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Case Report

ECMO support after arterial switch operation: A case report series from a single institution

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Extracorporeal membrane oxygenators (ECMO) after arterial switch operation (ASO) is a rescue therapy for patients who cannot wean from cardiopulmonary bypass as a bridge to recovery and/or transplantation. We, hereby, present a case report series of four patients from an institute who underwent ECMO after ASO for dextro-transposition of the great arteries (d-TGA). Two patients subsequently weaned off ECMO but only one patient survived to discharge. ASO is a complex surgery that requires an approach of a multi-disciplinary team where ECMO is a useful adjunct for recovery in difficult cases. Nevertheless, ECMO after ASO involves discriminating patient selection, prompt timing to initiate and good care team management to achieve acceptable results and survival.

Keywords: Extracorporeal membrane oxygenator (ECMO), arterial switch operation (ASO), transposition of the great arteries (TGA) and cardiopulmonary.

Application of extracorporeal membrane oxygenation (ECMO) in children after open heart surgery with low cardiac output is an important bridging modality for possible recovery.^(1, 2) An additional indication for ECMO are in infants beyond the ideal age for corrective arterial switch operations (ASO) where the degradation of the left ventricle was assessed as reversible.⁽³⁾ The first successful use of ECMO after transposition of the great arteries (TGA) repair was reported in 1972 where the patient went into profound cardiac shock after the operation and was supported by a modified heart-lung machine.⁽²⁾ This was the first successful use of ECMO for pediatric cardiac support. The overall survival rate of post-cardiotomy veno-arterial ECMO in congenital heart diseases in infants remains about 40 - 50% for the past decade from Extracorporeal Life Support Organization (ELSO) data. There is a multicenter study from 37 countries in North America that reported

a 48.1% survival rate to hospital discharge of ECMO postcardiac surgery.⁽⁴⁾ However, a number of recent reports on the use of ECMO for post-cardiotomy or pulmonary failure and pre-cardiotomy bridge to transplantation have shown encouraging results.⁽⁴⁾ The need for extracorporeal life support (ECLS) following repair of congenital heart disease is rare.⁽⁵⁾ ASO is a complex procedure with associated early morbidity and mortality although there have been some reports that mortality has fallen to the range of 0 - 6%.⁽⁶⁾ The objective of this study is to review our experience of patients undergoing ECMO after ASO from a single institution.

Case report series

Between 2007 - 2017, 143 ASO cases were operated at King Chulalongkorn Memorial Hospital (under The Thai Red Cross Society), and Faculty of Medicine, Chulalongkorn University. This is a descriptive study whereby medical data records are retrieved. The protocol was reviewed and approved by the Ethics Committee of the Faculty of Medicine, Chulalongkorn University.

There were 4 patients referred from other hospitals to our institution who underwent ASO and required ECMO support. All 4 male babies received

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veno-arterial ECMO following ASO. The patients' mean age at operation was 20 (12 - 41) days and mean weight was 3.3 (2.8 - 3.9) kilograms. The mean bypass time was 234 (187 - 308) minutes, and the mean cross clamp time was 88 (61 - 114) minutes. The mean ECMO duration time was 106 (70 - 194) hours. (Table 1) No coronary artery related untoward event was noted in all of the cases as a possible cause for low cardiac output post-operatively. Two babies (50%) survived after ECMO but only one was discharged alive.

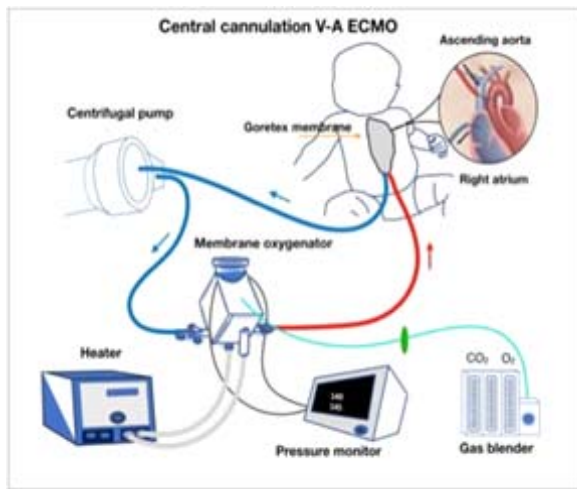
The ECMO circuit included the membrane and centrifugal head pump connected by coating tubing (Quadrox-iD pediatric, Rastatt, Germany) which was used for all patients. The ascending aorta was cannulated using an 8-Fr and 10-Fr aortic cannula (Medtronic, DLP) and the right atrium was cannulated using a 21-Fr venous cannula (Polystan) and 20-Fr venous cannula (Medtronic, DLP). The pump flow rate was set at 100 - 150 ml/kg/min to achieve adequate blood pressure and acceptable perfusion. CO₂ was kept within the normal range of 40 - 45

mmHg and 100% oxygenation of arterial blood gas. The ventilator was set as a lung protective setting mode at a tidal volume less than 5 mL/kg, inspiratory pressure a less than 30 cmH₂O, and a respiratory rate of 10 mm. The positive end-expiratory pressure (PEEP) was adjusted to maintain or to keep the alveolar open. Body temperature was maintained around 36 - 37°C by the integrated heater ECMO circuit and blanket. A heparin infusion was adjusted to maintain an activated clotting time (ACT) around 180 - 200 seconds. The platelet count was maintained above 100,000 per microliter and other blood components were adjusted as needed. We arranged bedside echos and chest X-ray films daily to check cardiac functions, effusions and cannula position. Once the heart improved and lung function was adequate, weaning ECMO was started by decreasing the pump flow and observing hemodynamics. After tolerating pump flows to 50 mL/kg/min with stable hemodynamic status, decannulation was then performed (Figure 1).

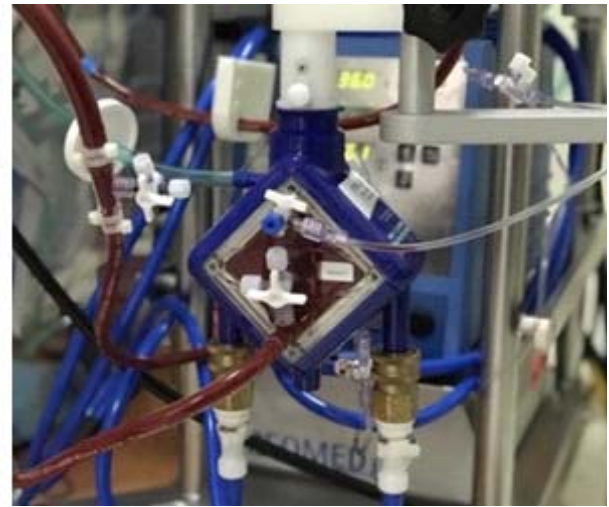
Table 1. Demographic data.

Case	Sex	Age (d)	BW (kg)	Diagnosis	Procedure	Pump time (min)	Clamp time (min)
1	Male	18	3.20	d-TGA, muscular VSD, ASD, PDA post Balloon atrial septostomy	ASO, direct closure VSD, closure ASD, PDA ligation	187	88
2	Male	41	2.81	d-TGA, left sided arch, Perimembranous VSD 8 -9 mm , ASD, large PDA 1.5 mm to right flow separated origin of coronary arteries	ASO, closure VSD, closure ASD, PDA ligation	238	91
3	Male	12	3.9	d-TGA, small VSD, Restricted ASD, Single ostium left coronary, PDA, dilate RV and LV	ASO, Closure VSD, closure ASD, PDA ligation	308	61
4	Male	14	3.3	d-TGA, muscular VSD, PDA and restricted ASD right side coronary aortic arch	ASO, closure VSD, closure ASD, PDA ligation	202	114

Abbreviations: d-TGA, Dextro-Transposition of Great Arteries; VSD, Ventricular septal defect; ASD, Atrial septal defect; PDA, Patent ductus arteriosus; ASO, Arterial switch operation; RV, Right ventricle; LV, Left ventricle.



(A)



(B)

Figure 1. ECMO circuit.

Case 1:

An 18-day-old baby weighing 3.2 kg was diagnosed with d-TGA, small muscular VSD 2 mm, ASD and PDA. There was marked cardiomegaly with an ascending aorta diameter of 10 mm, the thickness of the left ventricular posterior wall at end-systole (LVPWs) was 0.45 mm with normal coronary artery pattern. Balloon atrial septostomy was created before the operation. ASO with a VSD closure (cardiopulmonary bypass (CPB) time of 187 minutes and cross clamp time of 88 minutes with intermittent cold St. Thomas crystalloid cardioplegia) was performed 13 days after admission. Postoperatively, the patient presented with cardiac tamponade with echocardiography showing pericardial effusion. Immediate mediastinal exploration was done with findings of blood clots. However, the patient still had persistently low blood pressure due to poor ventricular contraction requiring ECMO. The chest remained opened and ECMO via central cannulation was started. The clinical condition of the baby during ECMO had persistent low cardiac output which required persistent volume replacement but was unresponsive to high dose inotropes. He also had anuria and subsequent azotemia that required peritoneal dialysis. The baby deteriorated further and subsequently died from multi-organ failure after 9 days on ECMO (Figure 2).

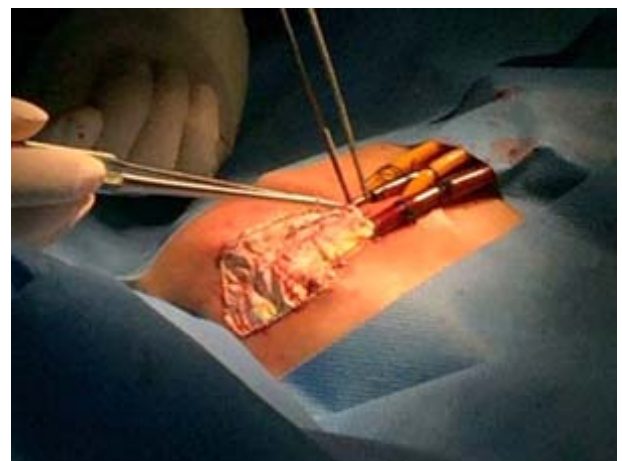


Figure 2. Open chest with ECMO cannulas.

Case 2:

A 41-day-old baby with 2.81 kg of weight was diagnosed with d-TGA, perimembranous VSD ASD and PDA, the LVPWs was 0.63 mm with normal coronary artery pattern. An ASO with VSD and ASD closure (CPB time 238 minutes and cross clamp time 91 minutes with crystalloid cardioplegia) was performed 5 days after admission. Two days after ASO, the patient developed cardiac tamponade requiring re-operation. The patient was noted to have poor heart contraction after mediastinal exploration despite high doses of adrenaline. ECMO was then instituted but his cardiac function had never recovered and expired after 4 days.

Case 3:

A 12-day-old baby weighing 3.9 kg, was diagnosed as d-TGA with small inlet VSD. There was a prominent conal septum causing mild subvalvular PS, ASD 2 mm (non-restrictive) and LVPWs was 0.34 mm with a single coronary ostium. The patient was also diagnosed with glucose-6-phosphate dehydrogenase (G6PD) deficiency with neonatal jaundice and anemia. ASO with VSD ASD (CPB 308 minutes and had cross a clamp time of 61 minutes utilizing the same cardioplegia technique) closure was performed 10 days after admission. The patient had a cardiac arrest after sternal closure and the decision to start ECMO was made. One day after ECMO, the patient was re-operated on for persistent bleeding. After 3 days, the patient was weaned off ECMO based on improvement in left ventricular contraction through 2D echo. The baby was extubated after 12 days of improvement and was discharged after 2 more weeks. The boy is now 2 years old and still well up to the last follow up.

Case 4:

A 2-week-old baby weighing 3.3 kg, diagnosed as d-TGA with muscular VSD 3.8 mm and a right side aortic arch. LVPWs was 0.65 mm with normal coronary pattern. ASO with VSD closure (CPB 202 minutes and cross clamp time 114 minutes with use of blood cardioplegia) was performed on the 12th hospital day. At the 1st intensive care unit day, the patient developed cardiac arrest due to blood clots in the chest resulting in cardiac tamponade. Despite exploration and evacuation of the clots the patient still required ECMO support via central cannulation for low cardiac output. The patient tolerated weaning from ECMO after 3 days of support. However, after 2 weeks, the patient developed fungal sepsis. The baby died one week later despite antibiotics and aggressive medical support.

Discussion

ASO for d-TGA is a complex procedure with high mortality and morbidity for neonates even with ECMO support. ECMO after ASO studies are sparse in medical literature. We found a 33%⁽⁷⁾ and 50%⁽⁸⁾ survival of ASO after ECMO from a small number of patients in some studies. In our experience, we have a 50% survival rate after ECMO but only one survived to hospital discharge. The indication for ECMO in our

institution is similar to other institutions with clinical findings of persistent hypotension or progressive decline in cardiac function despite maximal inotropic support. The main problems we found in our study were persistent hypotension, bleeding followed by renal dysfunction and infection which were the most frequently encountered ECLS-related complication.^(8,9) In 2018, the ELSO showed the result of neonate cardiac ECMO survival rate as 64% and those who survived to discharge as 41%. However, the cardiac procedure has to be definite about correcting any residual lesions before placing an ECMO for the patient.⁽⁷⁾ Although ECMO support after ASO is related to many complications, it is known that poor postoperative ventricular function brings the question of 3 major issues. These are poor myocardial preservation, technical issues with coronary transfer leading to postoperative ischemia and deconditioned left ventricular due to older age. We noted that 2 of the patients who died were older (18 and 41 days). Although LVPWs value was acceptable for operation, older patients might be sensitive to myocardial preservation. Perfusion strategies should be well performed (type, timing, temperature) in order to achieve a good myocardial preservation. A good repair of anatomic malformation is likewise essential to successful outcomes in case of poor ventricular contraction and requires ECMO in postoperative patients. Postoperative cardiac catheterization is also an objective modality for proving whether coronary transfer leads to postoperative ischemia. Even though ASO carries many complications, the rate of survival to hospital discharge was better in patients with biventricular repair than those with univentricular hearts.⁽⁹⁾ ASO carries a good prognosis as one study showed encouraging late survival of ASO 97.6% at 15 years.⁽⁶⁾ In our limited experience, we had a 12.2% mortality rate after ASO (139 patients for 10 years) which is much higher than most institutions that performed complex open heart procedures. The use of ECMO in our series of 0.02% and literature have shown its use to allow time and recovery of cardiac function and a possible “bridge to recovery”.^(5, 8, 10) Furthermore, the decision to utilize ECMO postoperatively should be based on patients’ reversible or reasonable criteria. This should include prompt and proper time to set up ECMO and management of complications during ECMO⁽⁸⁾ that children will hopefully survive their critical situation.

Conclusion

ECMO after ASO carries a high mortality but nevertheless is an invaluable support for a very difficult operation in a complex patient.

Conflict of interest

None of the authors has any potential conflict of interest to disclose.

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