

Editorial



Current Trends and Movements in Managing Pulmonary Arterial Hypertension in Korea

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Pulmonary hypertension (PH) is a group of diseases characterized by high pressure in the pulmonary arteries, leading to remodeling of the right ventricle. The most common initial symptoms of PH are exertional dyspnea and fatigue. Symptoms related to right heart disease, such as peripheral edema, abdominal pain with hepatic congestion, exertional syncope, or chest pain, may also appear. If left untreated, right ventricular failure progresses and can lead to death. Currently, a mean pulmonary arterial pressure of more than 20 mmHg measured by right heart catheterization at rest is defined as PH.¹⁾ This is different from the prior threshold of 25 mmHg or greater in acknowledgment of patients with a mean pulmonary artery pressure of 21 mmHg to 24 mmHg have a higher risk of hospitalization and mortality compared to 20 mmHg or lower.²⁾ Pulmonary arterial hypertension (PAH) is a 1 of the 5 groups of clinical classifications of PH. Mean pulmonary artery pressure greater than 20 mmHg, pulmonary artery wedge pressure of 15 mmHg or less, and pulmonary vascular resistance of at least 3 Wood units are the criteria for PAH (Group 1 PH).¹⁾ Despite the advances in the management of PAH, delayed diagnosis and the idea of untreatable PAH have made it difficult to treat this condition efficiently, and individuals with PAH continue to have a poor prognosis.

Current PAH treatments target 3 pathways: increasing prostacyclin effects on receptors, stimulating the nitric oxide–cyclic guanosine monophosphate biological pathway and antagonizing the endothelin pathway.³⁾ Randomized trials have demonstrated that initial combination therapies are superior to monotherapy in survival and clinical worsening.⁴⁾ Before the development of PAH-specific therapies, the prognosis of PAH was very poor. 1-year survival was 69%, 5-year survival was only 38%⁵⁾ contrast to 85% and 57%, respectively, with current therapies.⁶⁾ Although new drugs have been developed and survival rates have risen, there may be differences in survival rates between developing and developed countries due to healthcare burden.

To address current trends and movements in the management of PAH in Korea, Jang et al.⁷⁾ analyzed the nationwide database of National Health Insurance Service and specifically designed a 3-component algorithm to overcome the limitations of administrative health claims database and accurately extract PAH patients from the database. Due to a discrepancy between International Classification of Diseases, 10th Revision (ICD-10) and current World

Data Sharing Statement

The data required to reproduce these findings cannot be shared due to article type as it is an editorial.

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Health Organization (WHO) classification of PH, the authors comprehensively included several specific codes of PH and excluded those who had not been prescribed PAH-specific drugs and those who had not undergone RHC at least once.

There has been a remarkable rise in the prevalence (75-fold) and incidence (12-fold) of PAH annually from 2002 to 2018. The 5-year survival of incident PAH patients was 71.8%, which is comparable to those in recent Western registry data (REVEAL and COMPERA). This phenomenon might be attributable to the collaborative efforts of the Korean government and academic societies to advance healthcare policies and academic platforms for rare diseases in Korea.⁸⁾ Interestingly, the survival rate did not differ significantly between those on mono-, dual-, and triple-therapy, although monotherapy had been predominant and less than 30% of the patients received combination therapy in 2018. In fact, current international guidelines for PAH recommend upfront combination therapy for high-risk patients. In 2019, the Korean government approved rapid escalation to combination therapy in poor responders, expecting more improvement in survival rates in the future.

In Korea's current status of PAH, it has become a treatable disease. To increase the survival rate of PAH patients more, preemptive screening for early diagnosis and subsequent aggressive combination therapy are required in the future. In addition, lifestyle modification and better supportive care including exercise training and rehabilitation are also effect, cost-efficient and safe ways to enhance patient's life of quality.⁹⁾ However, there are still just a few alternatives available for the management of PAH, despite the efforts to develop novel therapeutics with a unique mode of action.¹⁰⁾ In this regard, current PAH registries and observational studies in Korea will provide a better knowledge of the prevalence of PAH, its prognosis, and the factors required for favorable outcomes and high quality of life for Korean PAH patients.

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