

Mechanical support for the failing single ventricle after Fontan



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Feature Editor's Note—*The rapidly growing population of patients with failing Fontan circulation brings new challenges to our specialty. As we all learned in the recent years, medical management alone of patients with failing Fontan circulation is often inadequate. Many patients with single-ventricle physiology eventually develop failure of the uni-ventricular circulation and require heart transplantation. Mechanical support of the failing single ventricle after the Fontan operation is complex and demands utmost attention to detail. A group of leading experts from Cincinnati Children's Hospital share their views on and discuss their approach to mechanical support of patients with failing Fontan circulation.*

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The 4 decades after the development of the Fontan procedure have seen serial improvements in the surgical and clinical care for children with single-ventricle physiology.¹ These gains have translated into improved mid- and long-term survival; approximately 90% survival is expected for the first 10 to 15 years after the Fontan in the current era.²⁻⁵ For those Fontan patients who reach adolescence, approximately 90% to 95% will survive to age 30 years and 80% will survive to age 40 years.⁴ The attrition that occurs in the 20s and 30s is multifactorial, but approximately one-third is due to heart failure (HF). For those undergoing the Fontan with HF and systolic dysfunction, ventricular assist device (VAD) therapy provides an effective means to manage HF as a bridge to transplantation or as destination therapy. We review the current indications, management, and outcomes associated with VAD therapy in the Fontan.



Chet Villa, MD (left), and David L. S. Morales, MD (right)

CENTRAL MESSAGE

Children with failing Fontan circulation, particularly those with predominant systolic dysfunction, can benefit from early consideration for VAD placement.

EVALUATION AND REFERRAL FOR VENTRICULAR ASSIST DEVICE THERAPY

Timely evaluation and referral are fundamental to achieve optimal outcomes for VAD therapy. While the exact timing of referral may vary on the basis of patient specifics including overall clinical trajectory and planned interventions, early referral and evaluation for patients at risk for decompensation are paramount. Patients receiving a device implantation in extremis (Interagency Registry for Mechanically Assisted Circulatory Support profile 1 or after extracorporeal membrane oxygenation cannulation) have significantly higher risk of early mortality after VAD therapy.⁶⁻⁸ Multiple studies have noted that patients with congenital heart disease (CHD) are considered for advanced therapies including VAD support, implantable cardioverter-defibrillators, and heart transplantation at lower rates than patients without CHD.⁹⁻¹² This pattern persists despite data suggesting these therapies, including VAD, can be effective when used in a timely fashion.^{10,13}

Comprehensive, longitudinal evaluation through a multidisciplinary “Fontan clinic” including an HF/transplant cardiologist is perhaps the most important step in ensuring timely referral for patients with recalcitrant HF and systolic dysfunction. This approach also facilitates

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the identification of potential anatomic and electrophysiologic substrates that may benefit from intervention to obviate the need for VAD therapy (or at least delay it given the long-term hazard for HF).^{14,15} This approach also may help identify and treat Fontan-related complications such as thrombosis, protein-losing enteropathy (PLE), and plastic bronchitis that may complicate VAD therapy. This approach in conjunction with national guidelines may help to standardize referral for evaluation. Multicenter groups have also begun to formulate “best practice” guidelines to supplement national guidelines. For example, Advanced Cardiac Therapies Improving Outcomes Network (ACTION) has created a document loosely based on the I-NEED-HELP indicators for HF referral included in the American College of Cardiology Expert Consensus Decision Pathway for Optimization of Heart Failure Treatment.¹⁶ While a step in the right direction, the specific indications for VAD therapy have not been elucidated given that VAD therapy has only become more widespread in the last few years or in light of constantly improving technology (eg, current generation of continuous-flow VADs) and implantable pulmonary artery pressure monitors (eg, CARDIOMEMS HF System, Abbott Medical, Inc). Given this, the authors will report our approach, noting the field is rapidly evolving.

EVALUATION FOR VENTRICULAR ASSIST DEVICE THERAPY

Cardiac Evaluation

Although symptomatic HF in the setting of isolated systolic dysfunction does occur, the Fontan circulation almost always fails at multiple levels from venous insufficiency, impaired passive flow through the cavopulmonary pathway, atrioventricular valve regurgitation, sinus node dysfunction, abnormal pulmonary vasculature, repeated subclinical pulmonary emboli, aorto-pulmonary collateral flow, and combined systolic and diastolic dysfunction. Multi-modality evaluation including echocardiography, cardiac magnetic resonance (CMR), cardiopulmonary exercise testing, and cardiac catheterization is needed to help discern the relative contributions of these factors and to inform surgical planning and perioperative management. In particular, this approach allows the identification of anatomic obstruction from the cavopulmonary circulation through to the ventricle that may impair VAD filling. The hemodynamic evaluation is especially important in assessing the multiple factors that contribute to Fontan failure, because a patient with significant venous insufficiency and inefficient passive flow through the cavopulmonary circuit in the setting of mild to moderate ventricular dysfunction and low filling pressures is unlikely to see a clinical and hemodynamic benefit from VAD therapy. However, it is important to note that assessing hemodynamics and venous pressures under multiple loading conditions may be needed. Volume loading a

patient who has been non per os before catheterization may be needed to unmask elevated filling pressures and diastolic dysfunction.¹⁷ Our center has generally used evidence of an end-diastolic pressure greater than 15 mm Hg with a low cardiac output (cardiac index <2.5 L/min/m²) as a rough guide to identify patients who may benefit from VAD. The authors acknowledge further data are needed to refine this approach and understand which hemodynamic profiles benefit most from VAD therapy and thus would hesitate to create “hard cutoffs” given the limited data on longitudinal hemodynamics in the Fontan after VAD. At the time of catheterization, we often place a CARDIOMEMS to help with fluid management and to help understand patient symptoms post-VAD. As in the case of Fontan patients without a VAD, further data are needed to clarify the benefits of this approach.¹⁸ Peripheral venous pressure assessment at rest and with exercise may also provide valuable data when assessing patient symptomatology and potential value for VAD therapy.¹⁹

Collaterals and Fenestration

Preoperative evaluation to assess for the presence and severity of aortopulmonary (AP) and veno-venous (VV) collaterals is especially important for operative planning, but also to facilitate postoperative management. We use a combination of catheterization and CMR, because CMR-based flow can identify cumulative effects of smaller collaterals that may be underestimated by catheterization. Patients with extensive AP collaterals may require preoperative catheter intervention to close large vessels. Alternately, when the AP collateral burden is more reasonable, elevated VAD flows may be used to ensure adequate systemic oxygen delivery to accommodate for pulmonary runoff. The ideal approach is likely patient-specific as increasing flows may result in elevation of pulmonary artery pressures if AP collateral flow is extensive. A combined catheterization and CMR-based approach may also help quantify fenestration flow.²⁰ Understanding the location and degree of fenestration flow in conjunction with VV collaterals will facilitate preoperative and postoperative planning. VV collaterals in particular may need to be addressed preoperatively to ensure adequate effective pulmonary blood flow after VAD (especially in circumstances where venous pressures remain more elevated).²¹

Whether to create or close a fenestration in the setting of a VAD remains a topic of debate. The authors generally recommend closing VV collaterals in the catheter lab, but leaving the fenestration or creating one if it does not exist. This will facilitate VAD filling in the perioperative period when positive pressure ventilation may limit pulmonary blood flow and when pulmonary vascular resistance may be labile. Many of our Fontan patients supported with a VAD have been supported for long periods of time, over 3 years in one case, and the fenestration is important if

they undergo procedures that require intubation or they have an upper respiratory infection. The position of the fenestration is always noted as well as whether we believe it can be addressed postoperatively should indications for closure arise (eg, thrombosis, severe cyanosis) and if not it is closed and created in a location where it could be closed if needed.

End-Organ Evaluation

Renal dysfunction and Fontan-associated liver disease (FALD) are known complications of Fontan circulation.^{22,23} Chronic venous pressure elevation and a limited cardiac output conspire to produce end-organ function, and in the case of FALD, risk for hepatocellular carcinoma. Monitoring renal and liver function is important for perioperative planning for VAD, but also when assessing the indications for VAD. Renal and liver dysfunction are known risk factors for perioperative mortality among Fontan patients and timely implantation may minimize this risk, while avoiding irreversible organ dysfunction.²⁴ To appropriately evaluate longitudinal changes in renal and liver function, a multimodality approach is generally needed. Noncreatinine-based measures of renal function as well as markers of tubular injury should be used to identify patients at risk for adverse outcomes given creatinine-based glomerular filtration rate estimates may be falsely high due to sarcopenia.^{22,25} Serial liver magnetic resonance imaging with elastography, computed tomography (CT), or ultrasound may identify patients with progressive FALD who can benefit from VAD therapy before the development of irreversible liver disease.^{23,26,27} Furthermore, given the association between inferior vena cava flow and liver fibrosis, VAD therapy may also provide the opportunity for “liver rehabilitation” before transplantation, although further study is needed to assess the changes in liver morphology and function with VAD therapy.²⁸ This is a particularly important point as the question of what is reversible and irreversible liver disease persists in the field.²⁹ A recent report documenting the evolution of liver imaging and histologic findings after heart transplantation underscores this point.³⁰ Finally, preoperative brain imaging (CT or magnetic resonance imaging as appropriate) is recommended given the frequency of neurologic injury in univentricular patients and in patients with HF in general.

Frailty

Frailty has been associated with adverse outcomes in patients with ambulatory HF as well as those patients who require VAD or transplant.³¹⁻³⁴ Emerging evidence suggests frailty and sarcopenia are important predictors of adverse outcomes in Fontan patients (and other patients with CHD).^{35,36} Understanding the physical, psychological, and metabolic state of the patient is also important, because adverse profiles may be amenable to cardiopulmonary

rehabilitation, counseling, and medications both preoperatively and after VAD.^{37,38} Thus, we would consider worsening frailty secondary to HF an indication for device implantation, as in the case of worsening end-organ function.

Fontan Lymphatic Complications

Fontan patients have a long-term risk for developing plastic bronchitis and PLE, and severe disease is an indication for heart transplantation.^{39,40} Although the development of each is clearly multifactorial, adverse hemodynamic profiles are clearly contributory given the resolution that occurs after heart transplantation and the response to anatomic/hemodynamic optimization. Importantly, elevated systemic ventricular filling pressures may not be elevated at all times, and in these patients a systemic VAD will not be helpful. Even when indicated, VAD therapy for patients with PLE can be challenging especially in regard to healing and anticoagulation.

Three-Dimensional Modeling

The diversity of thoracic and cardiovascular anatomic phenotypes in patients with a Fontan circulation necessitates patient-specific surgical plans. Although traditional cross-sectional imaging via CMR or CT is integral for surgical planning in every patient, additional benefit can be garnered from virtual reality–based systems that can facilitate optimization of inflow/outflow positioning.⁴¹ This approach is particularly important as the data continue to evolve regarding the impact of pump positioning on thrombosis/stroke.^{42,43} For pediatric patients, advanced imaging may also be needed because the recent withdrawal of the HeartWare HVAD system (HeartWare, Inc) has limited the device options for the 18- to 30-kg patients with small thoracic or intracardiac volumes in whom the HeartMate 3 (Abbott Medical, Inc) fit may be challenging or modified inflow position may be required. This approach may help understand when approaches such as systemic atrioventricular valve excision may be required or to help avoid this approach through alternate device/inflow positioning.⁴⁴

Consultation

The complications and physiologic derangements present in the failing Fontan circulation can lead to multiple complications, as noted earlier. With this in mind, routine consultation of a number of services is generally prescribed, with some services engaged as needed (Table 1). Our center has tried to engage specific subspecialists in the given field who have experience with the Fontan circulation. This is especially important regarding hepatology consultation given the rapidly evolving data on FALD.

OPERATIVE MANAGEMENT

Many of the Fontan patients are on Coumadin before surgery, and if time permits, we change to heparin and stop the

TABLE 1. Consulting services engaged for preoperative ventricular assist device evaluation

Clinical service	Indications for evaluation*
Consultations obtained in all patients	
Hepatology	FALD including hepatocellular carcinoma
Cardiac rehabilitation	Functional assessment, frailty
Palliative care	Advanced care planning
Consultation obtained as needed	
Nephrology	Chronic kidney disease, severe acute kidney disease
Hematology	History of thrombosis
Neurology	History of stroke, concerns on preoperative neurologic imaging
Pulmonology	Evaluation of plastic bronchitis

FALD, Fontan-associated liver disease. *Common considerations/reasons for evaluation are listed. Patient-specific concerns should be addressed accordingly.

heparin upon entering the operating room. If the patients are on Coumadin and the international normalized ratio is 3.5 or below, we do not necessarily reverse the anticoagulation it unless anesthesia feels uncomfortable placing central lines. When over 3.5, we would probably reverse with fresh-frozen plasma, starting it as the patient is going down to the operating room. We have a similar practice when taking VAD recipients to the operating room for transplantation.

The cannulation varies from a more straightforward VAD cannulation in only a few ways. Because we are not worried about preserving the right heart, we have no issues arresting the heart especially if we need to create a fenestration. If the aorta is difficult to circumferentially dissect out and the patient is quite sick, one can just fibrillate the heart.

If there is a Damus-Kaye-Stansel connection, we often find them dilated and very thin walled. Thus, we often sew a graft onto the innominate or subclavian artery even distal to a previous shunt location, ensuring no critical stenosis is present. At the end of the case, we clip this shunt just distal to the anastomosis and keep it long so we can use this at transplantation. This is especially helpful if a patient is undergoing heart-liver transplantation or will need arch reconstruction at transplant. We also attempt to place the VAD in the systemic ventricle for Fontan patients and do not think atrial-placed devices are as effective for long-term or chronic support. The ventriculotomy tends to be a bit more anterior in a systemic right ventricle. We always use a cut-and-sew technique for the apical cuff in a systemic right ventricle to ensure that we can easily explore the ventricle and remove any possible muscle bundles that could obstruct the inflow cannula. Creation of the fenestration can be challenging if the graft or patch is calcified or the atrium is particularly thick. However, as stated previously, we think this is a critical part of the procedure. We also

leave a pillow made of a sheet of Gelfoam sponge (Pfizer) sandwiched between 2 thin membranes of Gore-Tex mesh (WL Gore & Associates) that we place between the pump and chest wall when we lower the device into the left thorax and inferior to the left lower lobe, once apical cannulation is completed and the outflow bend relief is attached. In smaller patients, we have partially plicated the diaphragm (temporarily or permanently), ensuring not to injure the phrenic, to get the device to sit posterior to the diaphragm at the costophrenic angle. Also, one can increase space in the left thorax in the anterior posterior dimension but taking the anterior attachments of the left diaphragm down which allows costal margin and thus chest wall to move anterior. We have found “the pillow” to help with postoperative discomfort especially in patients especially smaller ones. Also, in Fontan patients who have a number chest wall collaterals, it may minimize the risk of bleeding due to device contact with the chest wall. This pocket also makes it easy to remove the device at the time of transplantation. We place all intracorporeal VADs in the extra pericardial space inferior to the left lower lobe. In Fontan patients, despite the vascular adhesions it is worth the time to enter the pleura, divide the pericardium at the diaphragm down to the phrenic, and mobilize the left lower lobe so the device can rest inferior to it. Small strips cut from Esmark bandages (Hartmann USA, Inc) are loosely placed around the inferior vena cava and superior vena cava as well as the pulmonary arteries and aorta if circumferentially dissected out. All of these small adaptations make initiating cardiopulmonary bypass at the time of the transplant efficient. For more information, we refer the reader to the instructional videos at <http://www.cardiovascular.abbott/us/en/hcp/products/heart-failure/left-ventricular-assist-devices/heartmate-3/education-training.html>. We also would again highlight our center approach regarding the creation of a fenestration (“Collaterals and Fenestration” section) if one does not exist.

POSTOPERATIVE MANAGEMENT

Immediate postoperative management generally focuses on 4 specific components: (1) obtaining rapid hemostasis, (2) management of vasoplegia, (3) early extubation to facilitate effective pulmonary blood flow and VAD filling, and (4) physiology-driven VAD titration. Obtaining effective hemostasis is especially important in Fontan patients to facilitate timely initiation of anticoagulation and early extubation, and to minimize the impact of the vasoplegia that often characterizes the end-stage Fontan circulation, especially when liver disease is present.⁴⁵ To facilitate VAD filling and a drop in venous pressures, patients are maintained on inhaled nitric oxide and early extubation is prioritized. Inhaled nitric oxide can generally be weaned within 2 to 3 days without the need for additional pulmonary vasodilators. We try to ensure that the patient should not require blood products once getting to the intensive care unit so that

extubation can be achieved in less than 24 hours, which we have done for the majority of our patients.

Given the published hemocompatibility outcomes with the HeartMate 3, we generally err on the side of later initiation of intravenous anticoagulation when compared with previous VADs given the risk of bleeding in this patient cohort. We typically use heparin with a subsequent transition to coumadin (goal international normalized ratio 2-3) and aspirin (81 mg daily), although we have used bivalirudin rather than heparin in selected patients, especially those with low AT3 or significant effusions.

After extubation, higher than normal VAD flows may be needed to optimize central venous pressures and systemic oxygen delivery given the presence of AP collaterals. A goal-directed approach is recommended given the individual anatomic and physiologic variability. Our center typically aims for a Fontan pressure of 10 to 15 mm Hg with adequate VAD filling at rest and with activity (eg, minimal pulsatility index events, no ectopy), arterial-venous oxygen saturation difference of approximately 25% to 30%, and normal end-organ function. Flow is often at a cardiac index of 4 or more with RPMs on the HeartMate 3 of 7000 or greater at the beginning, but we have noted with our chronically supported patients that as the collaterals regress, we need to turn down the RPMs.

Although the hemodynamic management is important, early mobility and initiation of rehabilitation are equally so. This will facilitate earlier hospital discharge, but also

begin the process of optimizing the clinical and functional state of patients whether they are headed toward transplant or will remain on support for chronic therapy. At our institution, we use a “flight plan” that tracks the individual patient’s clinical course as well as their expected clinical trajectory given comorbidities and clinical events (Figure 1).

CLINICAL OUTCOMES

The last 5 years (and the last 2 in particular) have seen a significant increase the use of VAD therapy in the failing Fontan circulation. This is likely a convergence of multiple factors including the ever-increasing number of patients living with a Fontan, the demonstration that VAD therapy can be effective in improving Fontan-related hemodynamics, and that hemocompatibility-related outcomes are manageable with the current generation, continuous-flow VADs. The first large, multicenter series from ACTION demonstrated VAD support can be effective.²¹ Approximately 80% of patients experienced a positive outcome at 1 year (70% transplanted, 9% alive on device). The results were encouraging, but also demonstrated opportunities to improve outcomes. Seventy percent of patients experienced at least 1 adverse event, including a relatively high rate of neurologic adverse events. This included 12 patients who had an ischemic or hemorrhagic stroke. All of these neurologic events occurred in patients on an HVAD, Centrimag (Abbott

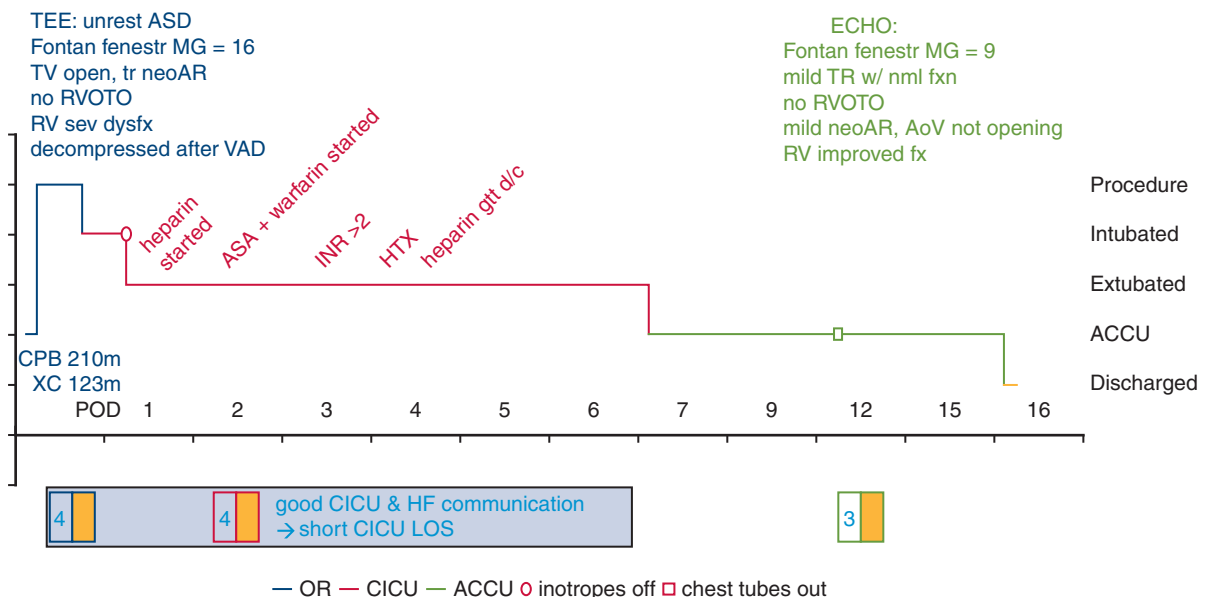


FIGURE 1. Perioperative “flight plan.” Example of the “flight plan” used by our institution that tracks each patient’s expected and actual clinical progression throughout the perioperative period. The patient is a 14-year-old with HLHS post-Fontan whose course was complicated by poor cardiac function and weight loss who underwent implantation of a HeartMate 3 VAD, resulting in clinical improvement and hospital discharge. TEE, Trans-esophageal echocardiogram; RVOTO, right ventricular outflow tract obstruction; VAD, ventricular assist device; ECHO, echocardiogram; OR, operating room; CICU, cardiovascular intensive care unit; ACCU, advanced cardiac care unit; ASA, aspirin; INR, international normalized ratio; HTX, heart transplant; d/c, discontinued; CPB, cardiopulmonary bypass; XC, crossclamp; HF, heart failure; LOS, length of stay.

Medical, Inc), or EXCOR (Berlin Heart). Given the superior hemocompatibility of the HeartMate 3 and with further maturation of the field, the authors expect this will decrease significantly.

The ACTION report also demonstrated the heterogeneity of hemodynamic responses to VAD support. Many patients, although not all, had improved hemodynamics (decreased Fontan pressures and common atrial pressures) post-VAD. This point is significant and highlights the importance of understanding the reasons for Fontan circulatory failure, as VAD therapy is effective when Fontan failure is due to systolic dysfunction with elevated end-diastolic pressure and low cardiac output. The hemodynamic benefit appears more mixed when the hemodynamic derangement is more mixed and includes predominant diastolic dysfunction, severe lymphatic disease, or cavopulmonary failure.

While the field does continue to mature and learn how to use VAD therapy optimally in the Fontan circulation, an optimistic approach does appear warranted. Recent data from the combined Pedimacs/Interagency Registry for Mechanically Assisted Circulatory Support databases demonstrate improving outcomes and increased device use.⁴⁶ This report documented a 95% freedom from stroke at 6 months and noted that more devices were implanted between 2018 and 2019 than were between 2013 and 2017.

CONCLUSIONS

VAD therapy is effective in supporting the patient undergoing the Fontan with predominant systolic dysfunction. Ongoing research will help identify patients who have multiple causes of Fontan failure and may still benefit from VAD therapy. As the VAD experience grows, fundamental questions such as how to approach Fontan fenestration and whether atrial cannulation is beneficial in smaller patients may also be answered.

Conflict of Interest Statement

D.L.S.M. is a consultant for Abbott Medical, Inc, Azyio, Inc, and Syncardia Inc; a consultant and member of the advisory board for Berlin Heart, Inc, Cormatrix, Inc, and Xeltis Inc. All other authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

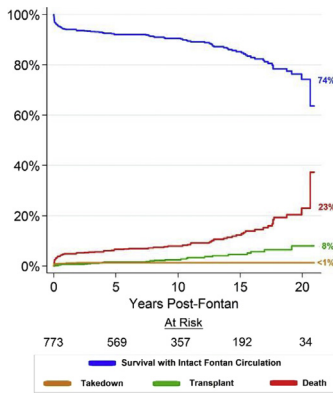
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Key Words: Fontan procedure, heart failure, heart transplantation, mechanical circulatory support, ventricular assist device

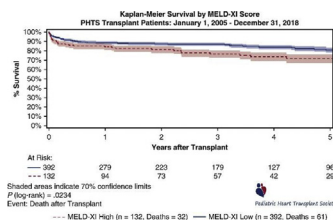
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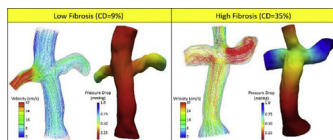


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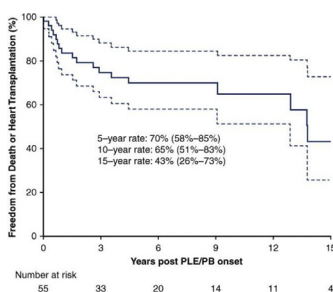
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