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Challenges During Repeat Extracorporeal Life Support in a Patient With Pulmonary Alveolar Proteinosis

Hemanth Lingadevaru^{*}, Matthew A. Romano^{†,‡}, Karen Fauman^{*}, Elaine Cooley[†], Gail M. Annich^{*,†}, and Timothy T. Cornell^{*}

^{*}Pediatric Critical Care Medicine, University of Michigan, Ann Arbor, Michigan

[†]Extracorporeal Life Support (ECLS) Program, University of Michigan, Ann Arbor, Michigan

[‡]Section of Cardiac Surgery, University of Michigan, Ann Arbor, Michigan

Abstract

Extracorporeal life support (ECLS) is used to support patients with pulmonary alveolar proteinosis (PAP) both during acute illness and during lung lavage therapies. We report the challenges encountered while providing ECLS for respiratory failure to a 12-year-old girl with PAP who had previously received ECLS as a toddler for a prior episode of respiratory failure due to PAP. She was placed on venovenous-arterial (VVA) ECLS and subsequently switched to venovenous (VV) ECLS with drainage from the right femoral vein and reinfusion by a long cannula placed into the right atrium. Our case illustrates standard cannulation may not be possible for children requiring a second ECLS course and the importance of considering alternative modes of cannulation and ECLS support when conventional methods are not possible.

Case Presentation

A 12-year-old girl weighing 40 kg with pulmonary alveolar proteinosis (PAP) and a history of requiring venoarterial (VA) extracorporeal life support (ECLS) at the age of 2 years presented to our hospital with pneumonia, hypoxemia, and respiratory distress. Pulmonary function tests performed 4 months before hospitalization demonstrated severe obstructive and restrictive lung disease. She required 1 L oxygen by nasal cannula at night, to maintain oxygen saturations >92% and had not received routine lung lavage.

She was admitted to the pediatric intensive care unit, and broad-spectrum antibiotics (vancomycin, piperacillin/tazobactam, and azithromycin), bronchodilators, and noninvasive ventilation were started. Total lung lavage was considered on admission, but because of tenuous respiratory status, it was deferred until more definitive respiratory support could be provided. Her work of breathing improved with noninvasive ventilation for the first 3–4 days, but she began to develop significant hypoxia despite maximal escalation of noninvasive support on hospital day 4. After significant desaturations (SaO₂ 70%) without recovery, she was intubated with FiO₂ requirements rapidly increasing to 100%. She was transitioned to high frequency oscillatory ventilation with a delta pressure of 58 mm Hg, mean airway pressure of 35 mm Hg, frequency of 8 hertz, and an FiO₂ of 100%, but her PaO₂ remained 44 mm Hg, SpO₂ remained in the low 80s, and oxygenation index (OI) was 58. We initiated ECLS (Baxter Centry roller pump, Quadrox D oxygenator, and polyvinyl

chloride tubing with super tigan raceway) to improve oxygen delivery and facilitate whole lung lavage.

Within 12 hours of ECLS initiation, initial whole lung lavage was performed, and four additional lavages were performed over the next 2 days. She was successfully decannulated from ECLS 9 days after initiation and was extubated 5 days after ECLS discontinuation. On day 27, she was discharged from PICU, and after 1 week of inpatient rehabilitation, she was discharged home.

ECLS Challenges

Challenges With Cannulation

Given her history of right internal jugular and internal carotid artery ligation as a toddler, the only sites available for cannulation were groin vessels. In our experience, VV ECLS using two femoral catheters in a child her size results in recirculation and inadequate ECLS support. Thus, the initial cannulation was VA by the right groin with 21 Fr 19 cm venous cannula and 15 Fr 19 cm arterial cannula. Her upper extremity saturations on VA ECLS remained in the 80s with SvO₂s in the 90s, and she remained too unstable for whole lung lavage. We surmised this as the result of normal native cardiac function preventing retrograde flow provided by the arterial cannula from reaching the aortic arch resulting in suboptimal ECLS support with discordant upper and lower body oxygenation (north/south syndrome). See Table 1 for maneuvers attempted to reduce north/south syndrome.

Under echocardiographic guidance, an additional 20 Fr 68 cm venous cannula was placed in the left femoral vein and positioned with the tip in the right atrium (venovenous-arterial [VVA] ECLS) in an attempt to more effectively decrease preload with the hope of decreasing cardiac output enough to improve the retrograde arterial cannula flow. Additionally, afterload reduction was achieved with nitroprusside and nicardipine to maintain relative hypotension with systolic blood pressure 85–95 mm Hg. These interventions resulted in improvement in the north/south syndrome with lactates decreasing to 1.4 mmol/L indicating improved oxygen delivery.

The use of VVA ECLS with additional afterload reduction stabilized the patient for the first 36 hours and allowed for the first lung lavage. However, after 36 hours of VVA support, upper extremity saturations fell to the high 70s and serum lactates increased to 3.1 mmol/L. Given her stable hemodynamics, we converted the ECLS to VV by isolating the arterial cannula from the circuit and using the right femoral venous cannula (low cannula, tip just above the junction of the iliac veins, and inferior vena cava) for drainage and the left femoral cannula (high cannula and tip in right atrium) for reperfusion.

Upper compartment saturations and discordance improved immediately on transition to VV support. Twenty-four hours after VV conversion with upper extremity SaO₂s of 80%–85%, SvO₂s of 70%–75%, and normal lactates, the arterial cannula was removed, and the vessel was surgically repaired while on ECLS.

Challenges With Acutely Decompensated Alveolar Proteinosis

Mainstay of therapy for patients with PAP includes whole lung lavage.^{1–3} In the acutely decompensated state of our patient, lavage was impossible without extracorporeal support, which has been reported for both children and adults.^{4–7} *N*-acetylcysteine and inhaled DNase were instituted before intubation, along with aggressive pulmonary toilet, in an effort to clear proteinaceous material, but our patient continued to have worsening hypoxemia. In addition, our patient had a superimposed adenoviral pneumonia resulting in further airway obstruction and increased oxygen. The dramatic response noted after the

initial lung lavage confirmed our suspicion that the PAP was the primary cause of the hypoxemia. The bronchoalveolar lavage fluid was abundantly periodic acid-Schiff (PAS) stain positive, consistent with PAP.

Discussion

Pulmonary alveolar proteinosis syndrome can lead to profound hypoxemia and requires whole lung lavage as treatment.^{1,2} As was the case with our patient, many patients during an acute respiratory illness are too unstable to tolerate whole lung lavage. In such a case, the use of ECLS can facilitate whole lung lavage.^{4–7} Our patient had a relative contraindication to ECLS because of her prior ECLS course 10 years prior in which her left internal jugular and carotid were ligated. We initially considered VV ECLS using both femoral vessels, but we were concerned that the degree of recirculation would not allow adequate ECLS support. We were able to initially place her on VA ECLS, then converted her to VVA, and subsequently converted to VV ECLS. VV ECLS through the groin vessels was successful, without significant recirculation, due to the innovative use of a long cannula, which redirected flow into the right atrium. The decrease in recirculation by directing flow into the right atrium has been described,⁸ but to our knowledge, this is the first report of redirecting flow using a long cannula. Although we were able to provide VV support for our patient using a long cannula, we acknowledge the increased risks of thrombosis and perforation, as well as the expertise required for long cannula placement.

Indications for ECLS support have expanded since its inception in 1975, and more complex patients are being placed on ECLS requiring employment of novel methods to meet the needs of more complex patients.

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Table 1
Parameters for Each Mode of ECLS and Maneuvers Used to Attempt to Reduce North/South Syndrome

Mode of ECLS	Duration of Mode (h)	SvO ₂ at the End of Each Mode of ECLS (%)	SaO ₂ at the End of Each Mode of ECLS (%)	Lactate at the End of Each Mode of ECLS (mmol/L)	Maneuvers Used to Attempt to Reduce North/South Syndrome
Before ECLS	N/A	68	79	2.6	N/A
VA	4	90	89	1.5	Nicardipine for afterload reduction
VVA	32	81	79	3.1	Nicardipine and nitroprusside for afterload reduction; temperature to 35°C; neuromuscular blockade
VV (with long cannula) *	168	N/A	N/A	N/A	N/A

* The arterial catheter was initially left in place but was removed and the arterial vessel repaired after 24 h on VV with good oxygenation. VVA, venovenous-arterial; VV, venovenous; ECLS, extracorporeal life support; N/A, not applicable; VA, venoarterial.