Emergency Central VA-ECMO and Pulmonary Artery Return in a Neonate With an MYH6 Mutation

Prolonged extracorporeal support after congenital heart surgery complicated by suprasystemic pulmonary hypertension and recurrent pulmonary hemorrhage

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Background

Neonates with complex congenital heart disease, pulmonary hypertension, and genetic cardiomyopathy may develop severe postoperative cardiorespiratory failure. Pulmonary hemorrhage may complicate extracorporeal support and anticoagulation.

Objective

To describe ECMO support, including emergency central cannulation during cardiac arrest and pulmonary artery return, in a neonate with an MYH6 mutation, hypoplastic aortic arch, and a large ventricular septal defect.

Methods

Single-patient case report.

Results

The patient was born in a quaternary maternity center after prenatal diagnosis of congenital heart disease. On day of life 5, unexplained pulmonary hemorrhage required intubation, mechanical ventilation, and treatment for pneumonia. On day 12, anatomical repair included ventricular septal defect closure and aortic arch reconstruction. Recovery was initially favorable, with delayed sternal closure and extubation on postoperative day 4.

At 28 days of life, recurrent pulmonary hemorrhage was associated with suprasystemic pulmonary artery pressure, right ventricular fractional area change of 12%, biventricular dysfunction, low cardiac output, systemic hypoperfusion, and refractory hypoxemia. Despite vasoactive support, the patient deteriorated. Peripheral VA-ECMO was not feasible because of small vessels. Cardiac arrest ensued, and emergency sternotomy with central VA-ECMO cannulation was performed. A 10-Fr arterial cannula was placed in the ascending aorta and a 16-Fr curved venous cannula in the right atrium, using a neonatal Eurosets circuit and ECMOLife console.

Within 24 hours, perfusion improved, the patient awakened, and vasoactive support was reduced. Recurrent pulmonary hemorrhage without laboratory evidence of coagulopathy led to heparin-free ECMO. By ECMO day 8, cardiac function had recovered, but severe pulmonary failure persisted. Because dual-lumen VV cannulation was not feasible, an additional return cannula was placed in the main pulmonary artery, followed by flow and ventilatory optimization.

On ECMO day 24, lung compliance improved and protective ventilation became possible. A pneumothorax was initially managed conservatively. On day 25, worsening pneumothorax required drainage, complicated by pulmonary laceration and massive hemorrhage. Thoracotomy failed to control bleeding, and the patient died.

Conclusion

Emergency central VA-ECMO restored perfusion during cardiac arrest, while pulmonary artery return provided an alternative when VV access was impossible. Severe pulmonary hypertension and lung injury shaped the course, while catastrophic hemorrhage determined the outcome.

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