ポスター発表 | IPAH

2025年7月11日(金) 15:00 ~ 16:00 ポスター会場（文化会館棟 2F 第1・2ギャラリー）2

ポスター発表（II-P02-2）
IPAH

座長：永井 礼子（北海道大学病院 小児科）
座長：中山 智孝（高知赤十字病院 小児科）

[II-P02-2-03] Experience of Transition from subcutaneous treprostinil to oral selexipag in pediatric PAH patients

○Yuan He, Qiangqiang Li, Chen Zhang, Hong Gu (Department of Pediatric Cardiology, Beijing Anzhen Hospital, China)
キーワード：pediatric PAH、pulmonary arterial hypertension、genetic testing

Objective: This study aims to evaluate the safety and efficacy of transitioning from subcutaneous treprostinil to oral selexipag in pediatric PAH patients.Methods: A retrospective study was conducted. At initial visits, all patients were evaluated in clinical high-risk status and started on PAH-targeted therapies including treprostinil. Upon clinical reassessment and transition to a low-risk status, all patients discontinued treprostinil and switched to oral selexipag.Results: A total of 8 pediatric patients were included, with a median age of 14.1 (10.9, 17.5) years. All 8 patients were diagnosed with idiopathic or heritable PAH, with 6 patients carried PAH-related genes. At the initial visits, all patients were classified as WHO-FC III-IV. After treatment, all patients' clinical symptoms improved, and all reached a low-risk clinical status . The drug transition involved tapering treprostinil dose while increasing the oral selexipag dose, with the entire process completed on an outpatient basis. The final dose of oral selexipag ranged from 0.8 to 1.6 mg. During follow-up after the drug transition (median follow-up time of 25 (10, 35) months), no patient died. However, three patients got clinically deteriorated and reused treprostinil, who were all PAH-related genes carriers (BMPR2 n=2, KCNA5 n=1). The other five patients were maintained low-risk status.Conclusion: For pediatric PAH patients with genetic mutations, transitioning from treprostinil to oral selexipag is risky, even if the patient has improved to a low-risk status.

© 2025 Japanese Society of Pediatric Cardiology and Cardiac Surgery