Intermediate-Term Outcome of 140 Consecutive Fontan Conversions With Arrhythmia Operations

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Background. **Atrial arrhythmias and progressive circulatory failure frequently develop in patients with a Fontan circulation. Improvement of flow dynamics and revision of the arrhythmia substrate may improve outcomes in selected patients. We sought to determine intermediate-term outcomes after Fontan conversion with arrhythmia operations and identify characteristics associated with decreased transplant-free survival.**

Methods. **The first 140 Fontan conversions with arrhythmia operations at a single institution were analyzed for predictors of cardiac death or transplant and incidence of arrhythmia recurrence.**

Results. **The median age at the Fontan conversion operation was 23.2 years (range, 2.6 to 47.3 years). Preoperative arrhythmias were present in 136 patients: right atrial tachycardia in 48 patients, left atrial tachycardia in 21, and atrial fibrillation in 67. Freedom from cardiac death or transplant was 90% at 5 years, 84% at 10 years, and 66% at 15 years. The median age at the last follow-up among survivors was 32 years (range, 15 to 61 years). By multivariable analysis, risk factors for cardiac death or heart transplantation were a right or indeterminate ventricular morphology, cardiopulmonary bypass time exceeding 240 minutes, ascites, protein-losing enteropathy, or a biatrial arrhythmia operation at the time of conversion. Freedom from recurrence of atrial tachycardia was 77% at 10 years. Among 67 patients with atrial fibrillation undergoing biatrial arrhythmia operations, none had recurrent atrial fibrillation.**

Conclusions. **Freedom from cardiac death or transplant for patients undergoing Fontan conversion with an arrhythmia operation is 84% at 10 years. The effects of atrial arrhythmia operations are durable in most patients. These outcomes may serve as useful benchmarks for alternative management strategies.**

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The atriopulmonary connection was introduced by Fontan and Kreutzer to separate the systemic and pulmonary circulations in patients with a functionally univentricular heart, with subsequent surgical modifications developed to optimize hemodynamic flow and limit arrhythmia development [1, 2]. Progressive exercise intolerance develops in patients with a Fontan circulation, particularly those with an atriopulmonary connection. By 15 years postoperatively, major adverse events develop in approximately half of patients, including thrombosis, atrial arrhythmias, or Fontan failure [3–5]. Timely intervention to correct hemodynamic abnormalities and address atrial arrhythmias may favorably alter the clinical course in these patients.

Conversion from an atriopulmonary connection to a total cavopulmonary connection combined with arrhythmia operations has been achieved with low surgical mortality and favorable short-term survival and freedom from arrhythmia recurrence [6–8]. However, the effect of this operation on intermediate-term outcomes compared with other management options for patients with a failing atriopulmonary Fontan circulation, including transcatheter ablation of atrial arrhythmias, atrial pacing, or cardiac transplantation, remains to be clarified. We conducted this study to identify risk factors for heart transplant or death and describe the incidence of atrial arrhythmia recurrence in a large cohort of patients undergoing Fontan conversion with arrhythmia operations.

Patients and Methods

Study Design

This single-center, retrospective study was conducted at Ann & Robert H. Lurie Children's Hospital of Chicago (formerly Children's Memorial Hospital). Institutional Review Board approval was obtained, and written informed consent was obtained from patients before requesting outside medical records for study purposes.

Study Patients

We included the first 140 consecutive Fontan conversions with arrhythmia operations performed in 139 patients at our center between 1994 and 2012. Preoperative patient

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Table 1. Preoperative Characteristics of 139[a] Consecutive Fontan Conversion Patients

Variables	No. (%) or Median (range)
Male gender	79 (57)
Ventricular morphology	
Single left ventricle	119 (86)
Single right ventricle	16 (12)
Indeterminate	4 (3)
Age at initial Fontan repair, y	5.6 (1.1–34.9)
Age at Fontan conversion, y	23.2 (2.6–47.3)
Interval from initial Fontan to conversion, y	16.7 (1.2–33.6)
Type of initial Fontan repair	
Atriopulmonary Fontan	109 (78)
Atrioventricular Fontan (Bjork modification)	19 (14)
Lateral tunnel (intracardiac)	11 (8)
Age at onset of atrial arrhythmia, y	15.0 (1.4–44.8)
New York Heart Association classification	
I–II	42 (30)
III–IV	97 (70)
Ascites	26 (19)
Protein-losing enteropathy[b]	4 (4)
Hemodynamic data	
Right atrial pressure, mm Hg (n = 134)	14 (6–25)
Pulmonary artery pressure, mm Hg (n = 133)	13 (5–25)
Ventricular end-diastolic pressure, mm Hg (n = 138)	8 (0–17)
Ventricular end-diastolic pressure ≥13 mm Hg (n = 138)	17 (12)
Cardiac index, L/min/m^2 (n = 127)	2.5 (1.1–5.4)
Cardiac index <2.0 L/min/m^2 (n = 127)	39 (31)
Ventricular systolic dysfunction ≥moderate	20 (14)

[a] Data for the patient who underwent two Fontan conversion operations were only reported once, at the time of his second Fontan conversion. [b] Albumin data were available for 91 patients.

clinical characteristics and comorbidities are noted in Table 1. The most common indications for Fontan conversion were refractory atrial arrhythmias in 136 patients and exercise intolerance (New York Heart Association Functional Classification III to IV) in 97 patients. Protein-losing enteropathy was defined as a baseline serum albumin of less than 3.5 g/dL in association with ascites.

Surgical Procedure

The technical details of Fontan conversion to an extracardiac total cavopulmonary connection, the modifications necessary for anatomic variations, and the evolution in arrhythmia surgical techniques have been previously reported [8, 9]. Our current surgical arrhythmia protocol is that patients with documented right-sided arrhythmia receive a modified right atrial maze procedure, as do those with no clinical arrhythmia history and no inducible arrhythmias on a preoperative electrophysiology study. A biatrial maze procedure is performed in patients with left atrial macroreentrant tachycardia or atrial fibrillation and in patients with heterotaxy, regardless of arrhythmia presence or type. This protocol evolved over time, and some patients received a different approach during its evolution.

Follow-Up

Cross-sectional follow-up was obtained for all patients between January 1, 2013, and March 6, 2014. A search of the Social Security Death Master File was performed when patients whose current vital status was unknown after a review of available medical records, communication with patients, their families, and referring physicians.

The two primary outcomes were a composite end point of heart transplantation or cardiac death and the late recurrence of atrial tachycardia. Cardiac deaths included those known or suspected to be directly related to the Fontan circulation or its sequelae, including end-stage heart failure, documented terminal arrhythmia, sudden death, liver or renal failure, liver cancer, and stroke. Patients who died of noncardiac causes (eg, automobile accidents, cancer) were censored at the time of death in survival analysis.

Late atrial arrhythmia recurrence was defined as those that developed or continued to occur 3 months after the Fontan conversion off antiarrhythmic medications. Included were symptomatic presentations and those detected on ambulatory 24-hour monitoring or at least annual pacemaker device interrogation.

Statistical Evaluation

Data are reported as median (range) or count (percentage). Associations between potential risk factors and the outcomes of interest were explored using univariable and multivariable proportional hazard models. Kaplan-Meier curves were constructed for freedom from cardiac death or transplant, and freedom from arrhythmia recurrence was stratified by the type of arrhythmia operation. The log-rank test was used to test for significant differences in time to arrhythmia recurrence based on type of arrhythmia operation . All analyses were conducted using SAS 9.3 software (SAS Institute Inc, Cary, NC).

Results

Operative and Early Outcomes

The median age at the time of Fontan conversion operation was 23.2 years (range, 2.6 to 47.3 years), at a median of 16.7 years (range, 1.2 to 33.6 years) after the initial Fontan operation. Tachycardia developed at a median of 8.8 years (range, 0 to 24.6 years) after the initial Fontan operation, and patients underwent Fontan conversion at a median of 7.8 years (range, 0.4 to 21.0 years) after the onset of tachycardia. Preoperatively, 48 patients had right atrial tachycardia, 21 had left atrial tachycardia, 67 had atrial fibrillation, and 4 patients had no clinical or inducible arrhythmia. A lateral tunnel was created in 7 patients (5%), and an extracardiac total cavopulmonary connection was performed in 133 (95%).