Ambulatory ECMO as a Bridge to Lung Transplant in a Previously Well Pediatric Patient With ARDS

abstract

Extracorporeal membrane oxygenation (ECMO) is increasingly implemented in patients with end-stage pulmonary disease as a bridge to lung transplant. Several centers have instituted an approach that involves physical rehabilitation and ambulation for patients supported with ECMO. Recent reports describe the successful use of ambulatory ECMO in patients with chronic respiratory illnesses being bridged to lung transplant. We describe the first case of a previously healthy pediatric patient with acute respiratory failure successfully supported with ambulatory ECMO as a bridge to lung transplant after an unsuccessful bridge to recovery. Although there are challenges associated with awake and ambulatory ECMO in children, this strategy represents an exciting breakthrough and a potential paradigm shift in ECMO management for pediatric acute respiratory failure. Pediatrics 2014;134: e583–e585
Extracorporeal membrane oxygenation (ECMO) is an important therapeutic modality for patients with refractory respiratory failure. Despite extremely high predicted mortality in patients with refractory respiratory failure, survival for some populations supported with ECMO exceeds 70%.1–3 However, in patients with irreversible pulmonary injury, lung transplant may be necessary. Traditional outcomes for ECMO patients bridged to lung transplant are poor,4–7 but there are increasing reports of successful bridge to transplant using awake and ambulatory ECMO approaches in patients with chronic respiratory conditions.8–19 We report the first successful bridge to lung transplant with ambulatory ECMO in a previously healthy pediatric patient with acute refractory respiratory failure.

**CASE DESCRIPTION**

A 16-year-old, previously healthy, physically active girl presented to a community hospital with right lower lobe pneumonia and respiratory failure (Fig 1). Her condition deteriorated rapidly, prompting transfer to our quaternary care center for possible ECMO. Upon admission to our institution, the patient’s chest radiograph demonstrated diffuse infiltrates, bilateral pneumothoracies, and pneumomediastinum (Fig 2). She was immediately placed on high-frequency oscillatory ventilation, but because of an oxygen index persistently >40, frequent desaturations, and worsening air leak, venovenous ECMO was initiated via a 27-French double-lumen right internal jugular vein cannula (Avalon Laboratories, Rancho Domingo, CA) ∼30 hours after admission. For the next 4 weeks she needed deep pharmacologic sedation, intermittent neuromuscular blockade, and mechanical ventilation in conjunction with ECMO to maintain gas exchange. During this 4-week period, her air leak resolved, but she continued to have persistent acute respiratory distress syndrome and no improvement. A transbronchial biopsy revealed no infectious etiology, and the pathology results demonstrated organizing and proliferative diffuse alveolar damage and neutrophilic alveolitis. This evolving pulmonary fibrosis, along with lack of clinical improvement, prompted discussion about the possible need for lung transplant.

On ECMO day 28, an open tracheostomy was performed at the bedside to facilitate rehabilitation for potential lung transplant. During the next week, sedation was minimized, and the patient began sitting in a chair position in bed. She stood on ECMO day 38, took several steps on day 46, and was listed for lung transplant on day 50 after ambulating 40 feet. Two days later, she received a bilateral orthotopic lung transplant. Final pathology on her native lungs revealed no infectious etiology and demonstrated severe fibrosing interstitial pneumonia and proliferative diffuse alveolar damage consistent with acute interstitial pneumonia.

The patient was decannulated from ECMO 6 hours after transplant, began tracheostomy collar trials on postoperative day (POD) 3, and was transferred to the stepdown unit on POD 8. After a brief readmission to the ICU for atrial flutter, her pulmonary rehabilitation continued, she was weaned from mechanical ventilation on POD 24, and her tracheostomy tube was removed upon discharge on POD 29.

**DISCUSSION**

Recent data suggest improved outcomes when patients bridged to lung transplant with ECMO are awake and participate in active physical rehabilitation.8,16 Several small case series demonstrate the successful use of ECMO as a bridge to lung transplant, but the majority of patients are adolescents and adults with chronic lung disease who are cannulated with the intent to transplant.4–17 Intent at the time of cannulation, based on the patient’s baseline pulmonary status, is an important consideration because data about outcomes for patients bridged to lung transplant after an initial attempt at a bridge to recovery are limited.12 Our report describes the first pediatric patient successfully transitioned to an ambulatory ECMO bridge to lung transplant after an unsuccessful bridge to recovery.

Our current institutional practice for ECMO patients is to list for lung transplant only those who are able to ambulate. This practice is based on the high mortality
rate in patients traditionally supported with ECMO as a bridge to lung transplantation in comparison with outcomes in patients supported with ambulatory ECMO. Therefore, patients cannulated with intent to transplant receive tracheostomies shortly after cannulation and are quickly weaned from pharmacologic sedation. Before the decision to pursue transplant, this patient needed prolonged mechanical ventilation and sedation infusions because of air hunger and agitation. The inability to awaken this patient was probably exacerbated by her history as a previously healthy teenager unaccustomed to pulmonary insufficiency. To date, all pediatric patients and most adult patients supported with ambulatory ECMO have had chronic pulmonary diseases, most commonly cystic fibrosis. Chronic respiratory insufficiency leads to greater tolerance for dyspnea, as evidenced by the patients supported with ambulatory ECMO who have been managed with minimal respiratory support or even extubated. The impact of weaning both sedation and ventilatory support in children without chronic respiratory disease must be addressed when ambulatory ECMO is considered for previously healthy children. This report indicates that ambulatory ECMO is a feasible strategy in pediatric patients without chronic respiratory illness presenting with acute respiratory failure. Despite challenges associated with awake and ambulatory ECMO in children, the early success of this strategy warrants additional investigation of this approach for refractory pediatric respiratory failure.

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