Published in final edited form as:

Semin Thorac Cardiovasc Surg. 2020 ; 32(3): 522–528. doi:10.1053/j.semtcvs.2020.02.018.

Identification of Risk Factors for Early Fontan Failure

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Abstract

Objective: Despite improvements in operative and perioperative care, the risk of significant morbidity and mortality for children undergoing Fontan procedures persists. Previous investigations have identified peri-Fontan characteristics that may predict early adverse events. The purpose of this study was to identify characteristics from throughout a patient's lifespan, including all perioperative stages, that might predict early Fontan failure — defined as death, Fontan takedown, or listing for cardiac transplantation before hospital discharge or within 30 postoperative days.

Methods: A single-center retrospective study of all patients undergoing a Fontan procedure was performed. Patient and intervention-related characteristics were examined from birth through Fontan. Data were described using standard summary statistics. Univariable, logistic regression was used to examine associations with early Fontan failure.

Results: In total, 191 patients met inclusion criteria. The incidence of early Fontan failure was 4% (n = 8: six deaths, two Fontan takedowns). Neonatal balloon atrial septostomy was the only patient characteristic significantly associated with Fontan failure. Patients who underwent balloon septostomy had 8.5-times higher odds of Fontan failure (CI 2.6 – 28.1, p < 0.001) than those who did not.

Conclusions: Children who require balloon septostomy as neonates remain at higher risk of Fontan takedown, listing for heart transplantation, or death in the early post-Fontan period.

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Conflict of Interest statement: None of the authors has any conflicts of interest to disclose.

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Graphical abstract

Incidence of operative death or Fontan takedown for patients with and without a history of neonatal balloon atrial septostomy.

Keywords

Congenital heart surgery; Fontan; Balloon atrial septostomy; congenital heart disease

Introduction

Early postoperative outcomes after Fontan have improved in the modern era. Yet the incidence of early Fontan failure (death, Fontan takedown, or listing for heart transplantation before discharge) in North America is currently still estimated to be approximately 3% [1].

In 1977, Drs. Choussat and Fontan published the “Ten Commandments” describing the ideal candidate for the Fontan procedure [2]. Over the last 40 years, cardiologists and surgeons have continued to examine and refine these criteria. Commonly considered risk factors for morbidity and mortality include heterotaxy [1,3], extracardiac Fontan [1,5], non-fenestrated Fontan [1, 4], elevated preoperative ventricular end diastolic pressure [1,8], elevated pulmonary arterial pressure on pre-Fontan catheterization [4,6,7,8], common atrioventricular (AV) canal [6,7], ventricular systolic dysfunction [8,10], and atrioventricular valve regurgitation [9,10].

Prior studies have described peri-Fontan patient characteristics as predictors of post-Fontan outcomes. There are fewer data on the predictive power of patient characteristics from earlier in a patient’s life, such as perioperative data from Stage 1 and Stage 2 palliations, for children who undergo Fontan palliation. The purpose of this analysis was to identify patient- and intervention-related characteristics from throughout a patient’s medical and surgical history that might predict early Fontan failure.

Patients and Methods

Patients

We performed a single-center retrospective chart review of all patients who underwent the Fontan operation at New York-Presbyterian/Morgan Stanley Children’s Hospital, Columbia University Irving Medical Center, January 1, 2006 to December 31, 2015, regardless of their

underlying anatomy. We excluded patients who had undergone Stage 1 or Stage 2 palliation at other institutions. One patient who had undergone a hybrid procedure at Stage 1 was excluded from analysis in order to improve the homogeneity of the study population.

Perioperative Patient Care

As per institutional clinical protocol, all children completing Stage 2 palliation and surviving to discharge were assumed eligible for either Fontan completion or biventricular conversion. The timing of Fontan surgeries was determined at the discretion of the pediatric cardiologist and operating surgeon, with the majority of referrals occurring between 2.5 and 4.0 years of age. For children with Trisomy 21, under shared decision making between parents and providers, timing of Fontan completion has historically been delayed at our institution, due to a perceived higher risk of Fontan completion in these patients. All children underwent routine transthoracic echocardiogram and diagnostic catheterization before Stage 2 and Fontan procedures. Interventional catheterizations were performed at the discretion of the medical team. Children with concerns for elevated pulmonary vascular resistance were often referred to our pulmonary hypertension team for medical optimization during both the pre- and perioperative periods.

For Stage 1, all neonates were cared for preoperatively in our neonatal cardiac intensive care unit (ICU). Decisions regarding interventions, such as balloon atrial septostomy (BAS) and surgical timing, were made on an individual basis at the discretion of the cardiology and interventionalist teams. Stage 1 palliation was ideally targeted between the first three to five days of life, pending scheduling and family preferences. There were no set criteria for BAS at our institution during the study period, though it was reserved for patients with echocardiographic evidence of intact or highly restrictive atrial septae, who, in the opinion of the cardiologists and surgeon, were unstably hypoxemic and could not wait for a scheduled operation date.

For Stage 2 palliation, patients at our institution underwent unilateral or bilateral bidirectional Glenn or Kawashima. Hemi-Fontans were not routinely performed at our institution. For Stage 3, the use of lateral tunnel or extracardiac Fontans was determined by patient anatomy and surgeon discretion. One of five surgeons rarely fenestrated; the other four preferred to fenestrate the majority of Fontans.

Standard cardiopulmonary bypass cannulation and perfusion strategies were used for all cases. Chests were routinely closed postoperatively following all three stages. All children were cared for postoperatively in our neonatal (Stage 1) or pediatric cardiac ICU (Stage 2 and 3) by dedicated cardiac nurses, cardiologists, and cardiac intensivists. All decisions regarding postoperative management, including timing of extubation and selection of inotropes and other medications, were made jointly by pediatric cardiologists, intensivists, and surgeons.

Patient and Operative Characteristics

We obtained data regarding patient characteristics and interventions by review of electronic medical records. Demographic data included age, sex, and dates of admissions, discharges, and procedures. Anatomic data included dominant ventricle, primary lesion/anatomic

subtype, and the presence of heterotaxy. Genetic syndromes were assessed jointly as a binary variable.

We assessed the following Stage 1-related characteristics: age at surgery, neonatal BAS, type of initial palliation (Norwood, isolated shunt placement, pulmonary artery banding, or none), and type of shunt placed (when applicable).

We assessed the following Stage 2-related characteristics: age, preoperative moderate to severe AV valve regurgitation or ventricular dysfunction on echocardiogram, preoperative mean pulmonary artery pressure (mPAP), transpulmonary gradient, and ventricular end diastolic pressure on diagnostic catheterization, preoperative interventional catheterization, type of Stage 2 repair (unilateral or bilateral bidirectional Glenn or Kawashima), concurrent cardiac procedures (such as pulmonary arterioplasty or AV valvuloplasty), duration of postoperative intubation and chest tubes, the presence of chylothorax, ICU length of stay, and hospital length of stay.

We assessed similar pre-Fontan catheterization and echocardiographic characteristics. We also assessed the effects of age at Fontan, Fontan type (extracardiac or lateral tunnel), the creation of Fontan fenestrations, concurrent cardiac procedures, and Fontan surgeon.

Outcomes

The primary outcome of interest was early Fontan failure, defined as death, Fontan takedown, or listing for heart transplantation prior to hospital discharge or within 30 postoperative days, if discharged prior to 30 days, in keeping with prior definitions used in analyses of the Society of Thoracic Surgeons-Congenital Heart Surgery Database [1]. All post-Fontan deaths or takedowns were reviewed and classified as 'high,' 'moderate,' or 'low' likelihood of attributability to failing Fontan physiology. 'High likelihood' was assigned in patients with persistent hypotension refractory to maximum pressor support and persistent hypoxemia refractory to maximum ventilatory support, without other explanation, and documented in the medical chart as presumed Fontan failure. For patients with prolonged hospital courses, 'high likelihood' was assigned in patients with persistent hypoxemia and two or more other sequelae typically associated with Fontan failure, such as abdominal ascites, persistent chylous effusions, or plastic bronchitis. 'Moderate likelihood' was assigned in patients with persistent hypotension refractory to maximum pressor support and/or persistent hypoxemia refractory to maximum ventilatory support, with possible alternate etiologies or without explicit documentation in the medical chart as presumed Fontan failure. 'Low likelihood' was assigned in all other patients.

Statistics

Data were described using standard summary statistics and univariable, logistic regression, clustering standard errors by surgeon to account for inter-surgeon practice variation. Variables with p-values ≤ 0.05 on univariable analyses were considered significant. Analyses were conducted using Stata software, version 13 (StataCorp LP, College Station, TX).

Results

Baseline Characteristics

In total, 191 patients met inclusion criteria. The most common underlying anatomy was hypoplastic left heart syndrome (HLHS) (42%, n = 81), followed by tricuspid atresia (16%, n = 31) and double-outlet right ventricle (14%, n = 27) (See Table 1 and Figure 1).

Stage 1 Perioperative Characteristics

Eight percent of patients (n = 15) underwent BAS shortly after birth. Of these, 73% (n = 11) had HLHS; two had pulmonary atresia with intact ventricular septae, one had tricuspid atresia, and one had double outlet right ventricle. All patients who underwent BAS had echocardiographic evidence of highly restrictive or intact atrial septae, as well as persistent hypoxemia. Two of the 15 patients required radiofrequency perforation of the atrial septum during the catheterization. A standalone surgical atrial septectomy was not performed in this cohort. Most patients underwent either a Norwood procedure (56%, n = 107) or isolated shunt placement (29%, n = 55) (Table 1).

Stage 2 Perioperative Characteristics

Pre-Stage 2 echocardiogram demonstrated moderate or severe AV valve regurgitation in 12% of patients (n = 22) and moderate or severe ventricular systolic dysfunction in 5% (n = 9). On pre-Stage 2 diagnostic catheterization, the median mPAP was 13 mmHg (IQR 11 – 15) and the median transpulmonary gradient was 6 mmHg (IQR 5 – 8).

One quarter of patients (n = 48) underwent cardiac intervention during pre-Stage 2 catheterization; the most common interventions were coiling of collateral vessels (n = 16) or balloon dilation of recurrent aortic coarctation (n = 13).

The vast majority of patients (98%, n = 187) underwent unilateral or bilateral bidirectional Glenn procedures for Stage 2 palliation; the remaining four patients underwent a Kawashima. Common concurrent procedures included pulmonary arterioplasty (54%, n = 104) and AV valvuloplasty (5%, n = 10). Median duration of intubation was 1 day (IQR 0–2), and duration of chest tubes was 4 days (IQR 3 – 5). Median postoperative ICU length of stay after Stage 2 was 3 days (IQR 2 – 5), with a median hospital length of stay of 6 days (IQR 5 – 9) (Table 1).

Fontan Perioperative Characteristics

Pre-Fontan echocardiogram demonstrated moderate or severe AV valve regurgitation in 12% of patients (n = 23) and moderate or severe ventricular systolic dysfunction in 5% (n = 10). Pre-Fontan AV valve regurgitation and pre-Fontan ventricular dysfunction were highly associated with pre-Stage 2 AV valve regurgitation ($p < 0.001$) and pre-Stage 2 ventricular dysfunction ($p = 0.007$). AV valve regurgitation persisted in 8 of 10 patients who underwent AV valvuloplasties at the time of Stage 2 palliation. Ventricular dysfunction persisted in 3 of 9 patients who had moderate to severe dysfunction at the time of Stage 2 palliation.

On pre-Fontan diagnostic catheterization, median mPAP was 11 mmHg (IQR 10 – 13) and median transpulmonary gradient was 4 mmHg (IQR 3 – 5).

Fifty-three percent (n = 101) of patients underwent cardiac intervention during pre-Fontan catheterization; the most common interventions included coil occlusion of collaterals (47%, n = 89), ballooning or stenting of branch pulmonary arteries (14%, n = 27), and ballooning of the Glenn anastomosis (9%, n = 17).

Of the 191 Fontan operations performed, 56% (n = 107) were extracardiac conduits and the remaining 44% (n = 84) were lateral tunnel procedures. Of note, the extracardiac Fontan was the preferred procedure at our institution in the latter half of the period studied, 2011–2015 (80 extracardiac, 2 lateral tunnel; $p < 0.001$). The Fontan was fenestrated in 48% of patients (n = 92). There was significant variation in the use of fenestrations and the type of Fontan by surgeon ($p < 0.01$ & $p < 0.001$). One surgeon fenestrated only 16% of cases; the others fenestrated 60–100% of cases. A fenestration was performed in 75% of lateral tunnel Fontans and 27% of extracardiac Fontans ($p < 0.001$). Concurrent procedures included pulmonary arterioplasty (17%, n = 33), shunt takedown (6%, n = 12), and AV valvuloplasty (4%, n = 8) (Table 1).

Outcomes

There were six operative deaths and two Fontan takedowns (4%). Four deaths and the two Fontan takedowns were classified as “highly likely” related to Fontan failure. Three deaths and the two takedowns occurred in patients with intractable hypoxemia and hypotension throughout the first postoperative week. One death occurred after a prolonged hospitalization, with refractory hypoxemia, chylous effusions, and plastic bronchitis. The other two deaths were classified as “moderately likely” related to Fontan failure. One occurred in a patient with postoperative hypotension who ultimately suffered significant Fontan thrombi and associated stroke. The other patient died of hypotension and hypoxemia that was not explicitly described as presumed Fontan failure. No patients were listed for cardiac transplantation before discharge. The earliest transplant listing occurred 13 months after Fontan.

The only patient-or intervention-related characteristic associated with early Fontan failure was a history of a neonatal BAS (Table 1). The odds of Fontan failure for those who underwent a balloon septostomy was 8.5 times the odds of Fontan failure for those who did not undergo balloon septostomy (CI 2.6 – 28.1, $p < 0.001$) (Figure 2 and Table 2). No other patient characteristic was significantly associated with Fontan failure. All deaths and Fontan takedowns in BAS patients were assessed as highly likely related to Fontan failure.

A history of neonatal BAS was not significantly associated with pre-Stage 2 or pre-Fontan hemodynamics at catheterization, including pre-Stage 2 mPAP (14 mmHg vs. 13 mmHg, $p = 0.233$), transpulmonary gradient (7 mmHg vs. 6 mmHg, $p = 0.603$), and ventricular end diastolic pressure (10 mmHg vs. 9 mmHg, $p = 0.112$), or pre-Fontan mPAP (14 mmHg vs. 11 mmHg, $p = 0.052$), transpulmonary gradient (6 mmHg vs. 4 mmHg, $p = 0.206$), or ventricular end diastolic pressure (10 mmHg vs. 9 mmHg, $p = 0.156$).

Comment

This study examines patient and management characteristics from across the lifespan as potential predictors of early Fontan failure (death, Fontan takedown, or listing for cardiac transplantation prior to discharge or within 30 postoperative days). This work attempts to address a clinical question consistently faced by cardiologists, surgeons, cardiac intensivists, and families as they prepare children for Fontan palliation: Is this child a good candidate and what is his or her risk of acute post-operative decompensation?

The incidence of early Fontan failure for children in our cohort was low overall (4%), similar to previous reports in the current surgical era, which have generally ranged from 2–6% [1, 3, 5]. Children who underwent a neonatal balloon atrial septostomy had an 8.5-times higher odds of early Fontan failure, though our data rely on a small number of events. No other patient or management characteristic was significantly associated with this outcome.

It is well-documented that children with single ventricles and restrictive or intact atrial septae requiring balloon atrial septostomy have poorer early outcomes after stage 1 palliation compared to those with unrestrictive septae. A prior report from our institution found that infants with hypoplastic left heart syndrome undergoing Stage 1 palliation who underwent preoperative balloon atrial septostomies had 14% higher probability of death prior to hospital discharge [11]. Other institutions have shown similar results, reporting up to 40% differences in the probability of neonatal death [12]. The deleterious effects of a restrictive atrial septum are concerning enough that some centers perform fetal atrial septoplasty or stenting in an attempt to halt the progression the pulmonary vascular disease as early as possible [13]. While this may improve neonatal stability, it is not yet known if fetal atrial septoplasty and/or stenting leads to sustained benefits and improved post-Fontan outcomes.

Our data suggest that a history of neonatal balloon atrial septostomy may remain relevant to a child's prognosis even among those who survive to Fontan. While our data were not able to elucidate the mechanisms behind this association, it may be due to long-term damage to the pulmonary vasculature caused by left atrial hypertension in fetal and neonatal life, which has been demonstrated on histopathology in patients with HLHS and an intact atrial septum [14]. Our study helps to quantify this risk at the time of Fontan and may assist in longitudinal counseling of families, though larger studies are needed to validate this association.

Limitations

This study is limited by its retrospective nature. Thus, while this study raises many mechanistic questions, we are unable to draw causal inferences. Perhaps the most important limitation is that it only includes patients who were deemed candidates for and reached the Fontan procedure. There exists a cohort of patients who died or underwent heart transplantation before reaching the Fontan, or who remained with Glenn physiology. A competing risk analysis of a cohort that included patients who died prior to Fontan would help assess the effects of potential selection bias. Notably, the presence of genetic syndromes, AV valve regurgitation, high transpulmonary gradient, and ventricular

dysfunction were not significantly associated early Fontan failure in our study. This may be due to censoring, medical or surgical optimization prior to Fontan, or simply low power. This study is limited by the small number of Fontan failures we observed under our selected definition. We used stringent criteria for Fontan failure, including hard end-points, so as not to confound our results by institutional or practitioner-related factors, such as length-of-stay in the ICU or duration of chest tube placement. This reduction in study power may explain why other patient characteristics often observed to be associated with Fontan failure did not meet statistical significance in our study. Finally, some other possible explanations for Fontan intolerance might include a technically inadequate repair or variations in postoperative management; unfortunately, we were unable to differentiate between these possible contributing factors to Fontan failure. Further investigations with larger numbers of Fontan failures and balloon septostomies are needed to validate these findings.

Conclusion

Children with a single ventricle physiology who have undergone balloon atrial septostomy as neonates may be at higher risk for early Fontan failure than their peers. Clinicians should incorporate this when counseling families, as this may help to set postoperative expectations. Further studies are needed to confirm these findings across institutions and to determine optimal pre-and post-operative management strategies that might mitigate these risks.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

Acknowledgments

Funding: Dr. Anderson received support for research from the National Center for Advancing Translational Sciences of the NIH (KL2 TR001874) and the National Heart Lung and Blood Institute of the NIH (K23 HL133454).

Glossary of Abbreviations:

AV	Atrioventricular
ICU	Intensive care unit
BAS	Balloon atrial septostomy
mPAP	mean pulmonary artery pressure

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Central message:

Single ventricle patients with a history of balloon atrial septostomy may be at increased risk for early Fontan failure. Such patients require particular vigilance in the immediate post-Fontan period.

Perspective statement:

Prior studies have described peri-Fontan patient characteristics as predictors of post-Fontan outcomes; there are few reports on data from earlier in children's lives. We found that a history of neonatal balloon atrial septostomy was associated with early Fontan failure. This work may help cardiologists and surgeons in setting appropriate expectations for families long before their surgical dates.

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Figure 1:
Anatomic subtypes of patients undergoing Fontan procedure.

Figure 2 (also Central Picture):

Incidence of operative death or Fontan takedown for patients with and without a history of neonatal balloon atrial septostomy. Patients who had undergone a balloon septostomy in the neonatal period had a 20% incidence of operative death or Fontan takedown, compared to just 3% in patients with no history of balloon septostomy.

Central picture:

Early Fontan failure for patients with and without a history of balloon atrial septostomy

Table 1:

Univariable analysis of patient characteristics with and without early Fontan failure

Patient Characteristics	Overall (n = 191)	No Early Fontan Failure (n = 183)	Early Fontan Failure (n = 8)	p-value
Demographics & Anatomy				
Sex (male)	116 (61%)	110 (60%)	6 (75%)	0.484
Genetic syndrome	15 (8%)	14 (8%)	1 (13%)	0.487
Heterotaxy	14 (7%)	12 (7%)	2 (25%)	0.108
Dominant left ventricle	68 (35%)	64 (35%)	4 (50%)	0.458
Primary cardiac lesion				0.849
Hypoplastic left heart syndrome	81 (42%)	78 (43%)	3 (38%)	
Tricuspid atresia	31 (16%)	29 (16%)	2 (25%)	
Double outlet right ventricle	27 (14%)	25 (14%)	2 (25%)	
Double inlet left ventricle	19 (10%)	19 (10%)	0 (0%)	
Pulmonary atresia	16 (8%)	15 (8%)	1 (13%)	
AV canal defect	10 (5%)	10 (5%)	0 (0%)	
L-Transposition of the great arteries	5 (3%)	5 (3%)	0 (0%)	
Truncus arteriosus	2 (1%)	2 (1%)	0 (0%)	
Stage 1 Characteristics				
Age at Stage 1 (days)	6 (4 – 8)	6 (4 – 8)	10 (5 – 21)	0.158
Neonatal balloon atrial septostomy	15 (8%)	12 (7%)	3 (38%)	< 0.001 *
Type of Stage 1 Norwood				0.273
Norwood	107 (59%)	104 (57%)	3 (38%)	
Isolated systemic-pulmonary shunt	55 (30%)	50 (27%)	5 (63%)	
PA banding	16 (9%)	16 (9%)	0 (0%)	
No stage 1 performed	13 (7%)	13 (7%)	0 (0%)	
Stage 2 Characteristics				
Age at Stage 2 (months)	6 (5 – 9)	6 (5 – 9)	6 (5 – 9)	0.643
<i>Preoperative evaluation</i>				
Pre-Stage 2 valve regurgitation (moderate)	22 (12%)	22 (12%)	0 (0%)	0.600
Pre-Stage 2 systolic dysfunction (moderate)	9 (5%)	8 (4%)	1 (13%)	0.325
Pre-Stage 2 mean PA pressure by catheterization (mmHg)	13 (11 – 15)	13 (11 – 15)	14 (8 – 17)	0.798
Pre-Stage 2 transpulmonary gradient by catheterization (mmHg)	6 (5 – 8)	6 (5 – 8)	6 (4 – 10)	0.840

Patient Characteristics	Overall (n = 191)	No Early Fontan Failure (n = 183)	Early Fontan Failure (n = 8)	p-value
Catheter-based intervention between Stage 1 and Stage 2	48 (25%)	44 (24%)	4 (50%)	0.111
Type of Stage 2				0.417
Unilateral bidirectional Glenn	161 (84%)	158 (85%)	3 (60%)	
Bilateral bidirectional Glenn	26 (14%)	24 (13%)	2 (25%)	
Kawashima	4 (2%)	4 (2%)	0 (0%)	
Concurrent procedures				1.000
Pulmonary arterioplasty	104 (54%)	101 (55%)	3 (38%)	
AV valvuloplasty	10 (5%)	10 (6%)	0 (0%)	
<i>Postoperative course after Stage 2</i>				
Duration of intubation (days)	1 (0 – 2)	1 (0 – 2)	2 (0 – 5)	0.323
Duration of chest tubes (days)	4 (3 – 5)	4 (3 – 5)	6 (3 – 7)	0.146
Chylothorax	29 (15%)	27 (15%)	2 (25%)	0.354
ICU length of stay (days)	3 (2 – 5)	3 (2 – 5)	5 (2 – 12)	0.222
Fontan Characteristics				
Age at Fontan (years)	3 (3 – 4)	3 (3 – 4)	4 (3 – 4)	0.227
<i>Preoperative evaluation</i>				
Pre-Fontan AV valve regurgitation, moderate	23 (12%)	23 (13%)	0 (0%)	0.600
Pre-Fontan dysfunction, moderate	10 (5%)	10 (5%)	0 (0%)	1.000
Pre-Fontan mean PA pressure by catheterization (mmHg)	11 (10 – 13)	11 (10 – 13)	11 (7 – 14)	0.658
Pre-Fontan transpulmonary gradient by catheterization (mmHg)	4 (3 – 5)	4 (3 – 5)	5 (3–5)	0.694
Catheter-based intervention between Stage 2 and Fontan	101 (53%)	95 (52%)	6 (75%)	0.285
Extracardiac Fontan	107 (56%)	100 (55%)	7 (88%)	0.080
Fenestrated Fontan	92 (48%)	87 (48%)	5 (63%)	0.485
Concurrent procedures				0.704
Pulmonary arterioplasty	33 (17%)	32 (17%)	1 (20%)	
AV valvuloplasty	8 (4%)	8 (4%)	0 (0%)	

Numbers represent medians (IQR) for continuous variables and counts (%) for categorical variables. AV = atrioventricular; PA = Pulmonary artery; ICU = Intensive care unit; BT = Blalock-Taussig.

* indicates p<0.05.

Table 2:

Characteristics of patients with early Fontan failure:

Anatomy	Neonatal balloon atrial septostomy	Pre-Fontan ventricular systolic function	Pre-Fontan AV valve regurgitation	Fontan type	Fenestration	Outcome	Cause of decompensation	Likelihood that failure due to poor Fontan physiology
Tricuspid atresia type IB (VSD, pulmonic stenosis, normally-related great arteries)	Yes	Normal	Trivial	Extracardiac	Fenestrated	Fontan takedown on POD #2	Hypotension & hypoxemia	Highly likely
Heterotaxy, LV-dominant AV canal and pulmonary atresia	No	Normal	Trivial	Extracardiac	Fenestrated	Fontan takedown on POD #3	Hypotension & hypoxemia	Highly likely
DORV with d-malposed great arteries, mitral atresia, pulmonary atresia	Yes	Mild dysfunction	Trivial	Lateral tunnel	Fenestrated	Death on POD #0	Hypotension & hypoxemia	Highly likely
HLHS (MA/AA)	Yes	Normal	Mild	Extracardiac	Non-fenestrated	Death on POD #3	Hypotension & hypoxemia	Highly likely
HLHS (MA/AA)	No	Normal	Trivial	Extracardiac	Non-fenestrated	Death on POD #8	Hypotension & hypoxemia	Highly likely
Heterotaxy, DORV, total anomalous pulmonary venous return	No	Normal	Trivial	Extracardiac	Fenestrated	Death on POD #25	Persistent hypotension and low cardiac output, clot in Fontan, stroke	Moderately likely
Tricuspid atresia type IB (VSD, pulmonic stenosis, normally-related great arteries)	No	Normal	Trivial	Extracardiac	Non-fenestrated	Death on POD #67	Hypotension & hypoxemia, not explicitly documented as presumed Fontan failure	Moderately likely
HLHS (MA/AA)	No	Mild dysfunction	Mild	Extracardiac	Fenestrated	Death on POD #173	Chylous effusions, plastic bronchitis, respiratory failure	Highly likely

VSD = ventricular septal defect; LV = left ventricle; AV = atrioventricular; DORV = double outlet right ventricle; HLHS = hypoplastic left heart syndrome; MA = mitral atresia; AA = aortic atresia.