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VA-ECMO in Managing Respiratory Failure and Severe Infection in an Low-Weight Infant Following Liver Transplantation: Case Report and Literature Review

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1. Abstract

1.1. Background

Pediatric liver transplantation (pLT) is the standard treatment for end-stage liver disease in children. Perioperative cardiopulmonary complications, including severe infection and respiratory failure, sometimes require extracorporeal membrane oxygenation (ECMO). We present the course of a 4-month-and-21-day-old , 5.5-kg infant with biliary atresia who developed refractory respiratory failure and bilateral pneumothoraces after living-donor left lateral segment transplantation. The infant received venoarterial (VA) ECMO for 30 days, was weaned successfully, but later died from progressive cerebral infarction and intracranial hemorrhage.

1.2. Case Summary

A 4-month-and-21-day-old male infant (weight of 5.5 k) with biliary atresia underwent a living-donor left lateral liver transplantation. Postoperatively he developed severe pneumonia with persistent hypoxemia and bilateral pneumothoraces requiring reintubation and chest drains. On POD 5 VA-ECMO was initiated for combined refractory hypoxemia, severe hypercapnia, and hemodynamic instability. After 30 days on VA-ECMO the patient was decannulated; however, imaging demonstrated cerebral infarction on ECMO day 24, and neurologic status worsened after decannulation, culminating in death on POD 50 from progressive cerebral infarction and cardiopulmonary collapse. Literature review :We reviewed 14 reported cases of infants <1 year receiving ECMO after liver transplantation. Most received VV-ECMO; only two received VA-ECMO. ECMO duration ranged from 0.5 to 26 days and overall mortality remained high.

1.3. Conclusion

VA-ECMO can provide temporary cardiopulmonary support in small infants post-liver transplant, but thrombotic, hemorrhagic, and neurologic risks are substantial. Multidisciplinary management and individualized anticoagulation and monitoring strategies are essential.

2. Introduction

Pediatric liver transplantation (pLT) is the definitive treatment for end-stage liver disease in children, yet managing perioperative complications remains a major challenge—particularly in extremely vulnerable populations such as very low birth weight or medically fragile infants [1]. Among these complications, severe infection and respiratory failure may necessitate the use of extracorporeal membrane oxygenation (ECMO), a high-risk intervention with limited evidence in this patient group [2]. We report the case of a 4-month-and-21-day-old infant weighing 5.5 kg, diagnosed with biliary atresia, who developed refractory respiratory failure and bilateral pneumothoraces after undergoing living-donor left lateral segment liver transplantation. The patient required venoarterial (VA) ECMO for 30 days, was successfully weaned from support, but ultimately died from progressive cerebral infarction and intracranial hemorrhage. To our knowledge, this is the longest recorded use of VA-ECMO in

an infant under one year of age after liver transplantation. This case underscores the unique challenges of ECMO management in pediatric liver transplant recipients, highlights the rarity and complexity of such presentations, and raises important considerations for perioperative care strategies and future research in similar high-risk populations.

3. Case Presentation

3.1. Timeline of Key Events

Dec 8, 2022 – Kasai portoenterostomy for biliary atresia.

Feb 8, 2023 – Admitted for fever and worsening jaundice.

Feb 14, 2023 (POD 0) – Living-donor left lateral segment transplant.

Feb 15, 2023 (POD 1) – Severe respiratory distress → reintubation; mNGS pending.

Feb 18–19, 2023 (POD 4–5) – Bilateral pneumothoraces, chest tubes placed; refractory respiratory failure.

Feb 19, 2023 (POD 5) – VA-ECMO initiated (right IJ venous and right carotid arterial cannulation).

Mar 11, 2023 (ECMO day 20 / POD 25) – Circuit thrombosis → oxygenator and pump exchange.

Mar 15, 2023 (ECMO day 24 / POD 29) – Neurologic decline; CT: cerebral infarction.

Mar 21, 2023 (ECMO day 30 / POD 35) – ECMO decannulated.

Apr 4–5, 2023 (POD 49–50) – Progressive neurologic deterioration and hemodynamic collapse → death on POD 50.

3.2. Initial Presentation and Pre-Transplant Course

A 4-month-and-21-day-old male infant (weight 5.5 kg) with biliary atresia had undergone Kasai portoenterostomy at 2 months of age. Postoperatively, he experienced recurrent cholangitis and progressive cholestasis. Despite multiple hospitalizations for sepsis control, his liver disease advanced to decompensated cholestatic cirrhosis with features of portal hypertension, coagulopathy, and malnutrition. Given the irreversible progression and failure of medical management, he was listed for liver transplantation. Differential diagnoses for worsening jaundice-including intrahepatic cholestasis from other metabolic disorders, graft-unrelated biliary obstruction, and hemolysis—were excluded through imaging, metabolic screening, and laboratory testing. On admission (Feb 8, 2023), he had intermittent fever, marked jaundice, and early signs of respiratory distress. Physical examination showed moderate malnutrition, diffuse jaundice of the skin and sclera, abdominal distension with ascites, hepatosplenomegaly, and no peripheral stigmata of chronic liver disease.

3.3. Laboratory Findings and Clinical Significance

Admission labs revealed leukocytosis (WBC 13.8 × 10⁹/L, ANC 10.02 × 10⁹/L), anemia (Hb 81 g/L), thrombocytopenia (139 × 10⁹/L), and elevated CRP (122 mg/L), consistent with systemic inflammation. Liver tests showed hyperbilirubinemia (total 154.05 μmol/L; direct 93.47 μmol/L), cholestatic pattern (GGT 198 U/L), and hypoalbuminemia (29.9 g/L). Coagulopathy was evident (PT 17.3 s, INR 1.62). Serum ammonia was mildly elevated (74 μmol/L). A rectal swab grew multidrug-resistant Klebsiella pneumoniae (MDR-Kp), guiding pre-transplant antimicrobial coverage. Doppler ultrasound confirmed cirrhosis, splenomegaly, moderate ascites, and intrahepatic bile duct dilation.

3.4. Transplant Surgery and Early Postoperative Course

On Feb 14, 2023, the patient underwent living-donor liver transplantation, receiving the left lateral segment from his father. Surgery was uneventful, and he was extubated three hours postoperatively. mNGS of tracheal aspirates was sent to assess for underlying pathogens given his MDR-Kp colonization. On POD 1, he developed severe respiratory distress (RR 80 breaths/min) with hypercapnia, requiring reintubation. Broad-spectrum antimicrobials polymyxin B, meropenem, and micafungin-were started for presumed severe pneumonia, along with tacrolimus for graft rejection prophylaxis. mNGS later detected K. pneumoniae, Legionella pneumophila, Pneumocystis jirovecii, Escherichia coli, and CMV-all with multiple drug resistance. Antimicrobials were escalated to include sulfonamides and azithromycin, but respiratory failure worsened. This patient received a reduced-size living donor liver transplant using the left lobe from his father. Adequate abdominal drainage was performed postoperatively, so respiratory failure was not considered to be related to liver volume or residual ascites.

3.5. Progression to ECMO

On POD 4, sudden hypoxemia developed. Chest X-ray revealed bilateral pneumothoraces, treated with emergency chest tube placement, which improved oxygenation but not hypercapnia. By POD 5, despite maximal ventilatory support (FiO_2 100%), oxygen saturation remained low, and ABG showed pH 7.15, PaO_2 56 mmHg, $PaCO_2$ 89.5 mmHg, OI 56 mmHg-meeting criteria for severe, refractory respiratory failure with persistent respiratory acidosis. Hemodynamic instability persisted despite high-dose vasopressors. Given the combination of life-threatening hypoxemia, hypercapnia, and shock, VA-ECMO was initiated with the goal of stabilizing gas exchange and perfusion while treating the underlying lung pathology. Cannulation was performed via the right internal jugular vein and right carotid artery. ECMO flow started at 0.7 L/min (127 mL/kg/min), with norepinephrine support titrated to maintain MAP 50–60 mmHg. Protective lung ventilation and systemic anticoagulation (ACT 160–220 s) were maintained throughout ECMO support.

3.5. ECMO Course and Complications

VA-ECMO stabilized circulation, allowing vasopressor weaning. On ECMO day 20, oxygenator and pump thrombosis occurred, requiring urgent circuit change. Bilirubin remained elevated, prompting intermittent therapeutic plasma exchange.

On ECMO day 24, the patient developed neurologic deterioration; CT revealed cerebral infarction. ECMO was gradually weaned, and decannulation was achieved on ECMO day 30 (POD 35).

3.6. Post-ECMO Course and Outcome

After decannulation, pulmonary function initially improved, but neurologic deficits progressed, with imaging showing worsening infarction. By POD 48, he developed circulatory collapse refractory to escalating vasopressors and died on POD 50 from progressive cerebral infarction and central cardiorespiratory failure. Chest X-ray during the course of the disease are shown in Figure 1.

3.7. Clinical Significance

To our knowledge, this represents the longest reported duration of VA-ECMO in an infant liver transplant recipient under 1 year of age. The case highlights the diagnostic and therapeutic complexity of managing perioperative respiratory failure in pediatric liver transplantation, the decision-making process for ECMO initiation in very low-weight infants, and the importance of balancing life-sustaining support with the risk of severe neurologic complications.

3.8. Literature Review

A literature review (2000–present) identified 14 infants (<1 year) who received ECMO after liver transplantation, as summarized in Table 1 [3-10]. Age ranged from 10 weeks to 10 months; primary diagnoses included biliary atresia (7), liver failure (2), cryptogenic cirrhosis (2), and unspecified (3). ECMO timing was mainly postoperative (12 cases), with 9 VV-ECMO, 2 VA-ECMO, and 3 unspecified. Indications were predominantly ARDS (11), pulmonary hemorrhage (2), and sepsis (1). ECMO duration ranged 0.5–26 days (IQR 1–15.6). Reported complications included hemorrhage, circuit thrombosis, and neurologic injury; mortality was 57.1% (8/14).

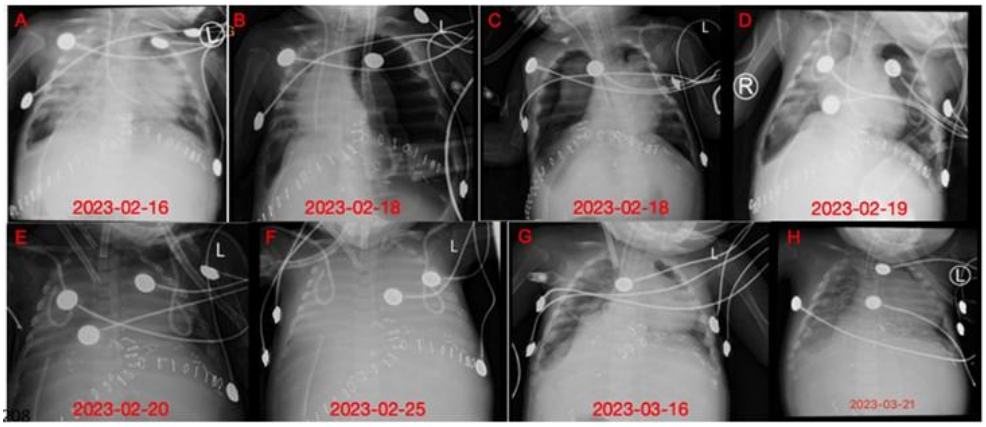

Figure 1: Changes in chest X-ray during the patient's course of illness.
A: Bilateral diffuse infiltrates (POD 2). B: Left-sided pneumothorax (POD 4). C: Right-sided pneumothorax after closed drainage (POD 4). D: Left-sided pneumothorax persists after bilateral closed drainage (POD 5). E: Bilateral diffuse infiltrates, day 1 of ECMO treatment (POD 7). F: Bilateral diffuse infiltrates, day 6 of ECMO treatment (POD 11). G: Bilateral diffuse infiltrates, significant improvement compared to previous days. Day 25 of ECMO treatment (POD 30). H: Bilateral diffuse infiltrates, ECMO weaning (POD 35).

Table 1: Perioperative ECMO Support in Liver Transplantation for Infants and Toddlers Under 1 Year of Age: A Literature Review

Investigator	Country/ Region	No. of Cases	Gender	Age	Underlying Disease	ECMO Timing	ECMO Mode	ECMO Indication	ECMO Duration (days)	ECMO Complications	Outcome
Itsuko Chih-Yi Chen et al. (2023) [3]	Taiwan, China	3	Male	4.8M	Biliary atresia	Post-op Day 7	VV	ARDS	2.81	None	Deceased
			Male	9.6M	Necrotizing hepatitis	Post-op Day 27	VV	Sepsis	4.58	None	Deceased
			Female	4.8M	Biliary atresia	Post-op Day 15	VV	ARDS	15.46	Intracranial hemorrhage	Deceased
Shimura et al. (2022)[4]	Japan	1	Female	5M	Biliary atresia	Intra-op	VV	ARDS/ Pulmonary hemorrhage	3	Blood clots in bronchi	Survived
Imam A et al. (2020)[5]	Turkey	3	Male	10M	Biliary atresia, Hepatopulmonary syndrome	Post-op	not available	ARDS	1	None	Deceased
			Male	10M	Cryptogenic cirrhosis	Post-op	not available	ARDS	1	None	Deceased
			Male	5M	Biliary cirrhosis	Postop	not available	ARDS	0.5	None	Deceased

Lobeck et al. (2018) [6]	USA	1	Male	5M	Biliary atresia	Pre-op	VA	ARDS	7	Catheter site bleeding and acute kidney injury	Survived
Gedik et al. (2015) [7]	Turkey	1	Male	9M	Biliary atresia	Post-op Day 3	VA	ARDS (Klebsiella pneumonia)	7	None	Deceased
Evashuk et al. (2008)[8]	Canada	1	Female	9.6M	Biliary atresia	Post-op Day 16	VV	ARDS (Metapneumovirus pneumonia)	26	None	Survived
Fujita et al. (2005) [9]	USA	1	Female	10W	Idiopathic acute liver failure	Post-op Day 27	VV	Pulmonary hemorrhage and respiratory failure	4	None	Survived
Mack et al. (2000) [10]	USA	3	Male	4M	N/A	Post-op	VV	ARDS (Parainfluenza pneumonia)	1	None	Deceased
			N/A	5M	N/A	Post-op	VV	ARDS (RSV pneumonia)	18	None	Survived
			N/A	9M	N/A	Post-op	VV	ARDS (Pseudomonas aeruginosa pneumonia)	16	None	Survived

4. Discussion

Pediatric liver transplantation has advanced considerably over the past four decades, with long-term survival rates now exceeding 85% at major transplant centers [2]. Despite these improvements, respiratory complications such as acute respiratory distress syndrome (ARDS) and pulmonary hemorrhage remain significant causes of morbidity and mortality in the peri-transplant period. Shimura et al. reported that 11.1% of pediatric liver transplant recipients develop ARDS, a complication associated with prolonged hospitalization and increased mortality [4].

Although extracorporeal membrane oxygenation (ECMO) has become an accepted rescue therapy for severe cardiopulmonary failure in critically ill children, its use following liver transplantation-especially in very low-weight infants-is rare [3,11-13]. Most published cases involve older children, and survival outcomes vary widely due to complications such as bleeding, sepsis, and multiorgan failure [14-16]. To our knowledge, this case represents one of the rarest reports of prolonged VA-ECMO in an infant under one year of age after liver transplantation.

5. Comparison with Previous Literature

A meta-analysis by Reid et al. [17] reviewing 19 pediatric liver transplant recipients on ECMO found that hepatopulmonary syndrome and ARDS were the most common indications (26% each), followed by infection-related respiratory failure (21%), cardiac causes (11%), and pulmonary edema (11%). In our patient, severe early postoperative ARDS developed despite circulatory stability, on a background of recurrent pre-transplant biliary infections and multidrug-resistant (MDR) pulmonary infection. While several reports describe successful VV-ECMO in pediatric liver transplant recipients [3,11-13], cases involving VA-ECMO in low-weight infants remain extremely uncommon.

5.1. Rationale for VA-ECMO in This Case

The decision to initiate VA-ECMO rather than VV-ECMO was driven by combined respiratory failure and hemodynamic instability unresponsive to maximal conventional therapy. VA-ECMO provided both oxygenation and circulatory support, improved graft perfusion, and reduced central venous pressure by draining from the right atrium—potentially supporting more effective graft regeneration. This approach aligns with Di Nardo et al. [2], who found VA-ECMO use in 67% of pediatric liver transplant ECMO cases. Given the patient's age, body weight (5.5 kg), and limited vascular access, right internal jugular vein and right internal carotid artery cannulation were selected, maintaining stable flows throughout the 30-day ECMO course.

5.2. Challenges in the Context of MDR Infection

Prolonged ECMO in the setting of MDR gram-negative bacteria, Pneumocystis jirovecii, and CMV presented unique management challenges. These included balancing broad-spectrum antimicrobial coverage with drug toxicity, preventing device-associated infections, and minimizing immunosuppression while maintaining graft function. This required close coordination among transplant surgery, critical care, infectious disease, and ECMO teams.

5.3. Complications and Risk–Benefit Considerations

Despite initial pulmonary improvement and successful weaning from ECMO, the patient developed progressive cerebral infarction, leading to circulatory and respiratory failure. Cerebral infarction during ECMO may result from thrombosis, suboptimal anticoagulation, hemodynamic instability, infection-related inflammation, or microcirculatory dysfunction [13]. In this case, possible contributors included ECMO-related platelet activation, microthrombus formation despite heparinization, vascular fragility, and transient hypotension. These findings echo the high complication rates reported by Di Nardo et al., with 26% thrombotic and 41% bleeding complications in pediatric liver transplant ECMO patients [2].

5.4. Clinical Implications

This case underscores that VA-ECMO can serve as a life-saving bridge in selected infant liver transplant recipients with combined respiratory and circulatory failure, but carries substantial risks, particularly neurological and thrombotic complications. Clinicians should weigh these risks against potential benefits, employ individualized anticoagulation strategies, ensure aggressive infection control, and maintain vigilant neurological monitoring. In summary, prolonged VA-ECMO after liver transplantation in a very low-weight infant is technically feasible but fraught with challenges, particularly in the presence of MDR infections. Sharing such cases is essential to refine patient selection criteria, optimize peri-ECMO management, and develop evidence-based guidelines for this rare but high-risk clinical scenario.

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