



# Five-year survival after an acute episode of decompensated pulmonary arterial hypertension in the modern management era of right heart failure

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**Novel medical, instrumental and surgical management of right heart failure translate to improvements in long-term survival of the youngest patients with acute decompensated PAH**

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## To the Editor:

Acute decompensated pulmonary arterial hypertension (PAH) is characterised by rapid worsening of clinical signs of right heart failure (RHF) with subsequent congestion and systemic circulatory insufficiency that can lead to multisystem organ failure [1–3]. Short-term outcomes of acute decompensated RHF are very poor and it remains the primary cause of mortality in PAH [4, 5]. Intensive care of acute decompensated PAH is based on treatment of triggering factors, careful fluid management, and strategies to improve cardiac function and reduce right ventricular afterload [1]. However, this medical strategy is not always sufficient to restore a long-lasting balance between the afterload imposed on the right ventricle and its capacity for compensation. In case of refractory RHF despite maximal medical treatment, the use of mechanical support should now be considered in selected candidates for lung transplantation, or less commonly as a bridge to recovery in patients with a treatable cause of right-sided heart failure [1]. Veno-arterial extracorporeal membrane oxygenation (ECMO) is currently the most widely used strategy to support the right ventricle in PAH patients. This strategy, combined with changes in organ allocation rules to prioritise patients with a short-term life-threatening condition, should contribute to the improved survival of eligible patients with end-stage PAH [6]. However, long-term survival of patients admitted to the intensive care unit (ICU) for severe acute RHF management has not been studied extensively in the modern management era of mechanical support and high-priority lung transplantation.