Commentary: Mechanical support for the subpulmonary circulation of Fontan physiology—A step in the right direction

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The Fontan operation is one of the greatest milestones in medicine and has saved the lives of numerous children born with single-ventricle heart disease.¹ Fontan was among the first to notice the palliative, rather than curative, nature of the operation. The palliative nature stems from the unique characteristics of the Fontan circulation, the lack of a subpulmonary ventricle, which leads to a range of cardiac and noncardiac complications. It is becoming apparent that such a peculiar physiology is destined for, in the long term, a failure of the circulation (failing Fontan circulation). This challenging condition has a high incidence of mortality with limited therapeutic options.²

In the hopes of “normalizing” the circulation, much effort has been expended to develop a method to reverse the “pumpless” subpulmonary hemodynamics.³ A novel support system reported by Lin and colleagues⁴ in this issue of the *Journal* is one such example. In their avant-garde concept, a percutaneously inserted multilumen cannula is coupled to a pump for subpulmonary assistance. The simple design allows the inflow lumen to draw flow from both caval veins while returning blood into the pulmonary arteries through separate, adjustable outflow cannulas. This strategy may not only provide downstream pressures gains but also minimize recirculation, which is a common concern for mechanical support for total cavopulmonary connection without compartmental separation of inflow and outflow ports.⁵

Proof-of-concept studies by computational methods are a good first step. Prototype development and rigorous in vitro testing are typical next steps needed before clinical translation. Because of the difficulty in creating an animal model of Fontan failure, testing with physiologically relevant mock circulation loops mimicking the failing Fontan circulation may be considered.^{6,7} Challenges to be overcome include the potential for inflow suction events of large devices in compliant and narrow venous channels, as well

VAD support for Fontan circulation, determined by primary reason for circulatory failure.

Central Message

With few medical or surgical options available for patients with failing Fontan circulation, innovative solutions are needed to improve patient outcomes.

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as the potential for inadvertent competing flows in the

contralateral superior vena cava in complex anatomies with bilateral superior cavopulmonary connections. Ideally, the proposal by Lin and colleagues⁴ would decrease central venous pressures while increasing pulmonary arterial flow to improve cardiac preload.

Patient selection is paramount. A major phenotype of failing Fontan circulation is systemic ventricular dysfunction, which can be supported well with a systemic ventricular assist device.⁸ When used appropriately, this type of support carries a high success rate, as evidenced by 95% 6-month survival.⁹ The concept of subpulmonary support would be relevant to a distinctly different phenotype, one that is characterized by flow limitation primarily in the pulmonary circulation. This phenotypic subset is more challenging for mechanical support, with only few sporadic reports of successful clinical cases and likely many more unreported failed attempts.¹⁰ This is the aspect that makes the study described by Lin and colleagues⁴ important to the field. As they appropriately recognize, the device is intended only for a relatively short duration (ie, months at most). Because of the insidious nature of the failing Fontan circulation, which typically progresses over decades, it is more prolonged support that would potentially reverse the inevitable disease processes. This could give patients with

long-standing failing Fontan circulations time to become better candidates for surgical interventions, including organ transplantation (cardiac transplant with or without other visceral organs). Although no such ideal device currently exists, promising technologic advances may one day achieve this goal.³

The report from Lin and colleagues⁴ demonstrates the need for ingenuity to progress the care of patients with a failing Fontan circulation. As has been repeatedly shown in medical history, it is this innovative spirit that brings about paradigm shifts in patient care. We eagerly anticipate that the article by Lin and colleagues⁴ will further stimulate the development of mechanical support options for this ever-growing population.

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