Fontan survival: As good as it gets?

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Survival for children with univentricular hearts has greatly improved since Fontan and Baudet¹ and Kreutzer and colleagues² described effective surgical palliation almost 4 decades ago, with initial early failure rates near 15%.^{3,4} Improvements in patient selection, surgical techniques, and perioperative management have resulted in contemporary early mortalities of less than 5%,^{3,5,6} with the focus now shifting toward identification of modifiable factors that may improve long term outcomes.

Downing and colleagues⁷ provide updated outcome data for a “modern” Fontan cohort, specifically including a substantial proportion of patients with single right ventricles, capturing eras during which Fontan completion strategy evolved from the lateral tunnel version to the extracardiac conduit approach. Survival was analyzed in 773 patients who underwent Fontan operations during a 20-year period, with spectacularly complete (>99%) follow-up obtained by combining local records with data from the National Death Index and the Scientific Registry of Transplant Patients. To demonstrate practice evolution, the cohort was divided into eras: 1992 through 1996, 1997 through 2002, and 2003 through 2009. A composite negative outcome variable included Fontan takedown, heart transplant, or death. Freedoms from this outcome were 94% at 1 year, 90% at 10 years, 85% at 15 years, and 74% at 20 years. Early outcomes improved as the eras progressed, but, disappointingly, late survival did not. Risk factors for early (<1 year) adverse outcomes include prolonged pleural drainage (>2 weeks), intensive care unit stay longer than 1 week, Fontan operation before 1997, preoperative mild or worse atrioventricular valve regurgitation (AVVR), and longer aortic crossclamp times, with adverse late outcome associated with mild or worse AVVR and post-Fontan intensive care unit stay longer than 1 week. Heterotaxy, ventricular morphology, and the presence of a common atrioventricular valve were not risk factors for adverse late outcome. Among patients undergoing pre-Fontan catheterization (87% overall, but only 76% in the most recent era), mean pulmonary arterial pressure greater than

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Central Message

Although early outcomes after the Fontan operation have improved, there remains substantial opportunity to improve late survival.

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15 mm Hg was a risk factor for early but not late adverse outcome.

The impact of AVVR was surprising and important given its high prevalence, increasing from 35% to 41% to 56% through the eras and accounting for 45% of the entire cohort. This is an interesting observation, given that half of patients with AVVR were in the mild category, not previously recognized as a high-risk subset.⁸ Indeed, evaluation of tricuspid valvuloplasty or replacement has focused on those with at least moderate regurgitation.^{9,10} Whether AVVR is a modifiable risk factor or simply a marker of worse Fontan candidacy remains to be elucidated.

The negative late implication of prolonged hospitalization was not unexpected but may raise concern at the institution of Downing and colleagues⁷ in light of the increases between the second and third eras in length of stay, length of pleural drainage, and incidence of drainage lasting longer than 14 days. Whether these changes reflect a 10-month older age at operation in the third era and the decision to omit catheterization in 25% of patients is not clear. It might be speculated that prolonged cyanosis results in more systemic to pulmonary collateral blood flow, longer effusions, and so on. Like all good studies, this report answers some questions, raises others, and points us toward possible improvement.

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