Cystic fibrosis is an autosomal recessive genetic disorder in which the defective gene produces a mutant protein that can attack the respiratory, digestive, and endocrine systems. This protein interferes with the body's ability to manage anionic and chloride, leading to a chloride deficit which thickens secretions throughout the body. In the lungs, this viscous buildup of mucus creates a risk of mucus plugs and leads to severely decreased lung functions and to life-threatening respiratory infections. In the digestive and endocrine systems, ducts become blocked stopping the flow of digestive enzymes which affects digestion and absorption. The restricted ability to break down food it consumes and the pancreas does not function properly, inducing diabetes. Currently, there is no cure for cystic fibrosis, only symptom management. As this disease progresses, the body's ability to breathe is reduced. ECMO is the best option for many cystic fibrosis patients is lung transplantation. With the higher demand for donor lungs than can be supplied, the use of Extracorporeal Membrane Oxygenation (ECMO) as a bridge to lung transplantation has become a viable therapy.

Clinical Summary

A 24-year-old woman who underwent bilateral sequential lung transplantation in 2013 for cystic fibrosis initially did well but unfortunately developed an upper respiratory failure. She presented in March of 2016 in chronic rejection with a progressive decline of her pulmonary function. She was subsequently admitted to the hospital with hypercapnic respiratory failure. She remained awake and alert, but her carbon dioxide levels persistently rose. The patient was counseled regarding the potential risks and benefits of veno-venous (VV) ECMO, as well as possible alternative approaches to the problem and gave informed consent.

She was taken to the catherization laboratory, placed under local anesthesia, and cannulated via the right internal jugular vein with a 29 French ProteckDuo Dual-Lumen Cannula with the distal port in the main pulmonary artery and the proximal port in the right atrium. The cannula was then connected to the ECMO circuit and veno-venous ECMO was initiated with a Thoratec Centrimag centrifugal pump and a Maquet Quadrox-i adult oxygenator.

Discussion

The use of ECMO as a bridge to lung transplantation as well as a rescue strategy post-transplant in recent years has gained significant attention. In a study of more than 900 patients from the UNOS database from 2005 to 2011, roughly 1% of pulmonary patients were bridged to transplant with ECMO support. (1)

These numbers continue to grow as centers across the country have demonstrated the utility and successful outcomes associated with ECMO as a bridging strategy to lung transplantation. Technological advances in the circuits; heparin coated circuits, polyethylene glycol oxygenators, centrifugal pumps, smaller circuits, dual-lumen cannula, have minimized anticoagulation needs and reduced hemolysis and blood component activation. As well, the benefit in weaning patients off of mechanical ventilation. Compared to the conventional mechanical ventilation strategy, patients who were spontaneously breathing without mechanical ventilation who received ECMO as a bridge to transplant and made it to transplantation had significantly better survival at 6 months (80% versus 50%) and had shorter postoperative hospital stays.

Conclusions

With the help of technological advances, this patient was able to be placed on VV ECMO as a bridge to transplant while remaining intubated during her entire ECMO course. This, along with being able to participate in rehabilitation exercises including being ambulated around the ICU, significantly improved her overall muscle strength resulting in an easier rehab process post ECMO and her second bilateral lung transplant. These two things alone have shown to result in patients having shorter post-transplant hospital stays and a better survival rates compared to those patients who remain mechanically ventilated and bedridden on ECMO. This patient was discharged 20 days post-transplant and currently continues to thrive at home.

References


Redo-Bilateral Lung Transplant of Veno-venous ECMO Patient

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