See Article page 227.

Commentary: Fontan candidacy in which all roads do not lead to Rome

Elizabeth H. Stephens, MD, PhD

Elizabeth H. Stephens, MD, PhD

CENTRAL MESSAGE

In the quest to optimize the care of single-ventricle patients, the characteristics required for Fontan candidacy continue to be refined, but not all patients will be Fontan candidates—what then?

Venna and colleagues[1] provide an excellent summary of our current understanding regarding optimizing Fontan completion. The famous "Ten Commandments"[2] related to Fontan completion proposed by Choussat and Fontan himself have, like many great propositions, been tested and refined with time and experience. This summary acts as a revision to those original concepts.

To briefly recap the highlights, although preserved systolic and diastolic function goes without saying, in recent years particular attention has been paid to the atrioventricular valve(s) (AVVs) and the importance of their competence for a successful Fontan circulation.[3-5] The importance of optimizing the pulmonary vascular bed, both the pulmonary arteries and venous return, is emphasized. The need for a pacemaker portends a particularly poor prognosis among patients with Fontan circulation, increasing mortality by 14-fold according to one study.[6] The paper further iterates potential actions to improve various aspects that may not be "ideal" for a given patient.

There are several clinical questions ripe for our field to investigate emanating from the concepts within this paper. The first is the timing of Fontan completion. Long term does unloading the single ventricle and AVV earlier improve the longevity of the AVV and ventricle, thereby prolonging successful Fontan circulation? Should Fontan completion be performed once an "adult-sized" graft can be placed if other aspects, such as AVV competency and ventricular function, are favorable for Fontan completion? Or is more time in a circulation with a cavopulmonary connection awaiting Fontan completion actually better long term compared to the added time with Fontan circulation if Fontan completion is performed earlier? Do the answers to these questions vary depending on anatomy? Perhaps those with a single ventricle with right ventricular morphology and a tricuspid valve have better outcomes if they are offloaded with a Fontan circulation sooner, but perhaps this isn't true for other single-ventricle anatomic subtypes.

As with the discussion of any intervention for a patient, we are taught to review the risks, benefits, and alternatives. Inherent to any patient's clinical management pathway when we choose one path we are by default choosing not to follow other paths. For the many patients who, even after the optimizations that Venna and colleagues have outlined, remain "not-ideal" Fontan candidates, the question becomes if we say no to Fontan completion, what will we say yes to? If they are bluer than ideal, do we choose to add some sort of shunt, thereby adding a volume load to a heart and/or AVV that already is struggling with a bidirectional cavopulmonary circulation? Or does this precipitate ventricular failure and need for transplant? Do we tolerate more hypoxia than we would like and refer to transplant once that worsens? Should some of these patients proceed to Fontan completion with the knowledge that transplant may be required sooner than other patients? How should

From the Department of Cardiovascular Surgery, Mayo Clinic, Rochester, Minn.

Disclosures: The author reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

Received for publication Aug 1, 2021; revisions received Aug 1, 2021; accepted for publication Aug 3, 2021; available ahead of print Aug 17, 2021.

Address for reprints: Elizabeth H. Stephens, MD, PhD, Department of Cardiovascular Surgery, Mayo Clinic, 200 First St, SW, Rochester, MN 55905 (E-mail: stephens.elizabeth@mayo.edu).

JTCVS Open 2022;9:235-6

2666-2736

Copyright © 2021 The Author(s). Published by Elsevier Inc. on behalf of The American Association for Thoracic Surgery. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

https://doi.org/10.1016/j.xjon.2021.08.003

the degree of sensitization inform our surgical decisions regarding their paths?

Ultimately, even for the "ideal" Fontan candidates, the Fontan circulation remains a palliation. These patients, in many ways, are living on "borrowed" time and the fact that so many of these patients live to adulthood is a testament to those physicians and surgeons before us who worked through the many failures and persevered. Certainly in coming years we will learn to further improve the care of these patients, but one of the most important aspects of their care that needs to be clarified is when patients should ideally be referred for transplant. We must optimize the lives of these patients within the context of a limited number of organs and frequent issues related to sensitization. All too often our single-ventricle patients are deemed "too well" for transplant but then reappear and are considered "too sick." We must find that sweet spot when patients gain the most benefit from transplant.

In ancient days, the saying was "all roads lead to Rome." We have learned that not all single-ventricle patients' paths lead to a Fontan, but the question remains how best to manage these "not ideal" Fontan candidates.

References

1. Venna A, Cetta F, d'Udekem Y. Fontan candidacy, optimizing Fontan circulation, and beyond. *J Thorac Cardiovasc Surg Open*. 2022;9:227-32.
2. Choussat A, Fontan F, Besse P. Selection criteria for the Fontan procedure. In: Anderson RA, Shindebourne EA, eds. *Pediatric Cardiology*. Edbinburgh: Churchill Livingstone; 1977:559-66.
3. King G, Ayer J, Celermajer D, Zentner D, Justo R, Disney P, et al. Atrioventricular valve failure in Fontan palliation. *J Am Coll Cardiol*. 2019;73:810-22.
4. Stephens EH, Dearani JA. Management of the bad atrioventricular valve in Fontan…time for a change. *J Thorac Cardiovasc Surg*. 2019;158:1643-8.
5. Stephens EH, Dearani JA, Niaz T, Arghami A, Phillips SD, Cetta F. Effect of earlier atrioventricular valve intervention on survival after the Fontan operation. *Am J Cardiol*. 2020;137:103-10.
6. Poh CL, d'Udekem Y. Life after surviving Fontan surgery: a meta-analysis of the incidence and predictors of late death. *Heart Lung Circ*. 2018;27:552-9.