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Editorial

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A 25-Year Journey in the Fight Against Pulmonary Arterial Hypertension at a Korean Center: What Has Changed and What Is Missing?

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▸ See the article "A Single Center Experience of Pulmonary Arterial Hypertension Management in Korea: A 25-Year Comparative Analysis Following the Introduction of Targeted Therapy" in volume 54 on page 636.

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Pulmonary arterial hypertension (PAH) is a chronic and progressive disease, clinically characterized by an elevation in pulmonary arterial pressure. If left untreated, this elevation in pressure can result in right heart failure and ultimately lead to increased mortality. Historically, PAH has been associated with a poor prognosis due to the limitation of available treatment options. However, the advent of targeted therapies in the early 2000s marked a pivotal transformation in the treatment landscape, significantly extending survival and instilling a renewed sense of hope among both patients and healthcare professionals.[1-3]

In the latest issue of the *Korean Circulation Journal*, Cha et al.[4] demonstrated the shift in treatment approaches through a single-center registry in Korea. They analyzed trends in PAH diagnosis, treatment patterns, and survival outcomes from 1980 to 2021. The study's findings are of particular importance, notably regarding the influence of targeted therapies and the advantages of prompt intervention regarding patient survival rates. The study offers valuable insights into how these advancements have redefined PAH management and provides a framework for understanding how timely therapy initiation can dramatically improve patient outcomes.

A significant finding is the considerable advancement in the management of PAH. The largest subgroup was PAH associated with congenital heart disease (CHD-PAH), accounting for 43% of cases, followed by connective tissue disease-associated PAH (CTD-PAH) and idiopathic PAH (IPAH). It is noteworthy that the proportion of CTD-PAH patients increased markedly after 2007, coinciding with a significant rise in the use of targeted therapies (95.4%). This shift toward early and aggressive treatment represents a pivotal turning point in PAH management, offering a more optimistic outlook for the future. The survival rates at 1, 5, and 10 years were 91.3%, 77.4%, and 65.8%, respectively. Patients with CHD-PAH exhibited superior survival compared to those with IPAH or CTD-PAH, underscoring the disparate prognoses across PAH subtypes.

One of the most significant findings of the study is the survival benefit associated with the early initiation of targeted therapy. The age-sex matching analysis revealed a notable

Conflict of Interest
The authors have no financial conflicts of interest.

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The data generated in this study is available from the corresponding author upon reasonable request.

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discrepancy in survival rates between patients who received immediate targeted therapy at diagnosis and those who were managed conservatively. This was particularly evident in patients with IPAH, emphasizing the critical importance of prompt therapeutic intervention in enhancing long-term outcomes for PAH patients.

The most recent guidelines for the treatment of PAH recommend a proactive approach that employs medications targeting 4 key pathways: endothelin-1, nitric oxide, prostacyclin, and bone morphogenetic protein/activin signaling. These agents are typically included in the initial combination therapy regimens. When combined with early and regular reassessment, this approach has the potential to significantly improve outcomes.[5)6)]

Notwithstanding these advances, a study by Jang et al.[7)] revealed that in 2018, less than 30% of PAH patients in Korea received combination therapy, despite its recommendation in international guidelines. Monotherapy remained the predominant approach, with survival rates among patients on mono-, dual-, and triple-therapy being comparable. This lack of a survival benefit from combination therapy may be indicative of delayed treatment initiation or a tendency to reserve more intensive therapies for patients with advanced disease.[7)]

The study conducted by Cha et al.[4)] is based on a single-center registry, so it may not fully represent the situation across Korea. However, it still offers important insights into the development of PAH treatment in Korea and its life-saving impact on patient outcomes. The introduction of targeted therapies in 2007 marked a significant turning point, leading to improved survival, especially for those who began treatment early. These findings highlight the necessity for prompt diagnosis and aggressive treatment initiation, particularly for IPAH patients, to optimize long-term outcomes.

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