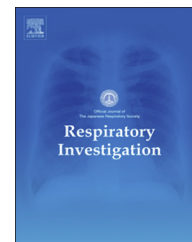




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## Original article

# Current trends in the management of pulmonary hypertension associated with respiratory disease in institutions approved by the Japanese Respiratory Society



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Abbreviations: PH, pulmonary hypertension; R-PH, pulmonary hypertension associated with respiratory disease; ESC, European Society of Cardiology; ERS, European Respiratory Society; RHC, right heart catheterization; PAH, pulmonary arterial hypertension; JRS, Japanese Respiratory Society; mPAP, mean pulmonary arterial pressure; COPD, chronic obstructive pulmonary disease; ASPIRE, Assessing the Spectrum of Pulmonary Hypertension Identified at a Referral Center

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## ABSTRACT

**Background:** Pulmonary hypertension (PH) often correlates with respiratory disease severity. Right heart catheterization (RHC) is recommended for the definitive diagnosis of PH associated with respiratory disease (R-PH). However, no previous studies have evaluated the perceived necessity for pulmonologists to use RHC for R-PH diagnosis, or the management of R-PH in Japan.

**Methods:** Questionnaires were mailed to 855 institutions, approved by the Japanese Respiratory Society. Questions included the prevalence and necessity of RHC and other methods in R-PH diagnosis, and current trends in the treatment of R-PH.

**Results:** Questionnaires were returned from 289 institutions (34%). Patients with R-PH were examined by pulmonologists in 89% of institutions; some pulmonologists performed echocardiography (15%) and some RHC (13%). Echocardiography was used to diagnose R-PH in 99% of institutions and RHC was used in 36%. RHC was considered in cases of suspected PH in 49% of institutions and prior to initiation of pulmonary arterial hypertension (PAH)-specific therapy in 57%. Of patients diagnosed with R-PH, 47% were treated with ambulatory oxygen therapy. Furthermore, 98 of 145 institutions used PAH-specific therapy to treat R-PH. Of the 1355 patients who underwent RHC as a part of PH evaluation, 29% were confirmed to have PH, and 8% had severe PH with a mean pulmonary arterial pressure of  $\geq 35$  mmHg.

**Conclusions:** The current diagnostic and treatment modalities for R-PH in Japan were evaluated. Although few pulmonologists perform RHC for R-PH diagnosis in Japan, more than half consider using RHC for patients before initiating PAH-specific therapy.

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## 1. Introduction

Pulmonary hypertension (PH) associated with respiratory disease (R-PH) correlates with worsening of respiratory disease and progression of hypoxemia. Patients with PH associated with lung disease and/or hypoxemia are classified as Group 3 of the clinical classification of PH (Dana Point, 2008) [1]. However, the current diagnostic and treatment modalities used in patients with Group 3 PH have not been extensively studied in Japan.

Although the European Society of Cardiology (ESC)/European Respiratory Society (ERS) guidelines recommend right heart catheterization (RHC) for the definitive diagnosis of R-PH [2], the prevalence of the use of RHC for the diagnosis of R-PH by pulmonologists in Japan has not been reported. Appropriate management of patients with R-PH might be improved if pulmonologists in Japan recognized the importance of RHC, especially before initiating pulmonary arterial hypertension (PAH)-specific therapy [3]. Long-term oxygen therapy is the standard treatment for R-PH [2]. PAH-specific therapy might also be appropriate to treat R-PH, particularly in severe cases, but this concept has yet to be accepted as the standard care worldwide. In the future, when the Japanese Respiratory Society (JRS) provides the medical community with theoretical and practical information on the management of patients with R-PH, the importance of using RHC to assess the pathophysiology of the disease should be emphasized. To assess the frequency and time point at which Japanese pulmonologists use RHC for R-PH diagnosis, as well as to investigate current treatment modalities for this condition, we conducted a research survey of specialized medical institutions ( $n=695$ ) and educational facilities ( $n=160$ ) approved by the JRS.

## 2. Methods

### 2.1. Questionnaires

Questionnaires were mailed to 855 institutions approved by the JRS in July 2012. Participants were asked about the prevalence and necessity of using RHC to diagnose R-PH, and about treatments used for the disease (Table 1a and b). We defined borderline PH, PH, and severe PH by mean pulmonary arterial pressures (mPAP) of 20–25 mmHg,  $\geq 25$  mmHg, and  $\geq 35$  mmHg, respectively.

### 2.2. Ethical consideration

By Japanese legislation, informed consent is not required to collect retrospective data. However, in this study, patients identities were concealed, and it complied according to the requirements of the Japanese Ministry of Health, Labor and Welfare, which is dedicated to privacy, information technology, and civil rights. The protocol was approved by the JRS board of directors in March 2012 and by the Research Ethics Committee of Chiba University School of Medicine (approval number 110; February 20, 2013).

### 2.3. Statistical method

Group differences were analyzed by a Chi-square test. A  $p$  value of  $<0.05$  was considered significant.

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