Still young at heart...

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Surgical palliation for single-ventricle (SV) physiology has continued to improve over the last 40 years. Since the original description of the Fontan operation for tricuspid atresia in 1971,¹ the procedure has undergone several modifications and is now used to palliate a wide range of SV defects. Most patients with SV lesions require some form of neonatal palliation to ensure unrestricted systemic and pulmonary blood flow and free mixing of blood within the heart. Invariably, first-stage palliation results in a volume-loaded ventricle, as pulmonary resistance decrease with age. Generally, the second-stage palliation, most frequently in the form of a Glenn, is undertaken between 4 and 8 months of age. This relieves the volume load on the SV and improves effective pulmonary circulation by diverting deoxygenated blood to the lungs. There has been a trend to transition earlier to the second stage, largely because of its greater stability, especially in single-right ventricle patients. A recent report of patients with hypoplastic left heart after Norwood procedure confirmed that 6 months is still an optimal age for Glenn and that moving this time point earlier does not rescue high-risk patients.²

In contrast, no study to date has formally evaluated the optimal timing for final-stage palliation. Fontan circulation establishes near-normal systemic arterial saturation levels. Given that almost all of the venous return passes through the lungs, a Fontan circuit is thought to decrease the formation of aortopulmonary collaterals and, by providing hitherto uncharacterized “hepatic factors,” reduce the incidence of arteriovenous malformations.³ These favorable changes result in improved exercise capacity.⁴

However, there are theoretical benefits to delaying the Fontan procedure until the child is older. Glenn circulation affords more stable hemodynamics, ensuring cardiac preload from the inferior vena cava even in the setting of reduced pulmonary venous return due to pulmonary pathology. By delaying Fontan, one can potentially delay the increase in hepatic afterload, and consequent congestive hepatopathy, that is inevitable in the setting of a Fontan. Lastly, when an extracardiac conduit is used (Figure 1), older age would allow the use of a larger conduit.

Extracardiac nonfenestrated Fontan.

Central Message

Fontan circulation can be safely accomplished in older children with single-ventricle lesions. However, ventricular function determines long-term outcomes.

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Currently, most centers, including ours, perform the Fontan operation on patients between 2 and 4 years of age.^{5,6} Delayed Fontan is usually the result of delayed diagnosis or lack of access to specialized surgical care. Rarely, patients may be lost to follow-up or surgery would be postponed due to “high-risk” status. In this issue of the *Journal*, Ono and colleagues⁷ describe their institutional experience with the Fontan operation in older patients. They arbitrarily dichotomize their patient cohort and analyze the outcomes separately in patients belonging to the top 10th percentile of age, viz., older than 9 years. With an impressive median follow-up of 10 years, the authors provide data from nearly 5000 longitudinal echocardiograms and exercise testing in approximately one quarter of the patients. The older patients required more extensive procedures and spent more time on the ventilator and in the hospital. Yet, mortality was no different, implying that the surgery itself can be safely accomplished. In contrast, they were more likely to have reduced ventricular function, exercise capacity, and survival during follow-up. Based on analysis of the few exercise tests performed, the authors suggest that 2.8 years may be the optimal time for Fontan completion. Along the same lines, other reports have also suggested inferior outcomes in patients who undergo Fontan later in life.^{8,9}

But is age the real culprit? When analyzed in multivariable models, age was not associated with either survival or postoperative ventricular dysfunction.⁷ The best predictor of postoperative ventricular dysfunction was preoperative dysfunction, which was more frequently encountered

FIGURE 1. Angiogram showing an extracardiac nonfenestrated Fontan with an 18-mm polytetrafluoroethylene conduit.

in the older cohort. All deaths in the older cohort were in patients who had ventricular dysfunction post-Fontan. And the relationship between ventricular function and exercise capacity is well recognized. Several factors were likely associated with poorer preoperative function in the older patients. More than one third of them did not undergo Glenn, and in the rest, Glenn was performed at about 6 years of age. They had a greater proportion of heterotaxy, and Qp:Qs pre-Fontan was 1.4, indicating that the SV was subjected to a prolonged period of volume overload.

A logical conclusion is that it is ventricular function, more than chronologic age, that determines outcomes after

Fontan. Is there then an inflection point in age up to which the benefits of a Glenn circulation can be extended while simultaneously ensuring that Fontan circulation is reached before the onset of ventricular dysfunction? Are there modifiable factors that can reduce the incidence of ventricular dysfunction both before and after Fontan? And, when ventricular dysfunction is encountered, knowing the worse outcomes with Fontan, is heart transplantation a better alternative? As more and more patients with complex SV anatomy survive early palliation, we continue to await studies that will allow us to answer these difficult questions and refine our approaches to ensure the most favorable long-term outcomes.

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