Commentary: How long until a new heart is a “normal” heart in transplanted single-ventricle patients?

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There are few existing data on expectations for hemodynamics changes following transplantation in children with failed single-ventricle (SV) palliation. Diastolic dysfunction manifested by elevated right atrial and wedge pressure (pulmonary capillary wedge pressure, or PCWP) is a known and frequent phenomenon observed during the early post-transplant period in patients.¹ Filling pressures improve over time, and most patients are asymptomatic with normal systolic function even before resolution of diastolic dysfunction. Whether normalization of posttransplant hemodynamics occurs similarly independent of pretransplant diagnosis is uncertain. The anatomic substrate and abnormal pretransplant physiology of patients with a failed SV place them at particular risk for intraoperative and perioperative posttransplant complications and might contribute to their greater posttransplant mortality.² Thus, understanding how preoperative diagnosis interacts with normalization of postoperative hemodynamics has the potential to provide important insights into dynamic changes in both allograft and patient physiology following transplantation. Furthermore, the timing of normalization of hemodynamics has important implications in understanding what should constitute “normal” posttransplant hemodynamics.

In this issue of the *Journal*, Stephens and colleagues³ provide an initial evaluation of how cardiomyopathy and SV patients' hemodynamics change over time. The authors found that, compared with patients with cardiomyopathy, those with failed SV palliation had delayed normalization of hemodynamics following transplantation. This is not surprising but also previously undocumented. Multiple factors inherent to this population, including pretransplant pulmonary architecture, the risk of silent pulmonary embolism, noncompliant augmented or stented aortas, underlying renal dysfunction, and a more complex intraoperative and perioperative transplant course, all likely contribute to delayed hemodynamics normalization of in this subset of patients.

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Central Message

Delayed normalization of hemodynamics is common following transplantation for failed single-ventricle palliation, but the underlying mechanisms and the impact on graft function and long-term outcomes are uncertain.

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Of the parameters measured by Stephens and colleagues (PCWP, pulmonary vascular resistance, and cardiac index), the PCWP had the longest period to normalization, suggesting a combination of diastolic dysfunction and potential volume loading as the result of collaterals. Other potential contributors to elevated PCWP in this complex population include the interaction between renal dysfunction and diastolic cardiac dysfunction (potentially exacerbated in these patients by longer cardiopulmonary bypass times and ischemic time),⁴ as well as abnormal pulmonary vascular resistance. Although acknowledged by the authors, it is important to reinforce that PCWP may not accurately reflect the true left ventricular end-diastolic pressure in patients with pretransplant congenital heart disease and a high risk for associated pulmonary vascular disease. Further details of both right heart and pulmonary vascular hemodynamic data would be important in fully understanding the post-transplant course in these patients, in whom the frequent need for pulmonary artery reconstruction and the potential for residual gradients may impact right graft function.

The authors have highlighted the delayed normalization of the PCWP in failed SV patients posttransplant but have also shown that this delay does not appear to affect short-term survival. Therefore, the study's principle utility may be in ameliorating family and provider anxiety and minimizing unnecessary invasive and noninvasive procedures.

Whether this delayed normalization has an impact on mid- to long-term graft function and survival remains unknown. Studies in adults suggest increased late mortality when either left or right diastolic dysfunction persists beyond 6 months posttransplant, but whether these findings apply to children, especially those with failed SV palliation, is less certain.⁵ Further studies assessing long-term transplant survival in this population would bring a new understanding to the field of pediatric transplantation. Future investigations should seek both to understand the etiology of elevated PCWP (collaterals, renal dysfunction, or intrinsic myocardial dysfunction) and to identify how abnormal hemodynamics influence long-term outcomes.

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