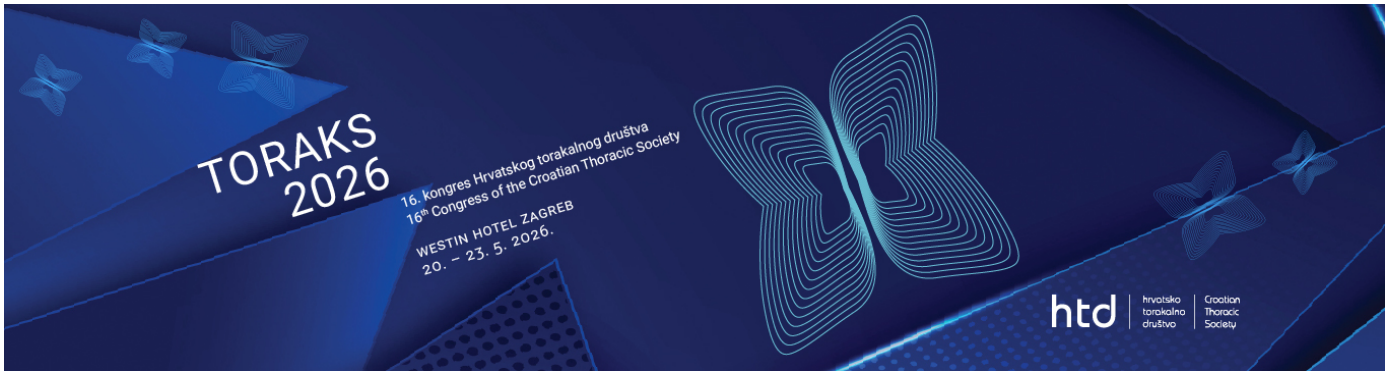

SOTATERCEPT IN THE INTENSIVE CARE UNIT FOR DECOMPENSATED PULMONARY ARTERIAL HYPERTENSION: A CASE REPORT

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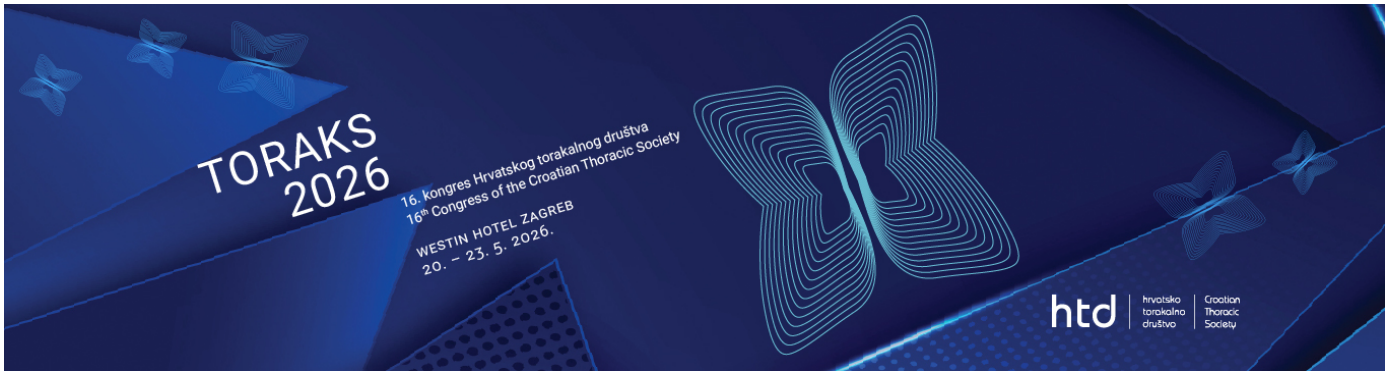
Background:

Pulmonary arterial hypertension (PAH) is a rare, chronic, and progressive disease characterised by pulmonary vascular remodelling and increased pulmonary arterial pressure causing right ventricular overload which can progress to right-side heart failure. Sotaterecept, an activin signalling inhibitor, is used in treatment of advanced PAH, usually as fourth line therapy in patients who do not achieve low risk status on triple therapy. We present a case of significant clinical and haemodynamic improvement in severe PAH and right heart failure following early, upfront initiation of sotatercept in the ICU.

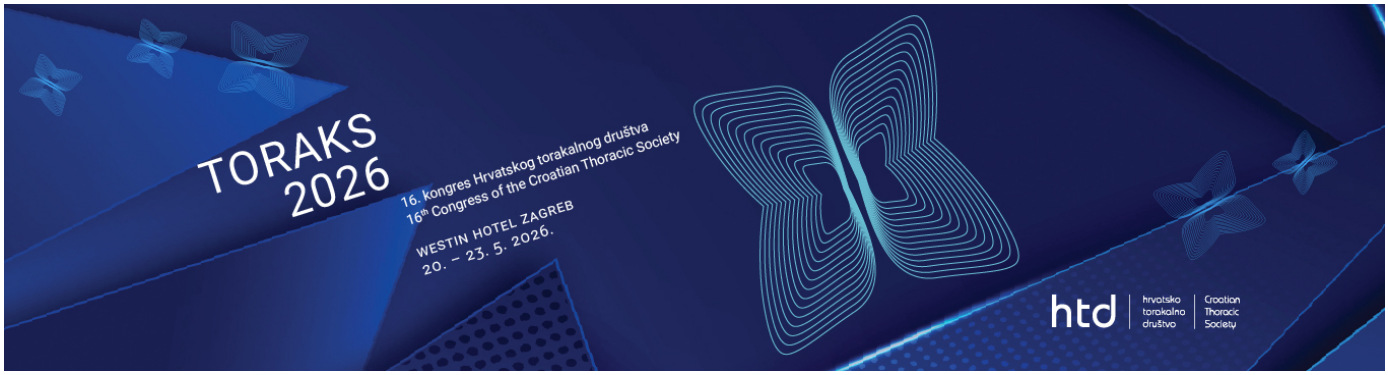


Conclusion:

This case highlights the potential role of sotatercept as a highly effective treatment option for PAH. Despite limited evidence of its early use in PAH patients with right heart failure requiring ICU treatment, sotatercept led to a striking clinical and haemodynamic improvement, suggesting potential expansion of its use beyond current practice.

Case:

A 57-year-old woman was admitted to the ICU due to advanced PAH with right heart failure, cardiogenic shock, anasarca and anuria. Her pulmonary hypertension was classified as WHO Group 1 secondary to connective tissue disease overlap syndrome (rheumatoid arthritis, limited cutaneous systemic sclerosis, Sjogren syndrome) for which she was receiving tocilizumab. Prior to this hospitalisation, the patient had been in follow-up for dyspnea and had been started on sildenafil and macitentan after echocardiography showed right ventricular dilatation and pericardial effusion without haemodynamic compromise. At admission, right heart catheterisation confirmed severe decompensated precapillary



pulmonary hypertension with a cardiac index (CI) of 1.80 L/min/m2, and pulmonary vascular resistance (PVR) of 15 WU, with markedly elevated NT-proBNP levels of 9191 ng/L. Treatment with sotatercept, treprostinil, dobutamine and furosemide was initiated. The clinical course was complicated by the development of chills and rigors. Blood cultures were positive for *Staphylococcus aureus*, and urine cultures for E*scherichia coli*, thus meropenem and cefazolin were introduced, leading to a decline in inflammatory markers. Four weeks after the initiation of sotatercept, repeat right heart catheterization demonstrated significant improvement with a CI of 2.63 L/min/m2 and PVR of 5.5 WU. Following haemodynamic stabilisation, the patient was discharged home on quadruple PAH therapy with sotatercept, treprostinil, sildenafil and macitentan. After 6 months of sotatercept treatment, control right heart catheterization showed further haemodynamic improvement, with a CI of 3.1 L/min/m2 and PVR of 3.8 WU, and normal NT-proBNP level of 60 ng/L.