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Commentary: If only we knew when we would fail

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Approximately 5 decades ago, Fontan and colleagues¹ gave a new lease on life to patients with single ventricle physiology by introducing a novel surgical procedure. Soon after the total cavopulmonary connection (TCPC) surgery was designed, it was realized that patient selection was critical for the success of the procedure; thus, the commandments of the Fontan procedure were introduced.² As the procedure underwent several modifications and perioperative management improved, contraindications of the procedure were liberalized.³

In this issue of the *Journal*, Quaila and colleagues⁴ have tried to address the problem of predicting early Fontan failure. They retrospectively reviewed the imaging and clinical data of 131 patients who had undergone total cavopulmonary circulation. In the first part of the study, they created and validated a formula to predict postoperative hemodynamic data using preoperative cardiac magnetic resonance (CMR) imaging. The calculated post-TCPC central venous pressure (CVP) correlated well with the

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CENTRAL MESSAGE

Early Fontan failure may be predicted early enough to improve outcomes of patients with single ventricle physiology.

actual postoperative CVP (correlation coefficient of 0.26; $P = .03$). An important caveat to this was that the calculated post-TCPC CVP overestimated the actual postoperative CVP by approximately 7 mm Hg. The correlation was even stronger in cases without a fenestration. In the second part of the study, they aimed to arrive at a calculated CVP that would predict Fontan takedown. The suggested threshold of predicted post-TCPC CVP of 33 mm Hg or more was highly predictive of Fontan failure requiring surgical intervention (fenestration, takedown, transplant) or mortality.

The authors have to be commended for the work on patients palliated with single ventricle circulation. With advances in the field of perfusion, perioperative care, and better understanding of single ventricle physiology, the risk of early failure after staged palliation of single ventricles has been minimized. The risk of early Fontan failure in this series was 5%, which is similar to other reports. The few cases of Fontan failure seen in clinical practice do not necessarily conform to conventional commandments, making prediction of Fontan failure challenging. Patients with normal pulmonary artery resistance and good single ventricular function may face Fontan failure. All patients in this study underwent preoperative CMR imaging

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before the TCPC. Quaila and colleagues⁴ devised a method based on CMR parameters to help physicians predict the risk of Fontan failure in the absence of classic harbingers. CMR is emerging adjuvant not only in pre-TCPC imaging but also in selected cases replacing conventional catheterization.⁵ However, many centers do not routinely perform CMR in the pre-Fontan workup. The variables used by the authors based on preoperative CMR to arrive at the predicted post-TCPC CVP are not available to those who do not routinely perform CMR before surgery. Thus, creating and validating similar formulae that predict post-TCPC CVP using cardiac catheterization would aid in surgical planning.

Engineers have recently come to the rescue of pediatric cardiac physicians in predicting postoperative hemodynamic variables using preoperative imaging, hemodynamic data, and state-of-art computational fluid dynamics.⁶ Recent studies on a small number of patients have shown a good correlation between predicted postoperative hemodynamics and calculated computational fluid dynamics data.⁷ Using these advanced computational fluid dynamics technologies to predict Fontan failure would be a desirable bench-to-bedside transition.

The prevention of a Fontan failure seems to be more important than its prediction. The lack of pulsatility is one factor that has been shown to result in endothelial dysfunction.^{8,9} This is one factor that could be producing Fontan failure despite normal Fontan pressures and ventricular function. In their retrospective comparing the long-term outcomes of the Fontan in patients with and without antegrade flow at the time of the Glenn procedure, Chen and colleagues¹⁰ found significantly better outcomes after pulsatile Glenn procedures, that is, a Glenn circulation with restricted antegrade pulmonary flow, which is still a topic of controversial discussion.

We congratulate the authors on their contribution to better understanding, predicting, and thus potentially preventing early Fontan failure after TCPC.

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