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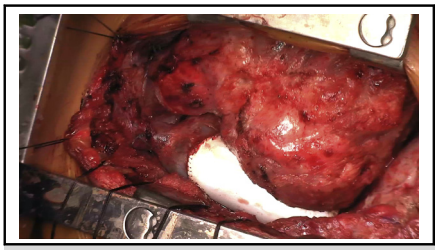
Commentary: The heart of the matter: Close clinical follow-up and exercise capacity in Fontan circulation

Andrew Well, MD, MPH, MSCHT, and Ziv Beckerman, MD



Extracardiac Fontan: 18-mm conduit.

CENTRAL MESSAGE

Exercise capacity is an important outcome in patients who have undergone the Fontan. A review presents the importance of continued hemodynamic and exercise testing. Critical assessment of exercise testing is required.

In this issue of the *Journal*, Patel and colleagues[1] evaluate the cross-sectional area (CSA) of extracardiac Fontan conduits and associated hemodynamic impact at various times during follow-up. The authors are to be commended for their compilation of a large number of patients with Fontan circulation (FC) with magnetic resonance imaging, catheter, and cardiopulmonary exercise testing (CPET) data. Their analysis revealed a median CSA decline of 68% compared with the original CSA of the conduit implanted. The decline in CSA was found to have no association with cardiac parameters; however, CSA indexed to body surface area was associated with percent predicted maximal oxygen uptake on CPET. These findings are consistent with previous computational modeling[2] and a retrospective review from South Korea.[3] It is insightful that no clear association was found between the implanted conduit size and the measured hemodynamic parameters. Surgeons frequently debate intraoperatively as to what size conduit to implant, with a bias toward trying to implant the largest conduit possible without causing distortion of surrounding structures.

Exercise capacity and CPET have been used as important outcomes in research of patients with FC. A prime example is the recent outcomes of the FUEL trial.[4] CPET requires careful analysis and interpretation. Many challenges in the conduct and evaluation of CPET in patients with FC are highlighted in the manuscript from Patel and colleagues.[1] CPET was performed by 2 methodologies: treadmill and cycle ergometry. There are inherent differences in these 2 methodologies with treadmill testing resulting in a mean of 9% and a maximum of 18% higher maximal oxygen uptake compared with cycle.[5] Given this disparity, it is not advisable to combine the results of the 2 methods. An important characteristic to evaluate in CPET is the respiratory exchange ratio (RER), the ratio of carbon dioxide produced to oxygen consumed. In practice, the RER represents the effort put forth during the CPET with a RER 1.1 or more representing adequate effort while a RER 1 or less represents poor effort and validity of the maximal oxygen uptake should be questioned.[6] As shown in the article by Patel and colleagues, the mean RER was 1.1, resulting in half of patients in the study having a CPET study in the sub-optimal range. It is important to note that a singular measurement of maximal oxygen uptake has not been consistently associated with

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mortality or hospitalizations in patients with FC.[7] Maximal oxygen uptake evaluation becomes prognostic in FC when performed serially and reveals a decline over time.[8]

Beyond any prognostic ability, in ongoing research, the ability to keep up with peers physically has been identified as an important outcome to patients with FC. Given the dual importance of exercise capacity, it should continue to be used as a key outcome. This article by Patel and colleagues emphasizes the importance of long-term continuous clinical and hemodynamic evaluation of patients with FC. Ideally, in a prospective manner, with strict adherence to standardized CPET methodology. Predictors of decline in CPET measures and interventions to prevent decline and improve measures can result in great impact in directing care of patients with FC.

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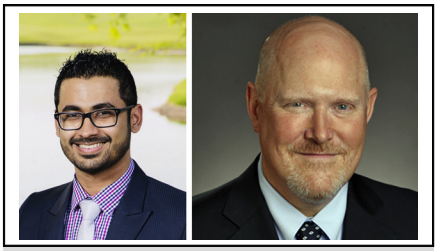
Commentary: As we learn more, we know little

Awais Ashfaq, MD, and James S. Tweddell, MD



Awais Ashfaq, MD, and James S. Tweddell, MD

CENTRAL MESSAGE

Optimal Fontan conduit size continues to be unknown.

The Fontan operation is the goal for patients with a single ventricle, and the procedure has undergone various modifications over the years. Originally introduced in 1990,[1] extracardiac conduit Fontan possesses some advantages such as improved flow dynamics, lower arrhythmias, and technically less challenging. Nonetheless, lack of growth potential and thrombogenecity are important drawbacks and lead to reinterventions. Patel and colleagues[2] have provided us with an important study looking at the intermediate outcome of the extracardiac Fontan. The authors studied 165 patients undergoing the extracardiac Fontan by magnetic resonance imaging over a span of 6 years. The results were discouraging. The Fontan cross-sectional area decreased in at least one-third of patients as early as 6 months postoperation, although there was no change thereafter. They found that patients had smaller than normal pulmonary arteries and there was an absence of pulmonary

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