Commentary: It isn't easy being single

Eric Purifoy, MD, and Iki Adachi, MD

Management of single-ventricle (SV) patients remains surgically and medically challenging. With increasing proficiency and experience in patient care, there has been substantial improvement in clinical outcomes for this population over the last several decades. However, despite advancements, numerous learning opportunities remain as we strive to make further progress. In this issue of the *Journal*, Stephens and colleagues reported an interesting observation characterized by differing timetables of hemodynamic normalization (defined as 2 consecutive catheterizations exhibiting the following: pulmonary capillary wedge pressure <12 mm Hg, pulmonary vascular resistance <3 indexed Woods units, and cardiac index >2.5 L/min/m²) between patients with SV physiology (Glenn or Fontan) and those with dilated cardiomyopathy. Although patients with dilated cardiomyopathy had normalization early after transplant, the SV population experienced delayed normalization (>12 months). Within the SV group, the delay in normalization for Fontan patients was more pronounced than those with Glenn physiology. This observation is fascinating and likely consistent with the impression of many clinicians. The critical question to be asked is, what are the implications of these findings? Among the plausible contributing factors, donor size selection was identified as potentially modifiable. The authors found that increased donor–recipient weight ratio contributed to the delay of hemodynamic normalization in SV patients. The authors attributed this finding to left ventricular hypertrophy found in patients who underwent a transplant with a donor–recipient weight ratio of >1.2:1. However, one may find this counterintuitive, as it is common practice to select a larger heart for SV patients, given the increased collateral burden due to chronic cyanosis. The quantity of pulmonary venous return is substantially increased in SV patients compared with those with structurally normal hearts. Thus, it would seem logical to use a larger heart to accommodate the increased preload so the myocardium can function within the normal operational range of the Frank–Starling curve. The collateral burden will likely resolve after establishing acyanotic biventricular physiology. In addition to cardiac adaptation, resolution of collateral burden likely plays a role in the delayed normalization of hemodynamics in SV patients. It may also explain, in part, the difference within the SV population (ie, Fontan normalize later than Glenn) due to the chronicity of SV physiology.

The landscape of pediatric heart transplantation continues to evolve as we advance our understanding of the complex interplay between anatomic and physiologic variability. As the number of palliated SV patients requiring transplantation increases, it is imperative to explore methods to improve outcomes. Each patient requires individualized evaluation, pretransplant optimization, and post-transplant monitoring. The authors’ work detailing post-transplant hemodynamics is certainly informative to the pediatric transplant community, particularly those caring for patients with complex SV physiology. It is not an
easy task to manage SV patients during the perioperative period, specifically post-transplant. It is also true that this patient population would receive the greatest benefit from transplant, given the dramatic conversion from cyanotic SV to acyanotic biventricular physiology. Ultimately, this is the goal for pediatric clinicians, to find solutions for these unique patients.

References