Commentary: The Fontan cardiac transplant—time to embrace change

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I enjoyed reading the article by Amdani and colleagues in which they objectify what seems intuitively obvious: For a Fontan cardiac transplant patient, a higher Model for End-Stage Liver Disease (MELD)-XI score (squared) portends worse survival. Although this is clinically relevant, I would like to discuss other nontrivial outcomes not emphasized by the authors.

**OBSERVATION 1**

Of 6518 pediatric cardiac transplant patients in the Pediatric Heart Transplant Society database, 783 Fontan patients were listed, and 565 were ultimately transplanted. This waitlist mortality of 28% is considerably higher than the published rate of 8% to 16% for the general population of listed pediatric patients. Worsening cardiac status and multiple system organ failure accounted for the majority of waitlist deaths.

**OBSERVATION 2**

One-year survival after transplantation was significantly higher for patients with dilated cardiomyopathy (DCM; 96%-97%) compared with Fontan recipients (85%-89%). This discrepancy existed for both eras (2005-2013 and 2014-2018). In those Fontan patients who died, 26% died of cardiac causes. Although the causes were likely multifactorial, this figure signifies that putting a new heart in a Fontan milieu is vastly different than doing so in the DCM setting. Unfortunately, single-ventricle or Fontan status is not incorporated in the risk models used to compute observed to expected survival—a figure that can have tremendous repercussions for programs. This needs to change!

**OBSERVATION 3**

Because there are numerous factors (some highly linked or interrelated) that contribute to mortality, I encourage the reader to not infer that use of a ventricular assist device (VAD) in a Fontan patient is bad. Good VAD support can definitely enhance recovery of end-organ dysfunction. Yes, as a specialty we need to better understand exactly which patients will benefit from a particular mode of support, and we need to refine some of our surgical techniques. In addition, we need a much better understanding of systemic-to-pulmonary artery collateral flow—applicable to both VAD placement and transplant.

In considering these 3 observations, my opinion is that as a specialty, we need to embrace change. I have yet to implant a VAD or transplant a Fontan patient and later felt that we pulled the trigger too soon. We need a collaborative effort to devise new selection criteria with the goal of either earlier VAD implantation and/or earlier listing. For either VAD implantation or transplantation, there is almost invariably some type of complication, albeit sometimes minor. A Fontan patient who meets the “traditional” criteria...
for intervention (transplant or VAD), at least in my opinion, does not have nearly the reserve of a typical biventricular patient regarding tolerance of intraoperative or postoperative insult and the therapy required to rescue. At the same time, we also need a better understanding and some level of specialty consensus on when to say “no.” It’s one thing when implanting off-the-shelf metal, but it’s another thing when using a limited resource donor organ. What projected transplant mortality warrants a “no” and still respects our primary duty to our patient? Ultimately, in the absence of a new supply of donor organs, and with improving technology, we will likely need to embrace a destination mindset for some (or many) of these patients.

**References**


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**Commentary: The MELD-XI score in Fontan patients: It’s about time**

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Total cavopulmonary connection and establishment of Fontan circulation has revolutionized the care of patients with single ventricle physiology.1 However, longitudinal follow-up shows that the functional health status of Fontan survivors decreases over time,2 and that a substantial number requires reintervention to maintain effective Fontan circulation.3 Late Fontan failure can result from primary single ventricle dysfunction and/or failure of Fontan circuit. The optimal management of patients with failing Fontan circulation remains a monumental challenge, and the appropriate timing of cardiac replacement therapy is one of the most difficult clinical decisions to make.

Many patients with Fontan circulation exhibit extra-cardiac organ dysfunction, frequently hepatic and renal pathology inherent to their chronically elevated venous pressures. The Model for End Stage Liver Disease (MELD) score was originally developed to quantify the degree of liver and kidney dysfunction using serum creatinine, bilirubin, and prothrombin time international normalized ratio (INR) values. However, many Fontan patients receive therapeutic anticoagulation for thromboembolic prophylaxis, fenestration patency, or mechanical support, which precludes use of the MELD score. The Model of End-Stage Liver Disease Excluding INR (MELD-XI) score has previously been shown to correlate