Commentary: Singling out single ventricles after Fontan

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The Fontan circulation.

Central Message

Novel therapeutics and interventions are needed to address the late morbidity and mortality risks associated with unfavorable Fontan circulation characteristics.

See Article page 2005.

The Fontan palliation for patients with single-ventricle physiology has undergone considerable modification since its introduction in 1971.¹ Over the past 2 decades, the atriopulmonary anastomosis has given way to the total cavopulmonary connection with an intracardiac lateral tunnel or an extracardiac conduit (Figure 1). Despite improvement in outcomes, the Fontan circulation remains compromised and long-term effects of this physiology, including venous congestion and decreased cardiac output, carry lifelong morbidity and mortality risks.²⁻⁴

Single-ventricle morphology and preoperative hemodynamic parameters have historically been the main predictors of deleterious late outcomes.^{5,6} More recently, prolonged postoperative intensive care unit (ICU) length

of stay (LOS) has been implicated in poorer prognosis.⁷ Complete characterization of the natural history of the Fontan circulation is essential in elucidating modifiable risk factors and developing beneficial interventions. Ono and colleagues⁸ report results of a single-center retrospective study of 483 patients over a 22-year period aimed at identifying morphologic, hemodynamic, and perioperative risk factors for extended ICU LOS after Fontan palliation and describe their effect on longer-term outcomes. Their first key finding is that anomalous systemic venous drainage; aortopulmonary collaterals; elevated preoperative

FIGURE 1. The Fontan circulation. A, Atriopulmonary anastomosis. B, Lateral tunnel. C, Extracardiac conduit. RPA, Right pulmonary artery; LPA, left pulmonary artery; SVC, superior vena cava; RA, right atrium; LA, left atrium; IVC, inferior vena cava; RV, right ventricle; LV, left ventricle, TCPC, total cavopulmonary connection. (Reproduced with permission of the © ERS 2018; *European Respiratory Review* 25(142):438-450; <https://doi.org/10.1183/16000617.0091-2016>. Published 30 November 2016.)

transpulmonary gradient; and postoperative pleural effusion, chylothorax, and ascites are predictors of prolonged ICU LOS. These factors either contribute to or result from the impediment to systemic venous flow, which is critical to adequate functioning of the Fontan circulation. Their second key finding is that ICU LOS was again associated with worse late outcomes.

These findings provide a set of potentially modifiable risk factors that span the life course of the Fontan palliation. Together, they should be viewed as another challenge to the multidisciplinary team treating these complex patients. Anomalous systemic venous drainage and aortopulmonary collaterals result in undesirable flow dynamics before the Fontan procedure, including inadequate pulmonary artery growth, venous hypertension, and elevated transpulmonary gradient, which all negatively influence Fontan circulation.^{9,10} Strong consideration should be paid to catheter occlusion of aortopulmonary collaterals, repair of pulmonary artery distortion, and pulmonary vasodilator therapy to reduce transpulmonary gradients before or at the time of Fontan completion. Development of novel methods to improve venous congestion and revisiting criteria for Fontan fenestration may also be warranted. Further, attention to venous flow patterns and hemodynamic parameters should extend beyond the perioperative period, when additional opportunities for early intervention may prevent late effects. In the study by Ono and colleagues,⁸ prolonged pleural drainage was the main contributor to increased ICU LOS. Respiratory insufficiency, inefficient Fontan circulation from elevated venous pressure, decreased oncotic pressure, and increased infection risk from protein and immunoglobulin loss may all mediate this negative association.^{11,12} Aggressive and systematic medical, interventional, and surgical management strategies to decrease pleural drainage should be implemented and have previously been shown to improve outcomes.^{13,14}

It is evident that there is a subset of patients who experience extended ICU LOS after undergoing the Fontan operation. The need to develop new or adaptable

interventions in this population is vital and will require careful investigation. However, the impetus to reduce ICU LOS using a multimodal approach throughout the perioperative period has never been clearer.

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