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.

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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 extracardiac organ dysfunction, frequently hepatic and renal

Fontan-associated liver disease.

CENTRAL MESSAGE

Refining long-term Fontan outcomes is the next challenging frontier in our battle against single ventricle disease.

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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.

Received for publication March 11, 2021; revisions received March 11, 2021; accepted for publication March 12, 2021; available ahead of print March 16, 2021.

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J Thorac Cardiovasc Surg 2022;163:1475-6

0022-5223/\$36.00

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<https://doi.org/10.1016/j.jtcvs.2021.03.036>

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 with survival following heart transplantation in adults.⁴ In

FIGURE 1. A, Cardiac catheterization showing a fenestrated Fontan (arrowhead, F) with a stent in the left pulmonary artery (arrow). B, Magnetic resonance imaging of the same patient showing nodular changes in the liver (arrows) secondary to Fontan circulation. The patient underwent successful heart-liver transplantation.

this issue of the *Journal*, Amdani and colleagues⁵ present data from the multi-institutional Pediatric Heart Transplant Society database demonstrating that the MELD-XI score reliably identifies pediatric Fontan patients at increased risk for mortality following heart transplantation. Patients with MELD-XI scores in the top 25th percentile at the time of heart transplantation had inferior 1- and 5-year posttransplantation survival.

On the face of it, these results imply that Fontan patients with worse liver and kidney function do poorly and, in that regard, are quite intuitive. Furthermore, because the authors used a percentile cutoff within their cohort to define high MELD score, how that value applies to an individual Fontan patient remains to be seen. In addition to high MELD-XI score, the authors report that a history of protein-losing enteropathy and the presence of a ventricular assist device (VAD) at transplantation also predicted poor outcome. These sobering data remind us once again that Fontan circulation is a suboptimal physiology with an ongoing hazard for multi-organ system dysfunction and death.

An interesting observation in this study is that VAD implantation was associated with improved MELD-XI scores during the waitlist period. VAD implantation comes with unique challenges in the single ventricle population⁶ and was encountered in only approximately 5% of the patients at heart transplantation in this study. Given the small numbers, VAD use did not correlate statistically with better heart transplantation outcomes; yet the ability to improve end-organ function using a VAD is encouraging and should further strengthen ongoing efforts to identify better devices to support the failing Fontan circulation. That said, this study, like previous smaller studies, shows that early identification of Fontan failure before progression to advanced liver and kidney dysfunction is paramount to maximize post-heart transplantation survival. Could proper utilization

of the MELD-XI score allow for timely referral for transplant evaluation? Or is there an earlier and more reliable predictor of progressive liver dysfunction? Studies of routine surveillance of Fontan patients suggest that virtually all of them will demonstrate some degree of liver disease by adolescence (Figure 1).⁷ Could we then predict even before reaching Fontan circulation who would progress to multiorgan dysfunction following Fontan completion? And if so, could we directly proceed with heart transplantation from the Glenn stage without transitioning through the Fontan circuit and the obligatory pressurization of the venous system? This study by Amdani and colleagues gives us strong reasons to pursue the answers to these daunting questions. Clearly, refining long-term Fontan outcomes is the next challenging frontier in our battle against single ventricle disease.

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