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Center Variation in Patient Age and Weight at Fontan Operation and Impact on Post-operative Outcomes

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Abstract

Background—The impact of age and weight on outcomes following the Fontan operation is unclear. Previous analyses have suggested that lower weight-for-age z-score is an important predictor of poor outcome in patients undergoing bidirectional Glenn. We evaluated variation in age, weight, and weight-for-age z-score at Fontan across institutions, and the impact of these variables on post-operative morbidity and mortality.

Methods—Patients in the Society of Thoracic Surgeons Congenital Heart Surgery Database undergoing the Fontan operation (2000–2009) were included. Center variation in age, weight, and weight-for-age z-score were described. Multivariable analysis was performed to evaluate the impact of age, weight, and weight-for-age z-score on in-hospital mortality, Fontan failure (combined in-hospital mortality and Fontan takedown/revision), post-operative length of stay, and complications, adjusting for other patient and center factors.

Results—A total of 2747 patients (68 centers) were included: 61% male; 45% right dominant lesions (38% left dominant, 17% undifferentiated). An extracardiac conduit Fontan (vs. lateral tunnel) was performed in 63%; 65% were fenestrated. Median age and weight at Fontan operation and proportion with weight-for-age z-score <−2 varied across centers ranging from 1.7–4.8 yrs, 10.5–16.1 kg, and 0%–30%, respectively. In multivariable analysis, age and weight were not significantly associated with outcome. Weight-for-age z-score <−2 was associated with increased in-hospital mortality (OR 2.73, 95%CI 1.09–6.86), Fontan failure (OR 2.59, 95%CI 1.24–5.40), and longer length of stay (+1.2 days, 95%CI 0.1–2.4).

Conclusions—Weight-for-age z-score <−2 is associated with significant morbidity and mortality following the Fontan operation independent of other patient and center characteristics.

Keywords

Congenital Heart Disease; Outcomes

Introduction

In 1978, Choussat published ten criteria describing optimal conditions under which to perform the Fontan operation, including age >4 years (1). In more recent years, the Fontan operation has been performed in younger and smaller patients at many centers. However, the impact of patient age and weight on outcome remains unclear. Some have reported satisfactory outcome following Fontan operation performed in children as young as 7 months (2). Others have reported that younger age at Fontan is associated with increased morbidity and mortality (3–5). There are also conflicting reports regarding weight with some studies suggesting that lower weight at Fontan operation (in particular weight <10 kg) is a risk factor for post-operative morbidity, while others have not shown this to be the case (6–8).

The majority of studies to date on this topic are single center reports limited by relatively small sample size (2–12). In addition, most studies have evaluated weight without taking into account weight relative to age as a predictor of outcome. A previous study has suggested that low weight-for-age z-score is an important predictor of poor outcome following the bidirectional Glenn operation (13). Further, recent data suggests that growth in single ventricle patients is modifiable, and may be optimized through a standardized feeding and monitoring program (14).

The purpose of this study was to evaluate variation across institutions in age, weight, and weight-for-age z-score at the time of Fontan operation, and to assess the impact of these factors on post-operative morbidity and mortality utilizing the Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database.

Patients and Methods

Data Source

The STS Congenital Heart Surgery Database contains de-identified operative, peri-operative, and outcomes data on more than 160,000 patients, and represents nearly three quarters of all US centers performing congenital heart surgery (15). Data quality and reliability are assured through intrinsic verification of data as well as a formal process of site visits and data audits (16). The Duke Clinical Research Institute serves as the data warehouse for the STS Databases. This study was approved by the Duke University Medical Center institutional review board with waiver of informed consent, and by the Access and Publications Committee of the STS Workforce for National Databases.

Patient Population

To maximize data integrity, analysis was restricted to 68 STS centers with >85% complete data for all study variables. From these centers, patients ≤18 years who underwent primary Fontan operation from 2000–2009 were eligible for inclusion. Patients undergoing Fontan conversion or repeat Fontan operation were excluded. In cases where it was unclear if the patient had undergone previous Fontan operation and was >6 years of age, the patient was excluded from analysis as it was felt to be less likely the patient was undergoing primary Fontan operation. Patients with missing or invalid data for key variables of age and weight

were excluded, along with those undergoing types of Fontan operation other than lateral tunnel or extracardiac conduit.

Data collection

Patient characteristics included age, weight, weight-for-age z-score, height, height-for-age z-score, and weight-for-height z-score (all calculated using standard growth curves) cardiac diagnosis (categorized as right dominant lesions, left dominant lesions, and undifferentiated), non-cardiac/genetic abnormality, number of prior cardiothoracic surgeries, preoperative length of stay, and other pre-operative factors including pre-operative neurologic deficit/seizures, arrhythmia, and complete heart block requiring pacemaker (these pre-operative factors represent those captured by the STS Database which occurred in >0.5% of patients undergoing the Fontan operation) (17). Center characteristics included annual Fontan operation volume and region. Operative characteristics included Fontan type (extracardiac conduit vs. lateral tunnel, and fenestration status as coded by the surgeon at the time of the operation) and secondary procedures performed at the time of Fontan operation.

Outcomes

The primary outcome was in-hospital mortality. Deaths occurring outside the hospital were not included in this analyses as these are not consistently captured in the STS Congenital Heart Surgery Database currently. Secondary outcomes included Fontan failure (combined in-hospital mortality, transplant, or Fontan revision/takedown), post-operative length of stay, and complications. Post-operative complications included any of those defined in the STS Congenital Heart Surgery Database (18).

Statistical Analysis

Patient, center, and operative characteristics were described. Age and weight-for-age z-scores were categorized for descriptive purposes based on the distribution of the data: age 0–2 years, 2–4 years, and >4 years; and weight-for-age z-score <–2, –2 to 0, and >0. Weight was categorized as <10 kg vs. ≥10 kg based on previous studies suggesting weight <10 kg was a significant risk factor for poor outcome (6).

Outcomes were compared across age, weight, and weight-for-age z-score groups in univariate analysis using the Chi-square and Kruskal-Wallis tests. The association of these variables with outcome was then evaluated in multivariable analysis utilizing hierarchical logistic and linear regression, with hospital specific intercepts to account for within center clustering. Age, weight, and weight-for-age z-score were evaluated both as continuous and as categorical variables. Multivariable analyses adjusted for other patient, operative, and center factors [sex, cardiac diagnosis, non-cardiac/genetic abnormality, other pre-operative factors as described above, preoperative length of stay, number of previous cardiac surgeries, Fontan type (extracardiac conduit vs. lateral tunnel, and fenestrated vs. non-fenestrated), other secondary surgical procedures performed at the time of Fontan operation, and center Fontan volume]. Length of stay was not normally distributed and was therefore log transformed for analysis. Results from logistic regression models are displayed as odds ratios (OR) and 95% confidence intervals and results from linear regression models as parameter estimates and 95% confidence intervals. Missing data were rare (< 0.5% for all variables). Patients with missing data for an endpoint were excluded from analysis involving that endpoint. All analyses were performed using STATA version 11.0 (StataCorp, College Station, TX). A p-value < 0.05 was considered statistically significant.

Results

Patient and operative characteristics

A total of 2747 patients from 68 centers (42.6% South, 22.1% Midwest, 19.1% West, 16.2% Northeast) were included (Table 1). Most patients were between 2–4 years, weighed 10–15 kg, and had a weight for age z-score between –2 and 1 at the time of Fontan operation (Figures 1–3). However, 13.1% were <2 years, 5.1% were <10 kg and 14.3% had a weight for age z-score <–2. Overall, both weight-for-age z-score and height-for-age z-score were low (Table 1). Evaluation of weight-for-height z-score suggested disproportionately lower weight in relation to height in the smallest patients (Table 1).

An extracardiac Fontan (vs. lateral tunnel) was performed in 63%, and 65% of all Fontans were fenestrated (Table 1). Thirty-nine percent (n=1058) of patients underwent a total of 1466 secondary procedures at the time of the Fontan operation. The most common secondary procedure was pulmonary arterioplasty in 10.8%; 4.8% underwent atrioventricular valve repair/replacement and 0.8% underwent arch surgery.

Center Variation

Median age and weight at Fontan operation and proportion with weight-for-age z-score <–2 varied from center-to-center with a range of 1.7 – 4.8 years, 10.5 – 16.1 kg, and 0% – 30%, respectively.

Outcomes

Unadjusted outcomes are displayed in Table 2. Of note, there were no in-hospital transplants performed following the Fontan operation, thus the data on in-hospital Fontan failure includes only mortality or Fontan revision/takedown. In multivariable analysis, age and weight were not significantly associated with outcome when analyzed as both continuous (data not shown) and categorical variables (Table 3). Lower weight-for-age z-score was associated with significantly increased in-hospital mortality, Fontan failure, and longer length of stay.

Comment

In this large multi-institutional cohort, we found substantial variation from center-to-center in age, weight, and weight-for-age z-score at the time of Fontan operation. In multivariable analysis, age and weight were not significantly associated with outcome, while lower weight-for-age z-score was associated with significantly increased in-hospital morbidity and mortality.

Previous studies have suggested that age <4 years was a risk factor for early Fontan failure (4,5,19). However, these reports included data from the time period when atriopulmonary and atrioventricular Fontan operations were favored techniques. More recent studies in the era of lateral tunnel and extracardiac Fontan operation have suggested that age is not significantly associated with post-operative outcome (8,11,12,20). Pizarro et al evaluated children <18 months and found that early mortality, Fontan takedown, and length of stay did not differ compared with older patients (20). Our study supports these findings in a large multi-institutional population. While our analysis focused on peri-operative outcomes, other studies have suggested that age at surgery may impact longer-term outcomes, such as aerobic capacity and hemodynamic status (21,22).

In our analysis of anthropometric data, we found overall impaired growth in regard to both height and weight in the population undergoing Fontan, similar to others (23). There was

substantial variation from center-to-center in patient weight at Fontan operation, but similar to age, we did not find weight to be significantly associated with post-operative outcomes. Ikai et al. evaluated 72 patients from a single institution and found that weight <10kg was a significant risk factor for prolonged length of stay (6). In contrast, our findings are consistent with several more recent studies suggesting lower weight at Fontan operation is not a risk factor for poor outcome (8,11,24–26).

We also found significant variation from center-to-center in weight-for-age z-score at the time of Fontan operation. In contrast to age and raw weight, we did find that lower weight-for-age z-score was significantly associated with post-operative morbidity and mortality independent of other patient, operative, and center factors. To our knowledge, the impact of weight-for-age z-score on outcome following the Fontan operation has not been evaluated previously. However, similar findings have been noted in an evaluation of 100 children undergoing bidirectional Glenn, where lower weight-for-age z-score was significantly associated with longer length of stay and a trend toward increased mortality and other morbidities (13).

We found that those in the lowest weight-for-age z-score group had disproportionately lower weight in relation to height. There are several factors which may impact weight gain, and in turn, impact outcome. Adequate nutrition can be impaired by feeding difficulties which are common in the single ventricle population (27). Poor nutritional status and low protein reserves could predispose to pleural effusions, infection, and poor wound healing (13,28,29). Impaired growth may also be present in patients with genetic syndromes (30). However, we found that the association of low weight-for-age z-score with outcome was independent of the presence of genetic syndrome/non-cardiac abnormality. Factors increasing metabolic demand or impairing the ability to meet metabolic demand can also impair growth. Vogt et al demonstrated a significant association between pre-operative cardiac medications and the presence of venous collaterals and lower weight-for-age z-scores at Fontan operation (23). Cardiac medication use may be a surrogate for congestive heart failure which may increase metabolic demand, and venous collaterals may be associated with cyanosis which can impair ability to meet metabolic demand. We did adjust for atrioventricular valve regurgitation and coarctation/arch hypoplasia requiring intervention at the time of the Fontan operation (among other secondary procedures at the time of Fontan) in our analyses, both of which may be associated with pre-operative heart failure and poor cardiac function. However there are likely other variables related to heart failure and cyanosis which we were unable to account for. Finally, lower weight-for-age z-score at the time of Fontan operation has also been associated with requirement for additional surgical procedures prior to the Fontan and older age at Fontan operation (23). However, we adjusted for age and number of previous cardiothoracic operations in our analysis, such that the impact of lower weight for age z-score on outcome in our study is likely independent of these factors.

Recent data from Hehir and colleagues suggest that growth is a modifiable factor in the single ventricle population (14). Through a comprehensive feeding and monitoring protocol, investigators demonstrated growth velocity in single ventricle patients similar to that of a normal child despite the presence of other comorbidities, and excellent outcomes through stage 2 palliation with an interstage survival of 98% (14).

Limitations

The limitations of this study are primarily related to the limitations of the STS Database and the observational nature of the analysis. Although this is largest study to date evaluating age and weight at Fontan operation, the STS Database does not include the entire US population and thus, may underestimate center variation.

In addition, pre-operative hemodynamic and echocardiographic variables are not available in the STS Database. Information regarding cardiologist and surgeon decision making concerning the timing of surgery is also not available. Therefore we were not able to account for these factors in our analysis. We were able to account for several factors captured by the database which may impact both growth and outcome, including the presence of genetic/non-cardiac abnormalities, diagnosis, previous interventions, other pre-operative comorbidities, as well as hemodynamic/anatomic abnormalities (atrioventricular valve regurgitation, coarctation, etc) requiring intervention at the time of the Fontan. The current state of the database also does not enable us to know with certainty whether all patients have undergone Fontan following a superior cavopulmonary anastomosis, as opposed to being done in a palliated or unoperated state. Thus, it is unknown whether a volume unloading procedure preceded the Fontan, or by how long. Somatic growth parameters may be reflective of a variety of types of hemodynamic burden, of which ventricular volume load certainly is one. Due to these limitations, we are not able to draw inferences regarding the exact mechanism by which weight-for-age z-score is related to outcome as discussed above. Further study is needed to evaluate the relationship with other variables including nutritional status, heart failure, and cyanosis. Recent data from the Children's Hospital of Wisconsin support the notion that growth velocity in single ventricle patients similar to that of a normal child can be achieved with a comprehensive feeding and monitoring plan despite the presence of these comorbidities (14).

We were also unable to analyze outcomes associated with varying practice patterns within an individual institution as this was precluded by the small number of patients and events. Finally, the STS Database currently only allows for the evaluation of in-hospital outcomes. Plans to expand data collection to include long-term morbidity and mortality will allow analyses of longer-term outcomes to be performed in the future.

Conclusions

In this large multi-institutional analysis, age and weight at Fontan operation did not significantly impact post-operative outcomes. However, lower weight-for-age z-score was associated with significantly increased post-operative mortality, Fontan failure, and length of stay, independent of other patient and center characteristics. Previous analyses have suggested that growth is modifiable with a comprehensive feeding and monitoring plan in single ventricle patients, and that poor growth is also a risk factor for poor outcome in patients undergoing bidirectional Glenn (13,14). These data, along with the results of the present analysis, support the implementation of strategies to optimize growth in this population and further investigation into mediating factors.

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Figure 1.
Age distribution

Figure 2.
Weight distribution

Figure 3.
Weight-for-age z-score distribution

Table 1**Patient, Operative, and Center Characteristics**

Variable	Overall (n=2747)	<-2 (n=393)	Weight-for-age z-score -2 to 0 (n=1580)	>0 (n=774)
Patient Characteristics				
Age, years	3.0 (2.3, 3.6)	3.0 (2.3, 4.0)	3.0 (2.3, 3.7)	2.8 (2.3, 3.4)
Male	1664 (60.6%)	213 (54.2%)	992 (62.8%)	459 (59.3%)
Diagnosis				
Right dominant lesions	1239 (45.1%)	195 (49.6%)	730 (46.2%)	314 (40.6%)
Left dominant lesions	1031 (37.5%)	128 (32.6%)	574 (36.3%)	329 (42.5%)
Undifferentiated	477 (17.4%)	70 (17.8%)	276 (17.5%)	131 (16.9%)
Anthropometric Data				
Weight, kg	13.2 (11.7, 14.8)	10.8 (9.8, 12.0)	12.9 (11.7, 14.2)	15.0 (13.8, 16.5)
Weight-for-age z-score	-0.7 (-1.5, 0.1)	-2.5 (-3.1, -2.3)	-0.9 (-1.4, -0.4)	0.6 (0.2, 1.0)
Height, cm	91 (86, 97)	87 (81, 93)	91 (86, 97)	93 (88, 99)
Height-for-age z-score	-0.8 (-1.5, -0.6)	-2.1 (-2.8, -1.4)	-0.9 (-1.5, -0.3)	0 (-0.6, 0.7)
Weight-for-height z-score	-0.2 (-1.1, 0.7)	-1.8 (-2.5, -1.2)	-0.4 (-1.1, 0.2)	1.0 (0.4, 1.6)
Pre-operative factors				
Neurologic deficit/seizures	82 (3.0%)	14 (3.6%)	50 (3.2%)	18 (2.3%)
Arrhythmia	50 (1.8%)	7 (1.8%)	29 (1.8%)	14 (1.8%)
Complete heart block/pacemaker	16 (0.6%)	5 (1.3%)	9 (0.6%)	2 (0.3%)
Any non-cardiac/genetic abnormality	536 (19.5%)	100 (25.5%)	298 (18.9%)	138 (17.8%)
Pre-operative LOS >2 days	58 (2.1%)	7 (1.8%)	37 (2.3%)	14 (1.8%)
Number of prior CT operations	2 (2, 3)	2 (2, 3)	2 (2, 3)	2 (2, 3)
Operative Characteristics				
Fontan type				
Lateral tunnel fenestrated	881 (32.0%)	154 (39.2%)	532 (33.6%)	195 (25.2%)
Lateral tunnel non-fenestrated	136 (5.0%)	16 (4.1%)	74 (4.7%)	46 (5.9%)
Extracardiac conduit fenestrated	907 (33.0%)	121 (30.8%)	513 (32.5%)	273 (35.3%)
Extracardiac conduit non-fenestrated	823 (30.0%)	102 (26.0%)	461 (29.2%)	260 (33.6%)
Secondary procedure at Fontan, n (%)	1058 (38.5%)	162 (41.2%)	611 (38.7%)	285 (36.8%)
Center annual Fontan volume, median operations/ year	10.8 (7.7, 31.4)	13.0 (8.8, 34.9)	10.8 (7.6, 31.4)	10.6 (7.7, 31.4)

Data for continuous variables are displayed as median (interquartile range)

LOS=length of stay, CT = cardiothoracic

Table 2

Unadjusted In-hospital Outcomes in Age, Weight, and Weight-for-age z-score Groups

A. Age				
Outcome	<2 years (n=361)	2–4 years (n=1918)	>4 years (n=468)	p-value
In hospital mortality	5 (1.4%)	30 (1.6%)	10 (2.1%)	0.63
Fontan failure	9 (2.5%)	49 (2.6%)	15 (3.2%)	0.72
Postoperative LOS, days *	9.7 (7.0, 13.0)	10.8 (7.0, 15.0)	11.5 (7.0, 15.0)	0.01
Postoperative complications	148 (41.0%)	772 (40.3%)	191 (40.8%)	0.95
B. Weight				
Outcome	<10kg (n=139)	≥ 10 kg (n=2608)		p-value
In hospital mortality	4 (2.9%)	41 (1.6%)		0.24
Fontan failure	6 (4.3%)	67 (2.6%)		0.21
Postoperative LOS, days *	10.9 (7.0, 15.0)	10.6 (7.0, 14.0)		0.80
Postoperative complications	63(45.3%)	1048 (40.2%)		0.23
C. Weight-for-age z-score				
Outcome	z-score <-2 (n=393)	z-score -2 to 0 (n=1580)	z-score >0 (n=774)	p-value
In hospital mortality	15 (3.8%)	20 (1.3%)	10 (1.3%)	0.001
Fontan failure	22 (5.6%)	34 (2.2%)	17 (2.2%)	<0.001
Postoperative LOS, days *	11.8 (7.0, 17.0)	10.6 (7.0, 14.0)	10.5 (7.0, 14.0)	0.33
Postoperative complications	175 (44.5%)	613 (38.8%)	323 (41.7%)	0.08

LOS=length of stay

* data are presented as 10% trimmed mean (interquartile range)

Table 3

Adjusted In-hospital Outcomes

In-hospital mortality	Adjusted OR (95% CI)	p-value
Age		
<2 years	0.97 (0.26, 3.67)	0.96
2–4 years	0.94 (0.42, 2.07)	0.87
>4 years	(reference)	
Weight		
<10 kg	(reference)	
≥10 kg	0.94 (0.24, 3.65)	0.93
Weight-for-age z-score		
<−2	2.73 (1.09, 6.86)	0.03
−2 to 0	0.98 (0.45, 2.16)	0.96
>0	(reference)	
Fontan failure	Adjusted OR (95% CI)	
Age		
<2 years	1.37 (0.48, 3.51)	0.60
2–4 years	1.03 (0.54, 1.96)	0.93
>4 years	(reference)	
Weight		
<10 kg	(reference)	
≥10 kg	0.99 (0.33, 2.98)	0.98
Weight-for-age z-score		
<−2	2.59 (1.24, 5.40)	0.01
−2 to 0	1.02 (0.56, 1.87)	0.95
>0	(reference)	
Complications	Adjusted OR (95% CI)	
Age		
<2 years	1.01 (0.69, 1.47)	0.96
2–4 years	0.97 (0.76, 1.25)	0.83
>4 years	(reference)	
Weight		
<10 kg	(reference)	
≥10 kg	0.76 (0.48, 1.21)	0.25
Weight-for-age z-score		
<−2	1.06 (0.78, 1.44)	0.73
−2 to 0	0.88 (0.72, 1.07)	0.20
>0	(reference)	
Length of stay	Estimate[*] (95% CI)	
Age		
<2 years	−0.01 (−0.11, 0.09)	0.84
2–4 years	0.04 (−0.02, 0.11)	0.22

In-hospital mortality	Adjusted OR (95% CI)	p-value
>4 years	(reference)	
Weight		
<10 kg	(reference)	
≥10 kg	−0.007 (−0.13, 0.12)	0.91
Weight-for-age z-score		
<−2	0.09 (0.01, 0.18) **	0.03
−2 to 0	0.02, (−0.03, 0.07)	0.41
>0	(reference)	

CI=confidence interval, OR=odds ratio,

* log days,

** estimate of difference in predicted length of stay in days from multivariable model in weight-for-age z-score <−2 group vs. >0 group = +1.2 days, 95% CI 0.1–2.4