

Variation in the use of Fontan fenestration in the current surgical era

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ABSTRACT

Fontan fenestration reduces chest tube duration and shortens post-operative length of stay but is not universally performed. Using a large national database, we sought to describe current use of the Fontan fenestration, including factors associated with placement.

The Virtual Pediatric System database was queried to identify patients with the Society of Thoracic Surgery code for a Fontan procedure during their admission from January 2009 until June 2016. Those undergoing a fenestrated Fontan were compared to those undergoing a non-fenestrated Fontan.

Of the 1695 patients, 1084 (64%) had a fenestration placed. There was variation between centers with the range of fenestration placement being 8 to 100%. Those with a fenestration placed had lower weight, despite similar age, and were more likely to have a single right ventricle. Those with a fenestration had lower systolic blood pressure and greater need for mechanical ventilation within the first hour of admission to the intensive care unit after Fontan.

Most patients still have a fenestration placed at the time of Fontan but this varies markedly by center. Fenestration placement is more common in higher risk patients including those with a systemic right ventricle. Further study is necessary to determine specific patient populations that benefit from fenestration placement at the time of Fontan.

1. Introduction

The Fontan procedure represents the final planned palliation for all forms of single ventricle heart disease. This operation completes the rerouting of systemic venous return directly to the pulmonary arteries, allowing the single ventricle to support systemic circulation exclusively. Since its first description in 1971 the Fontan operation has undergone numerous modifications. Fontan fenestration was first described in 1990 as a technique to improve post-operative outcomes for high risk patients with risk factors such as ventricular dysfunction, high pulmonary vascular resistance, and pulmonary artery distortion [1].

The fenestration reduces Fontan pressure, with the potential benefits being shorter duration of chest tube drainage and shorter hospital course [2]. Objections to fenestration include systemic desaturation and the risk of systemic embolization [3,4]. The paucity of comprehensive data in support of a single strategy likely contributes to the variable use of Fontan fenestration. Multicenter data from the Pediatric Heart

Network Fontan Cross-sectional Study showed that, of those undergoing Fontan between 1987 and 2002, 67% had a fenestrated Fontan performed. The percentage of patients getting a fenestration at the time of Fontan increased over time but varied significantly by center [5]. Using a large multicenter database, we sought to assess current use of the fenestrated Fontan and to evaluate factors that influence fenestration placement.

2. Methods

The Virtual Pediatric Systems (VPS) database was queried. This database includes data on consecutive intensive care admissions from 135 pediatric hospital systems in the United States. It includes standardized data definitions and data quality controls. This study protocol was evaluated by the institutional review board at the Children's Hospital of Wisconsin and determined to be exempt from review as data was de-identified. Waiver of consent was granted.

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Patients with a Society of Thoracic Surgery (STS) code for any type of Fontan procedure performed between January 2009 and June 2016 were identified. Only centers that input an STS primary diagnosis for the cardiac disease were included and subjects undergoing Fontan revision were excluded. Baseline characteristics, including age, weight, gender, race, and primary cardiac diagnosis, which was subsequently used to determine ventricular morphology (left, right or undetermined), as well as risk scores, were evaluated. The Pediatric Index of Mortality (PIM2) is a risk score used to predict mortality of patients admitted to an intensive care unit (ICU), and is calculated based on values for systolic blood pressure, pupillary response, PaO₂, base excess, mechanical ventilation, pre-admission surgical procedure, pre-admission cardiopulmonary bypass, and presence of a high or low risk diagnosis. The individual components of the PIM 2 score were evaluated rather than the calculated score to remove the PaO₂ component as this would be impacted by fenestration shunting [6]. For non-required variables (race, chest tube replacement and duration, and renal support), only centers that committed to collecting that variable for all cases were included in that particular analysis.

Categorical comparisons were made between those with and without a fenestration placed at the time of Fontan using Chi squared test or Fisher's exact test where indicated. Continuous variables were compared using the Wilcoxon rank sum test. Multivariable analysis using logistic regression with backwards elimination was used to determine factors associated with Fontan fenestration. Statistical analysis was performed using Stata 13 (Stata Corp, College Station TX) with $p < 0.05$ considered significant. Data are presented as number with percent of total, mean \pm standard deviation, or median with interquartile range (IQR).

3. Results

The cohort included 1744 patients from 41 centers. Baseline characteristics for the entire cohort can be seen in Table 1. Of the 1695 with data related to fenestration placement, 1084 (64%) had a fenestration placed at the time of Fontan. Fenestration placement varied by center with the frequency of use ranging from 8 to 100%, using only centers with ≥ 10 Fontan procedures in the database (Fig. 1). Comparison of patient factors by fenestrated versus non-fenestrated Fontan can be seen in Table 2. The fenestrated Fontan group had significantly lower weight, despite a similar age, was more likely to have a single right ventricle, had lower systolic blood pressure on admission to the ICU after Fontan and more frequently needed mechanical ventilation during the first hour of intensive care admission.

Table 1
Cohort characteristics ($n = 1744$).

Male, n	1029 (59%)
Race/Ethnicity, n	
White/Caucasian	680 (51%)
Black/African American	151 (11%)
Hispanic	314 (24%)
Other	188 (14%)
Cardiac diagnosis, n	
HLHS	646 (39%)
DORV	152 (9%)
DILV	172 (11%)
Tricuspid atresia	236 (14%)
AVSD	143 (9%)
PA/IVS or pulmonary stenosis	113 (7%)
AVSD + DORV	32 (2%)
Ebstein's anomaly	27 (2%)
Other single ventricle	116 (7%)
Median (IQR) age at Fontan, months	49.2 (36.3–54.2)
Median (IQR) weight at Fontan, kg	14.5 (13–16.5)

AVSD atrioventricular septal defect, DILV double inlet left ventricle, DORV double outlet left ventricle, HLHS hypoplastic left heart syndrome, IQR interquartile range, PA/IVS pulmonary atresia intact ventricular septum.

By diagnosis, there was variation in the frequency of Fontan fenestration with 68.7% of those with hypoplastic left heart syndrome having a fenestration, 63.3% with double outlet right ventricle, 65.4% with tricuspid atresia, 58.1% with double inlet left ventricle, and 55.5% with pulmonary atresia with an intact ventricular septum. In multi-variable analysis, significant factors associated with fenestration placement at the time of Fontan included center performing the surgery ($p < 0.001$), weight at the time of surgery ($p = 0.014$), and ventricular morphology ($p = 0.028$). Factors included in the model but not significantly associated with fenestration placement were presence of total anomalous pulmonary venous return ($p = 0.31$), age ($p = 0.67$), and gender ($p = 0.87$).

ICU mortality following Fontan surgery was 0.9% (16/1744). Median ICU length of stay was 3.8 days (IQR 1.9–6.8). From the ICU, 226 patients (13%) were discharged home, 1474 (84.5%) transferred to a lower level of care unit, and 25 (1.4%) transferred to another ICU, including at a different hospital. For the cohort, the median chest tube duration for chest tubes placed at the time of surgery was 68.6 h (IQR 44.3–117.5), with 159 (11.9%) requiring additional chest tube placement or chest tube replacement.

4. Discussion

In this national sample, Fontan fenestration is performed in the majority of cases. In particular, a fenestration is used in higher risk patients, as demonstrated by the higher frequency of a systemic right ventricle and lower weight despite similar age. However, most importantly, these data demonstrate that there is significant variation between centers in the use of Fontan fenestration. Some centers are always or routinely placing a fenestration while others rarely use a fenestration, likely in only the highest risk patients.

In this cohort from 41 centers in the United States, 64% of patients had a fenestration placed at the time of Fontan from 2009 to 2016. This percentage is remarkably similar to previous studies that have reported data on the frequency of Fontan fenestration during earlier surgical eras. Multicenter data from the Pediatric Heart Network Fontan Cross-sectional Study, which included patients who had their Fontan procedure completed between 1987 and 2002, showed that 67% of the cohort had a fenestrated Fontan performed, with marked variation of 13% to 91% among centers [5]. More recent data from the Society of Thoracic Surgery Database from 2000 to 2009 showed that fenestration was performed in 65% of Fontan operations [7]. Our study is unique in that we evaluated factors associated with Fontan fenestration in a multi-institution cohort. Other than center, the factors we found to be significant were ventricular morphology and weight which may be of statistical but limited clinical significance. The marked variation in use of fenestration among individual centers, as well as lack of change in fenestration frequency over multiple decades, suggests there is lack of consensus on the post-operative benefits of fenestration placement at time of Fontan operation for either standard or high risk patients.

After first publishing the initial description of Fontan fenestration, Bridges et al. performed a retrospective comparison of fenestrated Fontan patients with an historical control population of non-fenestrated Fontan patients and showed that despite being higher risk, patients with a fenestration had a significantly shorter duration of chest tube drainage and shorter hospital length of stay [8]. Other single center retrospective studies also identified fenestration as a protective factor for prolonged pleural drainage and hospital stay [9]. These post-operative benefits of decreased chest tube drainage and length of stay were confirmed in a prospective, single center randomized trial. This study by Lemler et al., representing the strongest data to date, included 49 standard risk patients undergoing the Fontan operation between 1997 and 2000. They demonstrated a reduction of 55% in chest tube output, 41% shorter hospital stay, and 67% fewer additional procedures in those randomized to the fenestration group [2].

Despite the results from these single-center studies, Fontan

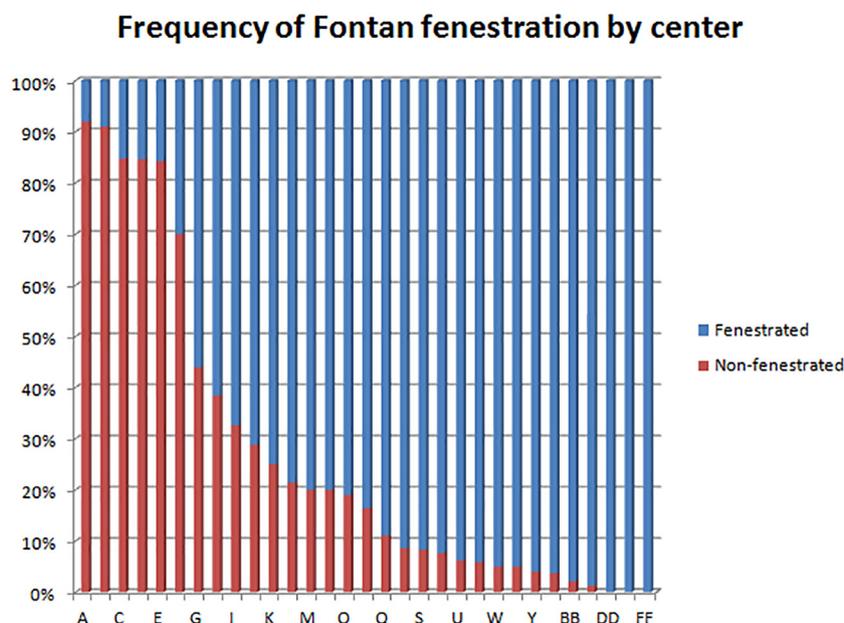


Fig. 1. Frequency of fenestrated versus non-fenestrated Fontan procedures at the 31 centers with ≥ 10 Fontan procedures in the VPS database.

Table 2

Comparison of risk factors between fenestrated and non-fenestrated Fontan patients ($n = 1695$).

	Non-fenestrated	Fenestrated	<i>p</i> value
Male	359 (59%)	643 (59%)	0.84
Ventricular morphology			0.04
Left	218 (38%)	327 (32%)	
Right	276 (48%)	553 (54%)	
Undetermined	77 (14%)	137 (14%)	
Heterotaxy syndrome	77 (13%)	128 (12%)	0.74
TAPVR	15 (2%)	38 (4%)	0.25
Weight (\pm SD), kg	16.0 (\pm 7.3)	15.3 (\pm 5.4)	0.003
Age (\pm SD), months	49.9 (\pm 29.7)	48.4 (\pm 25.0)	0.32
Systolic blood pressure on ICU admission (\pm SD), mm Hg	90.8 (\pm 18.4)	85.7 (\pm 17.6)	< 0.0001
Base deficit on ICU admission (\pm SD)	-2.0 (\pm 2.7)	-2.2 (\pm 3.4)	0.17
Ventilation in first hour after ICU admission	269 (44%)	561 (52%)	0.002

TAPVR total anomalous pulmonary venous return.

fenestration has not gained widespread acceptance, with several authors concluding it is unnecessary in standard risk patients and some even stating it is not necessary in those at increased risk [3,4,10,11]. Primary arguments against fenestration have been the systemic desaturation, risk of systemic embolization and need for a further procedure to close the fenestration. Schreiber et al. reported on 84 consecutive patients who underwent Fontan without fenestration, including 16 that were deemed high risk due to the presence of 2 or more risk factors preoperatively (\geq moderately impaired function, \geq moderate atrioventricular valve regurgitation, mean pulmonary artery pressure > 15 mm Hg, mean atrial pressure > 12 mm Hg, pulmonary artery distortion, or non-sinus rhythm). They did not find a significant difference between the high risk patients compared to those deemed lower risk in duration of mechanical ventilation, pulmonary artery pressure post-extubation, or duration of effusions, leading them to conclude that “routine fenestration is dispensable” for most patients. It can also be argued that a non-fenestrated Fontan can be performed without cross clamping or fibrillation, allowing for normothermic bypass with a beating heart which decreases oxidative stress and reduce myocardial perfusion injury [12,13].

In our study, we did not compare outcomes of fenestrated versus non-fenestrated Fontan's as our analysis is limited to only the available data collected in the VPN database. There are important clinical variables, such as pre-operative risk factors, surgical technique, or perfusion strategy, which were not included in our analysis that likely influenced post-operative outcomes and may have confounded the impact of fenestration itself. Beyond the post-operative period, the longer term benefits and risks of a persistent fenestration on clinical outcomes have not been determined. The Pediatric Heart Network study reported that those with a patent fenestration at medium term follow up (mean of 8 years after Fontan) had no increased risk of post-Fontan stroke, protein losing enteropathy or arrhythmia compared with patients with a closed fenestration. Additionally, there was no difference in measured exercise capacity or functional assessment as measured by the Child Health Questionnaire CHQ PF50 [5]. Our institution demonstrated no difference in event-free survival between those with or without a fenestration at most recent follow-up. However, this study readily admitted a selection bias in those that were felt to be candidates for fenestration closure [14]. Another retrospective non-randomized single-center study reported that persistent fenestration was associated with increased mortality and Fontan complications compared to closed fenestration at a median follow-up of 5 years [15]. A randomized, controlled follow up study, with adequate power, is needed to more accurately determine the longer term benefits and risk of fenestration after discharge.

4.1. Study Limitations

This study is limited by the data available in the VPS database which does not include all risk factors used to determine whether a fenestration is placed. Most notably, pre-operative echocardiographic and catheterization data is not included and likely has a significant impact on the decision to fenestrate. The database also does not include information on bypass strategy or lowest temperature during bypass to allow for assessment of the impact of these variables. The VPS database is also designed for the ICU stay without data on subsequent hospitalization time or re-admission. For these reasons, this study is not intended as a definitive evaluation of Fontan fenestration but rather an assessment of current use and variation in practice.

5. Conclusions

The rate of Fontan fenestration use remains stable at 64% with marked variation among centers. Fenestration use was more common in patients with higher pre-operative risk. The variation between centers in the rate of fenestration demonstrates the lack of data supporting best practice. Only through acquisition of further data will the short- and long-term benefits and risks of Fontan fenestration be identified.

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Conflicts of Interest

The authors have no conflicts of interest to report.

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