

Compassionate Use of Inhaled Treprostinil in Acute Right Heart Failure Due to CTEPH

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Background: Inhaled treprostinil (IT) is a prostacyclin analogue indicated to increase walk distance when treating PAH in patients with NYHA functional class III symptoms (Poms & Kingman, 2011). The use of inhaled prostanoid in the ICU is typically relegated to its analog epoprostenol, which can be delivered continuously through a ventilator, whereas inhaled treprostinil requires patient cooperation – a prerequisite not always probable in the critically ill patient, regardless of ventilatory status. Hence, inhaled treprostinil is not a likely candidate for use in the critical care setting. We describe a case where IT was successfully used on a compassionate basis.

Methods: A 50-year-old African American male with a history of Protein C deficiency, deep vein thrombosis, pulmonary emboli, cardiac cirrhosis, ESRD secondary to idiopathic focal segmental glomerulosclerosis, and WHO Group IV - CTEPH, was transferred from a neighboring facility with refractory cardiogenic shock secondary to acute right heart failure. Prior to transfer, his complex course can be summarized as spontaneous bacterial peritonitis, septicemia, and pericardial effusion with drain placement. CTEPH was diagnosed at our institution more than a year prior to this admission (RHC upon initial diagnosis showed mRAP 22, PAP 105/60, MPAP 75, PAWP18, CO/CI 5.24/2.83, PVR 10.8). He was on treatment with Sildenafil. However, unfortunately his follow up was erratic due to multiple hospitalizations. Upon admission to our ICU, he required intermittent non-invasive ventilation, inotropic, and vasopressor support. An initial echo confirmed right-sided heart failure without significant left ventricular involvement (RAP 15; RVSP 115-10 mmHg; LVEF >65%). Sildenafil 40mg TID and hydrocortisone 50mg TID were started, pharmaceutical management was optimized, and continuous renal replacement therapy was initiated for fluid removal. Although, the vasopressor and inotropic requirement decreased with medical management over the course of a few days, cardiogenic shock persisted and was unable to wean off from inotropes and pressors completely. Parenteral prostacyclin therapy was not considered due to overall poor prognosis and complex clinical circumstances. Patient and family fully understood the difficult situation, and he wished for more time with his family. Therefore, IT was started in the ICU on a compassionate basis which resulted in hemodynamic stability quickly, leading to a complete cessation of inotropes and pressors within 48 hrs. He was also switched successfully to intermittent hemodialysis. The patient's respiratory status also improved significantly and was able to transfer out of ICU.

Conclusions: While IT is not typically indicated for patients in Stage IV heart failure, it was effective in rescuing our patient from cardiogenic shock when used on a compassionate basis.

Poms, A. & Kingman, M. (2011). Inhaled Treprostinil for the Treatment of Pulmonary Arterial Hypertension. *American Association of Critical-Care Nurses*, 31(6). doi: 10.4037/ccn2011153



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