ポスター発表 | IPAH

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ポスター発表（II-P02-2）
IPAH

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[II-P02-2-02] Management of Severe Pediatric Pulmonary Arterial Hypertension: Initiating Targeted Therapy Prior to Diagnostic Right Heart Catheterization

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キーワード：pediatric PAH、Right Heart Catheterization、Treatment

Objective: This study aims to proposes a novel management strategy for severe PAH patients: initiating aggressive targeted therapy first and postponing RHC until the patient reaches a low-risk state. Methods: We retrospectively analyzed severe PAH patients who initially presented in a high-risk state. Instead of undergoing immediate RHC, these patients received intensive targeted therapy, once their clinical status improved to a low-risk state, they underwent RHC for further evaluation. Clinical parameters before and after RHC were collected.ResultsThis study included 13 patients, with a median age of 7 (4.4, 11.9) years at the time of their first visit. At the initial visit, all patients were in a clinical high-risk state and were immediately treated with aggressive targeted therapy, including treprostinil. The median time from the initiation of treatment to right heart catheterization was 52 (12, 426) days. Upon undergoing right heart catheterization, patients showed a significant decrease in nt-proBNP levels. Seven patients received intravenous anesthetics during the procedure, and no patient experienced a pulmonary arterial hypertension (PAH) crisis during the catheterization. The results indicated that pulmonary artery pressures and pulmonary vascular resistance were significantly elevated in these patints, with one patient testing positive for the acute vasoreativity test.ConclusionOur findings suggest that delaying RHC until after targeted therapy stabilization is a safe and effective strategy for managing severe PAH.