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Commentary: How to avoid early Fontan failure?

Michelle Mizrahi, MD, and Ziv Beckerman, MD

We congratulate Quail and colleagues¹ for their article in this issue of the *Journal* and their efforts to advance this complex field of single-ventricle palliation. Currently, the Fontan procedure, or total cavopulmonary connection (TCPC), serves as the last step of palliation for several single-ventricle congenital heart conditions.² It allows for the entire systemic venous return to passively flow to the pulmonary arterial tree without an interposed pump.³ Several requirements must be met for TCPC to be successful (some well-known, others still being explored), necessitating extensive and meticulous evaluation of pre-Fontan patients.

The current common practice of pre-TCPC evaluation is aimed at providing an in-depth physiological understanding of the prospective Fontan candidate, with one of the goals being to avoid, among several other complications, early Fontan failure (EFF). EFF is a devastating scenario following TCPC that presents with low cardiac output and elevated central venous pressure (CVP).⁴

During this evaluation, patients usually undergo echocardiography, cardiac catheterization, and computed tomography or cardiovascular magnetic resonance (CMR). Although some centers have a wide implementation of preoperative cardiac catheterization,² others do not.⁵ Cardiac catheterization provides precise hemodynamic and physiological parameters of the patient being evaluated.

In their study, Quail and colleagues attempted to omit the pre-TCPC catheterization, using a different approach to predict post-TCPC CVP (CVP_{TCPC}). It is generally accepted that elevated CVP after TCPC is associated with

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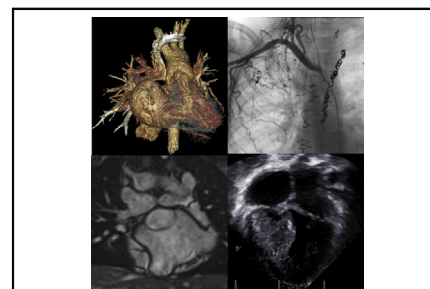
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The evaluation of the prospective Fontan candidate requires a multimodality approach.

CENTRAL MESSAGE

Our goal should be to achieve a zero percent rate of early and late Fontan failure.

Fontan failure.⁴ The authors used information gathered from a pre-TCPC CMR, along with the CVP measured via jugular venipuncture at the same time, to predict the CVP_{TCPC}.

Although the authors did not demonstrate a perfect correlation between the predicted and actual post-TCPC CVP, they did find a consistent association between the predicted CVP_{TCPC} and EFF. They concluded that estimated CVP_{TCPC} can help guide important clinical decisions, such as creating a preemptive TCPC fenestration (reserved by some institutions for high-risk patients).⁶ It should be mentioned that the rate of elective TCPC fenestration in this study was rather high (41%).

The authors should be congratulated for this important contribution to the body of knowledge, and for providing us with additional tools in our everlasting attempt to better define the ideal hemodynamics for Fontan patients. Owing to the critical importance of this palliative step and its potential morbid and/or fatal complications, it is our practice and recommendation to undertake elaborate and comprehensive preoperative evaluation for all patients. We recommend continuing to perform pre-TCPC catheterization, as well as taking a highly targeted approach to TCPC fenestration. The value of CVP_{TCPC} can very well be added to our armamentarium.

The ultimate goal should be to have an early Fontan failure rate of zero percent.

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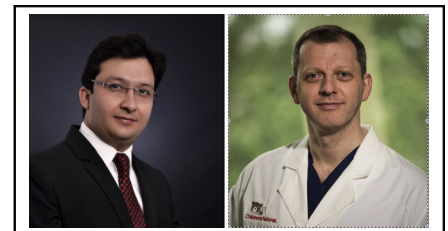
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Commentary: If only we knew when we would fail

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CENTRAL MESSAGE

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

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

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

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