CASE REPORT

Veno-venous Extracorporeal Membrane Oxygenation for Right Ventricular Failure with Atrial Septostomy After Corrective Repair of Tetralogy of Fallot

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Summary

Right ventricular (RV) dysfunction may occur after cardiac surgery and it is not rare after corrective repair of tetralogy of Fallot (TOF). If traditional treatments with volume management, infusion of inotropic agents, and use of pulmonary vasodilators cannot stabilize the patient, extracorporeal membrane oxygenation (ECMO) or a ventricular assist device (VAD) will be considered as the last resort. Here, we report a young infant patient with RV failure after corrective repair of TOF and without closure of an atrial septal defect (ASD), who was rescued by veno-venous (VV)-ECMO.

Key words: Right ventricular dysfunction, Inter-atrial communication, Mechanical circulatory support

Case Report

The patient was a 4-month-old male baby (body weight 6.7 kg) born to a G4P4 34-year-old healthy mother via vaginal delivery. The gestational age (GA) was 40 weeks and the birth body weight (BBW) was 3550 g. The delivery was smooth and the Apgar score was 8. However, cyanosis during crying was noted by his mother when he was three months old. Episodes of bluish color to the skin occurred frequently. Echocardiography was performed at a local clinic and TOF was diagnosed. He was subsequently transferred to our hospital for further management. After admission, heart computed tomography (CT) showed TOF and persistent left superior vena cava (SVC). Cardiac catheterization revealed the pulmonary arteries (PA) were of adequate size with a McGoon ratio of 1.9. Because of the frequent hypoxic spells and adequate PA size, we decided to perform corrective repair of TOF. Hypoxic episodes (SpO2 60%~75%) occurred intermittently despite medical treatment and oxygen supply pre-operatively. Surgical intervention including repair of ventricular septal defect (VSD), infundibulectomy and right ventricular outflow tract (RVOT) reconstruction with a trans-annular patch by using fresh autologous pericardium were performed smoothly under cardiopulmonary bypass (CPB) with aortic and bicaval cannulation. The CPB time was 208 minutes and the aortic cross-clamping time was 148 minutes. After CPB was stopped, RV pressure (38/15 mmHg) and main PA pressure (24/15 mmHg) were measured in the operating room by direct needle puncture before closure of the sternal wound.

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Ventilator support, inhaled nitric oxide (iNO), inotropic support, and antibiotics were given postoperatively. After surgery, his systolic blood pressure (SBP) was about 70~80 mmHg but desaturation persisted and his oxygenation saturation (SpO2) was 75~85% despite the use of a mechanical ventilator (FiO2 1.0) and iNO 20 ppm. On the next day after the cardiac surgery, SBP was still about 80 mmHg but central venous pressure was up to 20-22 mmHg and SpO2 decreased further to 60~75%. RV failure was diagnosed. Because acute RV failure had not improved despite maximal medical treatments, ECMO setup was considered. At that time, although desaturation persisted, BP could be maintained by volume management and the use of inotropic agents, and a right-to-left shunt through an ASD by echocardiography was also noted. Chest x rays (Figure 1) showed clear lung fields, which meant no pulmonary edema. Therefore, we decided to introduce VV-ECMO, which was performed smoothly with a double lumen catheter (MAQUAT, Rastatt, Germany) from cannulation through the right internal jugular vein into the right atrium (Figure 2). Afterwards, the unstable hemodynamics, respiratory distress, and oxygenation gradually improved, and thus inotropics, ECMO FiO2, and iNO were tapered day by day. On the 13th postoperative day (POD), the VV-ECMO, with a total supporting time of 288 hours, was removed. The usage of iNO was discontinued on POD 24 and extubation of the endotracheal tube was conducted on POD 34. The patient was discharged on POD 46 in stable condition.

Echocardiography was checked before discharge and showed a residual VSD of 0.2 cm, ASD, residual RVOT pressure gradient (PG) of 25 mmHg, moderate pulmonary regurgitation (PR), and left ventricular ejection fraction (LVEF) of 69%.

He grew and developed well, reaching a body weight of 18 kg when he was 5 years old. Echocardiography performed at that time showed a residual VSD of 0.2 cm, ASD, residual RVOT PG of 13 mmHg, mild-to-moderate PR, and LVEF of 85%. Therefore, ASD was occluded percutaneously with an Amplatzer occluder (Abbott Laboratories, Chicago, IL, USA). The patient is in functional class I of New York Heart Association and is currently drug free.

**Discussion**

RV dysfunction is not rare after corrective repair of TOF and various degrees of RV failure can occur. Acute RV failure may be caused by abruptly increased RV afterload (pulmonary embolus, hypoxia, acidemia) or decreased RV contractility (RV ischemia, post-cardiotomy syndrome). After cardiac surgery, there is a disruption in the native RV contractile pattern, which affects RV function in combination with the effect of cardiopulmonary bypass. During RVOT reconstruction in our patient, RVOT was not widely opened but just opened as necessary in order to create an unobstructed neo-RVOT. The length of the RVOT incision was limited to within 2 cm. The main causes of post-operative acute RV failure in our patient might be multifactorial and might include pre-operative hypoxia, right ventriculotomy, and the effect of cardiopulmonary bypass. RV failure may compromise heart function after cardiac surgery. Usually, volume management, infusion of inotropic agents, and the use of pulmonary vasodilators are the first modes of treatment. If the patient cannot be stabilized by any or all of these means, ECMO or VAD should be considered to save the patient’s life.

With respect to ECMO, VA-ECMO is usually used for patients with RV failure because it can unload RV, maintain systemic perfusion, and maintain adequate oxygenation. However, this time VV-ECMO was chosen for the reasons outlined below. In previous studies that created an atrial septostomy and used VV-ECMO to treat pulmonary and RV failure, it was concluded that right-to-left atrial shunting of oxygenated blood with VV-ECMO was capable of maintaining normal systemic hemodynamics and normal arterial blood gases during high RV afterload dysfunction. This concept with atrial septostomy and VV-ECMO was used successfully for patients with end-stage lung disease with RV failure as a bridge to lung transplantation. A systematic review of studies from 1990 to 2017 found that neurologic injury was overall more commonly reported in VA ECMO than in VV ECMO. If possible, it is
also our policy to choose VV-ECMO rather than VA-ECMO in consideration of potential neurologic complications. In our case, during corrective repair of TOF, the ASD was left open for potential RV failure in the postoperative period. Right-to-left shunt was expected if RV dysfunction was significant postoperatively. If RV failure occurs and the patient cannot be stabilized by traditional means, rather than VA-ECMO, VV-ECMO may be considered as an option to rescue this kind of selected case with atrial septostomy and RV failure after corrective repair of TOF.

Disclosure

Conflicts of interest: The authors have no conflicts of interest to declare.

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