

40-Year Follow-Up After the Fontan Operation

Long-Term Outcomes of 1,052 Patients



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ABSTRACT

BACKGROUND There are limited long-term, single-cohort, follow-up studies available about patients after the Fontan operation.

OBJECTIVES This study sought to determine the long-term outcome of all patients who had a Fontan operation at the Mayo Clinic.

METHODS Records of all patients who had a modified Fontan operation between 1973 and 2012 were reviewed. A follow-up questionnaire was mailed to all patients alive at the time of the study.

RESULTS Overall, 10-, 20-, and 30-year survival for 1,052 patients was 74%, 61%, and 43%, respectively. Factors associated with decreased overall or late survival in multivariate analysis included pre-operative diuretic use, longer cardiopulmonary bypass time, operation prior to 1991, atrioventricular valve (AVV) replacement at the time of Fontan operation, elevated post-bypass Fontan (>20 mm Hg) or left atrial (>13 mm Hg) pressures, prolonged chest tube drainage (>21 days), post-operative ventricular arrhythmias, renal insufficiency, and development of protein-losing enteropathy (PLE). Pre-operative and intraoperative sinus rhythm were associated with improved survival. Long-term survival was similar for patients regardless of ventricular morphology. The most common reoperations were pacemaker insertion/revision in 212 patients (20%), Fontan revision/conversion in 117 patients (11%), and AVV repair/replacement in 66 patients (5%). Clinically significant late atrial or ventricular arrhythmias occurred in 468 patients (44%). Ninety-five patients (9%) developed PLE, and 5-, 10-, and 20-year survival after diagnosis of PLE was 50%, 35%, and 19%, respectively.

CONCLUSIONS As the surgical techniques for the Fontan operation have changed over the last 40 years, survival has improved. However, development of PLE and arrhythmias and the need for reoperation during long-term follow-up pose significant management challenges. (J Am Coll Cardiol 2015;66:1700-10) © 2015 by the American College of Cardiology Foundation.

In 1971, Fontan and Baudet described a surgical technique for successful palliation of patients with tricuspid atresia (1,2). Subsequently, this technique has been applied to treat most forms of functional single ventricles (3-7). Theoretically, the Fontan operation separates the systemic and pulmonary venous returns to ameliorate the disadvantages of long-term hypoxemia, reduce thromboembolic events, preserve ventricular function, and prolong survival for patients with single-ventricle physiology. Although some of these beliefs have been fulfilled, a number of adverse results of the

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Fontan procedure have been recognized, including premature death, ventricular failure, thromboembolic disease, arrhythmia, liver disease, and protein-losing enteropathy (PLE) (8-14). In this study, we sought to determine long-term outcomes for all patients who had a Fontan operation at our institution.

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METHODS

In this institutional review board-approved, single-center, retrospective study, we reviewed the records of all patients (n = 1,052) who had their initial Fontan operation at the Mayo Clinic between October 1973 and June 2012 and who provided informed consent to participate in the study. Information regarding demographic, anatomic, pre-operative, operative, and post-operative variables, and Mayo follow-up was abstracted into a secure electronic database. Many of the patients had their follow-up care at other institutions. Any available correspondence regarding echocardiogram, cardiac catheterization, electrocardiogram, Holter/event monitor, laboratory tests, liver imaging, or surgical/procedural data was entered into the patient database.

A medical questionnaire was mailed to all patients not known to be dead at the initiation of the study. Nonresponders received second and third questionnaires, and if these were not returned or completed, an attempt was made to contact the patients by telephone. Quality-of-life surveys from patients rated their current health status on a scale of 1 to 4 (1 = excellent, 2 = good, 3 = fair, and 4 = poor). Data regarding death were updated using correspondence from physicians and patients/family members, chart review, and the Social Security Death Index. For the remaining patients, data curves were censored using the date of last available follow-up or date of transplant.

PLE was diagnosed based on documentation of enteric loss of alpha-1-antitrypsin or the presence of low serum total protein/albumin in addition to persistent or intermittent edema. Patients were excluded from subsequent PLE analysis if the timing of diagnosis was not known. Clinically significant arrhythmia was defined as the need for antiarrhythmic drug therapy (excluding digoxin), pacemaker placement, or electrical/pharmacological cardioversion. Patients with arrhythmias prior to the Fontan procedure were excluded from any analysis of post-operative arrhythmias. Cirrhosis was diagnosed based on liver biopsy/autopsy or characteristic findings on computed tomography, cardiac magnetic resonance imaging, magnetic resonance elastography, or ultrasound in

conjunction with clinical diagnosis by a gastroenterologist. Patients with isolated liver function or ultrasound abnormalities were not considered to have proven cirrhosis.

Variables used in the Cox regression analysis were initially analyzed as continuous variables, and then discrete cutoffs were selected based on the hazard ratios. The cutoffs for discrete variables used in univariate/multivariate analyses were defined as follows: pre-operative pulmonary artery pressure (PAP) (>17 mm Hg), pre-operative systemic ventricular end-diastolic pressure (>12 mm Hg), pre-operative pulmonary arteriolar resistance (>3 U × m²), post-bypass left atrial (LA) pressure (>13 mm Hg), post-bypass Fontan pressure (>20 mm Hg), and prolonged chest tube duration (≥21 days).

STATISTICAL ANALYSIS. Separate analyses were performed using either the date of the Fontan operation as time 0 (“overall survival”) or 30 days after the operation as time 0 (“late survival”). All deaths, regardless of cause, after the Fontan operation were considered in the survival analysis. Descriptive statistics for categorical variables were reported as frequency and percentage, and continuous variables were reported as mean ± SD or median (range) as appropriate. Time to PLE was compared between Fontan type groups using analysis of variance. Kaplan-Meier curves were derived to calculate 10-, 20-, and 30-year survival statistics. Cox regression models were used to determine univariate and multivariate predictors of survival and other long-term outcomes. The multivariable model considered significant univariate variables (p < 0.05) with model selection using the stepwise method. All statistical tests were 2-sided with the alpha level set at 0.05 for statistical significance. SAS version 9.3 (SAS Institute, Inc., Cary, North Carolina) was used for the analysis. The set of variables evaluated for association of survival and long-term outcomes and results of univariate Cox regression analyses are listed in [Online Tables 1 to 4](#).

RESULTS

Between 1973 and 2012, 1,052 patients had an initial Fontan operation at the Mayo Clinic ([Table 1](#)). At last follow-up, 426 patients (40%) were known to be dead. Of the 626 patients known to be alive, transplant-free survival was verified in 427 patients (68%) with follow-up information within 5 years of the study termination date. The mean age at initial Fontan procedure was 9.4 ± 7.5 years (median: 7 years; range: 7 months to 53 years). Mean follow-up after the Fontan operation was 15.3 ± 9.3 years

ABBREVIATIONS AND ACRONYMS

AVV = atrioventricular valve

LA = left atrial

PAP = pulmonary artery pressure

PLE = protein-losing enteropathy

SVEDP = systemic ventricular end-diastolic pressure

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