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18 Years of the Fontan Operation at a Single Institution

Results From 771 Consecutive Patients

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Objectives The aim of this study was to evaluate Fontan peri-operative outcomes for 771 consecutive patients.

Background Since the initial description by Fontan, mortality associated with the Fontan operation has declined substantially. However, postoperative effusions remain a significant challenge. Effusions are a key determinant of postoperative length-of-stay and have been shown to be associated with the development of protein-losing enteropathy and with decreased survival.

Methods This study was a single-center, retrospective review of 771 patients who underwent Fontan palliation from 1992 to 2009.

Results Patients were divided into 3 eras dictated by shift in clinical practice. Overall mortality was 3.5%, 1% since 1996. Importantly, age at Stage II palliation decreased from Era 1 to Era 3 (7.1 vs. 5.9 months; $p = 0.0001$), whereas age at Fontan increased (1.7 vs. 2.8 years; $p = 0.0001$). The proportion of patients with prolonged hospital stay (46.7% vs. 8.2% vs. 19.5%, $p < 0.001$) decreased substantially after Era 1. A diagnosis of hypoplastic left heart syndrome and longer operative support times were associated with prolonged pleural drainage (odds ratio [OR]: 2.17, $p < 0.001$; OR: 1.2, $p = 0.001$) and hospital stay (OR: 1.48, $p = 0.05$; OR: 1.34, $p < 0.001$). In patients who underwent invasive assessment, higher pulmonary artery pressure was associated with death (OR: 1.37, $p < 0.001$) and prolonged hospital stay (OR: 1.09, $p = 0.019$). Pulmonary arterial pressure ≥15 mm Hg was 90% specific for discriminating unfavorable outcomes.

Conclusions Mortality in the modern era is rare, whereas postoperative pleural drainage remains the dominant morbidity. Elevated pulmonary artery pressure seems to be a marker of unfavorable outcome. Continued investigation is warranted to determine whether medical interventions or alterations to operative strategy can alter peri-operative results and improve long-term outcomes. (J Am Coll Cardiol 2012;60:1018–25) © 2012 by the American College of Cardiology Foundation

The concept of rerouting systemic venous return directly to the pulmonary arteries for the palliation of congenital heart disease was first described by Fontan and Kreutzer in the early 1970s as an alternative approach for management of tricuspid atresia (1,2). Since the original description, the evolution of the "Fontan" has included numerous surgical advances, along with parallel advances in anesthesia management, perfusion strategies, and intra-operative and post-operative care. In sum, these changes have led to substantial reductions in early mortality (3–6) such that several recent reports demonstrate mortality rates of 0% to 5.5% (7–11). However, despite continued improvements, the incidence of postoperative pleural effusions remains high and is a key determinant of postoperative length-of-stay (7,9,11). Prolonged peri-operative course has been associated with the late development of protein-losing enteropathy and with decreased long-term survival, suggesting that improvements in peri-operative course might have both short- and long-term benefits (9). Factors previously shown to influence peri-operative morbidity and mortality include: anatomy, atrioventricular valve regurgitation, ventricular function, pulmonary artery pressure (PAP), support times, modified ultrafiltration (MUF), and Fontan type (12–15).

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In this study we review the peri-operative outcome for 771 consecutive patients after the Fontan operation at The Children's Hospital of Philadelphia from January 1, 1992, to December 31, 2009. We identify trends in patient characteristics, describe changes in surgical technique, and evaluate the impact of these variables on early morbidity and mortality with a goal of identifying factors that could be modified to reduce early postoperative morbidity.

Methods

Data from all patients who underwent a first-time Fontan operation during the study period were included. Demographic, anatomic, and procedural variables were recorded from the medical record. In reporting procedural variables, total support time (TST) represents the time circulation was supported or arrested, equal to cardiopulmonary bypass (CPB) plus deep hypothermic circulatory arrest (DHCA). If multiple periods of CPB and DHCA were used, the times were summed. Duration of pleural drainage was defined as total days from surgery to removal of final chest tube or drain or final thoracentesis, performed during initial admission or during a readmission within 30 days of surgery. Hospital length-of-stay was defined as total days of initial hospital stay plus any additional days during readmission(s) within 30 days of surgery. The Institutional Review Board at The Children's Hospital of Philadelphia approved this study.

Surgical technique. The surgical strategies for both the superior cavo-pulmonary connection and the Fontan procedure were not standardized and have evolved considerably over the time period of this study. Because 8 different surgeons were involved in the care of these patients, multiple different techniques have been used with regard to the creation of pathway fenestrations, the use of patch material to augment the branch pulmonary arteries, and the timing and style of cardiopulmonary bypass. Management of cardiopulmonary bypass and myocardial protection were at the discretion of each surgeon, as were the specific surgical techniques used.

Statistical analysis. All demographic, anatomic, clinical, operative, and postoperative variables were summarized with standard descriptive statistics and expressed as mean ± SD for normally distributed continuous variables, median (with range) for skewed continuous variables, and count (with percentage) for categorical variables. Differences across eras, described in the following text, were assessed with analysis of variance, Kruskal-Wallis (nonparametric analog of analysis of variance), or Pearson chi-square. Duration of hospital stay and chest tube drainage were converted to dichotomous variables on the basis of standards from previous literature and from review of the distribution of our data. Associations between all potential covariates of interest and dichotomous outcome measures were assessed in univariate testing with logistic regression. Multivariate logistic regression was then used to identify factors independently associated with outcomes. Covariates with a

Abbreviations and Acronyms

AVV = atrioventricular valve
CPB = cardiopulmonary bypass
DHCA = deep hypothermic circulatory arrest
ECC = extracardiac conduit
HLHS = hypoplastic left heart syndrome
IALT = intra-atrial lateral tunnel
MUF = modified ultrafiltration
OR = odds ratio
PAP = pulmonary arterial pressure
TCPC = total cavopulmonary connection
TST = total support time

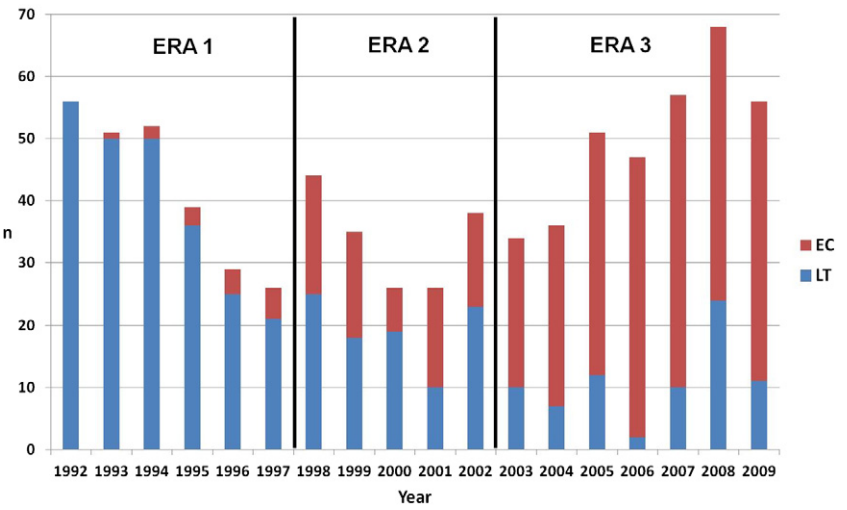

Figure 1 Number of Fontan Operations Performed/Year During Our Study Period

The **blue** represents intra-atrial lateral tunnel (LT) conduits, and the **red** represents extracardiac (EC) conduits.