The Use of ECMO During Total Lung Lavage in a Patient with Pulmonary Alveolar Proteinosis: A Case report

Author(s): M Mueller, CM Oermann, RS Vax, NL Glass, CJ Campos
Affiliation(s): Texas Children's Hospital, Houston, TX

Introduction:
Pulmonary Alveolar Proteinosis (PAP) is a rare disease of impaired surfactant clearance causing severe hypoxemia and respiratory distress. Autoantibodies against granulocyte- macrophage colony-stimulating-factors (GM-CSF) prevent the final maturation of alveolar macrophages and render them incompetent to clear surfactant lipids and proteins from the alveolar space (1). Symptomatic treatment involves repeated lavage of the bronchoalveolar tree to improve oxygenation and lung function.

Case report:
We report the case of a 4-year-old, 13 kg Hispanic girl with severe respiratory distress secondary to PAP who required arterio-venous extracorporeal membrane oxygenation (ECMO) to facilitate total lung lavage. She was born at 34 weeks, diagnosed with Turner syndrome and suffered from RSV pneumonia requiring mechanical ventilation for 2 weeks during the neonatal period. At age 3 she developed increasing respiratory distress; a lung biopsy helped diagnose PAP. Multiple attempts at lavaging individual lungs/segments resulted in ineffective therapy because of the difficulty of isolating one lung while ventilating the other. (Picture 1)

Improvement of her respiratory symptoms could only be achieved with large volume irrigation of both lungs simultaneously – under ECMO as previously described (2). Prior to surgery we confirmed the patency of the access vessels by ultrasound (Picture 2).
We induced general anesthesia, continued mechanical ventilation via a preexisting tracheostomy, and cannulated a radial artery. The surgeons obtained arterial access to the left femoral artery via an end-to-side gortex graft. Using the Seldinger technique, we then placed a 14 Fr venous cannula under fluoroscopic guidance into the right internal jugular vein and established ECMO circulation sufficient for adequate oxygenation. (Picture 3)

The pulmonologists irrigated both lungs through a 5.0 endotracheal tube, distributing the irrigation fluid evenly with the help of a percussion vest (Picture 4).
A total volume of 18 liters (in 350 ml increments) was used to irrigate the lungs and clear the bronchoalveolar tree of proteinaceous material (Picture 5).

A bronchoscopy done at the conclusion of the lavage confirmed clearing of the lung units. (Picture 6)
The patient’s lung function improved by the end of the procedure allowing us to wean her successfully from ECMO. She was transferred to the intensive care unit (ICU) where she continued to improve. She was ventilated mechanically for three days in the ICU and then transferred to a “step-down” unit. (Picture 7)

She was ventilated for five weeks and subsequently weaned to humidified oxygen delivered via a tracheostomy mask. At six weeks after the lung lavage, she has continued to do well and will be discharged home in a few days.
**Discussion:**
PAP is a devastating disease requiring frequent clearing of impacted alveolar material. Because of the patient’s size and age, providing effective oxygenation and ventilation, while infusing large volumes of fluid during bronchoalveolar lavage, was not possible using conventional means of ventilation. Only by using ECMO to bypass the lungs were we able to effectively improve the patient’s condition. Bronchopulmonary lavage is a temporary treatment for PAP and eventually has to be repeated. The number of times that ECMO can be performed in a patient is usually limited by progressive difficulty obtaining vascular access (scarring or thrombosis of veins etc). ECMO not only carries the potential for serious complications (limb ischemia, hemorrhage, neurological sequelae from stroke, pseudoaneurysm formation) but also poses logistical challenges. The successful completion of this procedure required the combined effort of all participating teams (surgeons, anesthesiologists, pulmonologists, perfusionists, nurses).

**References:**
1) Tazawa R. et al., *Respirology*, 2006