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Commentary: Fontan takedown: The journey off the beaten path

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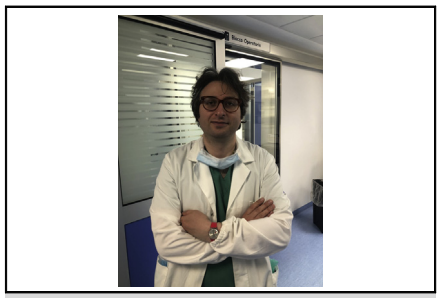

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CENTRAL MESSAGE

When Fontan circulation acutely fails, prompt surgical takedown is warranted. Although mortality has proven to be substantial, long-term outcomes for survivors are satisfactory.

Marathe and colleagues[1] queried the Australia and New Zealand Fontan Registry to evaluate early and late outcomes of patients who had undergone a Fontan takedown procedure.

Over a 43-year study period (1975-2018), 36 of 1540 (2.3%) patients had a Fontan takedown at a median age of 5.1 years (interquartile range, 3.7-7.0 years) with most takedowns occurring within 6 months after the index operation. A total of 16 patients died and data showed mortality was lowest when takedown occurred within 2 days after acute Fontan failure, and highest when done 3 to 24 weeks postcomplication. Eleven patients with an intermediate palliative circulation (8 with bidirectional superior cavopulmonary anastomosis) were alive at a median follow-up of 9.4 years. Five patients underwent a second Fontan operation after a median of 4.4 years and only 1 had a late failure. No remediable cause was identified in any patient before the second Fontan operation. Three other patients underwent biventricular or 1.5-ventricle repair.

Another publication reported that preoperative characteristics were not associated with an increased risk of failure and showed no correlation to the type of Fontan (atriopulmonary vs lateral tunnel vs extracardiac conduit).[2] Fifteen (41.6%) patients had a single-stage Fontan, although this was found not to be statistically significant. In addition, patients who underwent initial hybrid palliation were not included in the study, which represents a topic for further investigation.

Due to the high mortality risk associated with Fontan failure, 13 (27%) patients had 14 rescue procedures performed before takedown, including Fontan revision, enlargement of fenestration, coiling of aortopulmonary collaterals, pacemaker insertion, left pulmonary artery stenting, and removal of bronchial cast.

Early failure of Fontan circulation is rare (~2%), but remains associated with a high mortality rate due to the fact that these patients are often poor surgical candidates for a rescue procedure. The pathophysiology of acute failing Fontan circulation is characterized by a

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combination of increased systemic venous pressure and a low cardiac output state. Fontan takedown is warranted when medical therapy fails and treatments are exhausted. In some instances, when a poor hemodynamic state exists in the setting of elevated pulmonary artery pressure and low atrial pressure (ie, elevated transpulmonary pressure gradient), an attempt to establish a fenestration (if 1 is not present) might be sufficient to prevent Fontan takedown. In an acute setting, other surgical strategies have been proposed, such as institution of mechanical circulatory support[3] (either as a short-term bridge to recovery or as a bridge to transplantation), or rescue cardiac transplantation[4] without takedown or transcatheter takedown.[5] Nonetheless, morbidity and mortality associated with these options is still substantial.

When embarking on a traditional pathway of staged palliation, both the surgeon and the patient's parents must know that the treatment plan will be filled with its own unique challenges and that factors influencing the event of early Fontan failure remain unclear. Marathe and colleagues[1] further corroborate the idea that complex pediatric surgical procedures can fail even if everything goes forth in the correct manner. Although most operations go precisely as planned, there are occasions when surgeons must re-evaluate their approach due to less-than-ideal outcomes. In fact, a calculated retreat gives time to investigate failures and create solutions to optimize patient outcomes.

The authors are to be congratulated for this large experience and their clinical results. After reading this article, every surgeon should bear in mind 2 important things: Fontan takedown is a rare occurrence that when required, must be undertaken promptly; and even if mortality is high in the acute setting, alternative surgical options are still at hand (ie, heart transplant, redo Fontan later, or simply an intermediate palliative circulation).

When Fontan takedown seems imminent and necessary, surgeons and patients have options available to optimize long-term outcomes.

CONG

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