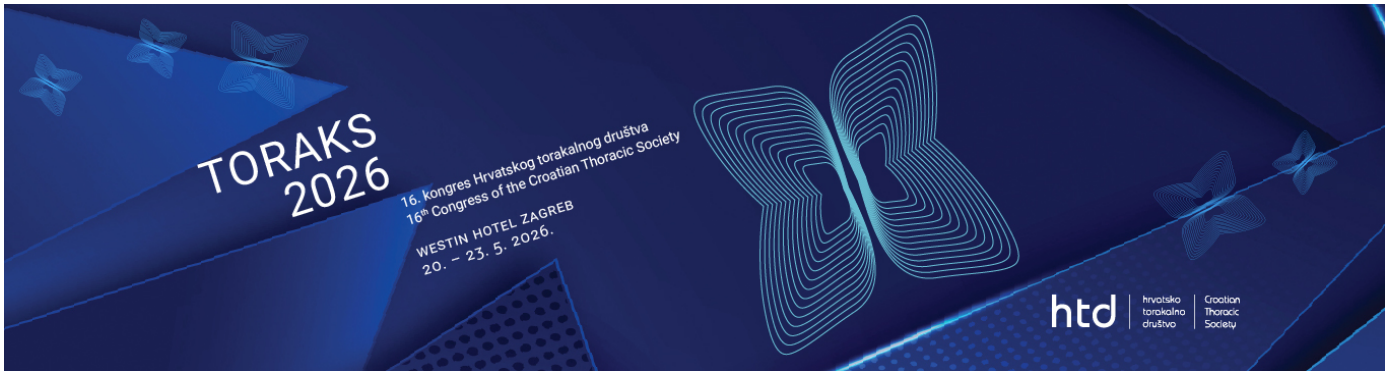


REAL-WORLD EXPERIENCE WITH SOTATERCEPT IN ADVANCED PULMONARY ARTERIAL HYPERTENSION: A SINGLE-CENTER CASE SERIES

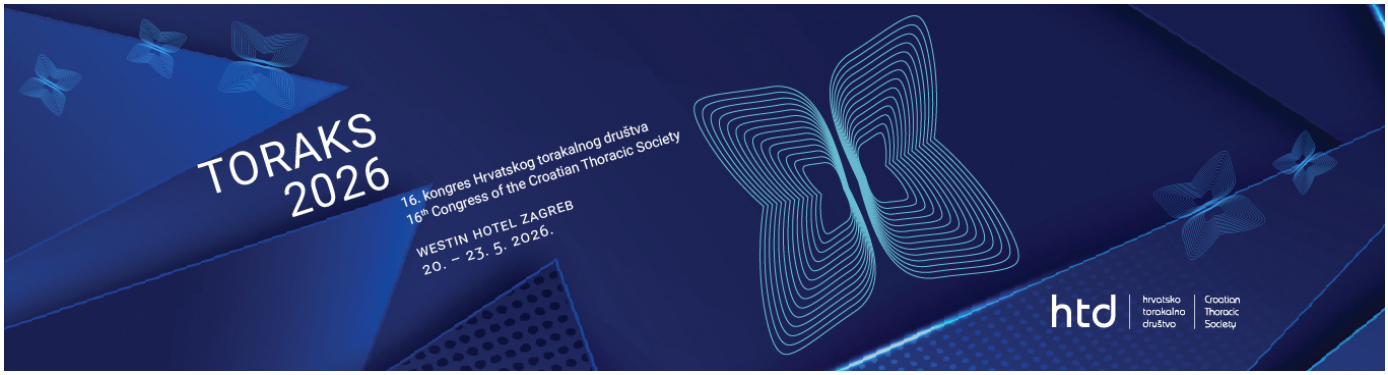
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Objective:

Pulmonary arterial hypertension (PAH) is a progressive disorder characterized by pulmonary vascular remodeling, increased pulmonary vascular resistance and elevated pulmonary arterial pressure, ultimately leading to right ventricular failure. Despite advances in therapy targeting the prostacyclin, endothelin, and nitric oxide pathways, long-term outcomes remain suboptimal. Sotatercept, a first-in-class activin signaling inhibitor, represents a novel therapeutic approach that addresses underlying vascular remodeling rather than vasodilation alone. Clinical trials have demonstrated improvements in hemodynamic and functional parameters with an acceptable safety profile, but real-world data remain limited.



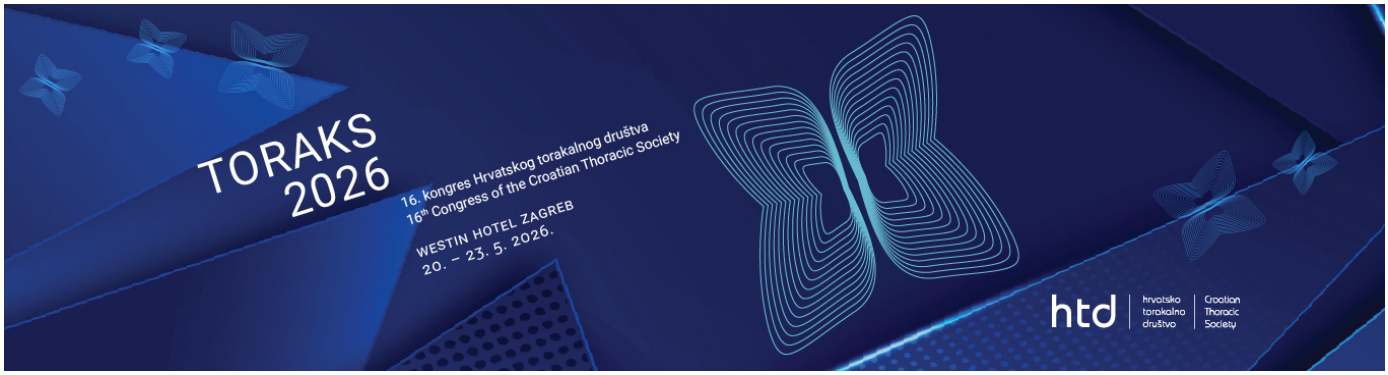
Methods:

This case series describes the initial Croatian experience with sotatercept in six female patients (aged 36–66 years) with advanced PAH of heterogeneous etiology, including idiopathic, connective tissue disease-associated and congenital heart disease-associated PAH. All patients were receiving stable triple therapy but had not achieved low-risk status prior to sotatercept initiation. Standardized assessments, including right heart catheterization, echocardiography, and functional testing, were performed at baseline, 6 months, and 12 months.

Result:

At 6 months, most patients demonstrated favorable responses. Three patients achieved approximately 50% reductions in pulmonary vascular resistance, with concomitant improvements in NT-proBNP levels and exercise capacity. These improvements enabled prostacyclin dose reduction in two cases and removal from the transplant list in one patient. Two additional patients showed moderate hemodynamic improvement, while one exhibited minimal response. At 12 months, sustained benefit was observed in two patients, whereas others showed partial decline compared with 6-month findings, though still improved from baseline.

Treatment was generally well tolerated; however, notable adverse events included erythrocytosis leading to treatment interruption and discontinuation in one patient, and progression of pericardial effusion in two patients. These findings highlight the importance of



careful monitoring, particularly in patients with advanced disease or pre-existing complications.

Conclusion:

Overall, this real-world experience supports clinical trial data, demonstrating that sotatercept can significantly improve hemodynamic and functional outcomes in selected patients with advanced PAH. However, treatment responses were heterogeneous, likely reflecting differences in disease stage and underlying pathology.

Limitations include the small sample size, single-center design, and lack of a control group. Nevertheless, this report provides valuable early insight into the clinical use of sotatercept. Further studies are needed to define optimal patient selection, timing of initiation, and long-term efficacy and safety.