

Recent outcomes of the extracardiac Fontan procedure in patients with hypoplastic left heart syndrome

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ABSTRACT

- Objective** : To investigate patient-related factors, echocardiographic, and anatomic variables associated with immediate and long-term clinical outcomes after extracardiac Fontan procedure at our institution.
- Materials and Methods** : Retrospective review of preoperative cardiac catheterizations and echocardiograms as well as medical records of all children with hypoplastic left heart syndrome (HLHS) who underwent Fontan between June 2002 and December 2018.
- Results** : Seventy-seven patients with HLHS were included (age 4 years [1.5–11.7]). Seventy patients (91%) received a nonfenestrated Fontan and 57 patients (74%) underwent Fontan without cardiopulmonary bypass (CPB). Presence of a Fontan fenestration ($P=0.69$) and use of CPB ($P=0.79$) did not differ between those with <2 weeks compared to those with ≥ 2 weeks of chest tube drainage. There were no differences in either pre- or intra-operative hemodynamics between patients who weighed <15 kg compared to those who weighed ≥ 15 kg at time of surgery; incidence of death, transplant, and transplant listing were similar between weight groups. Inferior vena cava (IVC) diameter z-score did not differ among patients with and without chylous chest tube drainage ($P=0.78$), with and without development of protein losing enteropathy ($P=0.23$), or death/heart transplant/transplant listing compared to survivors without transplant ($P=0.26$).
- Conclusion** : In HLHS patients undergoing Fontan, preoperative weight and IVC diameter appeared to have no influence on immediate postoperative outcomes. Performing the Fontan off CPB and with a fenestration also conferred no added clinical benefit. These observations should be considered when deciding optimal timing for Fontan completion.
- Keywords** : Fontan procedure, hypoplastic left heart syndrome, clinical outcomes, congenital heart disease

INTRODUCTION

Although the total cavopulmonary connection (or Fontan procedure) forms the accepted final palliation for the

single-ventricle heart, there remains controversy in the timing of the Fontan.^[1] While some institutions complete

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the Fontan circulation by the time children are 3 years old to minimize end-organ exposure to cyanosis, many other programs delay Fontan until the patient develops exertional cyanosis and/or achieves a certain weight threshold, typically completing the Fontan after 4 years of age.^[2-4]

In the current era, short-term survival after Fontan for hypoplastic left heart syndrome (HLHS) is excellent at >95%; however, the Fontan circulation results in several physiological consequences related to inefficient flow dynamics, chronic exposure to elevated central venous pressures, and lymphatic congestion.^[5-8] Moreover, the right ventricle acting as a systemic pump in patients with HLHS remains a risk factor associated with long-term mortality and heart failure after Fontan; recent data suggest that only two-thirds of patients with HLHS operated in the best tertiary care centers may reach adulthood.^[9,10]

Previous studies have investigated patient-related factors and hemodynamic determinants of clinical outcomes after Fontan;^[2,11-14] however, limited research exist on preoperative echocardiographic and anatomic variables such as inferior vena cava (IVC) diameter and the association with postoperative clinical outcomes.^[11,12] Our general surgical preference has been to wait until a weight of at least 15 kg or 3–4 years of age is achieved prior to elective Fontan completion, with the belief that patients of this size can accommodate an 18 or 20 mm extracardiac Gore-Tex conduit without difficulty related to the surgical anastomoses. Whether preoperative IVC diameter had associations with conduit size selected, performing the procedure off cardiopulmonary bypass (CPB), and/or poor postoperative hemodynamics remained unknown. This study investigated whether specific patient-related factors, echocardiographic, and anatomic variables were associated with worse immediate and long-term clinical outcomes after extracardiac Fontan completion at our institution, including protein losing enteropathy, heart transplant, or death.

MATERIALS AND METHODS

Patient population

The Stanford Children's Heart Center database was queried retrospectively to identify all children with HLHS with a preoperative cardiac catheterization and echocardiogram who underwent Fontan completion at our institution between June 2002 and December 2018. Patients with other subtypes of single-ventricle congenital heart disease including tricuspid atresia, double-inlet left ventricle, and double-outlet right ventricle with atrioventricular valve atresia or stenosis were excluded.

Data collected

Each patient's diagnosis, gender, date of birth, height, weight, body surface area (calculated using the DuBois

formula)^[15] at the time of surgery, dates of preoperative cardiac catheterization, dates of surgical procedures, types of surgical procedure including type and size of Fontan and whether the procedure was performed without CPB, and intraoperative hemodynamic data were obtained from the electronic medical record. Charts were also reviewed for postoperative clinical data including duration of chest tube drainage, presence of chylous effusions, extracorporeal membrane oxygenation (ECMO) support, length of hospital stay, date of last follow-up visit, presence of protein losing enteropathy, heart transplant listing/transplantation, and death.

The cardiac catheterization performed immediately prior to Fontan procedure was reviewed, and data collected included mean pulmonary artery pressure (mmHg), wedge pressure (mmHg), and Qp/Qs. Any interventions for residual arch obstruction and/or pulmonary artery stenosis were noted.

Echocardiographic studies were routinely performed in all patients as part of their preoperative evaluations. Images were acquired according to American Society of Echocardiography (ASE) guidelines and stored in our institution's secure server.^[16] The ultrasound equipment used for the echocardiographic studies was either the Siemens Sequoia C512 (Siemens Medical Solutions USA, Inc., Mountain View, CA, USA), Philips iE33 or Philips Epiq 7 (Philips Medical Systems, Bothell, WA, USA). The transthoracic echocardiogram performed immediately prior to Fontan was reviewed, and data collected included measures of right ventricular systolic function including fractional area change (FAC, %) and tricuspid annular plane systolic excursion (TAPSE) standardized to z-score,^[17,18] as well as degree of tricuspid regurgitation (graded by ASE standards),^[19] hepatic vein a-wave velocity (m/s), neo-aortic valve velocity time integral (VTI, m), IVC diameter (cm) measured at its largest from a subcostal sagittal plane just below the diaphragm [Figure 1a], and abdominal aorta diameter (cm) also from a subcostal sagittal plane [Figure 1b]. Z-scores for IVC diameter were recorded using published standardized normal IVC diameters for pediatric patients.^[20] The primary investigator made all offline measurements using the syngoDynamics workstation (Siemens Medical Solutions USA, Inc.; syngoDynamics Solutions, Ann Arbor, MI, USA).

A second investigator performed measurements of IVC diameter by the same method on a randomly selected subset of twenty patients (via random number generator on Microsoft Excel©) to determine interobserver variability. The second reader was blinded to the initial analysis, the two measurements were separated by at least 30 days, and the reader was free to independently choose frames for analysis within the designated study date.

Statistical analysis

All continuous data were presented as mean ± standard deviation. Parametric testing was used to compare data with normal distributions, such as age, body surface area, echocardiographic indices, and hemodynamic data. All unpaired comparisons were performed with the Student’s *t*-test. Nonparametric testing was used to compare data without normal distributions, such as duration of chest tube drainage and length of hospital stay. A Fisher’s exact test was used to compare presence of protein losing enteropathy or chylous effusions when stratified by patient-related factors.

Intraclass correlation analysis was used to compare IVC measurements between two investigators. *P* <0.05 was considered statistically significant. All statistical calculations were performed using IBM SPSS Statistics version 24 (IBM Corp., Armonk, NY, USA). The study protocol was approved by the Stanford University Institutional Review Board (Protocol #28556) and was performed in accordance with the Helsinki Declaration of 1975, as revised in 2000.

Surgical technique

Our surgical preference is to avoid the use of CPB when performing the Fontan procedure. The first consideration of whether the Fontan can be performed off CPB involves the proximal branch pulmonary artery and extracardiac conduit anastomosis. There must be adequate length along the branch pulmonary arteries to allow for conduit anastomosis while flow from the existing superior cavopulmonary connection can supply one lung. Typically, this involves diverting right superior vena cava flow into the right pulmonary artery, while the left pulmonary artery and hilum are clamped. The patient must be able to tolerate single-lung ventilation for a period of time. Passive lower body venous drainage is initiated by cannulation of the common atrium and IVC just above the entry of the hepatic veins. The atrium is divided from the IVC and carefully oversewn. The Gore-Tex tube graft is tailored to the length of the IVC and the distal end is then anastomosed to the diaphragmatic aspect of the IVC; the ability to perform



Figure 1: (a) Inferior vena cava (IVC) diameter measured at its largest in long axis from a subcostal sagittal plane just below the diaphragm (white line). (b) Abdominal aorta diameter (white line) measured at its largest in long axis from a subcostal sagittal plane. HV: Hepatic vein; RA: Right atrium

this anastomosis is the second consideration of whether the Fontan can be performed off CPB. There must not be too severe of a degree of apicocaval juxtaposition; if the heart mass cannot be effectively rotated while dissecting and suturing near the IVC, significant hemodynamic alterations can occur and necessitate CPB.

Conduit size is chosen by the surgeon performing the procedure. Our preference is to select a 20 mm extracardiac conduit when possible, but will use a 16 or 18 mm conduit when the 20 mm conduit appears too large for the IVC aperture.

Postoperative management

Our institutional goals are to extubate Fontan patients within the first postoperative day, transfer out of the cardiovascular intensive care unit before postoperative day 4, and ideally discharge home on postoperative day 10 [Figure 2].^[21] In our single-ventricle patients after Glenn and Fontan procedures, we maintain diuretics at a minimum of every 8 h, supplemental oxygen of 1 liter nasal cannula FiO₂ 100% while chest tubes are in place (regardless of oxygen saturation), and remove chest tubes when there is drainage of < 6 cc/kg/tube over any 24 h period with no radiographic evidence of significant pleural effusions.^[22,23] Patients are discharged home on oral diuretics often every 8 h, with a follow-up chest X-ray with their primary cardiologist 2–3 days postdischarge.

For those with chylous effusions defined as pleural triglyceride levels >100 mg/dL or cell counts >1000 cells/mm³ with predominance (>80%) of lymphocytes, intake is restricted to 80% maintenance volume of low-fat diet (and/or formula) in addition to diuretics given at a minimum of every 8 h. Chest tubes are removed if the pleural fluid transitions to serous, chest X-ray demonstrates no significant pleural effusion, and output is <4 cc/kg/tube over a 24 h period. Transitioning

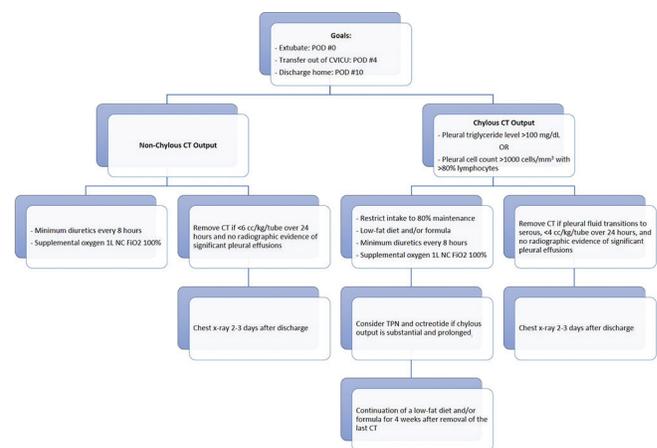


Figure 2: General postoperative management of chest tubes, diuretics and diet after Fontan at our institution.^[21–23] CT: Chest tube; L: Liter; NC: Nasal cannula; POD: Postoperative day; TPN: Total parenteral nutrition

to total parenteral nutrition and octreotide use is considered if chylous output is substantial and prolonged. We advise continuation of a low-fat diet (and/or formula) for 4 weeks after removal of the last chest tube.

RESULTS

Between June 2002 and December 2018, 77 patients with HLHS underwent Fontan completion at our institution. The median age of Fontan completion was 4 years (1.5–11.7), with 70 patients (91%) who received an extracardiac nonfenestrated Fontan and 57 patients (74%) underwent Fontan without CPB. Most patients underwent placement of an 18 mm extracardiac Fontan (56%). Subtypes of HLHS are included in Table 1. Males comprised 65% of the cohort, and 60% of the cohort was < 15 kg at the time of surgery.

On preoperative cardiac catheterization, mean pulmonary artery pressure was 10 ± 2 mmHg, wedge pressure 7 ± 2 mmHg, and Qp/Qs 0.6 ± 0.1 . Intraoperative

hemodynamics revealed a mean pulmonary artery pressure of 13 ± 2 mmHg and an atrial filling pressure of 6 ± 2 mmHg with postoperative transpulmonary gradient of 7 ± 1 mmHg. Twenty-six patients required coiling of veno-venous collaterals at the preoperative cardiac catheterization; no patients required balloon dilation for residual arch obstruction or pulmonary artery stenosis.

The average preoperative right ventricular FAC was $41.3\% \pm 6.9\%$, but TAPSE z-score of -6 ± 2 . Longitudinal IVC diameter z-score was 1.2 ± 1.0 .

Eighteen patients (23%) developed chylous chest tube output immediately postoperatively; the median duration of chest tube drainage was 13 days (4–112) and the median length of hospital stay was 16 days (7–112). Thirty-five patients (45%) had ≥ 2 weeks of chest tube drainage. There were two in-hospital deaths after Fontan; one patient exhibited progressive cardiac failure due to respiratory failure/acute respiratory distress syndrome from acute rhinovirus infection. The second patient had significant postoperative chest bleeding requiring mediastinal exploration and evacuation of an intrapericardial hematoma on the evening of surgery; the patient returned to the intensive care unit with elevated Fontan pressures (~ 20 mmHg) despite the presence of a fenestration, developed hepatic and renal failure, fluid overload and was cannulated onto veno-arterial ECMO but could not be weaned off support. In our study, one patient at 11 years old underwent extracardiac nonfenestrated Fontan; the patient remained in the hospital for 24 days after Fontan with 18 days of nonchylous chest tube output. Up until a recent 5-year follow-up, the patient had not yet developed protein losing enteropathy or required a heart transplant.

The average follow-up period for the cohort was 1.8 ± 2.6 years (range 0–9 years); six patients (8%) eventually developed protein losing enteropathy, one patient (1%) received a heart transplant in the follow-up period, while three patients (4%) were listed for heart transplant, and two additional patients died at 3 and 4 years after Fontan [Figure 3].

Preoperative inferior vena cava diameter Z-score compared to clinical outcome

Preoperative longitudinal IVC diameter z-score measured larger in the group requiring CPB at time of Fontan compared to the group who did not require CPB ($P = 0.01$) [Table 2]. IVC diameter z-score did not differ between patients who received either a 16 or 18 mm extracardiac conduit compared to those who received a 20-mm conduit ($P = 0.54$).

Longitudinal IVC diameter z-score measured similar in patients with ≥ 2 and < 2 weeks of chest tube drainage ($P = 0.96$) and ≥ 2 and < 2 weeks length of hospital stay ($P = 0.98$). IVC diameter z-score did not

Table 1: Patient characteristics

Characteristics	Patients (n=77)
Age (years)	4 (1.5-11.7)
Male (%)	50 (65)
Patients <15 kg (%)	46 (60)
Subtype of HLHS	
MA/AA (%)	42 (55)
MS/AA (%)	13 (17)
MS/AS (%)	22 (28)
Type of fontan	
Extracardiac nonfenestrated (%)	70 (91)
Extracardiac fenestrated (%)	7 (9)
Size of Fontan conduit	
16 mm extracardiac conduit	3 (4)
18 mm extracardiac conduit	45 (56)
20 mm extracardiac conduit	29 (36)
Echocardiographic measurements	
RV fractional area change (%)	41 ± 6.9
TAPSE (cm)	0.9 ± 0.3
TAPSE z-score	-6.3 ± 2
IVC longitudinal diameter (cm)	1.0 ± 0.2
IVC longitudinal diameter Z-score	1.2 ± 1
Abdominal aorta longitudinal diameter (cm)	0.9 ± 1
Preoperative hemodynamics	
PA pressure (mmHg)	10 ± 2
Wedge pressure (mmHg)	7 ± 2
Intraoperative hemodynamics	
PA pressure (mmHg)	13 ± 2
Atrial filling pressure (mmHg)	6 ± 2
Transpulmonary gradient (mmHg)	7 ± 1
Postoperative characteristics	
Chylous chest tube output (%)	18 (23)
Protein losing enteropathy (%)	6 (8)
Transplant listing (%)	3 (4)
Transplanted (%)	1 (1)
Death (%)	4 (5)
Duration of chest tube drainage (days)	13 (4-112)
Duration of hospital stay (days)	16 (7-112)

Data presented as mean \pm SD or median (range). AA: Aortic atresia; HLHS: Hypoplastic left heart syndrome; IVC: Inferior vena cava; MA: Mitral atresia; MS: Mitral stenosis; PA: Pulmonary artery; RV: Right ventricle; TAPSE: Tricuspid annular plane systolic excursion, SD: Standard deviation

differ among patients with and without chylous chest tube drainage ($P=0.78$), with and without eventual development of protein losing enteropathy ($P=0.23$), or death/heart transplant/listing for transplant compared to survivors without transplant ($P=0.26$). IVC diameter z-score also did not differ between patients who weighed <15 kg and ≥ 15 kg at the time of surgery ($P=0.86$). While smaller IVC diameters were associated with higher preoperative filling pressures (≥ 10 mmHg) ($P=0.04$), smaller IVC diameters were also associated with lower intraoperative Fontan pressures (<15 mmHg) ($P=0.04$).

Reliability of IVC diameter measurement was excellent between 2 readers, with an intraclass correlation coefficient of 0.92 (95% confidence interval: 0.80–0.97).

Fontan conduit size compared to clinical outcome

Size of the conduit had no implication on duration of chest tube drainage ($P=0.87$), length of hospital stay ($P=0.56$), presence of a chylous effusion ($P=0.86$), eventual development of protein losing enteropathy ($P=0.17$), or death/heart transplant/listing for transplant compared to survivors without transplant ($P=0.71$).

Patient weight compared to clinical variable

There were no differences in conduit size selection between patients who weighed <15 kg compared to

those who weighed ≥ 15 kg ($P=0.06$) [Table 3]. There were also no differences in duration of chest tube drainage ($P=0.24$), length of hospital stay ($P=0.43$), or pre- and intra-operative hemodynamics by patient weight. The number of patients requiring CPB to perform the Fontan was similar between patients <15 kg and ≥ 15 kg ($P=0.80$); incidence of death, transplant or transplant listing was also similar between groups ($P=1.0$). However, all the six patients who eventually developed protein losing enteropathy were <15 kg at the time of surgery.

Duration of chest tube drainage after Fontan

Age at time of Fontan procedure, weight, and BSA had no bearing on the duration of chest tube drainage [Table 4]. Preoperative echocardiographic measures of RV size and function, as well as degree of tricuspid regurgitation did not differ between patients with <2 weeks and ≥ 2 weeks of chest tube drainage. None of the pre- or intra-operative hemodynamic data differed between groups.

Three of 42 (7%) patients with a Fontan fenestration had chest tube drainage lasting <2 weeks compared to 4 of 35 (11%) with ≥ 2 weeks of chest tube drainage ($P=0.69$). Chylous effusions were present in 16 of 70 (23%) nonfenestrated Fontan patients and 2 of 7 (29%) fenestrated Fontan patients ($P=0.66$). All seven patients in our cohort underwent fenestration at the time of Fontan for elevated intraoperative Fontan pressures and transpulmonary gradients.

Performance of the Fontan on CPB

There was no significant difference in the duration of chest tube drainage between those who did and did not require CPB. While the median length of hospital stay appeared longer in patients who underwent Fontan with CPB at 19 days (7–67) compared to without CPB at 15 days (7–112), the difference was not statistically significant ($P=0.49$). Of the twenty patients who required CPB at the time of surgery, 4 (20%) died or required transplant/listing for transplant compared to 3 of 57 (5%) who did not require CPB ($P=0.07$).

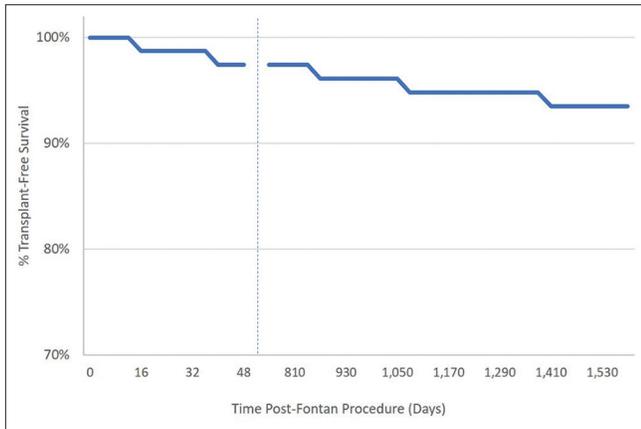


Figure 3: Kaplan–Meier curve displaying transplantation-free survival in our cohort after Fontan procedure

Table 2: Inferior vena cava diameter Z-score compared to clinical outcome

Operative variable	Mean±SD	Operative variable	Mean±SD	P
Chest tube drainage ≥ 2 weeks	1.2±0.9	Chest tube drainage <2 weeks	1.2±1	0.96
Hospital stay ≥ 2 weeks	1.1±0.9	Hospital stay <2 weeks	1.1±1	0.98
Chylous chest tube drainage	1.1±1	Nonchylous chest tube drainage	1.2±0.9	0.78
Protein losing enteropathy	0.65±0.9	No protein losing enteropathy	1.2±1	0.23
Weight <15 kg	1.2±0.8	Weight ≥ 15 kg	1.1±1.1	0.86
RV FAC <40%	1.1±1	RV FAC $\geq 40\%$	1.2±0.9	0.46
CPB required	1.7±0.7	CPB not required	1.0±1.0	0.01
16 or 18 mm extracardiac conduit	1.1±0.9	20 mm extracardiac conduit	1.3±1.0	0.54
Preoperative filling pressure ≥ 10 mmHg	0.6±1.2	Preoperative filling pressure <10 mmHg	1.3±0.9	0.04
Intraoperative fontan pressure ≥ 15 mmHg	1.9±0.7	Intraoperative fontan pressure <15 mmHg	1.0±0.9	0.04
Death, transplant or listing	1.5±0.7	Survived, no transplant	1.1±1	0.26

Bolded P value represents statistical significance. CPB: Cardiopulmonary bypass, FAC: Fractional area change, RV: Right ventricle, SD: Standard deviation

DISCUSSION

Much debate remains over optimal timing of Fontan completion.^[1] As mentioned earlier, our general surgical preference has been to wait until a weight of at least 15 kg or 3–4 years of age is achieved prior to elective Fontan completion, unless the patient presents with increasing cyanosis. However, as our data demonstrated, no obvious immediate clinical benefit was conferred by waiting until an arbitrary weight of 15 kg. One interesting finding in our study, however, was that all six patients who eventually developed protein losing enteropathy were <15 kg at the time of surgery despite having similar pre- and intra-operative systemic venous pressures as those who weighed >15 kg. While

the incidence of protein losing enteropathy in our cohort (8%) was not unusual based on previous studies, this observation likely warrants further prospective investigation given the profound morbidity associated with the condition.^[24,25]

From an anatomic perspective, smaller IVC diameter appeared to have no clear association with smaller conduit size selection, use of CPB, worse intraoperative hemodynamics or clinical outcomes. While smaller IVC diameters were associated with higher preoperative filling pressures ≥ 10 mmHg, smaller IVC diameters conflictly were also associated with better intraoperative Fontan pressures. A larger sample size may resolve these findings.

Our surgical preference has also been to perform the Fontan without CPB when possible, as Fontan completion off CPB has been demonstrated to attenuate pro-inflammatory markers and theoretically shorten length of hospital stay, and has been associated with less risk of early and late rhythm disturbance.^[14,26,27] Our decision to perform the Fontan off CPB takes into account a number of surgical considerations, including whether the patient's anatomic substrate will allow us to visualize the lay of the new conduit without hemodynamic compromise, and ability to divert pulmonary blood flow to a single lung while the other pulmonary artery is clamped to perform the proximal anastomosis. However, this study as well as others did not establish any difference in hemodynamics or postoperative clinical improvements when performing the procedure off CPB.^[28,29] While our study specifically investigated the use of CPB and its relationship to duration of chest tube drainage and length of hospital stay, Navabi *et al.* additionally evaluated duration of mechanical ventilation, incidence of arrhythmias, and postoperative hemodynamic instability and found no meaningful differences between those who did and did not require CPB.^[29]

Table 3: Patient weight compared to clinical outcome

	Weight <15 kg (n=46)	Weight \geq 15 kg (n=31)	P
Duration of chest tube drainage (days)	12 (4-66)	17 (5-112)	0.24
Length of hospital stay (days)	15 (7-67)	19 (8-112)	0.43
Chylous chest tube drainage (# patients)	11	7	1.0
Protein losing enteropathy (# patients)	6	0	0.07
Preoperative filling pressure (mmHg)	7 \pm 2	8 \pm 2	0.07
Preoperative PA pressure (mmHg)	10 \pm 2	10 \pm 3	0.10
Intraoperative filling pressure (mmHg)	5 \pm 2	6 \pm 2	0.67
Intraoperative Fontan pressure (mmHg)	13 \pm 2	13 \pm 2	0.77
20 mm extracardiac conduit (# patients)	13	16	0.06
CPB required (# patients)	11	9	0.80
Death, transplant or listing (# patients)	4	3	1.0

Data presented as mean \pm SD or median (range). CPB: Cardiopulmonary bypass; PA: Pulmonary artery, SD: Standard deviation

Table 4: Duration of chest tube drainage after Fontan

Demographic variable	Chest tube drainage <2 weeks (n=42)	Chest tube drainage \geq 2 weeks (n=35)	P
Age (years)	4.1 \pm 1.2	4.4 \pm 2	0.48
BSA (m ²)	0.66 \pm 0.2	0.65 \pm 0.1	0.78
Weight (kg)	18.2 \pm 22.9	15.3 \pm 3.8	0.45
RV area diastole (cm ²)	13.5 \pm 3.4	13.8 \pm 3.4	0.71
RV area systole (cm ²)	8.0 \pm 2.4	8.2 \pm 2.3	0.70
RV FAC (%)	41.2 \pm 7.0	40.9 \pm 6.6	0.84
Neo-aortic valve VTI (m)	0.19 \pm 0.06	0.19 \pm 0.04	0.97
Preoperative TR >mild (# patients)	6	3	0.50
Preoperative PA pressure (mmHg)	10 \pm 2	10 \pm 3	0.53
Preoperative wedge pressure (mmHg)	7 \pm 2	7 \pm 2	0.40
Intraoperative filling pressure (mmHg)	5 \pm 2	6 \pm 1	0.54
Intraoperative Fontan pressure (mmHg)	13 \pm 2	13 \pm 2	0.70
Intra-operative TPG (mmHg)	7 \pm 2	7 \pm 2	0.99
Fontan fenestration (# patients)	3	4	0.69
CPB required (# patients)	10	10	0.79

Data presented as mean \pm SD or median (range). CPB: Cardiopulmonary bypass, BSA: Body surface area, FAC: Fractional area change, PA: Pulmonary artery, RV: right ventricle, TPG: Transpulmonary gradient, TR: Tricuspid regurgitation, VTI: Velocity time integral, SD: Standard deviation

Similarly, the Fontan fenestration appeared to confer no advantage in preventing development of chylous effusion, meaningfully shortening duration of chest tube drainage or length of hospital stay in our institutional experience with HLHS patients. This finding is counter to a recent study by Kim *et al.* that found the presence of a fenestration to be associated with pleural effusions lasting <14 days.^[30] However, Kim's study included all subtypes of single ventricle patients who underwent Fontan, and in fact, found HLHS to be an independent risk factor for prolonged pleural effusion. As we investigated only HLHS patients in this study, it is possible that the effect of a fenestration is less evident within this population.

Age is also often used as a threshold for completing Fontan.^[1] In our study, one patient at 11 years-old underwent extracardiac nonfenestrated Fontan, and up until a recent 5-year follow-up, the patient had not yet developed protein losing enteropathy or required heart transplant. Thus, as we have known, age alone cannot be the sole factor in deciding optimal timing of Fontan.

Given the rare incidence of HLHS, our patient numbers remained limited. However, the benefit of restricting the cohort to only HLHS patients was the ability to exclude other subtypes of single ventricle disease (e.g. heterotaxy syndrome with right-dominant atrioventricular canal) as the etiology for the differences found in short and long-term outcomes. The retrospective nature of the study also limited our follow-up time for patients who recently underwent Fontan. Despite the relatively short follow-up period, a number of patients already developed protein losing enteropathy and were either listed for or underwent heart transplant. Moving forward, prospective studies will better elucidate risk factors for Fontan failure.

CONCLUSIONS

In HLHS patients undergoing Fontan completion, preoperative age, weight, and IVC diameter appeared to have no influence on immediate postoperative outcomes. Performing the Fontan off CPB and with a fenestration also conferred no added clinical benefit. These observations should be considered when deciding optimal timing for Fontan completion. Further prospective data may determine whether these findings remain true in all patients with single-ventricle congenital heart disease undergoing Fontan.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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