Management of portopulmonary hypertension and hepatopulmonary syndrome in patients undergoing liver transplantation

Hepatopulmonary syndrome (HPS) and portopulmonary hypertension (POPH) are pulmonary vascular complications of portal hypertension with or without cirrhosis. The prevalence among liver transplant candidates is roughly 5-32% for HPS and approximately 6% for POPH. Although these two conditions may initially present with dyspnea and are pathologically linked by the presence of portal hypertension, their pathophysiologic mechanisms are significantly different. HPS is characterized by low pulmonary vascular resistance secondary to intrapulmonary vascular dilatations and hypoxemia; on the other hand, POPH features elevated pulmonary vascular resistance and constriction of the pulmonary vasculature. Medical treatment for HPS has been disappointing overall. POPH patients can be treated with pulmonary artery-specific vasodilatory therapy. Whereas liver transplantation (LT) results in the resolution of HPS and is an indication per se for LT, its effect on POPH is highly unpredictable. LT poses a high risk of death in those with significant POPH, where pulmonary artery-specific vasodilatory therapy may improve functional status and allow successful LT in a small number of select patients. Modern strategies in managing HPS and POPH rely on a thorough screening and grading of the disease's severity, in order to tailor the appropriate therapy and select only the patients who will benefit from LT.