Case Report

# Underrated value of repeated right heart catheterization in pulmonary hypertension with heart failure—a case of persisted pulmonary arterial hypertension after treatment for biventricular failure

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**Abstract:** Pulmonary hypertension (PH) is a common complication of left heart disease and its presence in patients with heart failure predicts worse clinical outcomes. Specific agents targeting pulmonary arterial hypertension (PAH) have been developed over the last few years, but the efficacy of these agents in pulmonary hypertension due to left heart disease (PH-LHD) is uncertain. We report a case of idiopathic pulmonary arterial hypertension (IPAH) initially presented with biventricular failure, which was misdiagnosed as PH-LHD. A 31-year-old man who had a history of recurrent hemoptysis was referred to our center with biventricular failure. Right heart catheterization (RHC) showed elevated mean pulmonary arterial pressure (mPAP) and pulmonary capillary wedge pressure (PCWP). He was diagnosed as having PH-LHD, specifically combined post-capillary and precapillary PH (CpcPH). We treated him for 2 years with diuretics, a beta blocker, an angiotensin-converting enzyme (ACE) inhibitor, and sildenafil, which was added to treat CpcPH. A follow-up echocardiography showed that biventricular function had improved, but not PH. A second RHC revealed elevated mPAP and normal PCWP, which made us change the diagnosis to IPAH. In conclusion, it is important to perform repeated RHC in CpcPH patients after the improvement of left heart dysfunction to distinguish CpcPH from IPAH.

**Keywords:** Cardiac catheterization; idiopathic pulmonary arterial hypertension (IPAH); heart failure; pulmonary hypertension (PH)

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## Introduction

Left heart disease is accountable for about two thirds of pulmonary hypertension (PH) and PH in patients with congestive heart failure has negative impact on the survival (1). Pulmonary hypertension due to left heart disease (PH-LHD) is defined as a mean pulmonary arterial pressure (mPAP) ≥25 mmHg at rest and a pulmonary arterial wedge pressure (PAWP) >15 mmHg measured by right heart catheterization (RHC), which is essential for the evaluation of PH (2). A new clinical classification of PH-LHD was established on the 5th World Symposium on PH in Nice 2013 and was defined as isolated post-capillary PH (IpcPH) and combined post-capillary and precapillary PH (CpcPH) (3). CpcPH is hemodynamically defined as a mPAP >15 mmHg with a diastolic pressure difference (DPD = diastolic PAP – PAWP) ≥7 mmHg.

The pathophysiology of PH-LHD is not yet fully understood, but considering its negative impact on patient prognosis and the current lack of an optimal treatment protocol, new and more effective treatment modalities are required. Over the past years, specific agents targeting pulmonary arterial hypertension (PAH) have been

**Table 1** Comparison of echocardiographic parameters at admission with those after 2 years of treatment

| Echocardiographic parameters | Before treatment | After treatment |
|---|---|---|
| LV EF (%) | 26 | 65 |
| LV diastolic dimension (mm) | 60 | 44 |
| RV diastolic dimension (mm) | 38 | 29 |
| mPAP[†] (mmHg) | 34.9 | 30.4 |
| Peak systolic tissue velocity of tricuspid annulus (cm/sec) | 6.5 | 15.2 |

After 2-year treatment period, mPAP estimated by echocardiography is still elevated whereas the left ventricular function was restored (right column). [†], mPAP =79-0.45 (acceleration time at pulmonic valve). EF, ejection fraction; LV, left ventricle; RV, right ventricle; mPAP, mean pulmonary arterial pressure.

**Table 2** Changes in the parameters of RHC at admission and those after 2 years of treatment

| RHC parameters | Before treatment | After treatment |
|---|---|---|
| RAP (systolic/diastolic/mean, mmHg) | 27/22/24 | 11/8/9 |
| RVP (systolic/diastolic/mean, mmHg) | 127/12/27 | 84/5/12 |
| PAP (systolic/diastolic/mean, mmHg) | 135/73/93 | 89/43/61 |
| PCWP (mmHg) | 28 | 14 |
| Diastolic pressure gradient (mmHg) | 45 | – |
| Transpulmonary pressure gradient (mmHg) | 65 | 47 |
| Cardiac output[§] (L/min) | 2.84 | 3.1 |
| PVR (Wood units) | 22.89 | 8.15 |
| Pulmonary arterial vasoreactivity | – | Negative |

The second RHC shows elevated PAP with improved PCWP, and it confirms the diagnosis of idiopathic pulmonary arterial hypertension (IPAH) (right column). [§], cardiac output was measured using the indirect Fick method. PAP, pulmonary arterial pressure; PCWP, pulmonary capillary wedge pressure; PVR, pulmonary vascular resistance; RAP, right atrial pressure; RVP, right ventricular pressure; RHC, right heart catheterization.

developed, and several clinical trials have been conducted to measure the effect of PAH targeting agents on PH-LHD, but the results have been conflicting.

## Case report

A 31-year-old Asian man was referred to our center with hemoptysis and biventricular failure. He reported having some hemoptysis over a 2-year period previously and recently experienced two episodes of hemoptysis. He denied having symptoms such as dyspnea on exertion, fatigue, dizziness, or syncope, but confirmed having epilepsy for 23 years, which was well controlled with anti-epileptic medications, and had no history of alcohol abuse or illicit drug use, or family members or relatives with heart failure. His blood pressure was 126/88 mmHg and it remained within the normal range throughout the admission period. Cardiac enzymes and thyroid function tests were normal. Anti-nuclear antibody, rheumatoid factor, and anti-neutrophil cytoplasmic antibody tests were negative.

A chest CT scan showed bilateral enlargement of the main pulmonary arteries and diffuse ground glass opacities on both lungs, possibly resulting from aspirated blood. There was no evidence of thromboembolism or pulmonary vascular malformation. Echocardiography showed an enlarged left ventricle (LV) with decreased LV systolic function and the LV ejection fraction (EF) was 26%. The E/e' ratio was 18, indicating high LV filling pressure. The right ventricle (RV) was also dilated and systolic dysfunction was observed (*Table 1*). The heart valves were normal with respect to morphology and function. He experienced massive hemoptysis after admission and underwent bilateral bronchial arterial embolization.

RHC showed elevated mPAP and PAWP values, with a DPD of 45 mmHg (*Table 2*). A lung perfusion scan did not show any perfusion defects in either lung. We diagnosed

 

him as PH-LHD, specifically CpcPH, which we consider to have been caused by a dilated cardiomyopathy. He was treated with spironolactone 12.5 mg, bisoprolol 2.5 mg which was gradually increased to 10 mg, and losartan 50 mg which was increased later to 100 mg. Sildenafil, 25 mg 3 times a day, was also used to manage CpcPH because DPD was markedly elevated.

After 2 years of treatment, follow-up echocardiography revealed complete restoration of LV systolic function and the near restoration of RV systolic function, but PH remained elevated. Repeated RHC revealed an elevated mPAP with a normalized PAWP (*Table 2*). The diagnosis was subsequently changed from CpcPH to idiopathic pulmonary arterial hypertension (IPAH).

## Discussion

Cardiac catheterization is the gold standard for the evaluation of PH, despite advances in echocardiographic methods used to estimate PH (4). We initially diagnosed the patient as having CpcPH with a markedly elevated DPD, but we later changed the diagnosis to IPAH after treatment for heart failure and repeated RHC. It is important for CpcPH patients to undergo repeated RHC after restoration of LV dysfunction to eliminate other possible causes of PH, such as IPAH. Repeated RHC is recommended in heart transplant candidates (5), but the value of repeated RHC in CpcPH patients is underappreciated.

Our case report has some limitations. At the time of diagnosis, our RHC protocol did not included measurement of left ventricle end-diastolic pressure (LVEDP), the value could not be provided. Although the patient's RHC results meets the diagnostic criteria of IPAH, the diagnosis of IPAH relying solely on the PAWP may lead to misclassification of PH because there is a discrepancy between the PAWP and LV EDP (6). In this case, a left heart catheterization and additional imaging such as cardiac magnetic resonance (MR) would have provided better grasp of underlying pathology (7).

We added sildenafil to the treatment regimen to treat CpcPH in the current case, but the role of PAH targeting agents on PH-LHD is uncertain. Several studies showed that sildenafil improves exercise capacity and the quality of life in patients with heart failure (8,9). Other PAH targeting agents, such as bosentan, have also been shown to improve exercise capacity and functional status, but have failed to improve mortality (10,11). Another problem is that the optimal cut-off value of DPD for the initiation of therapy with these agents has not been properly studied. In conclusion, the use of PAH targeting agents in CpcPH patients requires further investigations.

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## Footnote

*Conflicts of Interest:* The authors have no conflicts of interest to declare.

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