Anesthetic Management for ECMO Cannulation in a Patient with Emery-Dreifuss Muscular Dystrophy

Ana Cox MD1, Lisa Wise-Faberowski MD MS1, Ahlia Kattan MD1, Calvin Kuan MD1, Katsuide Maeda MD2
Departments of Pediatric Anesthesiology1 and Cardiothoracic Surgery2, Stanford University School of Medicine, California, USA

Introduction:
• A 14 year old male with Emery-Dreifuss muscular dystrophy presented with recent URI and significant acute heart failure.
• His other past medical history was significant for history of a difficult airway requiring fiberoptic intubation. By report, he had a grade 4 view both with direct laryngoscopy and Glidescope.
• Pulmonary function test in 2015 demonstrated moderately restrictive and obstructive lung disease.
• His ECHO demonstrated worsening EF of 17%, down from baseline of 51%.
• He had LBBB on his current ECG with frequent PVCs.
• At the time of admission from an outside hospital, his heart failure was marginally compensated on dopamine 7 mcg/kg/min and milrinone 0.75 mcg/kg/min. He was on BIPAP 16/8 with rate of 12, and 30% FiO2.
• On exam, he had two finger breadth mouth opening and thyromental distance with limited range of neck motion. Access included a radial arterial line, left upper extremity double lumen PICC, and 18G PIVx2.
• Plan was for ECMO cannulation in the OR due to airway concerns and impending cardiac and respiratory failure.

OR Course:
• Midazolam and ketamine were titrated for sedation and to maintain spontaneous ventilation, but due to limited mouth opening, etomidate was given to allow for a William’s airway placement. Mask ventilation was easy, fiberoptic intubation was performed, and a 7.0 ETT was placed.
• ECMO cannulation was performed via median sternotomy to achieve ambulatory ECMO.
• The patient remained on ambulatory ECMO for two weeks before being taken back for LVAD placement.

Discussion:
• Emery-Dreifuss is a rare inherited muscular dystrophy which typically presents in the first decade of life with flexion contractures of the distal extremities and posterior cervical muscles.
• Cardiac involvement is variable, but usually occurs in the second or third decade of life with conduction disorders.
• Cardiomyopathy in these patients is not common. Only 12% of patients die of heart failure versus 46% who die as a result of arrhythmia.
• Anesthetic considerations include difficult intubation and line access due to muscle contractures and risk of cardiac complications due to conduction issues and decreased ventricular function.

Conclusion:
• Patients with Emery-Dreifuss muscular dystrophy pose significant challenges that must be kept in mind when forming an anesthetic plan.

Sources: