The journey toward improved hypoplastic left heart syndrome outcomes continues—another small step

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Effective, consistent, long-term surgical palliation for hypoplastic left heart syndrome (HLHS) remains a partially realized goal. Although survival associated with stage I (Norwood) palliation has improved, 5-year outcomes after multistage palliation continue to be marginal, with overall survivals just better than 50%. Achieving a durable, efficient Fontan circulation in these children can be a challenging proposition because of the compromised single right ventricle and the potential for distorted, poorly developed branch pulmonary arteries. Efforts to mitigate these confounding issues remain the focus of all involved in the care of patients with HLHS.

In this issue of the *Journal*, Bentham and colleagues¹ from Boston Children's Hospital add incremental refinement to our thinking about optimizing the opportunity for a durable Fontan circulation after HLHS palliation. Their efforts remind me of an aphorism from Lao Tzu I have admired, "A journey of a thousand miles must begin with a single step." While the journey toward staged HLHS palliation was begun several decades ago, and there have been many steps, it remains an arduous journey, fraught with frustration and disappointment. Even a small step of progress encourages further focus.

The use of a right ventricle–pulmonary artery (RVPA) conduit in the first stage of HLHS palliation was reintroduced to surgical methodology in 2004 by Sano and colleagues.² The single-ventricle reconstruction trial sought to discern outcomes differences in patients receiving an RVPA conduit in comparison to a systemic to pulmonary artery shunt.³ While the debate lingers as to which source of pulmonary blood flow results in superior outcomes, it is undeniable that many surgeons have adopted the RVPA conduit strategy as their primary method. In this context,

Bentham and colleagues sought in this study to refine their strategy further by examining outcomes after use of a ring-reinforced (RR) RVPA conduit in comparison with patients receiving a nonreinforced conduit. As others have observed, the use of a nonreinforced conduit is associated with a significant need for conduit reintervention as a result of stenosis. In their current article, Bentham and colleagues have documented a significant reduction in the need for reintervention after the use of a RR conduit. This is of course intuitive, in that the RR conduit should theoretically be less vulnerable to compression and distortion. An additional, and potentially very important, finding is that the patients receiving the RR conduit appeared to have superior branch pulmonary artery development.

Although avoiding the use of a nonreinforced conduit may seem to be a minor adjustment in technique, the potential for important long-term patient benefit cannot be disregarded. Improved branch pulmonary arteries should translate into improved Fontan candidacy, although that conclusion awaits further study. In the meantime, this does appear to be another small step forward in the journey.

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