Case report

Sepsis and acute respiratory distress syndrome requiring extracorporeal life support in an adolescent with mild cystic fibrosis

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ABSTRACT

Outcomes for invasive mechanical ventilation and extracorporeal membrane oxygenation (ECMO) to treat acute respiratory failure in patients with mild cystic fibrosis (CF) lung disease are not known. We present a case of the successful use of ECMO to treat acute respiratory failure secondary to staphylococcal sepsis in an adolescent CF patient with previously normal lung function. Her post-ECMO course was notable for severe airflow obstruction, hypoxemia, deconditioning, and growth failure. She had significantly improved at six months follow-up, though she continued to have moderate airflow obstruction on pulmonary function testing. This case illustrates that ECMO and prolonged intubation can prolong life in CF patients with mild lung disease who present with potentially reversible acute respiratory failure, though they are associated with significant morbidity.

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1. Introduction

Respiratory failure in cystic fibrosis (CF) patients treated with invasive mechanical ventilation (IMV) is associated with increased mortality [1]. Extracorporeal membrane oxygenation (ECMO) has been used as a bridge to lung transplant in CF patients. Extracorporeal Life Support Organization Registry data indicates that 56% of adult CF patients treated with venovenous (VV) ECMO survive, although the role of premorbid clinical severity in determining survival is not reported [2]. Data are limited regarding the use of ECMO to treat acute respiratory failure (ARF) in CF patients with mild lung disease. We present a case of the successful use of ECMO in a patient with mild CF lung disease and reversible respiratory failure due to sepsis, highlighting the prolonged recovery period and significant loss of lung function.

2. Case report

A 15-year-old female with homozygous delF508 cystic fibrosis presented to the emergency department with shortness of breath, hypoxemia, and a 10-kg weight loss. Six months prior to presentation, she had normal lung function with FVC 3.54 L (111% predicted) and FEV1 3.06 L (107% predicted), body mass index (BMI) at the 75th percentile, and mild bronchiectasis on CT scan. She had recently exi-
ted foster care and been reunited with her biological family. She had chronic endobronchial colonization by methicillin-sensitive Staphylococcus aureus (MSSA). On admission, the patient had labored breathing with moderate retractions. Her initial vitals were notable for a temperature of 38.8° Celsius, heart rate 160, blood pressure 113/ 66, respiratory rate of 46 breaths per minute, and SpO2 88% on pulse oximetry recording in room air, correcting to 99% with oxygen delivered by a non-rebreather mask. An arterial blood gas showed pH 7.25, pCO2 46, pO2 172, and base de

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expiratory pressure, frequent chest physiotherapy and suctioning, and pharmacologic muscle relaxation. She demonstrated worsening acidosis (pH ranging 7.24 to 7.13) and hypoxemia. SpO2 ranged 80–90% on FiO2 1.0, with arterial pO2 48–61. She was cannulated for VV-ECMO on HD three. Serial therapeutic bronchial lavage and frequent intrapulmonary percussive ventilation were performed for airway clearance. No other organisms were identified on bronchoalveolar lavage fluid culture. She improved and was decannulated on HD 24, then extubated on HD 27. Spirometry four weeks after decannulation showed severe airflow obstruction with FVC 1.30 L (41% predicted) and FEV1 0.82 L (28% predicted). Chest CT showed severe cystic bronchiectasis of the upper lobes (Fig. 1). She required supplemental gastrostomy tube feedings for weight loss and poor oral intake with intolerance of a nasogastric tube. She needed inpatient rehabilitation for physical deconditioning. She was discharged on HD 76 with supplemental oxygen due to hypoxemia with sleep and exertion. Her lung function slowly improved in the subsequent months, with FVC 2.52 L (78% predicted) and FEV1 1.81 L (62% predicted) six months after hospital discharge (Fig. 2). She no longer requires supplemental oxygen or tube feedings.

3. Discussion

The primary risk factor for poor outcome of IMV in CF patients is increasing age [1], though low BMI [3] and low FEV1 [3] may contribute. Small studies of children under age two with CF treated with IMV for ARF have not shown worse outcomes compared to age- and genotype-matched controls after five years [1]. Reported cases of adult CF patients requiring IMV have severe lung disease at baseline. Rates of survival of IMV are low but improving compared to a few decades ago [3].

Outcomes of prolonged IMV and ECMO for patients with mild CF lung disease are unknown. To our knowledge, two other cases describe pediatric patients with mild CF lung disease treated with ECMO for ARF. Neither case had a prior diagnosis of CF. Hirthler and Goldthorn reported a term infant with respiratory distress at birth treated with IMV and ECMO at 15 hours of life, then decannulated after 5 days. She had one episode of pneumonia before 5 months of age [4]. The second case, reported by Stauffer and Wallis, was a three year old with respiratory distress and *Pseudomonas aeruginosa* sepsis. She was treated with VV-ECMO for 19 days, and diagnosed with CF while on ECMO. She had near-normal lung function 12 years later [5]. In our patient, the decision to initiate ECMO was informed by her previously normal lung function and presumed reversibility of respiratory failure. There was no guide to help predict her prognosis, and she was an unlikely candidate for lung transplant based on her unstable social situation.

This case illustrates both survival and severe decline in lung function in an adolescent with mild CF lung disease after prolonged intubation and ECMO. ECMO may prolong survival but may also result in a loss of lung function and accelerated development of bronchiectasis. This patient showed slow improvement of lung function, oxygenation, and nutritional status in the six months after treatment with ECMO, though she has moderate airflow obstruction and evidence of irreversible damage on chest CT. ECMO should be considered in patients with mild CF lung disease who have acute respiratory failure of reversible etiology. The potential survival morbidity should be discussed with caregivers.

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**Fig. 1.** Chest CT images from before and after respiratory failure. Baseline images from 1.5 years before illness showed mild bronchiectasis (left panels). Repeat chest CT six weeks after separation from ECMO showed severe bronchiectasis predominantly in the upper lobes (right panels).
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Conflicts of interest

None.

Both authors, Dr. Lauren Elizabeth Faricy and Dr. Gwynne Church, have reviewed this work. Neither author has a potential conflict of interest to report. The patient and her legal guardian provided written informed consent for the submission of this case report. No closely related manuscripts have been published elsewhere. This case was presented at a poster session at the American Thoracic Society Conference in San Francisco in May 2016.

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