

# Development of Weight and Height Age z-Score after Total Cavopulmonary Connection

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## Abstract

**Objective** We aimed to analyze somatic growth of patients after total cavopulmonary connection (TCPC) as well as to identify factors influencing postoperative catch-up growth.

**Methods** A total of 309 patients undergoing TCPC at 4 years old or less between 1994 and 2021 were included. Weight for age z-score (WAZ) and height for age-z-score (HAZ) at TCPC and at postoperative time between 1 and 3 years were calculated. Factors influencing somatic growth were analyzed.

**Results** Most frequent diagnosis and initial palliation were hypoplastic left heart syndrome (HLHS) (34%) and the Norwood procedure (51%), respectively. Median age and weight at TCPC were 2.0 (IQR: 1.7–2.5) years and 11.3 (10.5–12.7) kg, respectively. Median 519 days after TCPC, a significant increase in WAZ (−0.4 to −0.2,  $p < 0.001$ ) was observed, but not in HAZ (−0.6 to −0.6,  $p = 0.38$ ). Older age at TCPC ( $p < 0.001$ , odds ratio [OR]: 2.6) and HLHS ( $p = 0.007$ , OR: 2.2) were risks for low WAZ after TCPC. Older age at TCPC ( $p = 0.009$ , OR: 1.9) and previous Norwood procedure ( $p = 0.021$ , OR: 2.0) were risks for low HAZ after TCPC. Previous bidirectional cavopulmonary shunt (BCPS) was a protective factor for both WAZ ( $p = 0.012$ , OR: 0.06) and HAZ ( $p = 0.028$ , OR: 0.30) at TCPC.

**Conclusion** In patients undergoing TCPC at the age of 4 years or less, a significant catch-up growth was observed in WAZ after TCPC, but not in HAZ. Previous BCPS resulted to be a protective factor for a better somatic development at TCPC. HLHSs undergoing Norwood were considered as risks for somatic development after TCPC.

## Keywords

- ▶ somatic development
- ▶ weight for age z-score
- ▶ single ventricle
- ▶ total cavopulmonary connection
- ▶ ventricular

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## Introduction

Children with univentricular heart represent a heterogeneous patients with various underlying diagnoses.<sup>1</sup> The Fontan procedures are performed empirically as definitive palliations in children with univentricular heart.<sup>2</sup> During childhood, growth is a fundamental measure of response to therapy.<sup>3,4</sup> Thus, the weight and height of children with univentricular hearts who have undergone Fontan procedure were studied to determine the effect of each intervention on growth.<sup>5-16</sup> It is generally known that early surgical and catheter intervention and early removal of volume loading and cyanosis lead to improvement of somatic growth.<sup>6-8</sup> However, many patients who have undergone Fontan palliation, which is often fraught with complications, might afterward suffer from impaired somatic growth. Previous follow-up studies examining somatic development in children with univentricular hearts showed inconsistent results, but currently suggest that somatic development is less favorable in early childhood and persist into adolescence.<sup>13-15</sup> The morphological, surgical, and hemodynamic factors associated with failure of somatic development in these patients are not well studied. It remains unclear which factor is the main determinant of somatic growth: the substrate of the univentricular heart by itself, the timing of the obligatory surgical steps from the first neonatal surgery to the total cavopulmonary connection (TCPC) with its inherent fluctuations in volume load and the degree of cyanosis, or the hazardous long-term performance of the univentricular heart.

Various factors have been implicated as underlying causes. However, studies have suggested that the key may lie in the timing of volume-unloading procedures staged by bidirectional cavopulmonary shunt (BCPS) or hemi-Fontan procedure.<sup>6-8</sup>

We hypothesize that the early removal of cyanosis and volume overload in the univentricular heart is not only beneficial for the early postoperative course but also provides the optimal basis for somatic development of the children. Therefore, we analyzed our early-term experience in children who underwent TCPC before the age of 4 years with regard to somatic development after early palliation. We also aimed to analyze the risk factors that could adversely affect postoperative catch-up growth.

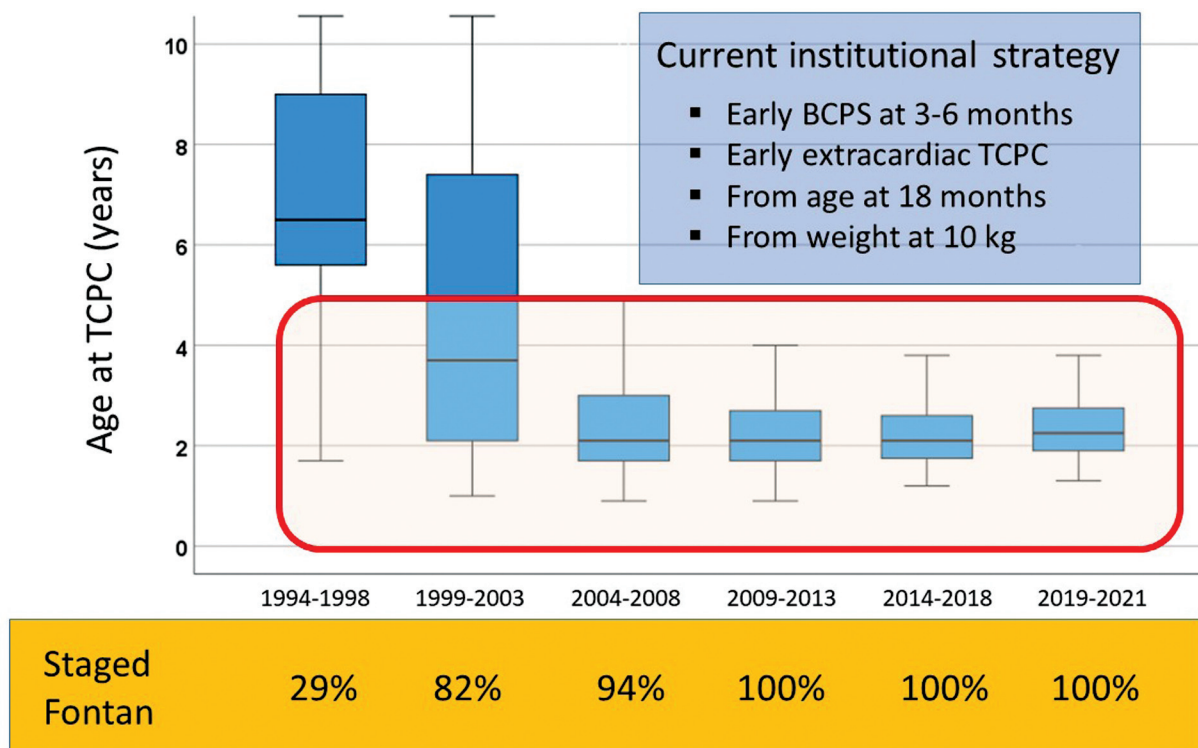
## Methods

### Ethical Statement

This study was approved by the Institutional Review Board of the Technical University of Munich (approved number of 2022-303-S-KH on June 27, 2022). Because of the retrospective nature of the study, the need for individual patient consent was waived.

### Patients and Data Collection

A total of 620 patients underwent TCPC at our institution between 1994 and 2021. Era distribution of age at TCPC and percentage of staged TCPC (previous BCPS) are shown in ►Fig. 1. Weight and height at the time of TCPC and in



**Fig. 1** Era distribution of age at TCPC and percentage of staged TCPC. Our current strategy is to perform BCPS at 3 to 6 months of age and to complete Fontan by extracardiac TCPC from the age at 18 months and the weight of 10 kg. Box-and-whisker dot plots showing era distribution of age at TCPC between 1994 and 2021, and percentage of staged Fontan completion with BCPS. The top and bottom whiskers mark the minimum and maximum values, the upper and lower borders of the box represent the upper and lower quartiles, and the middle horizontal line represents the median. BCPS, bidirectional cavopulmonary shunt; DHM, Deutsches Herzzentrum München; TCPC, total cavopulmonary connection.

the postoperative periods were collected in all available patients. Patients who underwent TCPC at the age of 4 years or younger (<1,825 days old) were selected. Among them, patients whose weight and height at the time of TCPC and a postoperative time between 1 and 3 years were available were included in this study. Medical records included baseline morphology and demographics as well as pre-, intra-, and postoperative data using electronic and paper chart reviews of each patient.

### Operative Techniques

The operative techniques for TCPC have been described in previous reports.<sup>17,18</sup> Lateral tunnel TCPC was performed in the early era. In January 1999, extra-cardiac TCPC was introduced, and has been our standard procedure since May 2002.<sup>18</sup> Fenestration was not routinely performed and was only used for high-risk patients.<sup>17</sup>

### Calculation of Age-Adjusted z-Score

Weight for age z-score (WAZ) and height for age z-score (HAZ) were calculated using WHO Anthro software version 3.2. (World Health Organization, Geneva, Switzerland). Body mass index (BMI) was calculated using the following formula:  $BMI = \text{weight}/\text{height}^2$ , and BMI for age z-score (BMIZ) was also calculated. Low weight, low height, or low BMI was defined as values less than the lower interquartile ranges (IQRs) of the entire group. As for the risk factor analysis for WAZ, HAZ, and BMIZ, a linear regression model was used with the cut-off value of lower IQRs of each value.

Delta WAZ (post-WAZ – pre-WAZ) was also calculated to analyze catch-up weight gain after TCPC. Using the same method, delta HAZ and delta BMIZ were also calculated, and risk factor analysis was performed using a linear regression model with the cut-off value of 25 lower IQR of delta WAZ, delta HAZ, and delta BMIZ.

### Statistical Analysis

Categorical variables are presented as absolute numbers and percentages. Chi-square test was used for categorical data. Continuous variables are expressed as medians with IQR. An independent sample *t*-test was used to compare normally distributed variables. The paired sample *t*-test was performed for the comparison of pre- and post-TCPC variables. The Mann-Whitney U test was used for variables that were not normally distributed. Risk factors for impaired somatic growth were assessed using a uni- and multivariate linear regression model with a cut-off value of lower IQR. For multivariate analysis, variables with a significance level of less than 0.1 in the univariate analysis were entered into the multiple linear regression models. Hazard ratios (HRs) with 95% confidence intervals were estimated. Final models were derived by the forward and backward stepwise selection procedure. *p*-Values <0.05 were considered significant. Data analysis was performed using SPSS version 28.0 for Windows (IBM, Ehningen, Germany) and R-statistical software (state package).

## Results

### Patient Characteristics and Operative Data

A total of 309 patients who met the inclusion criteria were identified. A flow chart of the patient selection is shown in **–Supplementary Fig. S1** (available in the online version). Patient characteristics are presented in **–Table 1**. The most frequent diagnosis was hypoplastic left heart syndrome (HLHS) in 105 (34%) patients, followed by univentricular heart in 55 (18%), tricuspid atresia in 50 (16%), double inlet left ventricle in 39 (13%), congenitally corrected transposition of the great arteries in 16 (5%), pulmonary atresia and intact ventricular septum in 15 (5%), and unbalanced atrioventricular septal defect in 12 (4%). Dominant right ventricle was observed in 172 (56%) patients. Most frequent stage I procedure was the Norwood procedure in 156 (51%), followed by systemic to pulmonary artery shunt in 87 (28%) and pulmonary artery banding in 31 (10%). BCPS was performed in 303 (98%) patients. Median age and median weight at TCPC were 2.0 (IQR: 1.7–2.5) years and 11.3 (10.5–12.7) kg, respectively. Intra-cardiac lateral tunnel TCPC was performed in 14 (5%) patients and extra-cardiac TCPC was performed in 295 (95%) patients. Median duration of cardiopulmonary bypass time was 61 (IQR: 46–83) minutes. Aortic cross-clamping was needed in 58 (19%) patients with a median duration of 43 (23–67) minutes. Fenestration was created in 13 (4%) patients. Median postoperative length of stay in the intensive care unit (ICU) was 6 (4–8) days and median hospital stay was 20 (14–27) days. Prolonged pleural effusion (drainage needed more than 7 days) was observed in 161 (52%) patients, and chylothorax in 78 (25%) patients. Re-admission to ICU was needed in 79 (26%) patients, and secondary fenestration was needed in 5 (2%) patients. There were six early deaths within 30 days and nine late deaths after TCPC.

### Weight and Height for Age z-Score

The median WAZ, HAZ, and BMIZ at pre-TCPC were –0.44 (–1.20, 0.27), –0.60 (–1.50, 0.28), and –0.08 (–0.95, –0.41), respectively. Pre-TCPC, 28 patients (9.1%) had WAZ of lower than –2.0, 43 patients (13.9%) had HAZ lower than –2.0, and 25 patients (8.1%) had BMIZ lower than –2.0. Post-TCPC data were obtained median 519 (IQR: 368–672) days following TCPC. The median WAZ, HAZ, and BMIZ at post-TCPC were –0.24 (–0.91, 0.54), –0.56 (–1.39, 0.34), and 0.31 (–0.41, 0.89), respectively. Post-TCPC, 8 patients (2.6%) had WAZ lower than –2.0, 36 patients (11.7%) had HAZ lower than –2.0, and 4 patients (1.3%) had BMIZ lower than –2.0. Comparison of pre- and post-TCPC WAZ, HAZ and BMIZ using paired sample *t*-test is shown in **–Fig. 2**. As the left graphic shows, WAZ was significantly increased after TCPC ( $p < 0.001$ ). However, as the central graphic shows, HAZ was similar between pre- and post-TCPC ( $p = 0.384$ ). As the right graphic shows, BMIZ was significantly increased after TCPC ( $p < 0.001$ ).

The results of factor analysis for impaired somatic growth are shown in **–Table 2** and **–Supplementary Tables S1 to S3** (available in the online version). As factors for low WAZ pre-TCPC, older age at TCPC ( $p < 0.001$ , odds ratio [OR]: 2.708),

**Table 1** Patient characteristics

Variables, N (%) or median (IQR)	N = 309
Primary diagnosis	
Hypoplastic left heart syndrome (HLHS)	105 (34.0)
Univentricular heart (UVH)	55 (17.8)
Tricuspid atresia (TA)	50 (16.2)
Double inlet left ventricle (DILV)	39 (12.6)
Congenitally corrected transposition of the great arteries (ccTGA)	16 (5.2)
Pulmonary atresia with intact ventricular septum (PAIVS)	15 (4.9)
Unbalanced atrioventricular septal defect (UAVSD)	12 (3.9)
Others	19 (6.1)
Associated cardiac anomaly	
Transposition of the great arteries (TGA)	95 (30.7)
Coarctation of the aorta (CoA)	46 (14.9)
Anomalous systemic venous connection	31 (10.0)
Double outlet right ventricle (DORV)	29 (9.4)
Dextrocardia/situs inversus	25 (9.1)
Heterotaxy	21 (6.8)
Anomalous pulmonary venous connection (TAPVC/PAPVC)	18 (5.8)
Dominant RV	172 (55.7)
Initial palliation	
Norwood/Damus–Kaye–Stansel (DKS)	156 (50.5)
Systemic to pulmonary shunt (SPS)	87 (28.2)
Pulmonary artery banding (PAB)	31 (10.0)
Prior bidirectional cavopulmonary shunt (BCPS)	303 (98.1)
Operative data	
Age at TCPC (y)	2.0 (1.7–2.5)
Weight at TCPC (kg)	11.3 (10.5–12.7)
Type of TCPC	
Intracardial	14 (4.5)
Extra-cardiac	295 (95.5)
Cardiopulmonary bypass (CPB) time (min)	61 (46–83)
Need aortic cross clamp (AXC)	58 (18.8)
AXC time (min)	43 (23–67)
Fenestration	13 (4.2)

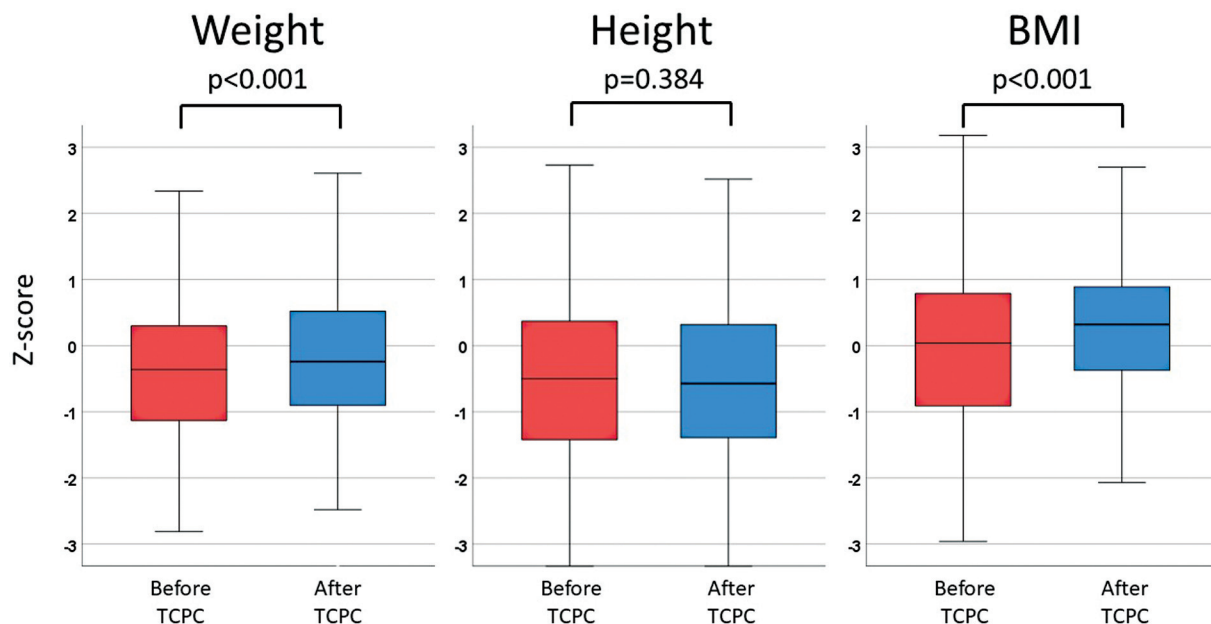
Abbreviation: TCPC, total cavopulmonary connection.

anomalous systemic venous return ( $p=0.011$ , OR: 2.960), and previous Norwood procedure ( $p=0.001$ , OR: 2.672) were identified as independent factors using multivariate analysis. Older age at TCPC ( $p < 0.001$ , OR: 2.575) and HLHS ( $p = 0.007$ , OR: 2.215) were identified as independent factors associated with low WAZ post-TCPC. As risk factors for low HAZ pre-TCPC, older age at TCPC ( $p = 0.002$ , OR: 1.673) was identified as an independent factor. As risk factors for low WAZ post-TCPC, older age at TCPC ( $p = 0.009$ , OR: 1.874) and previous Norwood procedure ( $p = 0.021$ , OR: 1.993) were identified as independent factors. As risk factors for low BMIZ post-TCPC, older age at TCPC ( $p = 0.021$ , OR: 1.713) was

identified as an independent factor. Previous BCPS was identified as a protective factor for WAZ pre-TCPC ( $p = 0.012$ , OR: 0.062), WAZ post-TCPC ( $p = 0.024$ , OR: 0.079), and BMIZ pre-TCPC ( $p = 0.036$ , OR: 0.159).

#### Factors Influencing Catch-Up Growth (Changes of Weight, Height, and BMI z-Score)

To analyze the factors influencing catch-up growth, changes in weight z-score were calculated as follows: delta WAZ (post-TCPC WAZ – pre-TCPC WAZ). Delta HAZ and delta BMIZ were calculated in the same way. Box-and-whisker dot plots of delta WAZ, delta HAZ, and delta BMIZ are shown



**Fig. 2** Box-and-whisker dot plots showing pre- and post-TCPC age z-score of weight, height, and BMI. The upper and lower whiskers mark the minimum and maximum values, the top and bottom borders of the box represent the upper and lower quartiles, and the middle horizontal line represents the median. BMI, body mass index; TCPC, total cavopulmonary connection.

in **Fig. 3**. Factors influencing delta WAZ, delta HAZ, and delta BMIZ are shown in **Table 3** and **Supplementary Table S4** (available in the online version). For changes in weight z-score, pre-TCPC Aortopulmonary collaterals (APCs) ( $p=0.025$ , OR: 1.958), fenestration at TCPC ( $p=0.037$ , OR: 4.840), and longer hospital stay ( $p=0.039$ , OR: 1.026) were identified as independent factors using a multivariate model. For changes in height z-score, age at BCPS ( $p=0.012$ , OR: 1.068) and re-ICU admission after TCPC ( $p=0.036$ , OR: 1.940) were identified as independent factors. As for changes in BMI z-score, tricuspid atresia ( $p=0.032$ , OR: 2.209) and fenestration at TCPC ( $p=0.049$ , OR: 4.677) were identified as independent factors. As the age at TCPC as a continuous variable was not identified as a risk for changes in weight z-score, or changes in height z-score, we analyzed the age at TCPC as a dichotomized variable using a cut-off value of 2 years old. Then, age at TCPC of 2 years or older was significantly associated with change of BMI z-score ( $p=0.032$ , HR: 0.536), but not associated with that for weight z-score ( $p=0.269$ , HR: 0.733), or height z-score ( $p=0.776$ , HR: 0.922).

## Discussion

The present study evaluated the somatic development of children with univentricular heart who underwent TCPC at the age of 4 years or younger. Our data showed that significant catch-up growth was observed in weight gain after TCPC, but not in height. Previous BCPS resulted as a significant protective factor for better somatic development at TCPC. Older age at TCPC was associated with reduced WAZ at TCPC and also at follow-up. Severely ill patients, such as those with HLHS, showed an impaired somatic development.

## Somatic Development in Patients with Univentricular Heart

All previous studies demonstrated that patients with univentricular heart have impaired somatic growth in infant age.<sup>5–16</sup> Several studies clearly demonstrated that this reduced somatic development was improved after stage II palliation by means of BCPS or hemi-Fontan procedure.<sup>4–9,11,15</sup> Although most of the patients had staged Fontan completion in this study, our results also showed that previous BCPS was a significant protective factor for the weight z-score at TCPC. However, catch-up growth after the Fontan procedure still remains controversial. Several studies have demonstrated significant catch-up growth in weight gain.<sup>4–8,12,16</sup> Day et al demonstrated that children with univentricular hearts grow more adequately following the BCPS than after the Fontan procedure, and that a decrease in growth after the Fontan procedure is associated with a poor prognosis.<sup>4</sup> Cohen et al demonstrated that weight improved by the time of the Fontan completion, and for the first 2 years after the Fontan procedure, but never normalized.<sup>5</sup> They also demonstrated that children with univentricular hearts who underwent Fontan procedure are significantly more often underweight and shorter than the general population and their siblings. François et al demonstrated that body growth is severely impaired in the Fontan population. They demonstrated that better catch-up growth at the time of TCPC was obtained when BCPS is performed earlier in the younger age.<sup>6</sup> Ovroutski et al reported that accelerated somatic growth, especially in small children, can be observed after the Fontan completion.<sup>7</sup> In this study, we did not observe an acceleration of somatic development after TCPC in younger patients, although younger patients demonstrated larger WAZ at TCPC and at follow-up. In the study from Ovroutski et al,

**Table 2** Risk factor analysis for weight, height, and BMI z-score before and after TCPC

Variables	P-Value	Univariate		p-Value	Multivariate	
		OR	95% CI		OR	95% CI
Weight pre-TCPC						
Age at TCPC	<0.001	2.489	1.754–3.534	<0.001	2.708	1.870–3.922
CoA	0.097	1.766	0.903–3.455			
Anomalous SVR	0.008	2.810	1.313–6.016	0.011	2.960	1.287–6.806
No. of palliation	0.088	1.295	0.962–1.744			
Previous Norwood	0.034	1.768	1.045–2.990	0.001	2.672	1.466–4.868
Previous BCPS	0.012	0.062	0.007–0.542			
Weight post-TCPC						
Age at TCPC	<0.001	2.358	1.514–3.674	<0.001	2.575	1.627–4.076
HLHS	0.024	1.891	1.087–3.290	0.007	2.215	1.239–3.961
No. of palliation	0.048	1.365	1.002–1.860			
Previous Norwood	0.059	1.697	0.980–2.939			
TGA	0.033	0.493	0.257–0.946			
Dextrocardia	0.058	0.240	0.055–1.047			
Previous BCPS	0.024	0.079	0.009–0.715			
Height pre-TCPC						
Age at TCPC	0.002	1.673	1.205–2.322	0.002	1.673	1.205–2.322
Type TCPC	0.037	0.317	0.108–0.934			
DILV	0.028	0.303	0.104–0.881			
Height post-TCPC						
Age at TCPC	0.023	1.694	1.074–2.672	0.009	1.874	1.169–3.004
No. of palliation	0.072	1.336	0.975–1.831			
Previous Norwood	0.054	1.745	0.992–3.070	0.021	1.993	1.110–3.580
Dextrocardia	0.053	0.234	0.054–1.021			
Previous BCPS	0.056	0.108	0.011–1.059			
BMI pre-TCPC						
Heterotaxy	0.055	2.426	0.981–6.004			
Previous BCPS	0.036	0.159	0.028–0.884			
BMI post-TCPC						
Age at TCPC	0.021	1.713	1.085–2.706	0.021	1.713	1.085–2.706

Abbreviations: BCPS, bidirectional cavopulmonary shunt; BMI, body mass index; CI, confidence interval; CoA, coarctation of the aorta; DILV, double inlet left ventricle; HLHS, hypoplastic left heart syndrome; OR, odds ratio; SVR, support vector regression; TCPC, total cavopulmonary connection.

they compared patients between age at TCPC under 4 years old and age 4 to 13 years old. However, we compared patients between age at TCPC under 2 years old and age 2 to 4 years old. Therefore, our results did not prove the rationale of early Fontan completion under 2 years old regarding the somatic catch-up growth.

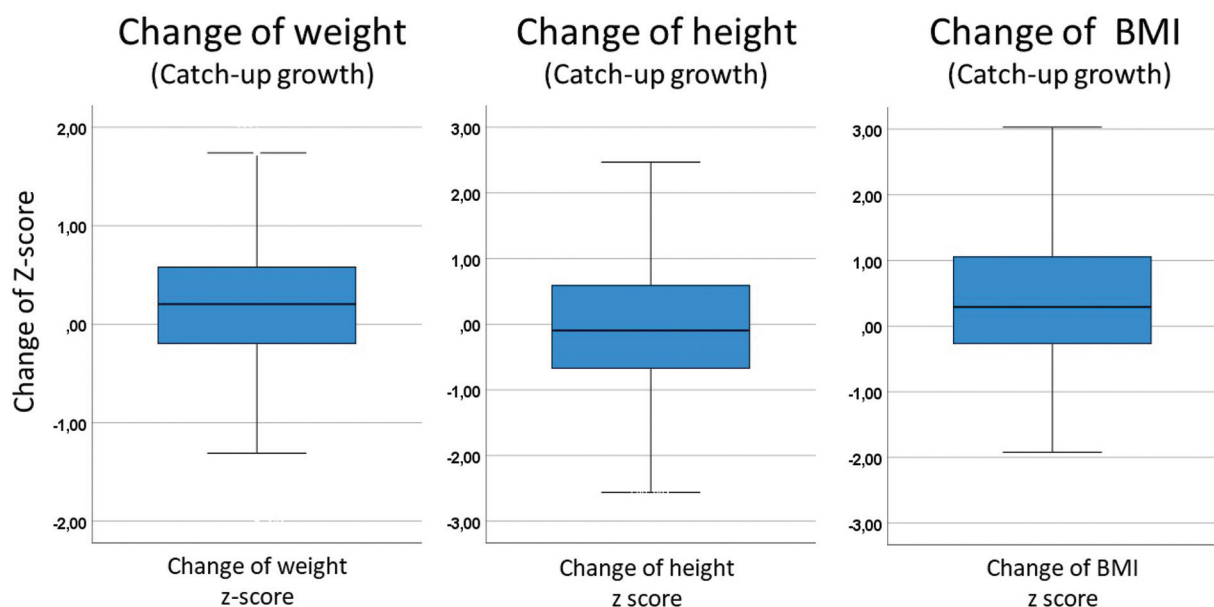
The z-score for height in this study differed from that for weight, and was similar to the results of Day et al.<sup>5</sup> They showed that no significant change in z-score for height was observed after the Fontan completion. Stenbøg et al demonstrated a small increase in height after the Fontan procedure.<sup>19</sup> These findings and our results suggest that patients with univentricular heart have impaired long-term growth

in height. Previous studies showed that patients with cyanotic congenital heart disease may have delayed bone age, which is affected by chronic hypoxemia after birth.<sup>20,21</sup> This delay in bone age might be correlated with lower z-score for height. Witzel et al also report that patients requiring the Fontan procedure remain significantly shorter than the normal population even into teenage, adolescence, and young adult.<sup>21</sup> They demonstrated delayed bone age density in these patients, resulting in the stunted growth. The chronic cyanosis presented in children with univentricular hearts might impair initial bone development at a crucial stage and impact growth for height even after the Fontan completion.

**Table 3** Risk factors for changes of weight, height, and BMI z-score

Variables	p-Value	Univariate		p-Value	Multivariate	
		OR	95% CI		OR	95% CI
Change of weight						
No. of palliation	0.065	1.338	0.962–1.823			
MAP	0.093	1.026	0.996–1.057			
APCs	0.005	2.250	1.275–3.971	0.025	1.958	1.087–3.529
Fenestration at TCPC	0.017	4.828	1.321–17.643	0.037	4.840	1.097–21.359
Hospital stay	0.023	1.023	1.003–1.044	0.039	1.026	1.001–1.052
Re-OP	0.083	2.213	0.902–5.428			
Re-intervention	0.047	1.489	1.006–2.205			
Change of height						
Age at PCPC (mo)	0.014	1.066	1.013–1.121	0.012	1.068	1.014–1.124
APS	0.026	1.624	1.058–2.493			
Hospital stay	0.083	1.017	0.998–1.036			
Re-ICU	0.065	1.767	0.966–3.234	0.036	1.940	1.045–3.603
Change of BMI						
Age at TCPC	0.063	0.604	0.355–1.027			
Age at PCPC (mo)	0.070	0.932	0.864–1.006			
TA	0.048	2.010	1.005–4.019	0.032	2.209	1.069–4.564
Fenestration at TCPC	0.045	3.952	1.029–15.174	0.049	4.677	1.005–21.767

Abbreviations: BMI, body mass index; CI, confidence interval; ICU, intensive care unit; OR, odds ratio; PCPC, partial cavopulmonary connection; TCPC, total cavopulmonary connection.



**Fig. 3** Box-and-whisker dot plots showing change of weight for age z-score, height for age z-score, and BMI for age z-score. The top and bottom whiskers mark the minimum and maximum values, the upper and lower borders of the box represent the upper and lower quartiles, and the middle horizontal line represents the median.

The z-score for BMI observed in our series is as expected, given the large improvement in z-scores for weight and the lack of improvement in z-scores for height during the staged palliation. It is of note that the z-scores for BMI are above

normal after the Fontan procedure, which might represent with increased adiposity in the setting of short stature and might also be related with the sedentary lifestyle recently described in this population.<sup>22</sup>

### Impact of Other Influencing Factors for Somatic Development

Our results suggest that reduced somatic growth is seen in patients who required more surgeries before TCPC. Several postoperative complications were also associated with postoperatively reduced somatic growth. The complexity and severity of the underlying diseases requiring more surgeries before Fontan completion and more complications after the Fontan procedure might be the reason why these patients had impaired somatic growth after TCPC.

Systemic ventricular function in patients with univentricular heart might affect the somatic development. Hessel et al reported that single ventricle children with a systemic left ventricle have better weight gain compared with children with a systemic right ventricle.<sup>12</sup> However, we did not note that systemic right ventricular dysfunction at TCPC was associated with impaired growth. Vogt et al demonstrated that venovenous collaterals might impede growth secondary to hemodynamic impairment. Embolization of these collaterals might allow for better growth potential.<sup>9</sup> Anderson et al reported that there were considerable variations in inter-stage growth between patients receiving care at these 16 surgical sites. Standardization of inter-stage nutritional management with focus on best nutritional practices might lead to improved somatic development in this high-risk patients.<sup>10</sup> Williams et al reported that aggressive nutritional support and earlier BCPS are modifiable factors associated with a favorable change in WAZ.<sup>11</sup> Therefore, nutrition should be an important consideration in the staged surgical reconstruction of patients with univentricular heart.<sup>23</sup> In this study, patients undergoing early Fontan completion (before 2 years old) demonstrated better WAZ at TCPC and at follow-up, but they did not demonstrate better acceleration of somatic development, compared with those undergoing late Fontan (>2 years old). The rationale of early TCPC as early as 18 months old should be examined in the further evaluation including protein losing enteropathy, plastic bronchitis, and Fontan-associated liver disease.

### Limitations

There are limitations due to the retrospective nature of this single-center study, and due to the nonstandardized measurements of weight and height. However, measurements were taken at each visit at our institution on standard equipment. The inclusion of only those patients who survived to the time of post-TCPC measurement might be a potential source of bias. Changes in surgical and medical management have occurred with time and might influence the catch-up growth. This study included only the patients who underwent TCPC at the age of 4 years or younger. This highly selected patient inclusion criterion is a limitation. For the postoperative evaluation for somatic development, we evaluated only one point after TCPC in the period of 1 to 3 years. This might cause bias for exact assessment of postoperative somatic development and limits the power of the potential results. This study is lacking in evaluation of hemodynamic criteria, oxygen saturation, medication, activ-

ity, or nutrition data, and they are also limitations. Given the incidence of inter-stage death and failing BCPS physiology, there might be a selection bias for this cohort.

### Conclusion

This study shows that patients with univentricular heart demonstrated a significant catch-up growth in WAZ after TCPC, but not in height-for-age-z-score. Previous BCPS resulted as a significant protective factor for a better somatic development. Several postoperative complications were associated with postoperatively reduced somatic growth.

#### Note

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#### Authors' Contribution

C.B.: conception and design of the work, data collection, analysis and interpretation of the data, drafting the manuscript. H.S.: data collection, drafting the manuscript. C.N.: data collection. T.S.: analysis and interpretation of the data, statistical analysis. M.C.: conception and design of the work. P.P.H.: analysis and interpretation of the data. M.B.: statistical analysis. N.P.: analysis and interpretation of the data. A.H.: critical revision of the manuscript. P.E.: conception and design of the work, critical revision of the manuscript. J.H.: conception and design of the work, critical revision of the manuscript. M.O.: conception and design of the work, drafting the manuscript.

#### Conflict of Interest

None declared.

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