The pendulum of Fontan fenestration

Antonio F. Corno^, Taylor S. Koerner, Jorge D. Salazar

Department of Pediatric and Congenital Heart Surgery, Children's Heart Institute, Memorial Hermann Children's Hospital, University of Texas Health Science Center in Houston, McGovern Medical School, Houston, TX, USA

Correspondence to: Antonio F. Corno, MD, FRCS, FETCS, FACC. Children's Heart Institute, Memorial Hermann Children's Hospital, University of Texas Health Science Center in Houston, McGovern Medical School, 6410 Fannin Street, MSB 4.144, Houston, TX 77030, USA. Email: tonycorno2@gmail.com

Keywords: Congenital heart defects; congenital heart surgery; fenestration; Fontan procedure; single ventricle

Submitted Oct 31, 2022. Accepted for publication Jan 10, 2023. Published online Jan 16, 2023.
doi: 10.21037/tp-22-562
View this article at: https://dx.doi.org/10.21037/tp-22-562

In the 50 years since the introduction of the Fontan procedure for single ventricle palliation (1), the clinical indications as well as the surgical techniques have substantially evolved.

Initially the indication for Fontan completion was following the "Ten Commandments": (I) age <4 years; (II) presence of sinus rhythm; (III) normal systemic venous return; (IV) normal right atrial volume; (V) mean pulmonary artery pressure <15 mmHg; (VI) pulmonary arteriolar resistance <4 Wood Units/m^2; (VII) pulmonary/aorta ratio >0.75; (VIII) single (left) ventricle ejection fraction >0.60; (IX) competent mitral valve; (X) absence of pulmonary artery distortion (2).

The preoperative selection criteria for Fontan completion have expanded from the original "Ten Commandments" to a more liberal application, accepting higher-risk patients with "functionally" univentricular hearts (3,4); also including children with a single lung (5).

The decision-making regarding a Fontan fenestration as part of completion has shifted as well, with the pendulum swinging back and forth between infrequent utilization to near universal use over time.

Hillel Laks (6) introduced the concept of an "adjustable atrial septal defect" to temporarily reduce the excessively elevated systemic venous pressure after a Fontan procedure and reduce immediate post-operative complications. The name "fenestration", termed by Nancy D. Bridges (7), was universally adopted to define a surgically created communication between the diverted systemic venous return and the lower pressure pulmonary atrium (*Figure 1*). With the goal of a shorter post-operative recovery, the use of a Fontan fenestration gained increased attention with both surgical techniques utilized for Fontan completion, lateral tunnel as well as extracardiac connection (8,9).

Immediately after its introduction, the indications for a Fontan fenestration were limited to high-risk candidates, as defined by pre-operative mean pulmonary arterial pressure ≥15 mmHg and/or presence of moderate or severe degree of systemic atrio-ventricular valve regurgitation. Fenestration reduced the systemic venous pressure, resulting in increased lymphatic drainage with an associated reduction in pleural effusions. A fenestration also provided adequate preload to the systemic single ventricle which reduced the post-operative low cardiac output state (8). The only available prospective randomized study comparing patients undergoing fenestrated versus non-fenestrated Fontan completion demonstrated a reduction in the length of stay in the intensive care unit and hospital (10).

Because of the positive outcomes, the indication for fenestration was then extended to almost all patients, regardless the level of pre-operative risk, and became commonplace for a Fontan procedure.

^ ORCID: 0000-0003-4374-0992.

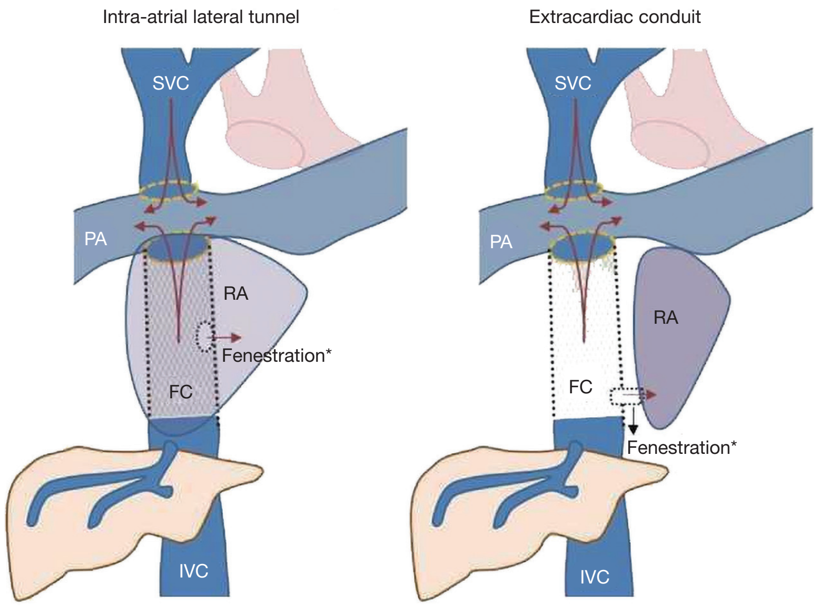

Figure 1 Drawing of the fenestration, constructed for the lateral tunnel (left) and for the extracardiac conduit (right). Modified from: Téllez L, Rodríguez-Santiago E, Albillos A. Fontan-associated liver disease: a review. Ann Hepatol 2018;17:192-204. SVC, superior vena cava; PA, pulmonary artery; RA, right atrium; FC, fenestrated conduit; IVC, inferior vena cava.

However, the benefits of fenestration in the early post-operative period were at the expense of late complications such as lower systemic oxygenation with prolonged cyanosis, risk of long-term systemic thromboembolism, and potential need for later intervention to close the fenestration. Fortunately, following an additional procedure and anesthetic exposure to close the fenestration, patients had improved resting and exercise oxygenation, lowered maximal heart rate during exercise, and increased exercise duration (11-13).

Despite the ability to mitigate the short-term effects, fenestration became limited, as at the beginning of its utilization, to patients with strict requirements, such as increased risks of complications in the immediate post-operative period, as reported by us (14).

Two systematic literature reviews and meta-analysis have focused upon the early outcomes of a Fontan fenestration, demonstrating a mix benefit in the immediate period, with reduced amount of chest drains and subsequent shorter stay in hospital as the only positive aspects (15,16). Our meta-analysis instead focused on late outcomes, with patients requiring either late closure or creation and/or reopening of a fenestration made at the time of Fontan completion (17). We found that, following fenestration closure, there was a significant increase in the mean arterial oxygen saturation of 7.9% [95% confidence interval (CI): 6.4–9.4%, P<0.01], at expense of a significant increase in the mean cavo-pulmonary pressure of 1.4 mmHg (95% CI: 1.0–1.8 mmHg, P<0.01) (17). The literature data for fenestration creation and/or reopening didn't allow any meaningful conclusion (17).

In addition to clinical study, mathematical and computational fluid dynamic models have also compared flow and hemodynamics for Fontan patients with and without a fenestration (18). These studies have sought to quantify the effects of different sizes of the fenestration (19) as well as alternative designs of Fontan circulation, different from the traditional surgical options (20).

The pendulum has since swung back. There has been a steady increase in the use of Fontan fenestration based on much broader indications. Largely related to better post-operative management, the option of Fontan completion has been extended to different patient populations, including patients undergoing completion at younger age, and with complex congenital heart defects, such as hypoplastic left heart syndrome (21), heterotaxy syndrome (22), and single ventricle physiology with associated total anomalous pulmonary venous connection (23). Because of these changes in the risk stratification of the patients accepted for surgery (24), the cohort of patients presented for Fontan completion are frequently at high risk for a complicated post-operative course. With the increasing complexity of patients undergoing a Fontan, surgical centers have begun to reconsider the use of fenestration and in some centers, especially those accepting the highest risk cases, it is used nearly universally.

As illustrated by the varying pattern of fenestration use, there is a substantial lack of high-quality scientific evidence supporting any therapeutic decision (17,25). Thus, in clinical practice, the decision to perform a fenestration and its size is based on the personal and institutional experience in relationship to the morphologic and pathophysiologic characteristics of a specific patient.

Acknowledgments

Funding: None.

Footnote

Provenance and Peer Review: This article was commissioned by the editorial office, *Translational Pediatrics* for the column "Pediatric Heart". The article has undergone external peer review.

Peer Review File: Available at https://tp.amegroups.com/article/view/10.21037/tp-22-562/prf

Conflicts of Interest: All authors have completed the ICMJE uniform disclosure form (available at https://tp.amegroups.com/article/view/10.21037/tp-22-562/coif). The regular column "Pediatric Heart" was commissioned by the editorial office without any funding or sponsorship. AFC serves as an unpaid Deputy Editor-in-Chief of *Translational Pediatrics* from July 2022 to June 2024. AFC and JDS served as the unpaid Guest Editors of the column. The authors have no other conflicts of interest to declare.

Ethical Statement: The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

References

1. Fontan F, Baudet E. Surgical repair of tricuspid atresia. Thorax 1971;26:240-8.
2. Choussat A, Fontan F, Besse P, et al. Selection criteria for Fontan's procedure. In: Anderson RH, Shinebourne EA. editors. Paediatric Cardiology. Edinburgh: Churchill Livingstone, 1977:559-66.
3. Corno A, Becker AE, Bulterijs AH, et al. Univentricular heart: can we alter the natural history? Ann Thorac Surg 1982;34:716-27.
4. Marcelletti C, Corno A, Giannico S, et al. Inferior vena cava-pulmonary artery extracardiac conduit. A new form of right heart bypass. J Thorac Cardiovasc Surg 1990;100:228-32.
5. Zachary CH, Jacobs ML, Apostolopoulou S, et al. One-lung Fontan operation: hemodynamics and surgical outcome. Ann Thorac Surg 1998;65:171-5.
6. Laks H, Haas GS, Pearl JM, et al. The use of an adjustable intraatrial communication in patients undergoing the Fontan and other definitive heart procedures. Circulation 1988;78:II357
7. Bridges ND, Lock JE, Castaneda AR. Baffle fenestration with subsequent transcatheter closure. Modification of the Fontan operation for patients at increased risk. Circulation 1990;82:1681-9.
8. Kuhn MA, Jarmakani JM, Laks H, et al. Effect of late postoperative atrial septal defect closure on hemodynamic function in patients with lateral tunnel Fontan procedure. J Am Coll Cardiol 1995;26:259-65.
9. Do-Nguyen CC, Kilcoyne MF, Gray P, et al. The evolution of surgical technique of the fenestrated Fontan procedure. J Card Surg 2020;35:1407-9.
10. Lemler MS, Scott WA, Leonard SR, et al. Fenestration improves clinical outcome of the fontan procedure: a prospective, randomized study. Circulation 2002;105:207-12.
11. Goff DA, Blume ED, Gauvreau K, et al. Clinical outcome of fenestrated Fontan patients after closure: the first 10 years. Circulation 2000;102:2094-9.
12. Meadows J, Lang P, Marx G, et al. Fontan fenestration closure has no acute effect on exercise capacity but improves ventilatory response to exercise. J Am Coll Cardiol 2008;52:108-13.

13. Boshoff DE, Brown SC, DeGiovanni J, et al. Percutaneous management of a Fontan fenestration: in search of the ideal restriction/occlusion device. Catheter Cardiovasc Interv 2010;75:60-5.
14. Salazar JD, Zafar F, Siddiqui K, et al. Fenestration during Fontan palliation: now the exception instead of the rule. J Thorac Cardiovasc Surg 2010;140:129-36.
15. Li D, Li M, Zhou X, et al. Comparison of the fenestrated and non-fenestrated Fontan procedures: A meta-analysis. Medicine (Baltimore) 2019;98:29-39.
16. Bouhout I, Ben-Ali W, Khalaf D, et al. Effect of Fenestration on Fontan Procedure Outcomes: A Meta-Analysis and Review. Ann Thorac Surg 2020;109:1467-74.
17. Greenleaf CE, Lim ZN, Li W, et al. Impact on clinical outcomes from transcatheter closure of the Fontan fenestration: A systematic review and meta-analysis. Front Pediatr 2022;10:915045.
18. Rijnberg FM, Hazekamp MG, Wentzel JJ, et al. Energetics of Blood Flow in Cardiovascular Disease: Concept and Clinical Implications of Adverse Energetics in Patients With a Fontan Circulation. Circulation 2018;137:2393-407.
19. Puelz C, Acosta S, Rivière B, et al. A computational study of the Fontan circulation with fenestration or hepatic vein exclusion. Comput Biol Med 2017;89:405-18.
20. Corno AF, Owen MJ, Cangiani A, et al. Physiological Fontan Procedure. Front Pediatr 2019;7:196.
21. Arunamata A, Tacy TA, Kache S, et al. Recent outcomes of the extracardiac Fontan procedure in patients with hypoplastic left heart syndrome. Ann Pediatr Cardiol 2020;13:186-93.
22. Baban A, Cantarutti N, Adorisio R, et al. Long-term survival and phenotypic spectrum in heterotaxy syndrome: A 25-year follow-up experience. Int J Cardiol 2018;268:100-5.
23. Vasquez Choy AL, Adebo DA, John S, et al. Essential role of cardiac computed tomography for surgical decision making in children with total anomalous pulmonary venous connection and single ventricle. J Card Surg 2022;37:1544-9.
24. Corno AF, LaPar DJ, Li W, et al. A narrative review of modern approach and outcomes evaluation in congenital heart defects. Transl Pediatr 2021;10:2114-22.
25. Daley M, Buratto E, King G, et al. Impact of Fontan Fenestration on Long-Term Outcomes: A Propensity Score-Matched Analysis. J Am Heart Assoc 2022;11:e026087.

Cite this article as: Corno AF, Koerner TS, Salazar JD. The pendulum of Fontan fenestration. Transl Pediatr 2023;12(1):104-107. doi: 10.21037/tp-22-562