# Ventricular assist device for Fontan: who, when and why?

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**Purpose of review**
Since the advent of the Fontan palliation, survival of patients with univentricular congenital heart disease has increased significantly. These patients will, however, ultimately develop heart failure requiring advanced therapies such as heart transplantation. As wait times are long, mechanical circulatory support (MCS) is an attractive therapy, both for bridge to transplantation and destination therapy in patients not suitable for transplantation. This review aims to summarize current thinking about how to determine which patients would benefit from a ventricular assist device (VAD), the optimal time for implantation and which device should be considered.

**Recent findings**
VAD implantation in end-stage Fontan is still in its infancy; however, case reports and research interest have increased extensively in the past few years. Mortality is significantly higher than in noncongenital heart disease patients. Implantation in patients with primarily systolic dysfunction is indicated, whereas patients with increased transpulmonary gradient may not benefit from a single-VAD solution. When possible, implantation should occur prior to clinical decompensation with evidence of end-organ damage, as outcomes at this point are worse.

**Summary**
Fontan patients demonstrating signs of heart failure should be evaluated early and often for feasibility and optimal timing of VAD implantation. The frequency of this procedure will likely increase significantly in the future.

**Keywords**
cardiac anaesthesia, cardiac surgery, congenital heart disease, Fontan, ventricular assist device

## INTRODUCTION

The number of patients living with a Fontan palliation for single ventricle or unbalanced double ventricle congenital heart disease has increased substantially over the past 40 years [1–3]. The first Fontan recipients are now entering into their fifth decade. Registry data ($n = 1006$) from Australia and New Zealand estimates survival at 15, 20 and 25 years after Fontan completion to be 93% [95% confidence interval (95% CI): 90–95), 90% (95% CI: 86–93) and 83% (95% CI: 75–89), respectively [1,4]. Although improvement in surgical and medical management of these complex patients has improved, leading to better life expectancy, it is still a palliation that then begs the question: what to do with a failing Fontan circulation?

For patients who survive the often arduous course to Fontan completion, morbidity and mortality are relatively low for the first 10–15 years, and a majority of patients are still alive at age 40. The number of complications and mortality rate rises substantially, however, throughout early adulthood; survival at age 40 is down to 80% (95% CI: 75–87) and estimated 5-year-mortality at this point approaches 20% (18.0%;

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## KEY POINTS

- Improved surgical techniques and perioperative care have led to good intermediate and long-term survival in Fontan palliation.
- Failure of the Fontan circulation is inevitable and with increased numbers of Fontan patients and a paucity of organs available for heart transplantation, MCS is a viable and necessary therapy.
- Careful preoperative workup is key to determining which end-stage Fontan patients would benefit from VAD implantation; primary systolic dysfunction is an indication for VAD, whereas patients with increased transpulmonary gradient do not qualify or require a bi-VAD or total artificial heart solution.
- Appropriate device selection is important; the withdrawal of Medtronic's HeartWare from the market, with its favourable sizing for paediatric patients, has significantly reduced the options for end-stage Fontan patients.
- Research and device development is ongoing, and the PumpKIN Trial is currently testing the implantable Infant Jarvik 2015 in patients between 8 and 20 kg.

95% CI: 12–25) [1,5–7]. Heart failure and sudden death are the predominant causes of mortality [1]. Patients also experience multiple comorbidities, including arrhythmias, thromboembolic events, protein-losing enteropathy, plastic bronchitis, cirrhosis and renal dysfunction [6,8▪▪]. These conditions can significantly impact quality of life and also lead to significant debilitation, which can complicate further therapy.

Orthotopic heart transplantation has been the therapy of choice for end-stage Fontan patients; however, due to scarcity of donor organs in many countries and high levels of antibody sensitization in this population, wait times are long and patients seldom survive until transplantation. Since Frazier *et al.* [9] in 2005 first described successful use of a ventricular assist device (VAD) as a bridge to transplantation in an adolescent with failing Fontan circulation, interest in mechanical circulatory support (MCS) has increased either as a bridge to transplantation or as destination therapy.

Both the timing and type of device implantation must be carefully considered for each patient, as symptoms, size and anatomy (i.e. 'right' or 'left' ventricle as the systemic ventricle) can vary significantly. Also, it is important to distinguish between failing myocardial function or failing pulmonary perfusion with preserved function as the principal cause of failure. The development of symptomatic heart failure is the start of an often rapid downward spiral, with 24% of patients dying within a year and 35% within 3 years after the first hospital admission [7,10]. VAD placement should be considered when medical therapy is being escalated and the patient continues to be symptomatic or show evidence of end-organ dysfunction (INTERMACS II or III); avoiding implantation in extremis has been demonstrated to improve outcomes [11▪]. There are case reports of successful deployment of VADs as a bridge to transplantation in patients with failing Fontan, although favourable results are uncommon in the documented literature [12,13].

The wide range of patients with failing Fontan circulation, from infants to adults, as well as the unique physiology, complicates the choice of device and implantation site. With Fontan physiology, it must also be determined if ventricular systolic dysfunction, increased pulmonary vascular resistance or both is the primary driver of heart failure. It is important to note that ventricular assist may not overcome failing Fontan physiology if low cardiac output is a result of inadequate preload through decreased passive pulmonary blood flow.

VAD for Fontan has been addressed in several reviews throughout the past 5 years [14–20]. We will provide a literature overview with the goal of breaking down a highly complex issue into digestible parts: who (which patients benefit), when (optimal timing for implantation) and what/why (device, implantation location, goal of therapy).

## WHO?

Most single ventricle lesions today undergo a staged surgical and interventional approach to a Fontan circulation, as 7-day survival after birth without an intervention is a dismal 39% (95% CI: 26–50) to 50% (95% CI: 25–63) [21]. Although the longer-term outcome of the Fontan patient population is remarkable given the pathophysiologic complexity, it is still a palliation and ultimately a terminal condition. An analysis of the Australian and New Zealand Fontan Registry ($n = 1561$) reported freedom from death or heart transplant after 10, 20 and 35 years post-Fontan surgery at 94% (95% CI 93–95), 87% (95% CI 85–90) and 66% (95% CI 57–78), respectively. The predictors for early mortality or transplant were male sex (hazard ratio 1.6, 95% CI: 1.1–2.2, $P = 0.01$), an atriopulmonary Fontan (hazard ratio 1 versus lateral tunnel hazard ratio 0.5 (95% CI: 0.3–0.7) or extracardiac conduit (hazard ratio 0.4, 95% CI: 0.3–0.7, $P < 0.001$), pre-Fontan atrioventricular valve intervention (hazard ratio 3.3, 95% CI: 1.7–6.4, $P = 0.002$) or prolonged pleural effusions (hazard ratio 2.3, 95% CI: 1.4–4.0). Time-dependent factors such as the development of atrial arrythmias (hazard

ratio 3.5, 95% CI: 2.2–5.5, $P < 0.001$), protein-losing enteropathy (hazard ratio 7.5, 95% CI: 4.4–12.6, $P < 0.001$) or late ventricular dysfunction (hazard ratio 15.8, 95% CI: 10.7–23.1, $P < 0.001$) were even stronger predictors of death or transplantation [46% (95% CI 34–67) at 15 and 56% (95% CI 43–67) at 25-years post Fontan)] [22]. Thus, despite creating a stable circulation for most single ventricle patients for many years, a significant number of Fontan patients will eventually present for advanced heart failure therapies including MCS and/or transplantation.

Failure of the Fontan circulation with subsequent development of heart failure symptoms and end-organ dysfunction is often multifactorial. Decision-making about VAD implantation (yes or no) is much easier when the primary problem is reduced systolic function of the systemic ventricle and/or significant atrioventricular valve regurgitation. The cause of ventricular dysfunction can include a combination of coronary ischemia, volume and pressure overload, chronic cyanosis and upregulation of the renin-angiotensin-aldosterone system as well as underlying genetic factors that caused the CHD [8▪▪]. A retrospective study of 45 Fontan patients in the Advanced Cardiac Therapies Improving Outcomes Network (ACTION) demonstrated a trend towards improvement in hemodynamic parameters following VAD placement; 69% of patients survived to transplantation, 21% died and 9% were still alive on device after 1 year [23▪]. These results were consistent with overall paediatric VAD use and encouraging for the future of VAD implantation in Fontan patients [23▪,24,25].

Unfortunately, in many patients, diastolic dysfunction and/or increased transpulmonary gradient (TPG) in the setting of preserved systolic function renders single VAD implantation insufficient. Fontan patients may not meet classic criteria for pulmonary hypertension, yet still have a PVR that is too high for adequate passive pulmonary blood flow. In single-ventricle physiology, preoperative criteria require a mean TPG of 6 mmHg or less. Post-Fontan, a clinically asymptomatic course with acceptable haemodynamics is seen with a PVRI of 3 WU x $m^2$ or less and a mean TPG of 6 mmHg or less. Stringent pulmonary hypertension therapy should be considered when the TPG rises above 6 mmHg or the patient demonstrates decreased exercise capacity [26,27]. Chronically elevated central venous pressure, TPG or PVR or any obstruction to venous or arterial blood flow can lead to heart failure or Fontan specific complications. These patients present with plastic bronchitis, protein-losing enteropathy or other signs of venous hypertension such as Fontan-associated liver disease (FALD) and are typically not candidates for VAD placement, even though successes in this situation using VAD have been described [28,29]. Before considering VAD therapy in this patient subset, procedures to improve blood flow, such as treatment of stenotic segments or Fontan conversion in disadvantageous Fontan variants, offer attractive alternatives [30]. Patients with the unfortunate combination of reduced ventricular function and increased TPG require simultaneous support of the pulmonary and systemic circulation; both a 'biventricular' solution such as two VADs or use of a total artificial heart have been described [31▪▪].

The severity of the overall condition of the Fontan patient at time of implantation plays an important role in post-VAD outcomes: children in cardiogenic shock (INTERMACS 1) and patients intubated despite underlying lower INTERMACS profiles have significantly increased postoperative mortality [11▪,32▪▪]. For these critically ill patients, options other than VAD implantation should be considered: further optimization of medical therapy for potential haemodynamic recompensation or even surgical Fontan conversion, Fontan fenestration or progression to heart transplantation [8▪▪].

## WHEN?

In general terms, heart failure means that the heart cannot generate enough blood flow to meet the body's demand during exercise at normal filling pressures. Essentially, all Fontan patients therefore have some kind of functional chronic heart failure from postoperative day one. These patients will deteriorate at some point in life, often developing typical symptoms of heart failure with fluid retention and exercise intolerance. In contrast to typical heart failure patients, those with a Fontan circulation decline more rapidly, showing increased cardiac and noncardiac morbidity. Agarwal *et al.* [10] reported that a cohort of CHD patients were more likely to have longer hospital length of stay, arrhythmias and in-hospital mortality compared to their non-CHD counterparts. A Dutch registry showed mortality in Fontan patients admitted with heart failure was 24% at 1 year and 35% at 3 years [33].

A major dilemma in this patient population is the heterogeneity and complexity of heart failure presentation (Table 1). Due to the underlying pathophysiology of passive blood flow through their lungs, they may have a challenging combination of systolic and diastolic ventricular dysfunction, structural cardiac or valvular disease, and arrythmias [34]. This often leads to a delayed referral for MCS, as clinical deterioration with severe end organ dysfunction has already occurred; at this point, they show clinical signs of hepatic and renal insufficiency, respiratory

**Table 1.** Symptoms of Fontan failure ordered by organ system

| Organ system | Manifestation |
|---|---|
| Constitutional | Exercise intolerance<br>Failure to thrive<br>Weight loss or gain |
| Neurologic | Depression<br>Transient ischemic attack<br>Stroke |
| Cardiac | Arrythmias<br>Reduced ventricular systolic function<br>Atrioventricular valve regurgitation<br>Venous congestion<br>NYHA functional class III or IV |
| Pulmonary | Plastic bronchitis<br>Cyanosis<br>Pleural effusions<br>Respiratory failure/Intubation<br>Pulmonary embolism |
| Gastrointestinal/Hepatic | Protein-losing enteropathy<br>Hyperbilirubinemia<br>Hypoalbuminemia<br>Ascites<br>Cirrhosis<br>Hepatocellular carcinoma |
| Renal | Renal insufficiency<br>End-stage renal disease |
| Haematologic | Thrombocytopenia<br>Thrombosis<br>Coagulopathy |

failure requiring intubation, low mixed venous oxygen saturation and lactic acidosis [8▪▪,31▪▪]. For the general practitioner, the inability to tolerate enteral feeds may be the first sign of inadequate cardiac output and oxygen delivery, and MCS should be considered at this point.

Evidence-based approaches and recommendations for optimal timing for MCS in Fontan patients are lacking, but repeated cardiopulmonary exercise testing may be a powerful tool for the creation of objective criteria in the future. Right now, it remains unclear which exercise measures are the most relevant and whether a threshold or trend offers better prognostic information [35]. A valid and useful clinical indicator might be the combination of a high PVRI ($> 2\,\mathrm{WU\,x\,m^2}$) and low cardiac index ($< 2.5\,\mathrm{l\,min\,x\,m^2}$), as this has been shown to be an independent risk factor for Fontan failure [36]. Apart from BNP, parathyroid hormone may be a useful biomarker of heart failure and for predicting outcomes and timing for MCS in Fontan patients [37]. Insufficient evidence currently exists for developing and grading recommendations for type and frequency of organ function tests. Nevertheless, surveillance testing for cardiovascular and end-organ function/dysfunction in Fontan patients seems reasonable, clinically important and to be encouraged as part of overall high-quality patient care [8▪▪].

A special case bears mentioning: there is some evidence that Glenn physiology represents a better setting for bridge to transplant due to the higher posttransplant mortality after early Fontan failure [38]. Thus, before undertaking a high-risk Fontan completion with severely reduced ventricular function, proceeding to transplantation or a VAD from a Glenn should be considered as a valid alternative. At present, the use of commercially available MCS therapy is limited to end-stage Fontan circulation failure as a bridge to transplantation. To reiterate an important point from above: outcomes are worse (in any patient) when a VAD is placed in the setting of critical illness. Planning should begin in earlier INTERMACS stages and ideally occur prior to decompensation with end-organ damage [11▪,15,39]. For patients who are in cardiogenic shock, temporary support with ECMO can also be considered to stabilize/reverse organ injury and evaluate for durable VAD placement [31▪▪]. Absolute contraindications to VAD placement include irreversible and nontreatable end-organ damage, especially severe neurologic injury, and active infection.

## WHY?

The current primary goal in VAD implantation in the Fontan population, similar to that of the overall paediatric population, is bridge to transplantation. The vast majority of 55 cases between 2012 and 2019 published in the PediMACS/INTERMACS database were bridge to transplant [40]. Although there was a trend toward increased VAD usage over this time (28 in 2018-19 versus 27 in 2012–2017), this is still a miniscule percentage of patients currently living with a Fontan palliation. As more than 1000 Fontan procedures are performed in the USA annually, this patient population requires other therapeutic options due to long wait times for CHD transplantation candidates. Patients who experience long wait times are often severely debilitated, which has been associated with worse outcomes. Riggs *et al.* [41▪] reported significantly reduced 1-year survival posttransplant in intubated patients with CHD in a study of paediatric heart transplantation (76 versus 95%); additional risk factors such as hepatic or renal dysfunction, present almost universally in end-stage Fontan circulation, also worsened expected survival. Increased usage of MCS while awaiting transplant may help prevent further clinical deterioration and organ dysfunction. VAD use, especially durable implantable devices, could also allow for rehabilitation, improving physical and nutritional status.

**Table 2.** Available VADs for paediatric patients

| Device | Type | Location | Min. patient size (approximate) | Additional information |
|---|---|---|---|---|
| HeartMate 3 (Abbott) | Single VAD<br>Continuous flow<br>Centrifugal pump | Intracorporeal | BSA $\geq$ 1.2 $m^2$ | |
| HeartWare (Medtronic) | Single VAD<br>Continuous flow<br>Centrifugal pump | Intracorporeal | BSA $\geq$ 1.0 $m^2$ | Implantation with BSA as low as 0.6 m2 has been described, but has a higher risk of pump thrombosis<br>Removed from market in June 2021 |
| EXCOR (Berlin Heart) | Single VAD<br>Pulsatile flow<br>Pneumatic pump | Extracorporeal | Weight > 2 kg | Available in 10, 15, 25, 30, 50, 60 and 80 ml pump sizes |
| SynCardia (SynCardia Systems) | Total artificial heart<br>Pulsatile flow<br>Pneumatic pump | Intracorporeal | BSA > 1.2 $m^2$ | Available in 50 and 70 ml pump sizes |

This would not only provide better quality of life, but also improve the chance of a good posttransplantation outcome.

When discussing the 'who, when, and why' of VAD implantation in Fontan patients, we must also address the 'what': which device for which patient (Table 2). This population is diverse in age, size and pathophysiology of heart failure, which makes device and location selection challenging. The use of implantable continuous pumps has increased, but they are limited by body size (BSA > 1.2 and 1.0 $m^2$ for Heartmate III and HeartWare, respectively). For paediatric patients with a BSA below this threshold, extracorporeal, pulsatile pumps have been utilized successfully. The paracorporeal Berlin Heart EXCOR, which exists in three different sizes, has been in use for over 20 years; the smallest version can be placed in patients with a BSA greater than 0.6 $m^2$ or weight greater than 2 kg. The FDA issued a warning on June 3rd, 2021 advising healthcare providers to stop implantation of the HeartWare (Medtronic, Dublin, Ireland) due to observed higher frequency of neurologic complications and mortality. Medtronic subsequently effectively removed the device from the market. This was a setback for the paediatric population, as the HeartWare is the most commonly implanted device in children due to its favourable sizing [42▪].

The intricacy of device placement and cannulation location is beyond the scope of this review, but C. Mascio [31▪▪] and I. Adachi [43▪] have both published articles in the past 2 years delving into this topic, both of which also provide helpful illustrations. There has been increased interest in a cavopulmonary assist device, especially in Fontan patients with preserved systolic function, but device development is still in its early phases [44▪,45,46]. The PumpKIN Trial investigating the Infant Jarvik 2015, which gained FDA approval for testing in humans, is currently ongoing [47].

## CONCLUSION

VAD implantation in the Fontan population is still in its infancy, and further experience is needed to determine optimal timing, patient selection and type of therapy (systemic, subpulmonary or BiVAD/TAH) and device. The current paradigm is aimed at bridge to transplantation, but as experience increases, destination therapy may become a viable option, especially in patients who do not qualify for heart transplantation. Another salient point is the importance of careful perioperative and long-term management of these patients. Most centres implant few paediatric or adult CHD VADs, and the Fontan population is one of the most complex subsets of these patients. Communication with larger CHD centres or possible transfer of care to teams with more experience should be considered to optimize outcomes and patient care.

The question of whether and how to utilize VADs in the complex pathophysiology of a failing Fontan circulation is unfortunately not one easily answered. As more patients survive and annual numbers increase, advanced therapies such as VAD implantation will likely play a larger role in supporting these patients while awaiting transplantation or as destination therapy.

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