Mechanical Circulatory Support for Patients With Adult Congenital Heart Disease

Maruti Haranal; Shuhua Luo, MD, PhD; Osami Honjo, MD, PhD

Advances in surgical and medical care of children born with heart defects have led to the emergence of a unique subgroup of young adults known as adults with congenital heart disease (ACHD). Heart failure (HF) is the leading cause of mortality and morbidity in this subset. Management of HF is challenging in these patients owing to inherent structural variations with their associated physiological consequences. Heart transplantation is of limited utility in this group either because of donor shortage or associated comorbidities that make these patients ineligible for transplantation. Mechanical circulatory support (MCS) devices have evolved as an alternative treatment modality in supporting the failing myocardium of this population, but are often used less frequently than in those with a structurally normal heart because of the unique anatomical and physiological variations. These variations create a need to gather adequate knowledge on how best to support the hearts of ACHD patients in order to reduce mortality and morbidity. This review presents clinical experience with MCS in ACHD patients.

Key Words: Adult congenital heart disease; Heart failure; Mechanical circulatory support

ADVANCES IN SURGICAL AND MEDICAL CARE OF CHILDREN BORN WITH HEART DEFECTS HAVE LED TO THE EMERGENCE OF A UNIQUE SUBGROUP OF YOUNG ADULTS KNOWN AS ADULTS WITH CONGENITAL HEART DISEASE (ACHD).1 Compared with the normal population, ACHD patients are more vulnerable to heart failure (HF), and a large population-based study showed chronic HF as the leading cause of mortality in the ACHD population2 (Figure 1). In addition, ACHD patients pose a significant challenge to medical treatment modalities compared with those with a structurally normal heart.3 Thus, many ACHD patients are waiting for heart transplants, but the shortage of donors has made this treatment strategy difficult. Often pulmonary vascular disease, high levels of allo-sensitization, or comorbidities such as liver cirrhosis make some of these cases ineligible for transplantation.4

Mechanical circulatory support (MCS) has emerged as an alternative method to support the failing myocardium in ACHD patients awaiting transplant, or it can be used as a definitive treatment modality. MCS acts by unloading the failing ventricle, thereby reducing myocardial oxygen demand, enhancing recovery, and enabling adequate systemic perfusion. Despite advances in device configuration, surgical techniques, and management, MCS is used less often in ACHD patients than in patients with acquired heart disease because of the inherent anatomic and physiological variations in the former.5 These variations create a need to gather adequate knowledge on how best to support the hearts of ACHD patients in order to reduce mortality and morbidity. This review presents clinical experience with MCS in ACHD patients.

HF in ACHD

Most patients who underwent repair for CHD in childhood have a risk of developing HF during adult life. A recent review by Stout et al clearly describes the possible mechanisms of late ventricular dysfunction in patients with repaired CHD.6 Further, there is a >20-fold increased risk of death in HF patients with CHD than in those with a structurally normal heart. The potential mechanisms of HF among ACHD patients include myocardial injury during prior procedures, ongoing ventricular overload (volume or pressure) due to residual lesions, altered coronary perfusion, pre-existing ventricular non-compaction, altered anatomy, and neurohormonal activation.7,8 Patients who have a morphological right ventricle (RV) as a systemic ventricle (i.e., patients who underwent an atrial switch procedure for dextro-transposition of the great arteries or those who underwent a physiologic repair for congenitally corrected transposition of the great arteries [cTGA]) are at a particularly high risk of late ventricular dysfunction. Furthermore, development of myocardial fibrosis, as seen in patients with repaired tetralogy of Fallot or in the RV in the systemic circulation, is strongly associated with the development of late ventricular dysfunction.9,10 The Fontan procedure is performed for various types of functional single ventricle lesions. Thus, by the time patients who have undergone a successful palliation procedure for a single functional ventricle reach adulthood, they have a Fontan circulation where systemic venous circulation lacks a pump and depends totally on low pulmonary vascular resistance. Because the Fontan circulation has no...
to stratify HF patients with ACHD requiring MCS. Using the INTERMACS scale, patients are classified based on the severity of symptoms and the trajectory of decline over time, which helps prognosticate those with advanced HF receiving MCS. Extracorporeal membrane oxygenation (ECMO) can be established either percutaneously or by an open technique (cut down). Cannulation can be either in the neck (internal jugular vein and the common carotid artery), groin (femoral vessels), or central (right atrial appendage and the aorta). Patients with venous anomalies and prior cavopulmonary anastomosis require special attention with regard to cannulation strategies. Ventricular assist devices (VADs) have been described as “mechanical pumps that take over the function of the damaged ventricle and restore normal hemodynamics and end-organ blood flow”. Inherent advantages of VADs include small priming volume, fewer anticoagulation and blood products, with a subsequent reduction in the infection rate and sensitization, the fact that patients can be extubated, and early mobilization. The classification of VADs is summarized in Table 2. MCS Strategies

MCS can be used as a bridge to decision, bridge to recovery, bridge to bridge (to subsequent long-term MCS), and bridge to transplant (BTT), or as destination therapy (DT). MCS can be used over the short, medium or long term, as described in Table 1.

The Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) scale is a helpful tool...
Mechanical Circulatory Support in ACHD

Ventricular assist devices (VADs) and total artificial hearts (TAHs) are used in congenital heart disease (CHD) to provide hemodynamic support. Prêtre et al. described the creation of a VAD-driven subpulmonic chamber following take-down of a Fontan circuit. Others have described total artificial heart (TAH) implantation in Fontan patients, which can be problematic due to the lack of normal compliance.

The TAH (SynCardia Systems, Tucson, AZ, USA) consists of right- and left-sided pumps and can be used as a BTT or as DT. TAH pumps can be configured in a variety of ways to address various physiologic (1 or 2 ventricles) and anatomical variations peculiar to CHD. The TAH provides optimal hemodynamic support in CHD patients with residual lesions compared with a VAD or biventricular assist device (BiVAD) alone; however, implantation of TAH is much more challenging than VAD implantation, especially in CHD patients considering the wide range of anatomical variations. One of the major advances in TAH implantation is the development of patient-specific virtual implantation based on cross-sectional imaging studies.

Numerous publications have reported details of surgical implantation. Either epicardial or transesophageal echocardiography is used to assist with cannula placement. The RV free wall and diaphragm surface are both alternative options in order to best orient the inflow cannula towards the tricuspid valve. Regardless of the epicardial entrance point, implantation of devices into a systemic RV may require resection of trabeculae to provide unobstructed inflow. To avoid issues with damage to the liver or bowel, or chamber compression upon closing the chest, the device may be placed back-to-front, closer to the midline, or through the right chest. Non-sternotomy approaches and non-aortic outflow graft positions have also been used.

| Short term (Days–weeks) | Mid and long term (Weeks–years) |
|-------------------------|---------------------------------|
| Extracorporeal membrane oxygenation | Pulsatile flow devices |
| Centrifugal ventricular assist devices | EXCOR |
| ROTAFLOW | PVAD/IVAD |
| PediMag | Continuous flow devices |
| CentriMag (St. Jude, Minneapolis, MN, USA) | DuraHeart |
| Impella | HeartWare HVAD |
| Tandem Heart | HeartMate II |
| Intra-aortic balloon pump | Jarvik 2000 |
| SynCardia TAH | |

HVAD, Heartware ventricular assist device; IVAD, implantable ventricular assist device; PVAD, percutaneous ventricular assist device; TAH, total artificial heart.

| Basis | Type |
|-------|------|
| Ventricle supported | LVAD, RVAD, BiVAD |
| Duration of support | Short term, intermediate, long term
| Purpose of support | BRT, BTT, DT |
| Device location relative to the body | Paracorporeal, extracorporeal, extracorporeal
| Generation of the device | First, second, third |
| Drive mechanism | Pneumatically driven, electrically driven |
| Pump mechanism | Axial, centrifugal |
| Flow characteristics | Continuous-flow devices, pulsatile-flow devices |

*Short term support ranges from 6 h to 7 days (e.g., Thoratec percutaneous ventricular assist device [PVAD], Levotronics Centrimag), intermediate support ranges from 7 days to 1 year (e.g., Thoratec PVAD, HeartMate II, Heartware), and long-term support is for >1 year (e.g., HeartMate II, Heartware). Examples of paracorporeal-, extracorporeal-, and intracorporeal-located devices are the Thoratec PVAD, Levotronics Centrimag, and HeartMate II (Heartware), respectively. BiVAD, biventricular assist device; BRT, bridge to recovery; BTT, bridge to transplant; DT, destination therapy; LVAD, left ventricle assist device; RVAD, right ventricle assist device.

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Table 1. Classification of Mechanical Circulatory Support

Table 2. Ventricular Assist Device Classification

neurologic impairment, a barrier to adequate anticoagulation, a constellation of congenital anomalies with poor prognosis, and major chromosomal aberrations generally preclude VAD therapy.

VAD implantation in CHD is different than in the structurally normal heart due to the altered mediastinal anatomy and cardiac physiology secondary to previous multiple operations, which may include systemic-pulmonary artery shunts and a disconnected inferior vena cava after a Glenn shunt or Fontan operations. Anatomic considerations related to prior complex repairs, abnormal size and location of the aorta, orientation of the ventricular chambers, heart position in the chest, devices designed for a morphologic left ventricle, and comorbidities of coagulopathy, malnutrition, and end organ dysfunction make VAD implantation technically challenging. Other considerations include the thickness of the ventricles, semilunar valve regurgitation and intracardiac shunts.

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An important change in VAD technology is the introduction of continuous-flow (CF) VADs. These can be implanted intracorporeally, which also allows for outpatient care. Over the past few decades there has been a major shift in use from pulsatile VAD to CFVAD, and studies
Tandem Heart systems can also be used to establish short-term ECMO circuits under different circumstances of cardiopulmonary failure.

MCS in ACHD Patients With Biventricular Physiology

MCS is challenging in ACHD patients with biventricular physiology because of ventricular morphology, residual or left heart. Tandem Heart systems can also be used to establish short-term ECMO circuits under different circumstances of cardiopulmonary failure.

Figure 2. Durable mechanical circulatory support in teenagers and adults with congenital heart disease. (A) Clinical course and vital status (at the time of report) by type of prior surgical repair. *The vital status was unknown for 1 patient each from the atrial switch and Fontan groups. (B) Comparison of assist device types between 1999–2010 and 2011 onwards. Intramics, Interagency Registry for Mechanically Assisted Circulatory Support; TAH, total artificial heart. (Reproduced with permission from Steiner JM, et al.)
Mechanical Circulatory Support in ACHD

Extracorporeal Life Support Organization data from ACHD patients requiring ECMO, Paden et al reported a 39% survival rate. Compared with ECMO, the use of VADs in ACHD patients is encouraging. A systematic review of durable MCS in teenagers and ACHD revealed the frequent utilization of MCS in patients with ACHD. Short-term survival rates in published series are approximately 70%, and the use of durable MCS as a BTT was 77% (Figure 2). In an INTERMACS analysis by VanderPluym et al, 21% of cases were successfully bridged to transplant and 51% were

lesions, systemic venous abnormalities, limited vascular access, previous multiple surgeries (sternotomies, thoracotomies), pulmonary hypertension, aortopulmonary collaterals, coagulopathy, and end organ dysfunction.

There are limited data on postoperative ECMO in ACHD patients as well as ECMO survival. The underlying cardiac anatomy, along with the associated comorbidities, complicates the decision to use ECMO in ACHD patients. A study from the Mayo Clinic showed a survival rate at discharge of 46% in high-risk ACHD patients requiring postcardiotomy ECMO support. Similarly, discharge survival rates in the studies of Maybauer et al and Schmidt et al were 50% and 42%, respectively. In an analysis of Extracorporeal Life Support Organization data from ACHD patients requiring ECMO, Paden et al reported a 39% survival rate. Compared with ECMO, the use of VADs in ACHD patients is encouraging. A systematic review of durable MCS in teenagers and ACHD revealed the frequent utilization of MCS in patients with ACHD. Short-term survival rates in published series are approximately 70%, and the use of durable MCS as a BTT was 77% (Figure 2). In an INTERMACS analysis by VanderPluym et al, 21% of cases were successfully bridged to transplant and 51% were
Heart, The Woodlands, TX, USA) is limited in ACHD patients. Fishberger et al demonstrated the usefulness of percutaneous RV support (Impella; Abiomed, Danvers, MA, USA) in providing hemodynamic stability during mapping and ablation of intra-atrial re-entrant tachycardia in patients after Mustard operation for TGA. 43 VAD to support a failing systemic RV with previous Senning or Mustard operation for TGA or ccTGA as a BTT is growing. Studies have shown that implantation of a VAD in a failing systemic RV has the potential to improve physiology and can be used as a bridge to heart transplantation. 44–46

Complex ACHD is not a contraindication to the use of MCS and should be considered as a treatment option early in the care of these patients. INTERMACS scores have trended towards lower acuity, suggesting implantation earlier in the course of advanced HF, as has been recommended

| References | Age (years) | Device | Duration of support (days) | Outcome |
|------------|-------------|--------|---------------------------|---------|
| Matsuda et al 44 | 10 | Toyobo (pulsatile) | 7 | Death |
| Sadeghi et al 45 | 8 | BVS 5000 (pulsatile) | 8 | Transplant |
| Frazier et al 46 | 14 | Centrifugal pump then Heartmate | 45 | Transplant |
| Nathan et al 47 | 4 | EXCOR (BiVAD) | 48 | Transplant, died, MOF |
| Newcomb et al 48 | 22 | Thoratec | 150 | Transplant |
| Calvaruso et al 49 | 10 | EXCOR | 7 | Transplant |
| Ricci et al 50 | 15 | Tandem Heart | 10 | Died |
| Russo et al 51 | 14 | Heartmate | 45 | Transplant |
| Prêtre et al 2 | 27 | EXCOR (LVAD) | 334 | Alive |
| Cardarelli et al 51 | 1.5 | EXCOR (LVAD) | 180 | Death, non-cardiac |
| Morales et al 52 | 15 | Heartmate II (LVAD) | 72 | Transplant |
| VanderPlum et al 53 | 3 | EXCOR (LVAD) | 174 | Transplant |
| Mackling et al 54 | 4 | EXCOR (LVAD) | 363 | Death, sepsis |
| Sanders et al 55 | 16 | EXCOR (LVAD) | 2 | Transplant |
| VanderPlum et al 56 | 2 | EXCOR+CMag (BiVAD) | 54 | Death |
| Valeske et al 57 | 19 | EXCOR (BiVAD) | 23 | Transplant |
| Niebler et al 58 | 4 | Heartware | 148 | Transplant |
| Rossano et al 59 | 13 | Syncardia, TAH | 61 | Transplant |
| Hoganson et al 60 | 4 | EXCOR (LVAD) | 26 | Transplant |
| Halaweish et al 61 | 14 | EXCOR (LVAD) | 179 | Transplant |
| Arnaoutakis et al 62 | 18 | HVAD (LVAD) | NA | Transplant |
| Arnaoutakis et al 63 | 14 | Syncardia TAH | NA | Transplant |
| Arnaoutakis et al 64 | 5 | EXCOR (LVAD) | NA | Transplant |
| Arnaoutakis et al 65 | 23 | Thoratec (LVAD) | NA | Death |
| Poh et al 66 | NA | EXCOR (LVAD) | 16 | Death |
| Poh et al 67 | NA | EXCOR (LVAD) | 17 | Death |
| Poh et al 68 | NA | EXCOR (LVAD) | NA | Transplant |
| Poh et al 69 | NA | EXCOR (LVAD) | 6 | Transplant, death |
| Poh et al 70 | NA | EXCOR (LVAD) | 150 | Transplant |
| Woods et al 71 | 11 | HVAD (LVAD) | 148 | Transplant |
| Woods et al 72 | 13 | HVAD (LVAD) | 272 | Transplant |
| Woods et al 73 | 17 | HVAD (LVAD) | 271 | Transplant |
| Lorts et al 74 | 22 | HeartMate III (LVAD) | NA | Ongoing |
| Chen et al 75 | 11 | Thoratec (LVAD) | 35 | Transplant |
| Chen et al 76 | 10 | HVAD (LVAD) | 76 | Transplant |
| Chen et al 77 | 10 | HVAD (LVAD) | 165 | Transplant |
| Chen et al 78 | 21 | HVAD (LVAD) | 69 | Transplant |

NA, not available. Other abbreviations as in Tables 1,2.
for patients with acquired disease. LVADs have shown promise in treating HF in ACHD patients, with similar survival benefits as in non-ACHD patients. The role of VADs as DT is increasing. The use of VADs to support a failing systemic RV is encouraging. Virtual implantation techniques using cross-sectional imaging may significantly alter device selection in ACHD patients.

**MCS in ACHD Patients With Single Ventricle Physiology**

The most important factor predicting the anticipated survival for any single ventricle patient requiring MCS is the stage of palliation. A patient’s current anatomy will affect their response to MCS. These patients typically have altered mediastinal anatomy due to numerous previous sternotomies, are chronically anticoagulated, and have hepatic and renal dysfunction related to the obligate elevated central venous pressure associated with long-standing single ventricle physiology and diastolic dysfunction with preserved systolic function.

ECMO in failing Fontan circulation is challenging and carries high mortality and morbidity rates. A survival to discharge rate of 35% was reported by Rood et al in Fontan failures supported by venoarterial ECMO. ECMO can be used in an acute setting as a bridge to decision or bridge to recovery. The stage of Fontan failure and patient selection determine the success of ECMO support in Fontan failure. The use of ECMO in advanced stages of Fontan failure is associated with even higher mortality rates due to the associated end organ damage.

There is growing evidence that VAD support can be successful in selected patients with a failing Fontan circulation. However, it remains unclear when VAD implantation should be considered and what percentage of patients will benefit from a systemic VAD alone. Patients with isolated or predominant ventricular systolic dysfunction are likely to benefit from VAD alone, but such cases are rare. A VAD will surely not help the clinical situation if the end-diastolic pressure of the systemic ventricle is not high (at least >12mmHg). However, patients with Fontan repair accounted for 14% of cases in the study of Steiner et al which showed increasing use of durable VADs in patients with ACHD with a short-term survival rate of 70%. Various clinical experiences with VAD in the failing Fontan circulation are summarized in Table 4. Fontan failure patients with marked end organ dysfunction, protein-losing enteropathy, and/or plastic bronchitis who are otherwise not good transplant candidates may benefit from TAH.

The feasibility of using a TAH in the failing Fontan circulation was demonstrated in a recent case report by Rossano et al. Fontan failure may be independent of ventricular function and often is driven by elevated pulmonary vascular resistance or pressure. Hence, VAD may or may not help in these patients, and cavopulmonary support may be necessary. Prêtre et al reported the first clinical experience with isolated cavopulmonary support in a case of failing Fontan as a BTT.

There are substantial research efforts into developing MCS for the failing Fontan circulation. Our group previously showed that a percutaneously implanted microaxial pump (Impella) for cavopulmonary assist in the failing classic Fontan physiology lowers the Fontan pressure, attenuates systemic venous congestion, and augments systemic oxygen delivery. Wang et al used a percutaneous Wang-Zwische double-lumen cannula (DLC) for cavopulmonary support for 2h in a failing sheep model. Derk et al showed the feasibility of an axial pump (Jarvik 2000) to restore baseline hemodynamics and cardiac output in Fontan circulation in a pig model. Rodefeld et al used a 3-dimensional computational model and showed that a single viscous Impella pump augments cavopulmonary blood flow and can be used as a bridge to recovery or transplant in patients with established univentricular Fontan circulation. Our group is currently investigating a novel multilumen cannula to support the failing Fontan circulation by a computational fluid dynamic simulation model.

Improvements in palliation of single ventricle physiology have given rise to an increase in the number of single ventricle patients susceptible to HF later in life, which necessitates the use of some form of MCS. Supporting the failing myocardium in single ventricle patients is a complex challenging problem. Even though the literature shows the feasibility of MCS in patients with a failing single ventricle, the outcomes are mixed: MCS outcomes are better in early stages of failure than in the advanced stage. With technological advances in MCS devices and better understanding of single ventricle pathophysiology, more research is needed to determine the medium- and long-term outcomes of MCS in these patients.

**Conclusions**

With the advent of MCS devices it has become feasible to support ACHD hearts despite the inherent surgical challenges present in this population. The earlier MCS devices used as a salvage had suboptimal outcomes. Recent advances in MCS technology have provided more compact, durable, and better-profile devices. The use of short-term MCS devices has been extended to high-risk catheter laboratory procedures or cardiac interventions. Percutaneous MCS, the most recent advancement in MCS, can now be used in the resuscitation of patients in cardiogenic shock. Outcomes of long-term MCS in the ACHD population are encouraging and may be improved further when timing is optimized. The subgroup of ACHD patients with failing Fontan circulation is an area of ongoing clinical research. Even though the failing Fontan subgroup represents a significant proportion of the ACHD population, the best available modality to support these hearts is yet to be determined. With further research and technological advancements, it may be possible to improve prognosis in this subset of patients.

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**Conflict of Interest**

None declared.

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