Trends in Fontan surgery and risk factors for early adverse outcomes after Fontan surgery: The Australia and New Zealand Fontan Registry experience

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Objectives: This study examined changes in practice and analyzed risk factors for adverse early outcomes after Fontan surgery through use of a binational, population-based registry.

Methods: Demographic, preoperative, and perioperative data were collected from all participating institutions of the Australia and New Zealand Fontan Registry. Patient and operative characteristics were analyzed with multivariable logistic regression for impact on early mortality, early Fontan failure (death, takedown, or mechanical support), effusions (prolonging hospital stay >30 days or requiring surgical reintervention), and stay longer than 30 days.

Results: Overall mortality was 3.5% (37/1071) and declined throughout the study period, from 8% (1975-1990) to 4% (1991-2000) and 1% (2001-2010). There were no differences between the extracardiac and lateral tunnel modifications for any outcome. After 2006, the extracardiac conduit was performed exclusively, with 1.3% mortality. The proportion of patients with hypoplastic left heart syndrome rose to 17% in the current era, and this group had more effusions (odds ratio, 3.0; 95% confidence interval, 1.4-6.6) and stayed on average 2 days longer in the hospital. Hypoplastic left heart syndrome was also an independent risk factor for composite adverse early outcome (death, failure, prolonged effusions, or prolonged stay >30 days; odds ratio, 2.6; 95% confidence interval 1.4-4.8 respectively).

Conclusions: The extracardiac conduit is now the exclusive Fontan modification performed in Australia and New Zealand. Even with a higher proportion of high-risk cases, perioperative outcomes are excellent in the modern era. Hypoplastic left heart syndrome confers a higher risk of prolonged pleural effusion and early composite adverse outcome. (J Thorac Cardiovasc Surg 2014;148:566-75)

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After Fontan and Baudet's initial description of the atriopulmonary connection,[1] subsequent modifications were introduced in 1988 by de Leval and colleagues[2] and in 1990 by Marcelletti and associates.[3] The atriopulmonary connection has now been abandoned because of the survival attrition of these patients and the progressive decline in functional capacity.[4-6] It has long been assumed that the lateral tunnel and the extracardiac conduit techniques would yield equivalent early outcomes, and that differences between these techniques would only become obvious in the analysis of long-term outcomes after Fontan surgery. In a recent report from the Society of Thoracic Surgeons Congenital Heart Surgery Database, patients undergoing an extracardiac conduit were found to have a higher risk of early reintervention and failure and were likely to stay in the hospital longer than those undergoing the lateral tunnel.[7] Along with the modification of techniques, the population offered Fontan surgery has evolved during the course of the last 2 decades. In particular, the improved survival after Norwood surgery has been responsible for an increase in the

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Abbreviations and Acronyms
CI = confidence interval
HLHS = hypoplastic left heart syndrome
IQR = interquartile range
OR = odds ratio

proportion of patients with hypoplastic left heart syndrome (HLHS) reaching Fontan surgery.[8]

In 2008, the Australia and New Zealand Fontan Registry was established to clarify long-term expectations of patients undergoing Fontan procedures and to enable recruitment of larger numbers of patients into prospective studies. During the creation and recruitment of patients into this registry, the preoperative and perioperative data of all patients undergoing Fontan procedures in Australia and New Zealand were collected. We thus sought to determine trends in indications and demographic characteristics of patients undergoing the Fontan procedure since its introduction in the region in 1975 and to analyze the impact of these trends on early outcomes, with a focus on the surgical techniques used. This analysis represents the entire experience of Fontan surgery in Australia and New Zealand.

MATERIALS AND METHODS

Institutional approval to collect data at all participating centers was obtained, and the need for consent was waived because of the retrospective nature of the study. All Fontan procedures performed in Australia and New Zealand were identified by consulting the cardiac surgical and cardiology databases at each institution (Appendix Table 1). Preoperative, perioperative, and early postoperative data collected are described in Appendix Table 2. Operative reports, echocardiographic reports, and discharge summaries were reviewed. Patients for whom insufficient data existed to satisfy the minimum data set (age at Fontan, Fontan type, and discharge status) were excluded. Fontan conversions were excluded from the data set, with the exception of 3 patients who had previously undergone a Björk procedure and subsequently had a Fontan completion.

Outcomes

The primary outcome was early mortality, defined as death within the first 30 postoperative days or before hospital discharge if the stay was longer than 30 days. Secondary outcomes were early failure (takedown, major reintervention not bleeding related, including Fontan revision, fenestration creation, enlargement or closure, mechanical circulatory support, or percutaneous reintervention), prolonged effusions (lasting >30 days or requiring reoperation for transcatheter intervention, pleurodesis, or Fontan revision), and a composite adverse early outcome (death, failure, prolonged effusions, or prolonged stay >30 days).

Statistical Analysis

Patient, operative and outcome data were described according to era and Fontan type (Tables 1 and 2), with normally distributed, nonnormally distributed, and proportional data described as mean ± SD, median with interquartile range (IQR), and number and percentage, respectively. Logistic regression analysis models were constructed for each outcome, and multivariable models were designed a priori to compare lateral tunnel versus extracardiac Fontan types and morphologically left versus right ventricles while adjusting for sex, age, and morphology as categoric variables. Remaining covariates were analyzed in univariable models to identify potential confounders and entered into the multivariable models if they were associated with a large effect size (odds ratio (OR) $\geq$2.5 or $\leq$0.4) or there was evidence against the null hypothesis of no association (2-tailed $P \leq .05$). Because of collinearity between Fontan type, era, staging with bidirectional cavopulmonary shunt, and fenestration, we included only Fontan type in multivariable models. Additionally, southern hemisphere respiratory syncytial virus season (the months of May to September) was analyzed for its effect on pleural effusions. The variables selected for multivariable models and their regression parameters are reported in Table 3.

Exposure groups in which no failures or deaths occurred could not be entered into logistic regression models. Numeric covariates were analyzed as continuous variables unless a nonlinear relationship could be demonstrated between covariate and outcome. Linearity was examined by performing a likelihood ratio test comparing regression models with continuous covariates against models in which the numeric variable had been categorically transformed. Age at Fontan operation had nonlinear associations with mortality (Figure 1) and the other end points and was analyzed as a categoric variable.

All analyses were performed with the Stata 11.2 statistical software package (StataCorp, College Station, Tex).

RESULTS

Description of Cohort

In all, 1095 Fontan procedures were identified, of which 1071 had sufficient data to be included for analysis.

A total of 1989 previous surgical palliations were performed in 990 patients. Previous staging with bidirectional cavopulmonary shunt occurred in 636 patients (59%): 4% who underwent the atriopulmonary technique, 16% who underwent lateral tunnel, and 96% who underwent extracardiac conduit. Mean pre-Fontan pulmonary arterial pressure was lowest among patients with extracardiac conduits (11.1 ± 2.7 mm Hg for extracardiac, 12.5 ± 4.5 mm Hg for lateral tunnel, and 13.1 ± 4.3 mm Hg for atriopulmonary; $P < .001$; $R^2 = 0.05$). The proportion with detected aortopulmonary collaterals increased with each era (3 patients [3%] before 1991, 47 patients [16%] from 1991 to 2000, and 115 patients [30%] after 2000), and 46 of 165 patients (28%) had these embolized before the Fontan procedure by transcatheter coiling. At 1 center, internal thoracic to pulmonary artery collaterals were routinely divided at operation if visualized.

Moderate or severe atrioventricular valve regurgitation was present before the Fontan procedure in a total of 65 patients (7.5%). Of the whole cohort, 40 patients (4.6%) had previously undergone atrioventricular valve repair or replacement; 18 (45%) were left with recurrent regurgitation by the time of the Fontan procedure, but re-repair was only attempted in 9 patients. Atrioventricular valve repair was undertaken concomitantly with the Fontan procedure in 28 patients (3%, 9 re-repairs and 19 de novo repairs).

Extracardiac conduit was the only modification performed after July 2006. Fenestration was performed in 50% of lateral tunnel and 44% of extracardiac connections.

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