

Symposium

Symposium 6 (II-S06)

Treatment strategy for failed Fontan

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Sat. Jul 8, 2017 1:50 PM - 3:35 PM ROOM 1 (Exhibition and Event Hall Room 1)

1:50 PM - 3:35 PM

[II-S06-01 【Keynote Lecture】]Treatment for the failed Fontan

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For the child with single ventricle anatomy the Fontan operation is the anticipated goal of palliation and is one of the great success stories of congenital heart surgery. Nevertheless, sustained elevation of central venous pressure and chronically low cardiac output are uniform consequence of the total cavopulmonary connection. As a result, a series of end-organ complications are now increasingly recognized. Among the increasingly recognized problems are; liver fibrosis and cirrhosis, protein losing enteropathy, impaired lymphatic drainage, decreased bone density and renal dysfunction.(1) Standardized follow-up and screening for individuals with Fontan physiology through childhood and beyond is indicated to optimize management. Even the individual with the " perfect" Fontan faces an uncertain future likely marked by progressive right heart failure and risk of premature mortality.(2-5) Strategies to improve long term outlook may include maintaining optimal Fontan architecture, minimization of thromboembolic complications, control of arrhythmias, resynchronization therapy and potentially procedures to improve lymphatic drainage.(6-10) Ultimately, heart transplant may offer the best hope but mortality remains higher among patients transplanted for a failing Fontan due to challenges of reoperation with aortopulmonary collaterals and the comorbidities of liver and renal dysfunction.(11, 12) Risk assessment and the best timing for transplant may require not only assessment of cardiac dysfunction but also careful assessment of the potential for hepatic dysfunction. Mechanical circulatory support tailored to the Fontan circulation will provide better bridge to transplant and may also permit recovery of liver and renal dysfunction.(13, 14) A comprehensive team approach that considers all organ systems is essential.(15)

1. Rychik J. The Relentless Effects of the Fontan Paradox. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2016;19(1):37-43.
2. Gewillig M, Brown SC. The Fontan circulation after 45 years: update in physiology. Heart. 2016;102(14):1081-6.
3. Pundi KN, Johnson JN, Dearani JA, Pundi KN, Li Z, Hinck CA, et al. 40-Year Follow-Up After the Fontan Operation: Long-Term Outcomes of 1,052 Patients. J Am Coll Cardiol. 2015;66(15):1700-10.
4. Paridon SM, Mitchell PD, Colan SD, Williams RV, Blaufox A, Li JS, et al. A cross-sectional study of exercise performance during the first 2 decades of life after the Fontan operation. J Am Coll Cardiol. 2008;52(2):99-107.
5. Giardini A, Hager A, Pace Napoleone C, Picchio FM. Natural history of exercise capacity after the Fontan operation: a longitudinal study. Ann Thorac Surg. 2008;85(3):818-21.

6. Lastinger L, Zaidi AN. The adult with a fontan: a panacea without a cure? Review of long-term complications. *Circ J*. 2013;77(11):2672-81.
7. Deal BJ, Costello JM, Webster G, Tsao S, Backer CL, Mavroudis C. Intermediate-Term Outcome of 140 Consecutive Fontan Conversions With Arrhythmia Operations. *Ann Thorac Surg*. 2016;101(2):717-24.
8. John AS, Johnson JA, Khan M, Driscoll DJ, Warnes CA, Cetta F. Clinical outcomes and improved survival in patients with protein-losing enteropathy after the Fontan operation. *J Am Coll Cardiol*. 2014;64(1):54-62.
9. van Melle JP, Wolff D, Horer J, Belli E, Meyns B, Padalino M, et al. Surgical options after Fontan failure. *Heart*. 2016;102(14):1127-33.
10. Sojak V, Mazic U, Cesen M, Schrader J, Danojevic N. Cardiac resynchronization therapy for the failing Fontan patient. *Ann Thorac Surg*. 2008;85(6):2136-8.
11. Bernstein D, Naftel D, Chin C, Addonizio LJ, Gamberg P, Blume ED, et al. Outcome of listing for cardiac transplantation for failed Fontan: a multi-institutional study. *Circulation*. 2006;114(4):273-80.
12. Michielon G, van Melle JP, Wolff D, Di Carlo D, Jacobs JP, Mattila IP, et al. Favourable mid-term outcome after heart transplantation for late Fontan failure. *Eur J Cardiothorac Surg*. 2015;47(4):665-71.
13. Imielski BR, Niebler RA, Kindel SJ, Woods RK. HeartWare Ventricular Assist Device Implantation in Patients With Fontan Physiology. *Artif Organs*. 2017;41(1):40-6.
14. Rossano JW, Goldberg DJ, Fuller S, Ravishankar C, Montenegro LM, Gaynor JW. Successful use of the total artificial heart in the failing Fontan circulation. *Ann Thorac Surg*. 2014;97(4):1438-40.
15. Stout KK, Broberg CS, Book WM, Cecchin F, Chen JM, Dimopoulos K, et al. Chronic Heart Failure in Congenital Heart Disease: A Scientific Statement From the American Heart Association. *Circulation*. 2016;133(8):770-801.