Lung transplantation in six patients with idiopathic pulmonary artery hypertension

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Background: Idiopathic pulmonary artery hypertension (IPAH) is an incurable and invariably fatal disease. Lung transplantation is a useful therapeutic option in patients who are unresponsive to medical treatment; however, lung transplantation performed for pulmonary hypertension is associated with significantly high perioperative mortality rates.

Methods: We report case series of six patients who underwent lung transplantation for IPAH between October 2008 and June 2021.

Results: Patients' median age was 28.5 years, and the study included 5 of 6 female (83%). Pre-transplantation hemodynamic parameters showed mean right atrial pressure of 12.0±7.1 mmHg and mean pulmonary artery pressure of 62.2±29.5 mmHg. Two of six patients received extracorporeal membrane oxygenation (ECMO) therapy as a bridge to transplantation over 14 and 17 days, and four patients underwent elective transplantation. Two patients required prolonged postoperative venoarterial (VA) ECMO support. Grade 3 primary graft dysfunction occurred in one patient; however, the clinical course improved following prolonged VA ECMO therapy. Five patients (83.3%) required intervention for postoperative bleeding control; one of these patients died of uncontrolled bleeding concomitant with disseminated intravascular coagulation, on the 14th postoperative day, and we observed no other perioperative deaths. One patient died of carbapenem-resistant *Acinetobacter baumannii* bacteremia, a year postoperatively. The 1-month, 6-month, and 1-year survival rates were 83.3%, 83.3%, and 66.7%, respectively.

Conclusions: In view of the poor prognosis of IPAH, lung transplantation (1-year mortality rates <40%) merits consideration as a useful therapeutic option in this patient population. However, postoperative bleeding tends to occur in most patients; therefore, close monitoring is important during post-transplantation management.

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