

Title:

Diffuse microvascular ischemia after lung transplantation in a patient with pulmonary arterial hypertension.

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Introduction:

Hemodynamic management (HM) during lung transplantation (LTx) for pulmonary arterial hypertension (PAH) is complex. Switching or discontinuation of IV prostanoids entails potential adverse effects. We present a case of critical bilateral limb ischemia in the setting of LTx after abrupt changes of IV prostanoid therapy and its resolution after LTx.

Case:

A 28 yo female with familial PAH initially treated with with Arbisentan, Sildenafil and IV Treprostinil presented with right ventricular (RV) dysfunction. 7 years after diagnosis she was referred for LTx evaluation but rapidly progressing RV failure prompted urgent listing for LTx. IV prostanoids were switched urgently (Treprostinil to Epoprostenol). Deterioration of cardiogenic shock required peripheral VA-ECMO support. Significant O₂ extraction was noted on ECMO with need for inotropes and vasopressors. Worsening central cyanosis and peripheral microvascular malperfusion and critical lower extremity ischemia ensued in absence of peripheral arterial disease. Bilateral sequential LTx was performed on cardiopulmonary bypass and post-transplant conversion to central VA-ECMO. Prostanoids were discontinued during anesthesia due to severe systemic hypotension. Bilateral lower extremity fasciotomies did not immediately result in adequate reperfusion. Despite significant post-operative improvement in vasopressor requirements, all sacral tissues and lower limb musculature remained malperfused, IV Treprostinil was restarted (10th of therapeutic dose) to promote microvasculature re-perfusion. Subsequent inspection showed dramatic resolution within 48 hours. Patient required a tracheostomy with successful discontinuation of ventilation, intense rehab for profound critical illness myopathy and transient replacement therapy for acute renal failure.

Discussion:

Abrupt switch or cessation of IV prostanoids before or after lung transplantation for PAH can lead to microvascular tissue malperfusion. Especially in acute deterioration or in the post-LTx phase weaning of IV prostanoids over several days is recommended.