PAH. It should be noted that since 2016, following the publication of PH guidelines by the European Society of Cardiology (ESC) and the European Respiratory Society (ERS) [18], the time to diagnosis has noticeably decreased for both conditions, although the number of medical facilities visited prior to diagnosis has remained unchanged.

The most common tests performed at the initial medical facility that the patient visited regarding suspected PH included chest X-ray (107 patients, 66.9%), electrocardiogram (104 patients, 65.0%), blood tests (93 patients, 58.1%), and echocardiography (81 patients, 50.6%). Right heart catheterization was performed in 23.1% of cases (Table 4).

Before receiving a PAH/CTEPH diagnosis, the most frequent interim diagnosis was "no diagnosis" (54 patients, 33.8%), followed by "heart failure" (31 patients, 19.4%). The rate of heart failure diagnoses in patients ultimately diagnosed with CTEPH (7 patients, 17.1%) was similar to that in patients ultimately diagnosed with PAH (24 patients, 20.2%).

The Impact of PH on Employment and School Status

Figure 2 illustrates employment and school status before diagnosis, immediately after diagnosis, and at the time of the survey. The percentage of individuals employed or studying (full-time or part-time) dropped from 110 out of 160 patients (68.8%) before diagnosis to 71 out of 160 patients (44.4%) immediately after diagnosis, and further to 59 out of 160 patients (36.9%) at the time of the survey. The proportion of those not working or attending school due to health issues increased from nine out of 160 patients (5.6%) before diagnosis to 15 out of 160 patients (9.4%) immediately after diagnosis, and further to 33 out of 160 patients (20.6%) at the time of the survey. The observed decline in employment or school attendance was similar across both mild (WHO-FC I, II) and severe PH (WHO-FC III, IV) groups at the time of survey completion (Fig. 2b). This trend was also seen in separate analyses for PAH and CTEPH (Electronic Supplementary Material [ESM] Fig. S1a–d).

Communication About Treatment Goal

Patients' perceptions of communication about treatment goals and physicians' experiences with such discussions are shown in Fig. 3. Among patients with PAH, 76 (63.9%) reported discussing treatment goals such as work, school, travel, and sports with their physicians, compared to 126 physicians treating PAH patients (59.7%). In patients with CTEPH, 34 (82.9%) reported these discussions, compared to 87 physicians treating CTEPH patients (41.2%). These results indicate a significantly higher perception of communication among patients.

Patients with WHO-FC I/II PH had a higher consultation rate (83 patients, 70.9%) compared to those with WHO-FC III/IV (17 patients, 63.0%) PH (ESM Table S1). Patients living within 1 h of a PH treatment facility had higher consultation rates (77 patients, 71.3%) than those living further away (33 patients, 63.5%). Physicians at PH centers reported higher consultation rates (79 physicians, 75.2%) than those not affiliated with such centers (55 physicians, 51.9%). Physicians who had treated > 5 patients with CTEPH in the past year also had higher consultation rates (ESM Table S2).

Health-Related Quality of Life

The HRQoL results as assessed by the emPHasis-10 are shown in Table 5. The median HRQoL scores for patients with PH, PAH, and CTEPH were 21.0, 21.5, and 18.0, respectively. HRQoL in patients with PH worsened with increasing WHO-FC severity (WHO-FC I: 14.0; II: 21.0; III: 36.0; IV: 31.5). This trend was observed in both patients with PAH and those with CTEPH. HRQoL scores based on the etiology of PAH were similar.

Medication Adherence

The results of the 12-item Medication Adherence Scale are given in Table 6. The overall score was 47.0 (SD 7.2), with patients with PAH scoring

Table 2 Characteristics of the participating physicians at the time of survey response

Characteristics of participating physicians ($N = 211$ physicians)	N (%)
Age (years)	
21–40	82 (38.9)
41–60	105 (49.8)
61–80	24 (11.4)
Sex	
Male	189 (89.6)
Female	21 (10.0)
No answer	1 (0.5)
Medical specialty	
Cardiology	73 (34.6)
Respiratory medicine	53 (25.1)
Rheumatology	30 (14.2)
Gastroenterology	19 (9.0)
Pediatrics	13 (6.2)
Cardiac/respiratory surgery	3 (1.4)
Dermatology	2 (1.0)
Orthopedics	2 (1.0)
Others	16 (7.6)
Years treating PAH or CTEPH patients	
< 1	15 (7.1)
1–3	35 (16.6)
3–5	30 (14.2)
≥ 5	131 (62.1)
Number of patients with PAH in the previous year	
≤ 5	153 (72.5)
6–10	22 (10.4)
11–20	19 (9.0)
≥ 21	17 (8.1)
Number of patients with CTEPH in the previous year	
≤ 5	184 (87.2)
6–10	10 (4.7)
11–20	5 (2.4)
≥ 21	12 (5.7)
Work location	
Northern Japan	31 (14.7)
Central Japan	106 (50.2)
Western Japan	74 (35.1)
Working at a PH center	
Yes	105 (49.8)

CTEPH chronic thromboembolic pulmonary hypertension, *PAH* pulmonary arterial hypertension, *PH* pulmonary hypertension. Values are expressed as N (% of the total number of physicians)

46.9 (SD 7.6) and those with CTEPH scoring 47.1 (SD 6.2). Among the four subscales (15 points per 3 items), the highest and lowest scores were for medication adherence (mean 14.0, SD 2.1) and for obtaining and using knowledge on medication (mean 10.2, SD 2.7), respectively. Patients with PAH or CTEPH displayed similar trends across all items.

Analysis of the relationship between patient-physician communication and medication adherence revealed that patients who reported discussing with their physicians on treatment goals, work, school, travel, and sports had a higher adherence score (mean 48.3, SD 6.9) than those who did not have such a discussion (mean 44.1, SD 7.2) (Table 7). This trend was consistent for both PAH and CTEPH patients. Despite high scores in the medication adherence section of the Medication Adherence Scale, 47 patients (29.4%) responded "almost always/often/sometimes" to the question "Have you ever thought it would be easier to follow your treatment plan if you had fewer pills?". Among patients with PAH, 40 (33.6%) responded this way, suggesting that one-third of patients with PAH were seeking improved dosing regimens (ESM Table S3).

Table 3 Journey of patients with pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension

Patient journey	Patients with PAH or CTEPH		Patients with PAH		Patients with CTEPH	
	N	Mean ± SD	*N*	Mean ± SD	*N*	Mean ± SD
Number of medical facilities before a definitive diagnosis						
Overall	158	2.3 ± 1.2	119	2.2 ± 1.3	39	2.3 ± 1.0
Definitive diagnosis before 2015	79	2.3 ± 1.4	63	2.2 ± 1.4	16	2.7 ± 1.4
Definitive diagnosis after 2016	77	2.2 ± 1.0	54	2.3 ± 1.1	23	2.0 ± 0.6
Diagnostic timeline						
Months to a definitive diagnosis						
- Overall	145	18.0 ± 43.5	106	20.2 ± 49.6	39	12.2 ± 17.5
- Definitive diagnosis before 2015	70	21.0 ± 55.8	54	22.0 ± 62.2	16	17.6 ± 24.8
- Definitive diagnosis after 2016	75	15.2 ± 27.5	52	18.2 ± 32.2	23	8.5 ± 8.4

CTEPH Chronic thromboembolic pulmonary hypertension, *PAH* pulmonary arterial hypertension, *SD* standard deviation

DISCUSSION

This study is the first large-scale questionnaire-based survey targeting patients with PH and their treating physicians in Japan. The key findings are: (1) the mean number of medical facilities visited by patients from symptom onset before a PH diagnosis was reached was 2.3 (PAH patients: 2.2; CTEPH patients: 2.3), and the mean time to definitive diagnosis was 18.0 months, with longer times for patients with PAH than for patients with CTEPH (PAH patients: 20.2 months; CTEPH patients: 12.2 months); (2) employment and education rates declined postdiagnosis compared to pre-diagnosis, with further decreases reported after treatment initiation, particularly in patients with classified with WHO-FC III/IV compared to those classified with WHO-FC I/II; (3) there were significant discrepancies in perceptions of communication (e.g., treatment goals) between patients and physicians; and (4) HRQoL for patients with PH worsened with increasing WHO-FC. These findings highlight the current status and challenges in PH treatment in Japan and provide key insights for improving future clinical practice.

Patient Journey in PH

The results of this study provide insights into the full patient journey of individuals with PH in Japan. We found that an average of 2.3 medical facilities were involved in the diagnosis of PH. These findings suggest the development of an effective referral network in Japan, which is in line with recent ESC/ERS guidelines that emphasize collaboration between general practitioners and PH centers [19]. However, the mean time from symptom onset to confirmed diagnosis was 18.0 months for the overall patient population, 20.2 months for patients with PAH, and 12.2 months for patients with CTEPH.

A 2014 press release from a patient association reported an interval of 3.5 years from symptom onset to diagnosis [20], a time interval that exceeds our findings. Our shorter interval for diagnosis suggests that there have been improvements in current understanding of PH and increased disease awareness, supported by guideline updates, leading to faster PH diagnoses in Japan. It also implies that, in line with updated guidelines [18, 19, 21], accumulated knowledge on PH and increased awareness have contributed to more timely diagnoses

Table 4 Medical tests and disease assessments before pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension diagnosis

Medical tests and disease assessments	Patients with PAH or CTEPH (*N* = 160)	Patients with PAH (*N* = 119)	Patients with CTEPH (*N* = 41)
Medical tests performed at the initial health care facility visited by the patient			
Consultation only	39 (24.4)	28 (23.5)	11 (26.8)
Physical examination	76 (47.5)	59 (49.6)	17 (41.5)
Electrocardiogram	104 (65.0)	75 (63.0)	29 (70.7)
Chest radiography	107 (66.9)	75 (63.0)	32 (78.0)
Pulmonary function test	43 (26.9)	33 (27.7)	10 (24.4)
Echocardiogram	81 (50.6)	60 (50.4)	21 (51.2)
Blood test	93 (58.1)	68 (57.1)	25 (61.0)
Right heart catheterization	37 (23.1)	32 (26.9)	5 (12.2)
Unknown	15 (9.4)	13 (10.9)	2 (4.9)
Diagnosis before visiting the current facility			
None	54 (33.8)	40 (33.6)	14 (34.1)
Heart failure	31 (19.4)	24 (20.2)	7 (17.1)
Symptoms associated with underlying disease	23 (14.4)	21 (17.6)	2 (4.9)
Asthma	19 (11.9)	14 (11.8)	5 (12.2)
Valvular disease	5 (3.1)	4 (3.4)	1 (2.4)
Epilepsy	1 (0.6)	1 (0.8)	0 (0)
Others	58 (36.3)	41 (34.5)	17 (41.5)

Values in table are the number of patients (*N*), with the percentage (%) of the total number of patients in each group given in parenthesis

CTEPH Chronic thromboembolic pulmonary hypertension, *PAH* pulmonary arterial hypertension

[a]A patient who gave multiple responses was counted twice or more across categories

in Japan. The onset of increased pulmonary arterial pressure (PAP) typically occurs only after approximately 70% of the vascular bed is compromised [22]. Consequently, when PH is suspected and a patient is referred to a specialized PH center, pathological vascular remodeling has often progressed significantly. Thus, in terms of the pathogenesis of this condition, there is a need for further improvement, with the mean duration from symptom onset to diagnosis being 18.0 months. PH is occasionally misdiagnosed as other conditions, such as asthma or congestive heart failure, due to the presence of nonspecific symptoms [23]. Indeed, our results showed that about 40% of patients received such misdiagnoses before being diagnosed correctly with PH. Additionally, barriers to diagnosing PH and accessing appropriate care may exist in Japan, particularly at the general practitioner level [24–29]. Delayed diagnosis and treatment initiation, including guideline-directed medical therapy, have worsened the prognosis of heart diseases, including

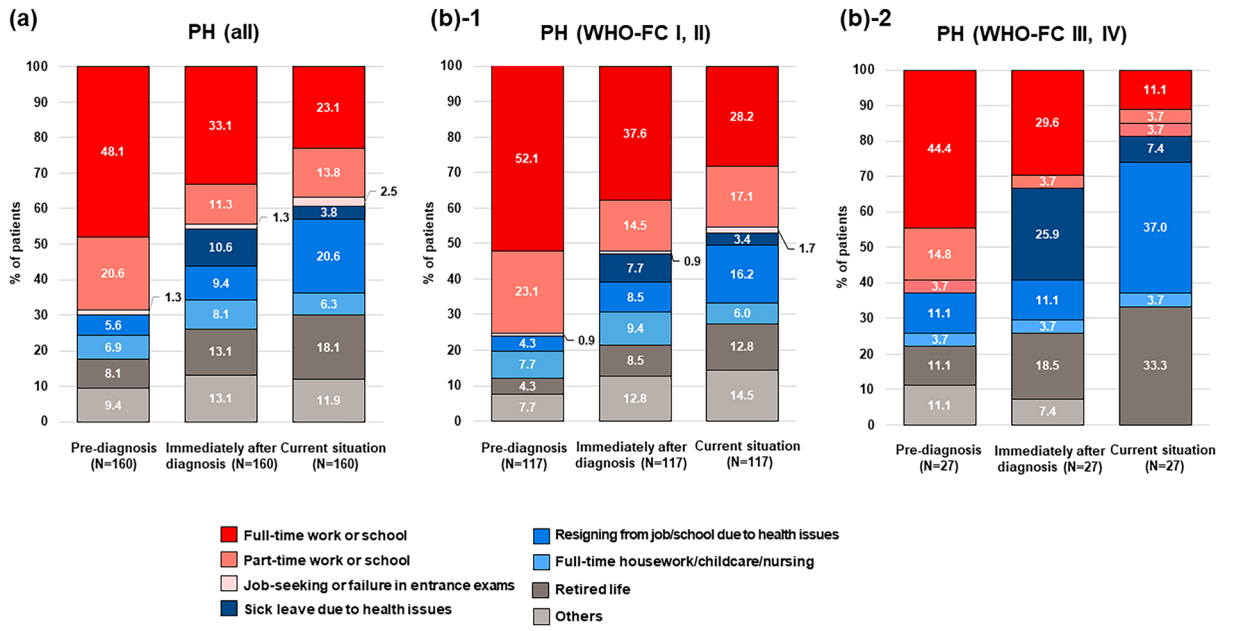

Fig. 2 Changes in work and school status in patients with PH. Change in work or school status for patients with PH from pre-diagnosis, immediately after diagnosis, to current situation (% of the total number of patients) is shown for patients with PH (**a**) and those stratified according to WHO-FC I and II (**b-1**) and III and IV (**b-2**). *PH* Pulmonary hypertension, *WHO-FC* World Health Organization functional class

PH [6, 30, 31]. As indicated by previous studies [6, 30, 31] and the actual number of medical facilities visited by the patients in our study before a definitive diagnosis was made (mean 2.3), it is evident that challenges persist at the level of general practitioners. Efforts should focus on identifying factors contributing to diagnostic delays and on developing strategies to improve early diagnosis across Japan's healthcare system.

Impact of PH Burden on Employment and Education

The impact of PH on patients' employment and educational opportunities is severe and long-lasting, as our study demonstrates. Employment and school attendance rates dropped sharply from 68.8% before diagnosis to 44.4% immediately after diagnosis, further declining to 36.9% at the time of the survey. Additionally, our study showed that even patients classified with mild PH (i.e., WHO functional class I–II) faced significant challenges in continuing employment or education, highlighting the considerable socioeconomic burden of PH. The WHO-FC-dependent decline in employment and school rates observed in our study aligns with findings from a previous international survey [32]. Regardless of severity, the diagnosis of PH itself imposes a significant psychological burden, such as anxiety and depression, which in turn affects the patient's ability to work, especially as some patients require hospitalization [33]. The decline in employment status results not only from factors related to PH severity (e.g., 6-min walk distance, WHO functional class) but also from educational level [34]. Furthermore, unemployment or missed educational opportunities are linked to reduced social engagement and subsequent poor quality of life (QoL) in individuals with PAH [34, 35]. Recent studies have noted a decline

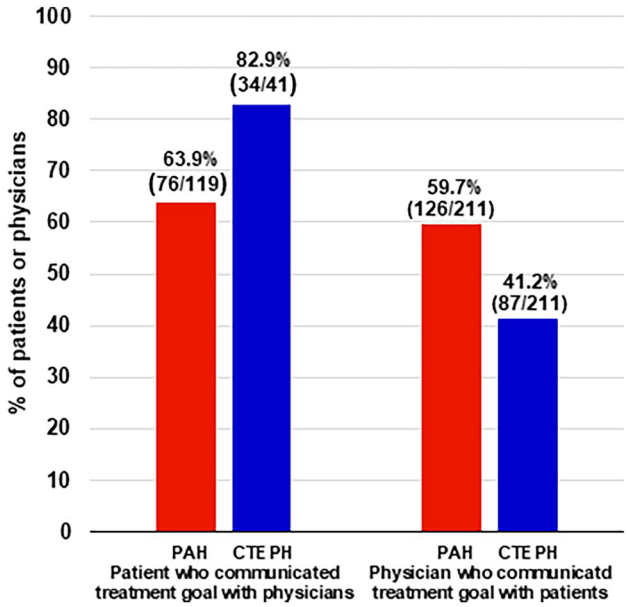

Fig. 3 Communication regarding treatment goals between patients and physicians. The question regarding the consultation of patient's goals was: "Are you able to consult with your primary physician for pulmonary arterial hypertension regarding the goals you want to achieve through treatment, such as work, school, travel, or sports?". The question regarding physician's experience of receiving consultation regarding goals was: "Have you ever received a consultation regarding what the patient wants to achieve through treatment, such as return to work, school, travel, or sports?". Vertical axis shows the percentage and number of patients or physicians who answered yes to each posed question. *CTEPH* Chronic thromboembolic pulmonary hypertension, *PAH* pulmonary arterial hypertension

in socioeconomic status (SES) due to loss of employment and educational opportunities among patients with PAH [36]. The impact of low SES on PH care (e.g., delays in diagnosis, difficulties accessing treatment, and prognosis) varies across healthcare systems in different countries [37, 38]. Therefore, as suggested by the decline in employment and school attendance status among patients with PH in our study, further research is needed to understand why these statuses remain low even after treatment, and to determine whether low SES may affect the prognosis of patients with PH in Japan.

Communication Between Patient and Physician

In our study, the communication rates for treatment goals in PH varied among patients and physicians. Among patients with PAH and physicians treating PAH, 63.9% and 59.7%, respectively, reported discussing treatment goals; among patients with CTEPH and physicians treating CTEPH, 82.9% and 41.2%, respectively reported discussing treatment goals. Discussing treatment goals is crucial in terms of respecting shared decision-making (SDM). Previous studies have shown that effective communication significantly impacts SDM. Furthermore, SDM leads to increased self-efficacy, self-management, well-being, treatment decision-making, and improved health outcomes [39]. A recent study of patients with PAH found that better coping abilities are associated with higher satisfaction regarding the information provided about treatment and medical tests [11].

Despite the clear need for such conversations and the substantial benefits they offer patients, effective communication is not reliably integrated into clinical practice [40, 41]. Our study revealed that physicians affiliated

Table 5 emPHasis-10 questionnaire score based on the severity and types of pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension

Severity and types of PAH or CTEPH	Patients with PAH or CTEPH		Patients with PAH		Patients with CTEPH	
	N	Median (min–max)	*N*	Median (min–max)	*N*	Median (min–max)
Overall	158	21.0 (0–50)	118	21.5 (2–50)	40	18.0 (0–45)
WHO functional class						
I	37	14.0 (0–39)	26	14.0 (2–39)	11	11.0 (0–27)
II	79	21.0 (1–45)	57	22.0 (3–45)	22	21.0 (1–42)
III	24	36.0 (3–45)	21	36.0 (3–44)	3	36.0 (33–45)
IV	2	31.5 (22–41)	1	41.0 (41–41)	1	22.0 (22–22)
Classification of PAH						
Idiopathic/heritable			66	21.5 (2–45)		
Associated with CTD			32	22.0 (2–44)		
Associated with CHD			11	22.0 (10–50)		
Others			9	15.0 (9–44)		

Values in the Median (min–max) columns of the table are the emPHasis-10 questionnaire scores

CHD Congenital heart disease, *CTD* connective tissue disease, *CTEPH* chronic thromboembolic pulmonary hypertension, *min–max* minimum–maximum, *PAH* pulmonary arterial hypertension, *WHO* World Health Organization

with PH centers and those with extensive case experience communicated treatment goals more frequently with their patients, supporting SDM. Although differences in communication practices depend on physician affiliation and experience, full implementation of these practices remains lacking, which aligns with previous reports [40, 41]. This suggests that patients' preferences are not adequately communicated to HCPs and are not reflected in treatment strategies. Thus, our results highlight important issues regarding communication between patients and physicians, including SDM in the PH treatment landscape.

HRQoL and Medication Adherence in Patients with PH

In this analysis, we provided a nationwide picture of HRQoL in Japanese patients with PH using the emPHasis-10 questionnaire. Our findings showed that declining HRQoL, as measured by the emPHasis-10 questionnaire, was positively correlated with higher WHO-FC—i.e., HRQoL was increasingly impaired with higher WHO-FC. This observation is consistent with the results of previous Japanese studies [16]. The emPHasis-10 score, which reflects physical and psychological distress in patients with PH, indicated significant impairment in HRQoL even among those with mild symptoms (e.g., WHO-FC I-II). Additionally, in our study we noted the impact of the patient journey on SES and the necessity for improved communication between patients and physicians. Accordingly, a more comprehensive approach is needed to address the direct and indirect factors associated with HRQoL decline in patients with PH.

This study reveals the perceptions of patients with PH in Japan on medication adherence for the first time. The overall adherence score closely aligns with that of previous studies involving patients with chronic illnesses [17]. Notably, there are consistent trends in four key components: high scores for medication adherence, acceptance of medication, and how taking medication fits into patients'

Table 6 Medication adherence based on the 12-item Medication Adherence Scale

Medication Adherence Scale	Patients with PAH or CTEPH ($N = 157$)	Patients with PAH ($N = 117$)	Patients with CTEPH ($N = 40$)
Medication compliance	14.0 ± 2.1	13.8 ± 2.3	14.5 ± 1.2
1. Over the past 3 weeks, I have taken the prescribed daily dosage of my medication	4.8 ± 0.7	4.7 ± 0.8	5.0 ± 0.2
2. Over the past 3 weeks, I have followed the instructions about when or how often to take my medication	4.6 ± 0.8	4.5 ± 0.9	4.8 ± 0.5
3. I have stopped taking medication based on my own judgment (not including times when I forgot to take my medication)	4.6 ± 0.9	4.6 ± 0.9	4.7 ± 0.9
Collaboration with healthcare providers	10.6 ± 3.1	10.7 ± 3.1	10.5 ± 3.2
4. I feel comfortable asking my healthcare provider about my medication	3.3 ± 1.3	3.4 ± 1.3	3.1 ± 1.5
5. My healthcare provider understands when I tell him/her about my preferences in medication taking	3.4 ± 1.4	3.4 ± 1.4	3.3 ± 1.5
6. My healthcare provider understands when I explain to him/her about my past medication including previous allergic reactions	4.0 ± 1.3	3.9 ± 1.3	4.1 ± 1.2
Willingness to access and use information about medication	10.2 ± 2.7	10.5 ± 2.6	9.5 ± 2.8
7. I understand both the effects and side effects of my medication	3.9 ± 1.0	3.8 ± 1.0	3.9 ± 1.1
8. I report side effects, allergic reactions, or unusual symptoms caused by the medication	3.8 ± 1.3	3.9 ± 1.2	3.3 ± 1.4
9. I personally search for and collect information that I want about my medication	2.6 ± 1.2	2.7 ± 1.2	2.3 ± 1.2
Acceptance to take medication and how taking medication fits patient's lifestyle	12.2 ± 2.2	12.0 ± 2.2	12.7 ± 2.1
10. I accept the necessity of taking medication in the prescribed manner to treat my illness	4.5 ± 0.7	4.5 ± 0.7	4.6 ± 0.5
11. Taking medication is part of my everyday life, just like eating or brushing my teeth	4.5 ± 0.9	4.4 ± 0.9	4.7 ± 0.7
12. I sometimes get annoyed that I have to keep taking medicine every day	3.1 ± 1.3	3.0 ± 1.3	3.4 ± 1.3
Total score	47.0 ± 7.2	46.9 ± 7.6	47.1 ± 6.2

Values in table are the mean (± standard deviation) Medication Adherence Scale scores

CTEPH Chronic thromboembolic pulmonary hypertension, *PAH* pulmonary arterial hypertension

lifestyles, alongside comparatively lower scores for collaboration with HCPs and willingness to access and use information on medication. Evidently, misinformation and disinformation in the healthcare domain pose significant challenges to all patients, including those with PH

Table 7 The 12-item Medication Adherence Scores stratified according to a yes/no answer to the question of consulting about their treatment goals with a physician

Answer	Patients with PAH or CTEPH		Patients with PAH		Patients with CTEPH	
	N	Mean ± SD	*N*	Mean ± SD	*N*	Mean ± SD
Consultation regarding treatment goals with a physician, which include factors such as return to work, school, travel, or sports						
Yes	108	48.3 ± 6.9	75	48.4 ± 7.5	33	48.0 ± 5.4
No	49	44.1 ± 7.2	42	44.3 ± 7.1	7	42.6 ± 8.0

CTEPH chronic thromboembolic pulmonary hypertension, *PAH* pulmonary arterial hypertension, *SD* standard deviation

[42]. Consequently, there is an urgent need to enhance communication with HCPs and ensure access to accurate information. As treatment regimens for PH continue to evolve, incorporating therapies for concurrent conditions, medication adherence will become a critical issue [43]. Therefore, patient expectations regarding the number of medications in our study reflect this evolving trend.

Joint Perspectives from Patients and Physicians

This study reports the perspectives of both patients and physicians on PH across Japan, providing wide-ranging insights. By targeting both PAH and CTEPH, we highlight notable similarities and differences regarding diagnostic delays, communication practices, and employment situation. This patient survey aimed to clarify challenges and treatment goals extending from diagnosis to daily life. The results revealed that diagnostic delays persist and that social participation, such as employment or schooling, remains restricted. In contrast, the physician survey explored how institutional affiliation and clinical experience influence patient-centered care. The results showed that physicians at PH centers or those with longer experience were more likely to discuss goals such as work or education, thereby supporting SDM. However, the limited sample sizes in both surveys reduced statistical power for subgroup analyses based on patient and physician characteristics. Future research should employ larger samples to enable deeper insights into these subgroups and provide more robust conclusions.

Clinical Implications

From a clinical perspective, it is important to emphasize that the results of this study clearly indicate the need to shorten the time to a definitive diagnosis of PAH and highlight the current issues, such as a decline in SES and impaired HRQoL. Effective communication between physicians and patients regarding treatment goals, as well as discussions aimed at improving SES and HRQoL, are essential for establishing and adapting individualized patient care pathways. Given the role that effective communication plays in important health outcomes, multidisciplinary teams, including physicians, need to consider these findings and make necessary adjustments in their communication with patients.

Limitations

There are several limitations to this study. First, since data collection relied on patient and physician self-reporting, the possibility of self-report bias cannot be ignored. This is particularly relevant when considering the apparent underreporting of right heart catheterization procedures. In fact, despite Japanese guidelines requiring right heart catheterization for a definitive diagnosis of PAH or CTEPH [21], relatively few patients in this study reported undergoing right heart catheterization. This does not necessarily imply an absence of right heart catheterization in clinical practice but is likely influenced by factors inherent to patient-reported outcomes (PROs), such as imperfect recall or unfamiliarity with medical terminology. Second, because

participants were primarily recruited through specific medical facilities and patient associations, the study may not fully represent the overall population of patients with PAH/CTEPH and physicians in Japan. Furthermore, patients who are part of these associations tend to have a higher interest in their condition and treatment and are more likely to reside in areas or facilities with better access to healthcare. Third, we did not collect data on income or survival, thereby limiting our ability to assess socioeconomic impacts and long-term outcomes. Moreover, the limited number of patients and physicians impeded detailed subgroup analyses. These factors restrict the generalizability of our findings and underscore the need for further comprehensive studies. Finally, the cross-sectional design limits the availability of long-term follow-up data, as data collection occurs at a single point in time, making it difficult to evaluate the long-term effects of treatment and changes in patient QoL over time. Future studies should include larger and more diverse patient cohorts, adopt prospective designs, and incorporate comprehensive assessments that integrate objective clinical data with PROs.

CONCLUSION

This is the first study to thoroughly elucidate the nationwide PH treatment situation in Japan from the patients' perspective. The findings reveal delays in diagnosis, significant impacts on employment and education, communication challenges between patients and physicians, and effects on HRQoL. This study emphasizes the need for a comprehensive approach to PH care that encompasses not only medical treatment but also psychological and social support. Registries and community-based randomized studies will be further needed to enhance understanding of the patient experience and improve the overall quality of care in PH management.

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Declarations

Conflict of Interest. Noriko Murakami, Junichi Omura, Megmi Watanabe, Seitaro Nomura, Hiroaki Kitaoka, and Yuichi Tamura have nothing to disclose. Daiki Asano and Natsuko Tokushige are employees of Janssen Pharmaceutical K. K.

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