ECMO after Liver Transplantation in Pediatric Patient with Hepatopulmonary Syndrome

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Introduction

Hepatopulmonary syndrome (HPS) is a severe but common disease characterized by decreased arterial oxygenation in the presence of intrapulmonary vascular dilations (IPVD) in the setting of hepatic disease. Current understanding of HPS is limited and there is no effective medical treatment available. Liver transplantation (LT) is the only therapy capable of reversing the condition and improving survival rates. In this case, we present a pediatric patient with very severe HPS whose post-LT course was complicated by progressive hypoxemia, requiring extracorporeal membrane oxygenation (ECMO). To our knowledge, this is only the second case report of a pediatric patient with HPS requiring ECMO after liver transplantation.

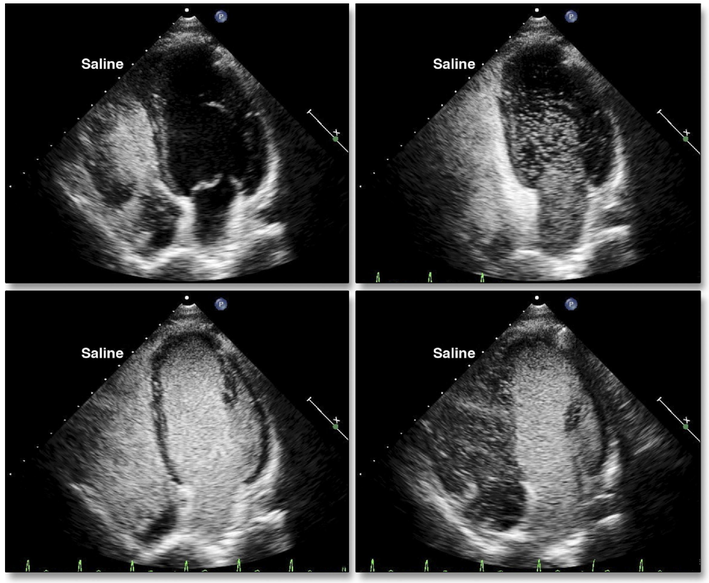

Figure 1. Saline microbubble transthoracic echocardiography (MTTE) in ESLD patient with severe HPS [5].

Case Presentation

Our patient is a 13-year-old male with idiopathic non-cirrhotic hepatoportal sclerosis, portal hypertension and HPS. He presented with platypnea, cyanosis and orthodeoxia. His PaO2 on room air was 37 mmHg, with an alveolar-arterial gradient of 65. At rest he required 15 L/min O2 via mask with intermittent need for BiPAP. Cardiac catheterization revealed arterial-venous malformations resulting in severe intrapulmonary shunting. He subsequently underwent an uneventful LT. Postoperatively he experienced progressive hypoxemia, complicated by bilateral pneumothoraces and multiple reintubations. Medical interventions included aggressive pulmonary toilet, chemical paralysis, proning, inhaled nitric oxide, and a sildenafil infusion, all resulting in minimal improvements in oxygenation. He was eventually placed on veno-venous (VV) ECMO, which he required for three months as his pulmonary function slowly improved. He was decannulated and discharged home one month later on room air.

Discussion

HPS is a common, yet severe pulmonary vascular complication of liver disease. LT is the only effective therapy for reversal of HPS. As a perioperative clinician, it is vital to understand the diagnosis, staging and prognosis of the disease, as well as the various treatment options available for treating HPS-related hypoxemia following LT.

HPS can present with progressive dyspnea, cyanosis, fatigue, orthodeoxia, platypnea and spider nevi [1]. The diagnostic triad includes liver disease, an elevated A-a gradient on room air (>15 mmHg) and IPVD. Microbubble transthoracic echocardiography (MTTE) is the diagnostic gold standard [2,5]. For prognosis and the timing of LT, HPS is classified by stage: mild (PaO2 ≥ 80 mmHg), moderate (PaO2 60 to < 80 mmHg), severe (PaO2 50 to < 60 mmHg), and very severe (PaO2 < 50 mmHg) [1]. Patients with severe HPS have an increased risk of death after LT. Three year post-LT survival is 84% for patients with PaO2 of 44.1-54.0 mm Hg vs 68% for those with PaO2 ≤ 44.0 mm Hg [2].

Reversal of HPS after LT usually occurs within one year [3]. Treatment of hypoxemia in these post-LT patients before HPS resolution is difficult, and recommendations are derived from case reports and expert opinion. Treatment algorithms have been proposed, which focus on decreasing right-to-left shunting via IPVD and include: Trendelenburg position, phosphodiesterase inhibitors, inhaled nitric oxide and/or epoprostenol, methylene blue, embolization of IPVD, and ultimately ECMO [4]. More prospective studies are needed to optimize treatment strategies.

References

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