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Review

# Extracorporeal Membrane Oxygenation in Pediatric Patients With Congenital Heart Disease: Surgical Considerations

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**ABSTRACT**

**The use of extracorporeal membrane oxygenation (ECMO) in the paediatric population has increased over time, with the ability to rescue pulmonary and cardiovascular deterioration. ECMO can be utilised by neonates and children with congenital heart disease in both preoperative and postoperative settings to improve survival and minimise morbidity. ECMO cannulation strategy must be tailored to the age, weight, and physiologic state of the patient. Careful patient selection and rapid deployment of ECMO may improve survival and morbidity in patients with congenital heart disease.**

**RÉSUMÉ**

**L'oxygénation extracorporelle sur oxygénateur à membrane (ECMO ou *extracorporeal membrane oxygenation*) est de plus en plus utilisée dans la population pédiatrique, et permet d'éviter une détérioration pulmonaire et cardiovasculaire. L'ECMO peut être utilisée en contexte pré- et postopératoire chez les nouveau-nés et les enfants atteints d'une cardiopathie congénitale pour améliorer la survie et réduire au minimum la morbidité. La stratégie de canulation de l'ECMO doit être adaptée à l'âge, au poids et à l'état physiologique du patient. Une sélection rigoureuse du patient et un déploiement rapide de l'ECMO peuvent améliorer la survie et réduire la morbidité chez les patients atteints d'une cardiopathie congénitale.**

Mechanical circulatory support (MCS) is an integral part of the management of paediatric patients with congenital heart disease (CHD). Mechanical circulatory support in the acute short term typically utilises extracorporeal membrane oxygenation (ECMO), which offers both circulatory and respiratory support, whereas more chronic cardiac disease processes are managed with the use of ventricular assist devices. ECMO is used to support paediatric patients with severe cardiovascular and/or pulmonary collapse and importantly, ECMO plays an important role in cardiopulmonary resuscitation (ECPR). ECMO provides effective gas exchange and oxygenation and achieves adequate systemic perfusion to aid end-organ recovery. In children with congenital heart disease, ECMO can play an important role in preoperative stabilisation and perioperative recovery. This review will discuss the current use of ECMO in neonates and children with CHD.

## ECMO in Neonates and Children

Use of ECMO has increased over the years and has been documented by the multi-institutional registry managed by the Extracorporeal Life Support Organisation (ELSO). In the most recent ELSO registry data (2009-2022), there have been more than 45, 000 paediatric ECMO runs, with approximately 25,000 in children and 21,000 in neonates (pulmonary, cardiac, ECPR) (Fig. 1).[1,2] Cardiac veno-arterial (VA) ECMO accounts for approximately 7000 neonates and 11,500 paediatric ECMO runs, with 7800 patients requiring ECPR.[1] CHD accounts for 80% of neonates and 60% of paediatric patients requiring ECMO for cardiac failure, whereas the remainder is composed of those with cardiomyopathy, irretractable arrhythmia, and acute myocarditis.[3] The Society of Thoracic Surgery (STS) registry reports that 2.8% of patients require ECMO in the perioperative setting, with risk factors identified as patients undergoing higher complexity surgery (STS−European Association for Cardio-Thoracic Surgery congenital heart surgery [STAT] categories 4 and 5) and neonatal age group.[4,5] Survival after cardiac ECMO is approximately 45% in neonates and 54% in paediatric patients, compared with 70% and 60%, respectively, for neonates and paediatric patients requiring isolated respiratory ECMO. Survival to discharge for patients undergoing ECPR is approximately 40%.[1]

ECMO configurations include veno-venous ECMO for pure respiratory failure peripheral or central VA ECMO supports cardiac or cardiopulmonary failure (Fig. 2). Veno-venous ECMO involves cannulation of systemic veins (typically femoral-femoral or internal jugular-femoral) to support ventilation and oxygenation in patients with respiratory failure. In contrast, VA ECMO is typically utilised in the setting

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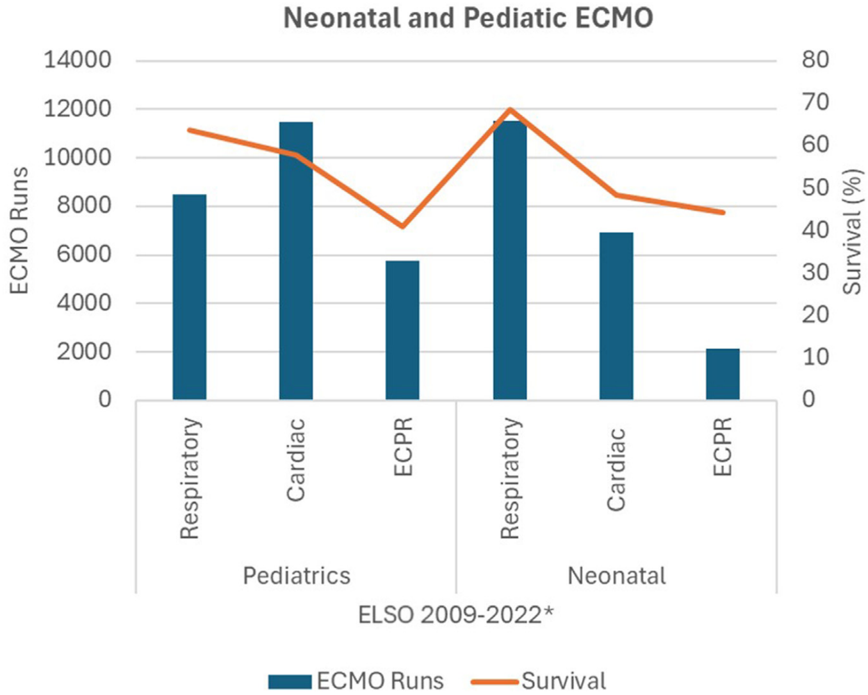


**Figure 1.** Aggregate Extracorporeal Life Support Organization (ELSO) data for paediatric and neonatal ECMO runs and survival to Hospital Discharge. Graphical representation of ELSO data from registry data.

of cardiogenic shock unresponsive to maximal medical management. In the context of CHD, VA ECMO can be used for perioperative stabilisation as a bridge to surgical correction, support after failure to separate from cardiopulmonary bypass, support for postoperative cardiopulmonary failure, and, with the use of ECPR, cardiac arrest.

## Considerations for Initiation of VA ECMO

Initiation of VA ECMO in the setting of CHD is a temporising measure that aims to achieve either 1) stabilisation to recovery or surgical intervention, 2) bridge to long-term MSC or transplantation, or 3) bridge to further decision making. The optimal timing of ECMO initiation remains ill defined but is dependent on unique patient clinical factors. Early initiation, before severe tissue and end-organ compromise, is thought to improve survival and minimise long-term morbidity. It is unclear if early initiation in the operating room vs intensive care demonstrates a clear advantage. One retrospective study demonstrated a survival benefit for children cannulated in the operating room compared with the intensive care unit,[6] however this is not consistently reported in the literature. Contraindications to initiation of ECMO have been outlined by ELSO and include prolonged cardiogenic shock, prematurity < 32 weeks, extreme low birth weight (< 1.5 kg), uncontrolled bleeding, severe intraventricular hemorrhage, severe brain injury, severe chromosomal abnormalities, and irreversible primary disease without feasible long-term palliation.[7] Weight < 2.0 kg and prematurity of < 35 weeks are regarded as relative contraindications and require careful consideration of the risks vs benefits.

Risk factors for ECMO mortality include premature infants, complex CHD such as single-ventricle physiology, low birth weight, indicators of poor cardiorespiratory status before initiation of ECMO, and increased duration of ECMO support.[4,5,7-9]

In the setting of CHD, adoption of a multidisciplinary heart center approach with anticipatory parental counselling allows for preemptive understanding of goals of care within the context of the patient's long-term clinical trajectory. Understanding candidacy for ECMO and ECPR outside of the immediate perioperative period should be discussed and documented in the patient's chart.

## VA ECMO in CHD Patients: Survival and Complications

ELSO registry data demonstrates survival after ECMO of approximately 60%.[2] For children requiring VA ECMO in the postoperative period, overall mortality is 53%, which contrasts with the average mortality of those undergoing surgery for CHD (1%-20%) across various STAT categories.[10] Complications related to VA ECMO include neurologic injury (embolism, hemorrhage), renal dysfunction, hemorrhage, and infection.[2] ELSO data demonstrate that approximately 30% of patients require renal replacement therapy while on ECMO, and seizures and stroke are seen in approximately 20%. In cardiac VA ECMO patients, neurodevelopment scores are below those matched to non-ECMO control subjects, suggesting long-term consequences on gross motor, cognitive, and language domains.[11] Long-term outcome metrics such as indices quantifying neurologic injury, impact of critical illness, and quality of life are required to fully understand the impact of ECMO and development of strategies to support these patients as they grow.[11-15]

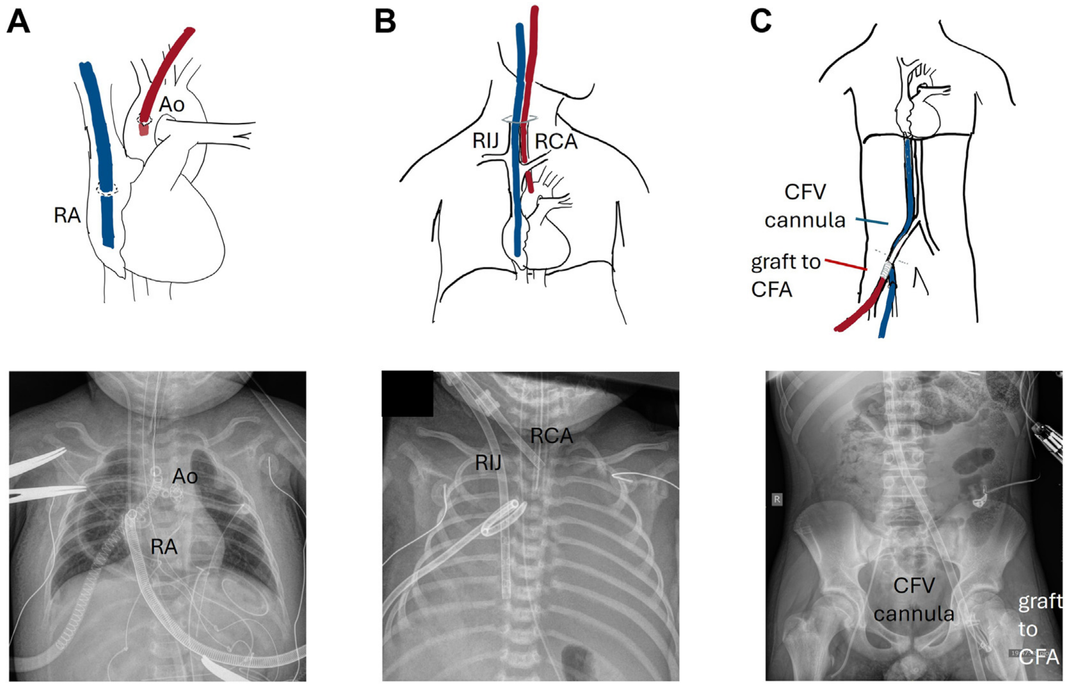


**Figure 2.** Infant cannulation strategies for VA ECMO. (**A**) Cenral cannulation with open chest via direct cannulation of the ascending aorta and right atrium. (**B**) Peripheral neck cannulation via right common carotid artery and right internal jugular vein. The venous cannula should sit with the tip at the inferior caval junction and the arterial cannula sits in the proximal aortic arch/ascending aorta. Pigtail drain in the right chest. (**C**) Femoral cannulation, venous cannula in the common femoral vein, advnced so that the tip sits in the right atrium, the arterial cannula is in the common femoral artery and advanced toward the external iliac, or connected to a graft placed end-to-side onto the common femoral artery. Ao, aorta; CFA, common femoral artery; CFV, common femoral vein; RA, right atrium; RCA, right common carotid artery; RIJ, right internal jugular vein.

## Use of VA ECMO in CHD

### Preoperative stabilisation in unrepaired CHD

Use of VA ECMO to stabilise unrepaired CHD, particularly in the neonate, can be used to bridge surgical repair and aid end-organ recovery. In an STS registry report, approximately 0.5% of patients utilised ECMO stabilisation before surgical repair.[3] CHD lesions such as obstructive total anomalous pulmonary veins, tetralogy of Fallot with absent pulmonary valve, and dextro-transposition with pulmonary hypertension may present with severe cardiopulmonary collapse shortly after birth, necessitating VA ECMO to stabilise and facilitate surgical repair. Other lesions, such as circular shunts in neonatal Ebstein anomaly, may be suspected prenatally and require multidisciplinary teams to facilitate planned delivery with ECMO standby. With increased fetal echocardiography programs and antenatal counselling, referral to centres affiliated with cardiovascular surgery and ECMO capabilities is paramount to minimising hemodynamic compromise in these at-risk patients.

### Postoperative use of VA ECMO and identification of residual lesions

Use of VA ECMO in the postoperative period can occur after a failure to wean from cardiopulmonary bypass in the operating room or because of low cardiac output or incessant arrhythmia in the intensive care unit. Analysis of the STS registry demonstrates that postoperative ECMO occurs in 2% to 3% of CHD patients.[4,5] Operations associated with higher mortality risk are associated with higher need for postoperative ECMO after 48 hours. Mortality is highest in patients undergoing Norwood procedure (17%), complex arterial switch operation (14%), Truncus arteriosus repair (9%), and Ross Konno operation in infancy (9%).[4,5]

Intraoperative initiation of VA ECMO due to failure to wean from cardiopulmonary bypass is more likely to occur in neonates with important preoperative risk factors such as previous cardiac arrest and need for higher-complexity surgery.[16-18] Low cardiac output and arrhythmia in the immediate postoperative period (24-48 h) are common and typically respond to medical management. However, persistent end-organ dysfunction (eg, oliguria), widening arterial-venous saturation differences, and elevated lactate unresponsive to conventional medical management necessitate VA ECMO (where appropriate). During the ECMO course, it is imperative to understand the burden of residual lesions, with low threshold for imaging, such as diagnostic catheterisation, to identify residual lesions, allow surgical or interventional correction, and facilitate successful ECMO weaning. This is particularly important in the setting of arrhythmia, whereby coronary ischemia may be a culprit lesion after arterial switch or Ross procedure. Studies have shown that correction of residual lesions leads to shorter duration of ECMO and potential improved survival.[19-22]

### ECMO during isolated transcatheter procedures

Patients placed on VA ECMO in the postoperative period may require diagnostic transcatheter procedures to identify residual lesions requiring surgical revision or transcatheter interventions to allow for successful weaning of ECMO support. In addition, transcatheter procedures can be associated with risk of cardiac arrest or low cardiac output that may necessitate initiation of ECMO in the catheterisation lab. Formalized ECMO standby protocols to facilitate communication of risk and coordinate line placement, circuit preparation, and surgical personnel during the time of catheterisation are paramount to minimising end-organ dysfunction in the event of hemodynamic instability or cardiac arrest. Our institution has both an ECMO-aware and an ECMO standby triage system (Table 1) to balance resource allocation and ECMO readiness.

### ECMO in the context of CPR

Initiation of ECMO during cardiopulmonary resuscitation or within 20 minutes after return of spontaneous circulation is considered to be ECPR.[23,24] Early initiation should be considered in patients with lesions that may limit effective cardiac output or pulmonary blood flow (and oxygenation) with chest compressions and where cerebral perfusion may be compromised. Examples include pulmonary hypertension, critical aortic stenosis, and single-ventricle physiology.[23] Timely initiation of ECPR is critical in patients after CHD repair because of the potential for limited physiologic reserve and altered circulation in some populations. Tertiary centres will typically have in-house surgical and perfusion personnel to facilitate rapid deployment of ECPR.[23] Patients < 2 weeks after surgery typically require open chest central cannulation, whereas those > 2 weeks after chest closure typically use peripheral cannulation (neck or femoral vessels). Given multiple team involvement, resuscitation and ECPR simulation play a key role in coordinating resuscitation and cannulation with the goal of minimising time from initiation of ECPR alert to achieving full flows (< 30 min).[24,25]

## Surgical Considerations in VA ECMO for CHD

### Cannulation considerations

Cannulation for VA ECMO requires cannulation of systemic venous return and systemic arterial access to achieve target flows of around 100 mL/kg. Typically, in children weighing less than 15-20 kg, peripheral cannulation can be initiated preferably through the right common carotid artery and the right internal jugular vein via an incision ∼ 1 cm above the clavicle over the anterior border of the sternocleidomastoid muscle of the right neck. Alternative access through the left neck can occur if there is concern for right internal jugular patency owing to thrombosis or previous cannulation. For children weighing more than 20 kg with adequate femoral vessel size to accommodate flows required, femoral cutdown for femoral vessel access can be initiated. Percutaneous femoral cannulation can be feasible in larger adolescents (see below for discussion on distal limb perfusion). In postsurgical patients, central cannulation via the aorta and right atrium is typically a rapid process if previous surgery was < 2 weeks before. For children with severe sepsis or decompensated heart failure, initiation of sternotomy for central VA ECMO may be required to facilitate high ECMO flows. It requires coordination with intensive care physicians for patient selection.

A successful ECMO run requires cannula size selection in the context of anticipated ECMO flow requirements. Patients with open systemic-pulmonary shunts or severe sepsis can require 150-200 mL/kg ECMO flow, which can be challenging in centrifugal-pump ECMO circuits if the arterial cannula size (as opposed to venous drainage) is not maximised (Table 2). If venous drainage is inadequate, additional venous cannulas can be added to improve flows. Cannulation strategy can be affected by the patient's previous surgical procedure history and institutional preferences.[26] Previous diagnostic imaging, vascular Doppler recordings, and surgical history should be reviewed to identify any potential issues with vascular anatomy and vessel patency. Documentation of vascular integrity is critical to successful cannulation in an emergency setting. Whenever possible, ligated carotid vessels or thrombosed/stenosed neck and femoral vessels should be flagged in the preoperative consultation and the patient's chart.

Difficulty in peripheral cannulation can occur in the setting of indwelling central venous lines (looped lines, hematomas, thrombosis) and necessitates review of the chest x-ray and vascular Doppler recordings (when available) before cannulation. In children weighing < 20 kg with unilateral

**Table 1.** The Hospital for Sick Children ECMO readiness for the catheterisation lab procedures for paediatric patients with congenital heart disease

| | ECMO-aware | ECMO standby |
|---|---|---|
| Triage | • Low to moderate risk level<br>• Stable physiology with risk of destabilisation<br>• Low to moderate procedural complexity | • High risk level<br>• Higher procedural complexity<br>• Risk of acute decompensation |
| Catherisation Team | • Triage ECMO level for case<br>• Notifies ECMO standby group<br>• *Preprocedural huddle:* plan, access, special consideration, ICU disposition | |
| Anaesthesia | *Preprocedural huddle:* line placement, head position, resuscitation readiness | |
| Perfusion | • Cannulas and circuit in catherisation lab<br>• Blood ordered<br>Aware: assigned perfusionist mot in the catherisation lab | Standby: perfusionist in the catherisation lab |
| Cardiovascular surgery | • ECMO consultation<br>• *Preprocedural huddle:* positioning, access, cannulas confirmed, equipment<br>Aware: assigned primary surgeon and fellow not in the catherisation lab | Standby: Primary surgeon assigned, and cardiovascular surgery fellow in the catherisation lab |

ECMO, extracorporeal membrane oxygenation.

**Table 2.** The Hospital for Sick Children approximate arterial cannula size for peripheral ECMO using centrifugal circuit

| Weight, kg | Size, F | Approximate flow rate, mL/min |
|---|---|---|
| Up to 3 | 8 | 600 |
| 3-5 | 10 | 800 |
| 5-8 | 12 | 1000 |
| 8-15 | 14 | 1500 |
| 15-25 | 15 | 2500 |
| 25-35 | 17 | 3500 |
| 35-50 | 19 | 4500 |
| 50-60 | 21 | 5500 |
| > 60 | 23 | 6000 |

Venous cannulas are per manufacture guidelines. Central cannulation follows cardiopulmonary bypass cannula sizes.

ECMO, extracorporeal membrane oxygenation.

occluded cerebral vessels, alternative cannulation strategies (eg, central ECMO) should be considered. Central cannulation may be difficult in patients with extensive aortic arch reconstructions (eg, Norwood procedure); in patients with a clipped innominate polytetrafluoroethylene (PTFE) graft (used for cannulation in the operating room), this can be used to facilitate rapid initiation of ECMO. Patients with a Le Compte procedure postarterial switch operation may also have difficult aortic access requiring careful exposure. Thorough knowledge of the patient anatomy and surgical history is required for successful ECMO cannulation.

## Circuit maintenance

Proper anticoagulation and management of ECMO circuits is paramount to a successful ECMO run. During the ECMO run, cannulas should be inspected for fibrin and clot build-up. In particular, neonates with smaller cannulas require tight monitoring of heparin levels to decrease probability of clot formation. One must be prepared to do circuit changes or cleaning of cannulas, because clots risk circuit failure or embolisation of mobile clot. In addition, the duration of ECMO should be considered when considering ECMO cannulation strategy. If longer duration of support is anticipated, peripheral ECMO may be preferred, either as the primary cannulation strategy or by conversion if there is failure to wean from ECMO. If longer central ECMO is required, tunnelling of cannulas to facilitate chest closure is ideal.

## Left ventricular decompression

By convention, VA ECMO unloads the right heart through right atrial venous drainage. In the setting of adequate left ventricular (LV) function (eg, pulmonary hypertension), venting of the LV may not be required. However, in the setting of LV dysfunction, myocardial recovery is aided by LV decompression to decrease the metabolic requirements of the myocardium. Signs of potential susceptibility for left-side distention include lack of pulsatility of the arterial line, radiographic evidence of pulmonary edema, echocardiographic evidence of elevated LA pressure, LV distention, and associated stasis. Sequelae include LA hypertension with concomitant pulmonary hemorrhage, myocardial irritability, and intracardiac thrombosis. Decompression of LV mitigates the need for inotropes to maintain LV ejection and facilitates LV recovery. If LV distention is suspected, rapid LA decompression is necessary, and shorter time to decompression has been associated with improved outcomes.[27,28] LA venting strategies differ for open- and closed-chest ECMO (Fig. 2). In closed-chest (peripheral) ECMO, atrial septostomy by the catheterisation lab is preferred.[26] In larger paediatric patients, alternative options, such as peripheral placement of cannula, can be directed across the atrial septum, or Impella (placement across the aortic valve) can be used. In open-chest (central) ECMO, an LA vent can be placed via the right upper pulmonary vein or, less commonly, the LA appendage (Fig. 3). Pulmonary artery (PA) venting or venting through the LV apex is rarely used at our institution. Careful attention to the size of vent is necessary to prevent vent thrombosis. Monitoring through use of a flow probe on the LA vent limb of the venous circuit should be considered to alert changes in levels of LV decompression.

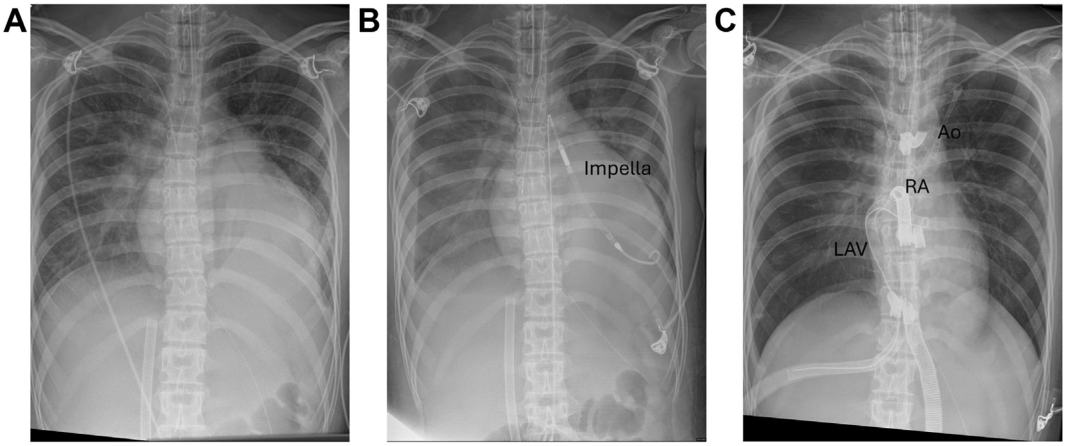


**Figure 3.** Left ventricular decompression strategies. (**A**) Peripheral cannulation via femoral artery and femoral vein in patient with dilated cardiomyopathy. (**B**) Attempted decompression with Impella 2.5 with persistent cardiomegaly. (**C**) Conversion to central ECMO (aorta and right atrium) with left atrial venting (LAV) via the right superior pulmonary vein. Cannulas were tunnelled and the skin closed. RA, right atrium.

### Peripheral cannulation and vascular integrity

In femoral peripheral cannulation, cannulation can result in distal limb ischemia or congestion. When time facilitates, use of a chimney graft (6 or 8 mm) on the femoral artery ensures distal perfusion. If direct cannulation of the femoral artery is required and there is concern for poor distal perfusion, distal limb perfusion can be initiated by connecting either a vascular access sheath or small arterial cannula (8 or 10 F) in the superficial femoral artery to the ECMO circuit.[26] Venous congestion has been reported in the literature, with some placing a distal venous cannula to decompress the leg.[29] In smaller adolescent patients, cannulation of femoral artery with contralateral femoral venous cannulation can mitigate limb ischemia/perfusion issues. Regardless, monitoring limb perfusion and drainage through near-infrared spectroscopy, Doppler, and saturation monitoring should be done.

Cannulation of peripheral vessels places vessels at risk for thrombosis and stenosis after decannulation. Maintenance of arterial patency should be achieved by vessel reconstruction after decannulation, avoiding ligation whenever possible. Thrombosis prophylaxis should be considered within the clinical context of the patient, and vascular surveillance (vascular Doppler) should be included in follow-up surveillance.

## Special Populations: Single-Ventricle Physiology

### Stage 1 palliation: Norwood operation

Single-ventricle physiology was once seen as a contraindication to VA ECMO, but in the recent era, up to 20% of palliated single ventricles have been supported by ECMO.[30] Hypoplastic left heart syndrome and its associated variants undergo stage 1 palliation (Norwood procedure), which is establishment of unobstructed systemic outflow (arch reconstruction and Damus-Kaye-Stansel anastomosis), adequate intracardiac mixing, and secured pulmonary blood flow through either a modified Blalock-Taussig-Thomas (mBTT) shunt or right ventricle (RV)—PA conduit. Risks for requiring ECMO after stage 1 palliation include lower birth weight, aorta < 2 mm, and long cardiopulmonary bypass time.[4,31] In the Single Ventricle Reconstruction Trial, the mBTT shunt was associated with increased probability of needing CPR but the need for ECMO was similar compared with the RV-PA conduit.[31] In patients who have undergone a Norwood procedure, ECMO can be initiated after failure to separate from cardiopulmonary bypass or postoperatively because of low cardiac output, cardiac arrest, or shunt thrombosis. A single-centre study demonstrated that those requiring ECMO for shunt thrombosis have better outcomes than those with low cardiac output or hypotension.[9]

The ECMO cannulation strategy in the early postoperative phase is central VA ECMO with either direct cannulation of the reconstructed arch or use of the clipped PTFE innominate graft and the right atrium (Fig. 4A). Cannulation using peripheral neck cannulation can be done more than 2 weeks postoperatively. In the setting of an mBTT shunt, ECMO flows are typically 150-200 mL/kg with the shunt left open and rest ventilatory settings. Attention is paid to systemic perfusion in the setting of potential diastolic run-off, with clipping the shunt to downsize or even occlude the BTT shunt diameter, if needed. If ventricular function is good, the BTT can be left open if perfusion pressures are good. If ventricular function is reduced, then occluding the BTT shunt maximises coronary perfusion and end-organ perfusion. Note that an RV-PA conduit Norwood procedure does not require clipping or occlusion because it does not cause any diastolic run-off. Survival to hospital discharge after ECMO varies from 29% to 80% across series.[4,9,31,32]

For many centres, patients who undergo the hybrid procedure (ductal patency with bilateral PA bands) are higher-risk patients (low birth weight, prematurity, or contraindications to heparinisation for cardiopulmonary bypass) and thus have contraindications for ECMO initiation. Cannulation requires careful consideration of progressing to a more stable physiology with a Norwood procedure, albeit with increased risk of postoperative ECMO.

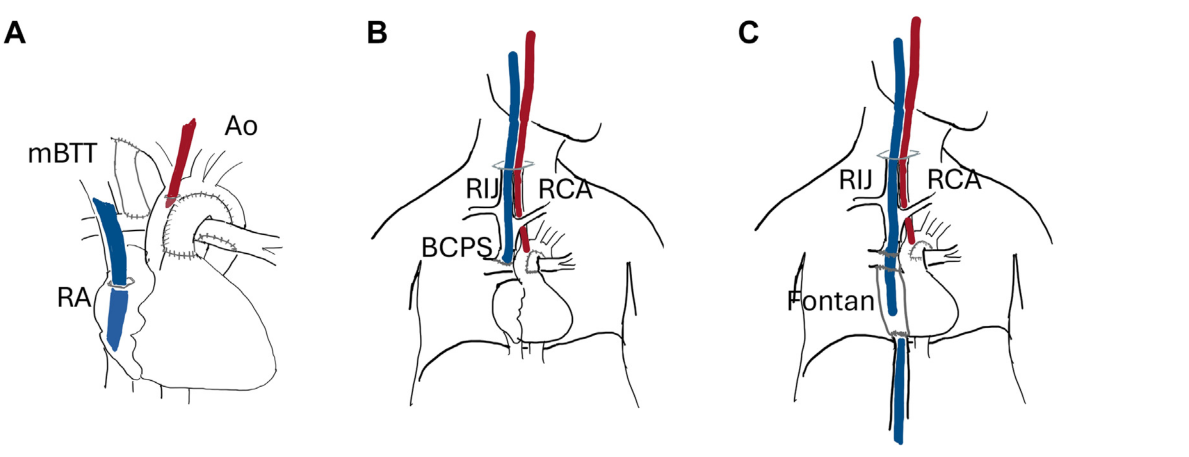


**Figure 4.** Potential ECMO Configurations for single-ventricle patients. (**A**) Central cannulation: right atrium (RA) and aorta (Ao) after first-stage palliation with Norwood and modified Blalock-Tausing-Thomas shunt (mBTT). Care must be taken to ensure aortic cannula position does not lead to pulmonary overcirculation in Norwood mBTT patients. (**B**) Peripheral cannulation: right carotid artery (RCA) and bidirectional caval pulmonary shunt via the right internal jugular vein (RIJ). (**C**) Peripheral cannulation of extracardiac Fontan through right common carotid artery, RIJ, and additional drainage cannula introduced in the right caval pulmonary shunt with advancement into the inferior vena cava.

### Stage 2 palliation: Bidirectional caval pulmonary shunt

Patients typically undergo stage 2 palliation at 4-6 months of age and weight around 5-6 kg. The bidirectional caval pulmonary shunt (BCPS) involves redirection of the superior vena cava venous blood to the pulmonary circulation through direct anastomosis to the PA. ECMO initiation is rare in this population, representing < 1% of those reported in the STS registry.[4] Care must be taken when considering cannulation strategy in this population. Owing to separation of upper and lower systemic venous blood flow, failure to decompress the BCPS circuit may result in impaired cerebral perfusion due to venous congestion resulting in neurologic injury or failure to achieve full flows. In the early postoperative phase, central cannulation should adequately decompress the BCPS circuit. If peripheral cannulation is the primary mode, peripheral cannulation of the neck vessels is preferred with potential conversion to central, if needed (Fig. 4B). Centres have reported IVC access via extraperitoneal dissection, use of a dual-lumen cannula in the right atrium, and percutaneous transhepatic cannulation of the IVC.[29] Survival to hospital discharge is reported for approximately 40%.[33]

### Stage 3 palliation: Total caval anastomosis (Fontan operation)

The completion of the Fontan operation typically occurs between the ages of 2 and 4 years and involves incorporation of the IVC venous flow directly to the pulmonary circulation, typically through a PTFE conduit. This Fontan circuit bypasses the ventricle and results in passive pulmonary flow into the systemic ventricle. Failure of the Fontan circulation can be acute or chronic with higher mortality associated with late-phase failure. Acute Fontan failure may require ECMO, and the goal should be decompressing the Fontan circuit while permitting some flow in the extracardiac conduit to prevent stasis. Central cannulation in the early postoperative phase can provide adequate support, but peripheral cannulation outside the perioperative period may require upper and lower venous drainage to achieve decompression and full flows (Fig. 4C).[23] Some centres consider veno-arterial venous EMCO, with 1 drainage cannula (femoral vein) and 2 return cannula (right carotid artery and internal jugular vein [stasis prevention]).[25] Survival to hospital discharge is 35%-50% for those requiring ECMO.[34,35] Those presenting with chronic Fontan failure with multiple resultant comorbidities, such as protein losing enteropathy and plastic bronchitis, have significant mortality risk when contemplating ECMO.

## Special Populations: Heart Transplantation

Use of ECMO as a bridge to transplant is not generally used owing to inferior outcomes compared with durable MCS.[36] Nevertheless, VA ECMO can be used to bridge alternative devices with the end goal of transplantation. In situations of acute decompensation in the setting of myocarditis or cardiomyopathy, VA ECMO can stabilise patients and allow implantation of temporary MCS (Impella, Centrimag) to facilitate rehabilitation and myocardial recovery.

## Conclusion

VA ECMO improves survival of neonates and paediatric patients with CHD. Use in the preoperative, intraoperative, and postoperative settings can bridge patients to recovery, surgical repair, or further decision making. ECPR is an important modality for patients with CHD. Overall, patient selection and clear goals of care are critical when utilising cardiac ECMO, with careful consideration of potential complications and outcomes. Assembly of multidisciplinary teams are essential to successful ECMO programs and improved outcomes in patients with CHD.

## Ethics Statement

The authors confirm that this article adheres to the relevant ethical guidelines.

## Patient Consent

The authors confirm that patient consent is not applicable to this review article.


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## Disclosures

The authors have no conflicts of interest to disclose.

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