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Commentary: Successful pairing of the Norwood and bidirectional Glenn in select older infants

Joseph B. Clark, MD

In the stepwise palliation of single ventricle disease with systemic obstruction, the Norwood operation is typically performed in the first week of life and followed at 3 to 6 months by superior cavopulmonary connection. The optimal timing of the staged operations has been extensively studied and has shaped current practice.^{1,2} Nevertheless, there may be the rare occasion when a patient presents unusually late and needs a Norwood operation. This scenario was recently described in a large series from Saudi Arabia in which patients underwent delayed Norwood surgery.³ Although pulmonary blood flow was routinely accomplished with a Blalock-Taussig shunt or Sano conduit, 7 patients were successfully managed with a cavopulmonary shunt (CPS) (ie, bidirectional Glenn).

Elmahrouk and colleagues⁴ present an expanded series of 16 patients treated with combined Norwood CPS at a mean age of 138 days. No patient had prior intervention and all had evidence of increased pulmonary blood flow. Cardiac catheterization was performed in the majority and showed severe pulmonary overcirculation with increased but reversible pulmonary vascular resistance. Not unexpectedly, postoperative CPS pressures were elevated (mean 20 mm Hg) compared with other reports.⁵ Inhaled nitric oxide was used to manage postoperative pulmonary hypertension, but the effectiveness was not detailed. Previously, inhaled nitric oxide has shown variable efficacy at decreasing CPS pressures and improving oxygenation.^{6,7} Of the 2 hospital

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CENTRAL MESSAGE

This novel experience shows that a combined Norwood cavopulmonary shunt is a viable option with comparable survival for select late-presenting infants with increased pulmonary blood flow.

deaths, one occurred in the oldest patient (366 days) with the highest pulmonary vascular resistance (3.5 WU/m² on 100% oxygen) who died from pulmonary hypertensive crisis on the second day. Given this outcome, the authors recommend an upper threshold of 2.5 WU/m² on 100% oxygen for Norwood CPS.

As the authors comment, hospital mortality in this series (12.5%) mirrored the comprehensive stage 2 operation (12.4%) in the hybrid pathway.^{4,8} Although both operations share aortopulmonary amalgamation, arch reconstruction, and CPS, the patient groups possess distinct anatomy and physiology. In hybrid-treated patients, the pulmonary arteries have been banded resulting in restricted blood flow and a protected pulmonary vasculature. When the bands are removed, arterioplasty may be required to treat residual stenosis.⁸ On the contrary, late-presenters in this series have lived with unobstructed pulmonary blood flow and massive pulmonary overcirculation. Although these patients may realize some benefit from larger pulmonary arteries, they are disadvantaged by aggravated pulmonary vascular resistance.

Although the late timing of the Norwood is noteworthy, the early timing of the CPS in 5 patients younger than age 3 months may merit caution. Because studies have shown increased risk for early CPS, the optimal timing of surgery is reported as age 3 months or older.² Similarly, hybrid proponents have recommended avoiding the comprehensive

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stage 2 operation before 3 months of age.⁹ Although the youngest patients in this series all survived, a Blalock-Taussig shunt or Sano conduit may have been a better option.

In the end, the study provides an interesting look at an unusual scenario uncommon to most centers. This novel experience shows that a combined Norwood CPS is a viable option in late-presenting infants with increased pulmonary blood flow. Although the postoperative course may be rocky in this challenging situation, often requiring delayed sternal closure and pulmonary vasodilator therapy, intermediate survival appears comparable to the conventional sequence of staged operations.^{1,2,4}

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Commentary: The right procedure for the right patient

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The Achilles' heel of single-ventricle palliation is the inherently unstable interstage circulation following both the Norwood procedure with systemic-to-pulmonary artery shunt or stage I hybrid palliation. This is reflected in the high mortality reported in patients with hypoplastic left heart syndrome before completion of a cavopulmonary shunt and the subsequent flattening of survival curves after stage II palliation.¹ Primary construction of

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CENTRAL MESSAGE

Primary combined Norwood cavopulmonary shunt procedures are feasible, but only in a highly selected group of patients.

a cavopulmonary anastomosis is impeded by high pulmonary vascular resistance (PVR) in neonates and young infants. Attempts to achieve a primary in-series circulation showed that this strategy was challenging, although not impossible.²⁻⁴

Elmahrouk and colleagues⁵ report their experience with primary combined Norwood and cavopulmonary shunt

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