Cavopulmonary assist: Bridge to Fontan repair of single-ventricle circulation

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The Toronto group is courageously tackling the intractable problem of neonatal single-ventricle palliation. Despite the clinical uptake of significant alternative procedures (right ventricle—pulmonary artery shunt, hybrid procedures), their impact has been incremental and outcomes remain a challenge. An entirely different approach may be necessary, providing the impetus behind the study of Honjo and colleagues¹ in this issue of the *Journal*.

In healthy neonates with biventricular circulation, pulmonary vascular resistance (PVR) drops nearly to adult values within hours after birth. Hypoxemia is absent, and PVR remains low—as long as there is no reactive stimulus. The risk for reactivity diminishes during transitional pulmonary maturation (approximately 2-4 months). In neonates with single-ventricle circulation, a systemic-to-pulmonary arterial shunt is used as a high-pressure source of pulmonary blood flow, ostensibly to overcome elevated PVR. Under “optimal” physiologic conditions, however, PVR should theoretically be low.

Here lies the enigma of neonatal single-ventricle palliation. The shunt paradoxically induces and sustains the pathophysiology that mandates it. High-pressure flow and hypoxemia are potent stimuli of pulmonary hypertension. The parallel arrangement of the systemic and pulmonary circulations is inherently unstable. The hypoxia-stressed ventricle is volume overloaded. Pulmonary vascular maladaptation likely worsens eventual Fontan candidacy and long-term status. Not surprisingly, clinical courses are correspondingly difficult and, all too expectedly, unexpectedly end poorly. If the problematic shunt could be avoided, these problems could be resolved.

A series circulation is inherently stable, with normoxia and normal volume. Neonatal pulmonary perfusion from a systemic venous source (neonatal Glenn or Fontan circulation) will, however, require a modest pressure boost (about 6-12 mm Hg) from a subpulmonary power source for some period. This requirement led to

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Central Message

Outcomes of neonatal single-ventricle palliation remain a serious challenge. Alternative approaches, such as mechanical cavopulmonary assist, may be necessary to make a meaningful impact.

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the concept of bridge support with mechanical cavopulmonary assist.²

To this end, the study of Honjo and colleagues¹ provides acute physiologic insight into mechanically assisted bidirectional Glenn shunt (combined stages I and II) as initial palliation. A superior cavopulmonary shunt with mechanical assist is used in lieu of a systemic-to-pulmonary arterial shunt, producing a theoretically more stable “series” circulatory arrangement. It is only a “partial series” circulation, however; inferior vena caval flow continues into the systemic circulation, and hypoxemia persists. Although an assisted Glenn shunt may reduce instability risk, it may provide an incomplete solution.

A mechanically assisted Fontan circulation (combined stages I, II, and III) is theoretically a complete solution.³ The pulmonary and systemic circuits are fully separated, and hypoxemia and volume overload are eliminated. This may seem unrealistic, but it would emulate normal biventricular circulatory physiology (imagine a stable Norwood circulation with alveolar P_{O_2} >400 mm Hg) and may yield the best long-term outcomes. Technology that can safely

and reliably mechanically assist Glenn and Fontan flows is emerging, but it has yet to be clinically translated.⁴

Although cavopulmonary assist is an exceedingly complex technical challenge, it has the potential to provide meaningful advances in single-ventricle repair, including the possibility of correction rather than palliation. This study is an important step in that direction. Otherwise, we are condemned to keep repeating the past. Unconventional approaches are difficult; however, patients with single-ventricle anatomy fare poorly in the current pathogenic paradigm and stand to benefit greatly from a paradigm shift.

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