

Perioperative Anesthetic Management of Living Donor Liver Transplantation in a Child with Extrahepatic Portal Vein Obstruction & Hepatopulmonary Syndrome: A Case Report

Submitted: 05 Sep 2026

Revised: 14 Sep 2026

Accepted: 20 Sep 2026

Published: 30 Sep 2026

Authors:

Vijay Shankar, (ORCID ID: 0000-0002-6446-9030), Consultant Anaesthetist, Medical Trust Hospital – South Block, Cochin.

Tista Sarkar, (ORCID ID: 0009-0009-3403-9740), Fellow in Transplant Anesthesia, Indraprastha Apollo Hospital, New Delhi.

Access This Article Online



Website:
theanaesthesiologist.com

Corresponding Author:

Vijay Shankar, Consultant Anaesthetist, Medical Trust Hospital – South Block, Cochin.
vijaysnair95@gmail.com

How to cite this article: Vijay Shankar, Tista Sarkar. Perioperative Anesthetic Management of Living Donor Liver Transplantation in a Child with Extrahepatic Portal Vein Obstruction & Hepatopulmonary Syndrome: A Case Report. TAISAK 2026; 2 (2): 102-105

ABSTRACT

Hepatopulmonary syndrome (HPS) in children with extrahepatic portal vein obstruction (EHPVO) is a rare but life-threatening complication associated with severe hypoxemia and elevated perioperative risk during liver transplantation. We report the perioperative anesthetic management of a 6-year-old male with EHPVO complicated by HPS, presenting with room air oxygen saturation of 77–79%, who underwent living donor liver transplantation (LDLT). An integrative approach incorporating meticulous oxygenation optimization, invasive hemodynamic monitoring, targeted reperfusion management, and planned postoperative mechanical ventilation resulted in successful extubation on postoperative day 1 and intensive care unit discharge on day 6 undergoing Modified Radical Mastectomy.

Keywords: Extrahepatic portal vein obstruction, Hepatopulmonary syndrome, Liver transplantation, Pulmonary arteriovenous malformation

Introduction

The clinical triad of portal hypertension, intrapulmonary vascular dilatation, and arterial hypoxemia defines hepatopulmonary syndrome (HPS). While recognized as a complication of chronic liver

disease, HPS in the setting of extrahepatic portal vein obstruction (EHPVO) is exceedingly rare in children[1]. EHPVO, a primarily vascular condition without intrinsic hepatocellular dysfunction, can nonetheless lead to portal hypertension and, in uncommon cases, HPS with severe hypoxemia. The

concurrent presence of pulmonary arteriovenous malformation (PAVM) further complicates the pathophysiology and substantially increases anesthetic risk [2]. Liver transplantation is the only definitive cure for HPS; however, perioperative management in this rare pediatric subset presents unique and formidable challenges. We report our experience with the perioperative anesthetic management of a child with EHPVO & HPS undergoing living donor liver transplantation (LDLT). Informed parental consent was obtained for publication of this case report.

Case Description

The patient is a 6-year-old male child weighing 17 kgs and with a height of 118 cm who is undergoing LDLT in view of End-Stage Liver Disease (ESLD) due to EHPVO and complicated by HPS. He initially presented with platypnea, orthodeoxia, cyanosis during crying episodes, and progressive peripheral cyanosis involving the fingertips. He subsequently developed portal hypertension, hepatosplenomegaly, and jaundice, consistent with chronic liver disease complicated by HPS.

Preoperative evaluation included saline contrast echocardiography, which was positive within 4–5 cardiac cycles, suggesting PAVM. Pulmonary angiography demonstrated adequate opacification of the pulmonary artery and intraparenchymal arterial branches. A technetium-99 m macroaggregated albumin (MAA) scan revealed a shunt fraction of 11%. Computed tomography of the liver demonstrated intrahepatic biliary radicle dilatation with portal biliopathy, hepatomegaly, splenomegaly, and a dilated splenic vein.

Disease progression was marked by worsening cyanosis, platypnea, mild dyspnea, orthodeoxia, and arterial oxygen saturation (SaO₂) of 79% on room air, with a P/F ratio of 190, necessitating liver transplantation. Pulmonary function tests and chest radiography were normal. Electrocardiography revealed normal sinus rhythm, and transthoracic

gas analysis in supine and sitting positions confirmed orthodeoxia. Preoperative hemoglobin, hematocrit, platelet count, and liver function tests were within acceptable limits. The Child–Pugh score was B, and the Pediatric End-Stage Liver Disease (PELD) score was 6.9.

The patient was classified as ASA physical status III with Mallampati Grade II airway. Following informed written parental consent, the patient was transferred to the operating room. Pre-induction monitors, including electrocardiogram, non-invasive blood pressure, and pulse oximetry, were attached. After preoxygenation with FiO₂ >90% for 3 minutes, anesthesia was induced with propofol 2 mg/kg, fentanyl 2 µg/kg, and atracurium 0.5 mg/kg. A 4.5 mm cuffed endotracheal tube was placed. Pressure-controlled mechanical ventilation was initiated targeting SpO₂ of 88–92%, with inspiratory pressure 12 cmH₂O, respiratory rate 22 breaths/min, FiO₂ 50%, and PEEP 8 cmH₂O.

Invasive monitoring was established via a left radial arterial catheter for blood sampling, a right femoral arterial catheter for continuous hemodynamic monitoring, and a right internal jugular venous catheter. Anesthesia was maintained with atracurium infusion at 0.05 mg/kg/hr, dexmedetomidine at 0.8 mcg/kg/hour, and fentanyl at 1 mcg/kg/hour, along with 50 % oxygen and 50 % air. We used bispectral index monitoring throughout and titrated the agents to maintain the value between 40 and 60.

Given the inadequate oxygenation, we maintained restrictive fluid management. We used ½ DNS at 4 ml/kg/hour and 5% albumin at 6 ml/kg/hour. We accurately measured blood loss and replaced it with 600 mL of packed red cells. The anhepatic phase was characterized by a wet surgical field and a hypocoagulable thromboelastogram (TEG). This was corrected with fresh-frozen plasma at 10 mL/kg

Reperfusion was uneventful, requiring phenylephrine at 2 mcg/kg and 100 mL of 5% albumin. The implanted graft weighed 259 gm, yielding a graft-to-recipient body weight ratio (GRWR) of 1.52%. The surgery lasted 10 hours and required 800 mL of 5% albumin, 500 mL of ½ DNS, 600 mL of packed cells, and 200 mL of fresh-frozen plasma. Given the severity of postoperative hypoxia, it was decided to ventilate the patient electively. Sedation was achieved with dexmedetomidine at 0.25 mcg/kg/hour. Mechanical ventilation was continued with a tidal volume of 150 mL, rate of 20, FiO₂ of 0.5, and PEEP of 6.

Postoperative fluid management included ½ DNS at 44 mL/hour and 20% Human Albumin at 5 mL/hour. The drains were inspected hourly and replaced hourly using normal saline. Urine output was adequate (consistently >0.5 mL/kg/hr) & hemodynamic stability was maintained. Ventilatory support was gradually weaned, maintaining an oxygen saturation between 90% and 92%. The patient was extubated uneventfully on postoperative day 1. He was transferred from the intensive care unit on postoperative day 6 following an otherwise uncomplicated postoperative course.

Discussion

The epidemiology of HPS in children is distinctly different and historically underrecognized. Earlier estimates suggested HPS was rare in children; however, with the adoption of ERS/ILTS diagnostic criteria and routine arterial blood gas screening, the prevalence in cirrhotic children has been reported to be as high as 42.5 %⁴. Unlike hypoxemia, which is amenable to conventional oxygen therapy, the shunt physiology in HPS renders the conventional efforts to achieve normoxia largely ineffective. We knew that aggressive efforts to achieve normoxia may be largely ineffective and, in fact, be deleterious, leading to complications such as barotrauma and hemodynamic instability. In this context, we adopted a deliberate strategy of permissive hypoxemia, accepting an arterial oxygen saturation (SpO₂) of

of 88–90%. This approach is well supported in the literature, acknowledging that supranormal FiO₂ and escalating ventilator pressures confer little benefit in the setting of fixed intrapulmonary shunts while introducing significant barotraumatic and oxidative risks^[5]. Targeting this saturation range ensured adequate end-organ oxygen delivery while avoiding the hazards of overzealous respiratory intervention in a physiologically compromised pediatric patient.

A restrictive fluid strategy was maintained throughout all three phases of transplantation to mitigate the risk of fluid overload, which can exacerbate intrapulmonary shunting, compromise respiratory mechanics, and precipitate right ventricular strain. Total Intravenous Anesthesia was preferred over volatile agents due to its minimal effects on hypoxic pulmonary vasoconstriction and shunt fraction. Blood transfusion was strictly guided by TEG and inspection of the surgical field, which further helped us restrict overall fluid intake. We continued the intraoperative ventilatory and fluid strategy in the post-operative period as well.

The patient's successful extubation on postoperative day (POD) 1 is a noteworthy outcome that underscores the effectiveness of the described intraoperative strategy. Early extubation after pediatric OLT is associated with lower rates of ventilator-associated pneumonia, shorter intensive care unit stays, improved patient comfort, and earlier enteral nutrition—all of which are particularly consequential in the pediatric context. The resolution of HPS following liver transplantation is well-documented, though the timeline varies; intrapulmonary vascular dilatations typically regress over weeks to months as portal hypertension resolves and hepatic synthetic function is restored. Early extubation in this case reflects the cumulative benefit of lung-protective ventilation, judicious fluid management, and stable hemodynamics achieved through TIVA—

collectively minimizing postoperative respiratory burden.

Conclusion

Hepatopulmonary syndrome is not uncommon in pediatric liver transplant and can pose significant perioperative challenges during liver transplantation. This case reinforces that a protocolized, pathophysiology-driven anesthetic approach — accepting permissive hypoxemia, limiting fluid accumulation, and preserving pulmonary mechanics — can yield favorable short-term outcomes even in pediatric HPS patients presenting with significant preoperative hypoxemia.

Conflict of interest : NIL

Financial support : NIL

Reference

1. Gupta D, Hussain M, Soin AS, Sahi B, Eapen CE, Nundy S. Prevalence of hepatopulmonary syndrome in cirrhosis and extrahepatic portal venous obstruction. *Am J Gastroenterol*. 2001;96(12):3395-3399.
2. Fauconnet P, Klopfenstein CE, Schiffer E. Hepatopulmonary syndrome: the anesthetic considerations. *Eur J Anaesthesiol*. 2013 Dec;30(12):721-30. doi: 10.1097/EJA.0b013e328365bb6f. PMID: 24161936.
3. Lee HJ, Lee JM, Jung CW, Lee J. Anesthetic management of a pediatric patient with pulmonary arteriovenous fistula undergoing liver transplantation: A case report. *Pediatr Transplant*. 2016; 20: 711-716.
4. Alberto LD, Fagundes EDT, Rodrigues AT, Queiroz TCN, Castro GV, Ferreira AR. HEPATOPULMONARY SYNDROME IN PEDIATRIC PATIENTS WITH PORTAL HYPERTENSION - AN INTEGRATIVE REVIEW.
5. Nayyar D, Man HS, Granton J, Lilly LB, Gupta S. Proposed management algorithm for severe hypoxemia after liver transplantation in the hepatopulmonary syndrome. *Am J Transplant*. 2015 Apr;15(4): 903-13. doi: 10.1111/ajt.13177. Epub 2015 Feb 3. PMID: 25649047; PMCID: PMC5132094.

of 88–90%. This approach is well supported in the literature, acknowledging that supranormal FiO₂ and escalating ventilator pressures confer little benefit in *Arq Gastroenterol*. 2024 Sep 2;61:e24040. doi: 10.1590/S0004-2803.24612024-040.PMID: 39230090.