Commentary: Between the devil and the deep blue sea: A new palliation for high-risk neonates with hypoplastic left heart syndrome

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In 1980, William Norwood reported his first successful first stage palliation for hypoplastic left heart syndrome (HLHS).¹ Despite variations in technique, patients with HLHS undergoing the first-stage operation retain a significant risk of mortality perioperatively, with estimates of 7% to 39%.² Much of the variation depends on center experience, but another important feature is inherent patient characteristics. Specific subgroups of neonates with HLHS have been identified as being at risk for mortality, including patient factors such as small size, prematurity, or a genetic syndrome.³ There are also high-risk anatomic subsets, including those patients with an intact atrial septum, right ventricular dysfunction, atrioventricular valve regurgitation, and the presence of left ventricular dependent coronaries.^{3,4}

What is the best option for patients who are at high risk for mortality when undergoing stage 1 procedure or hybrid procedure? If primary transplant appears to be the best long-term option, what is the best strategy to bridge these patients until a donor heart is available? Different approaches to these high-risk neonates have been suggested, including using a hybrid technique as a bridge to transplant or bridge to decision.^{5,6} A small case series by Philip and colleagues⁷ describes a novel approach: marrying a hybrid palliation, including placement of bilateral pulmonary artery bands, a patent ductus arteriosus stent, and an atrial septectomy, with the placement of a Excor pulsatile ventricular assist device (Berlin Heart Inc, The Woodlands, Tex) (from the right atrium to the main pulmonary artery).⁷ They were able to palliate these complex patients for several months (range, 56 to 162 days) with 100% 30-day survival and with 2 of 5 patients successfully transplanted. This is an early experience with limited follow-up. No patients in this series had clinically apparent cerebral vascular accidents, although the high risk of neurologic events has been well described

Between the devil and the deep blue sea.

Central Message

Neonates with HLHS and certain high-risk factors remain at high risk for mortality and cardiac transplantation following attempts at palliation. This report provides a novel strategy for palliation.

See Article page e59.

with the Excor,⁸ especially in patients with single ventricle anatomy.

Ultimately, neonates with HLHS and certain patient factors and anatomic features remain at very high risk for mortality following all attempts at primary palliation, as well as on the waitlist for cardiac transplantation. The authors and their entire team should be congratulated on their outcomes following this novel strategy that may provide another option to families and physicians stuck between the devil and the deep blue sea.

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