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Commentary: Form improves function: The importance of a well-constructed neo-aortic arch

Mohan J. John, MD, and Travis J. Wilder, MD

The importance of a well-constructed neo-aortic arch during a Norwood procedure is not just because it is aesthetically pleasing. According to Schafer and colleagues,¹ well-constructed neo-aortic arch may have long-term hemodynamic implications for children with hypoplastic left heart syndrome (HLHS) and related variants requiring single-ventricle palliation.¹

Survival among neonates with HLHS has dramatically improved since the Norwood operation was first reported in 1983.² Hospital survival that was reported around 52%-68% in early studies has improved to between 80% and 85%.³ Furthermore, the 10-year survival published in reports from 2000 to 2005 was 40% to 50% compared with contemporary survival reported to be between 60% and 70%.³⁻⁵ With the expectation that many patients with HLHS will live into their second and third decade of life—perhaps longer—clinicians are focused on improving the long-term durability of the right ventricle when subjected to single-ventricle physiology.

The Fontan circulation is associated with nonpulsatile pulmonary blood flow, chronic systemic venous hypertension, and low cardiac output.⁶ Although many of the complications associated with single-ventricle physiology are a result of these hemodynamic derangements (ie, plastic bronchitis and protein-losing enteropathy), over time, all patients with HLHS will eventually develop heart failure.^{7,8}

Smooth versus tapered arch.

CENTRAL MESSAGE

Suboptimal arch reconstruction during Norwood procedure causes energy loss and decreased RV power. Diminished RV efficiency may lead to worse long-term outcomes in patients with hypoplastic left heart syndrome.

Although it is somewhat intuitive to expect a systemic right ventricle to fail, improved mechanical efficiency of the systemic and pulmonary circulations may delay the onset of cardiac failure after single-ventricle palliation.⁹

Given the importance of optimizing cardiac efficiency after single-ventricle palliation, Schafer and colleagues¹ set out to identify the association between neo-aortic arch geometry and flow-related viscous energy loss (E_L). Specifically, they used state-of-the-art, 4-dimensional magnetic resonance imaging to characterize dynamic aortic flow patterns in patients after Norwood palliation. They went on to classify the degree of neo-aortic tapering (ie, variation of aortic size along its length) and aortic morphology in an effort to determine the relationship between arch geometry and E_L . Further, they investigated the effect of flow-related mechanical E_L on right ventricle power. Thus, the study indirectly links the quality of neo-aortic reconstruction with cardiac efficiency of the single right ventricle.

The study included 26 patients with HLHS after stage-1 Norwood procedure. The stage of palliation was varied and included 5 patients in the post-Norwood/pre-Glenn inter-stage period, 12 patients after cavopulmonary shunt, and

From the Division of Congenital Heart Surgery, Texas Children's Hospital, Baylor College of Medicine, Houston, Tex.

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Address for reprints: Travis J. Wilder, MD, Division of Congenital Heart Surgery, Texas Children's Hospital, Baylor College of Medicine, Legacy Tower, Floor 19, 6651 Main St, Mail Code 19345H, Houston, TX 77030 (E-mail: tjwilder@texaschildrens.org).

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9 patients post-Fontan. A median aortic tapering value (defined as the standard deviation of aortic diameter measured at 4 prespecified locations along the aorta) was calculated for all patients. A median value of 0.52 cm was considered the cutoff between a high degree of tapering (Group H) and low degree of tapering (Group L). Key findings showed that patients in Group H demonstrated increased flow related E_L in all measured aortic regions. The study also showed that patients in Group H were susceptible to increased right ventricle power loss compared with Group L. Finally, they showed that independent of group, the magnitude of arch reduction from transverse arch to aortic isthmus, correlated directly with the amount of E_L across the entire measured aorta. Notably, there was not a significant association between palliation stage and E_L , suggesting that initial arch geometry—independent of arch growth—heavily influences flow dynamics.

The clinical importance of these findings is rooted in the fact that delaying Fontan failure is among the most difficult challenges faced by clinicians treating patients with single-ventricle physiology. Intuitively, it is reasonable to assume that optimal ventricular-aortic coupling and improved right ventricle efficiency would perhaps help delay Fontan failure. The current study is among the first to show that suboptimal arch reconstruction leads to flow related E_L and diminished right ventricle power. As such, it provides a

pathway to improve right ventricle efficiency and potentially improve long-term survival for patients with HLHS.

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