Hitting “turbo” after the race is done: Subpulmonic support in failed Fontan circulation

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Central Message

If subpulmonic ventricular assist device support of the Fontan circulation is the new “fourth stage,” perhaps it should be instituted sooner than we can at this time.

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The article by Broda and colleagues¹ represents an excellent compilation of the previous efforts at improving the subpulmonic blood flow after the Fontan procedure. As the authors describe, there has been a significant number of investigations into the logical concept of re-creating what evolution has created over millions of years, a 2-ventricle heart for humans. However, there are a multitude of unique issues, which the Fontan circulation presents, and any device that requires deconstructing the cavopulmonary connection to augment forward flow and decrease venous congestion represents a significant surgical burden. Unfortunately up to now, most in vivo devices have failed to achieve any real benefit, but several existing concepts show future promise of being able to augment pulmonary blood flow for long durations of time. Although there is currently only one instance of a subpulmonic assist device successfully supporting a failing Fontan circulation to reverse end-organ dysfunction before transplantation, Broda and colleagues¹ agree that elective placement of subpulmonic “support could become the ‘fourth stage’ of single ventricle palliation.”^{2,3} If this is to become the next step of treating single-ventricle pathologies, then not only is device development and implantation strategy critical, but we must also consider the optimal timing for this intervention.

The primary issue with the concept of assisting subpulmonic flow is that the Fontan circulation typically fails at multiple levels as the body develops several maladaptive responses to the unusual passive flow circulation. These changes lead to an abnormal pulmonary vasculature, compromising blood flow into a systemic ventricle, which often develops a restrictive physiology from years of underfilling. Long-term studies have shown Fontan survivors to have preserved systolic function in 73% of patients and diastolic dysfunction in 72% further emphasizing that this group is quite unique from patients we commonly place on mechanical

circulatory support.⁴ The diastolic dysfunction is likely due to a chronic preload deprivation and impaired ventricular filling, which worsens as patients get further from their Fontan surgery.⁵ Therefore, when the features of circulatory failure become present, forcing blood through a pulmonary vasculature that has already developed chronic pathology into a restricted ventricle would appear to be a futile attempt to fix a broken system after it's too late. This definitely would not appear to be an effective strategy for chronically supporting these patients. Conversely, instituting subpulmonic support early after the Fontan procedure could provide ample blood flow to prevent some of the maladaptive responses and pathophysiological changes that occur. If we could find a means of safely augmenting subpulmonic blood flow for long durations of time, perhaps soon after Fontan completion, then we could “turbocharge” the Fontan. A “turbo Fontan circulation” has the potential to avoid the issues from chronic passive flow by manufacturing the state evolution had intended, a “biventricular circulation.”

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