

# Therapeutic Approach for PAH and CTEPH in Korea



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### Educational background

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### Professional experience

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Pulmonary arterial hypertension (PAH) and chronic thromboembolic pulmonary hypertension (CTEPH) are rare but progressive pulmonary vascular diseases. After the 7th World Symposium on Pulmonary Hypertension, current approaches emphasize the roles of early and comprehensive risk stratification using multiparametric tools, multidisciplinary approach and the proactive establishment of therapeutic strategies. In Korea, the pulmonary hypertension treatment has rapidly evolved, yet significant challenges remain, including the limited availability of therapeutic agents and restrictions on upfront combination therapy.

For PAH, upfront combination therapy with an endothelin receptor antagonist and a phosphodiesterase-5 inhibitor is firstly recommended for low-to-intermediate risk patients, while rapid escalation by adding prostacyclin pathway agents is required if treatment goals are not achieved. High-risk patients are recommended to receive upfront triple therapy including parenteral prostacyclin therapy without delay. However, owing to reimbursement limitations in Korea, there are considerable difficulties in implementing treatments as recommended by international guidelines. Recently, riociguat has been reimbursed for patients with pulmonary arterial hypertension, providing an additional therapeutic option. Recently, novel agents targeting the activin signaling pathway, such as sotatercept, have demonstrated promising efficacy and are expected to expand therapeutic options in Korea.

Pulmonary endarterectomy is considered as the standard treatment for operable CTEPH patients, offering the potential for cure. In patients deemed inoperable, or with residual or recurrent pulmonary hypertension following surgery, balloon pulmonary angioplasty is actively implanted. Although the use of riociguat is permitted, it is not covered by insurance benefits.

In Korea, substantial progress has been made in the diagnosis, evaluation and treatment of PAH and CTEPH. Nonetheless, limitations in available resources and policy-related barriers remain. Given the rapid progress in the treatment of PAH and CTEPH, continued efforts are required to ensure that evidence-based practices are implemented in clinical settings.