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Commentary: Management of patients with failing Fontan: More questions than answers

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Rajab and Jagers¹ review heart transplantation for Fontan failure. Their article raises unresolved questions. When should a Fontan patient be referred for transplantation? Their Table 1¹ outlining indications is disappointingly lacking in concrete guidelines. How does one determine that a Fontan patient has failed all standard therapies? What level of hepatic dysfunction warrants transplantation? To avoid the risk of combined heart/liver transplantation, one must predict when the nearly universal hepatic dysfunction associated with the Fontan circulation will become irreversible.

Despite improved results with heart transplantation in Fontan patients, the challenges of peri-transplant care should not be minimized. These include multiple redo sternotomies, difficulty calculating pulmonary vascular resistance, management of arteriovenous collaterals, bleeding diatheses, need for pulmonary artery reconstruction and re-routing of systemic and/or pulmonary venous connections, preformed antibodies, and underlying debility due to protein-losing enteropathy, hepatic or renal dysfunction, muscle wasting, and so on.

What is the fate of patients who undergo heart transplantation for Fontan failure? Patients transplanted for congenital heart disease do not fare as well as those with idiopathic cardiomyopathy. The median survival for all transplant patients aged 11 to 17 years (most Fontan transplant recipients are teenagers or young adults) is only 14.3 years.² This includes

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CENTRAL MESSAGE

The care of patients with failing Fontans is difficult. Transplantation is not an ideal solution. A widespread collaborative effort is necessary to answer important questions about this problem.

all transplants, not just patients with congenital heart disease or previous Fontan operations. Certainly, the transplanted Fontan patient cannot be expected to do even this well.

Mechanical circulatory support for Fontan patients is in its infancy, currently used as a bridge to transplant. More than 1000 Fontan procedures are done annually according to the Society of Thoracic Surgeons Congenital Heart Surgery Database.³ Yet the number of transplants for patients aged 11 to 17 years has remained constant at 200 to 300 per year (including all idiopathic cardiomyopathy patients and non-Fontan congenital heart disease patients).² Also, congenital heart disease accounts for only 2% to 3% of adult transplants.⁴ Due to donor limitations, transplantation cannot be the answer for all patients with failing Fontan, particularly considering Rajab and Jagers¹ suggest that nearly all patients with Fontan physiology may require eventual transplantation. Long-term mechanical support of these patients hopefully is the future. Can these patients be supported with a ventricular assist device for the systemic ventricle alone? Is an implantable total artificial heart necessary? Can an intravascular mechanical long-term support device be perfected?

To answer these important questions, a multi-institutional registry of Fontan patients is desirable. Longitudinal follow-up can aid understanding the onset of Fontan failure, helping ascertain the optimal timing for transplant listing or mechanical assistance. Following these patients after transplantation or mechanical support will define long-term outcomes. Armed with these data, we should improve the fate of

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Fontan patients with a 3-pronged approach: improved medical therapy reducing or delaying the onset of Fontan failure, improved posttransplant management with a goal (possibly unobtainable) of immune tolerance, and the development of reliable and safe long-term implantable mechanical support. The future for patients with failing Fontan might be promising. Remember, there are a lot of Fontan survivors out there!

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