

# Univentricular heart and body growth: are we timing the Fontan stages properly?



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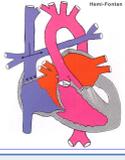
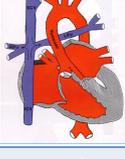
**Objective** The **staged Fontan procedure** for univentricular heart palliation is designed to gradually reduce volume overload and cyanosis during infancy and early childhood. The **optimal timing** of this staging and **its impact on somatic growth** is still a matter of debate. We explored the trends in body growth related to subsequent surgical and interventional procedures and the need for heart failure treatment.

**Methods** We reviewed **64 consecutive patients** that ultimately underwent a total cavopulmonary connection (TCPC) in our centre since 1992. Serial **anthropometric parameters** (weight, height) were recorded from birth to latest follow-up (mean patient follow-up 12,5 ± 6,1 years) and at each intervention (neonatal surgery, bidirectional cavopulmonary anastomosis (BCPA), TCPC, catheter treatment), and converted to z-scores. The influence of oxygen saturation, heart failure treatment and interval between surgeries on body growth was determined.

## Patient characteristics and diagnosis

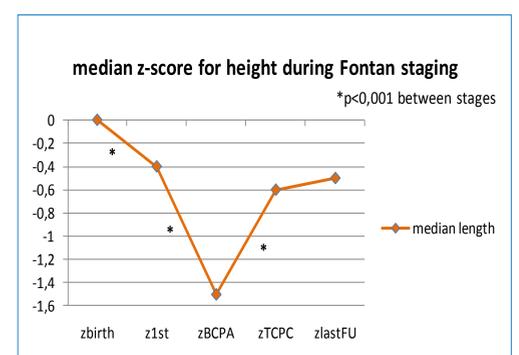
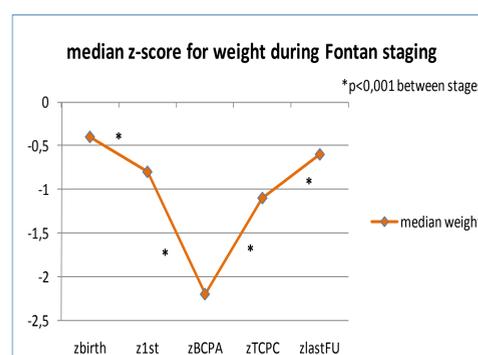
Variable	Value	Missing data
Gender		0
male	31 (48%)	
female	33 (52%)	
Median birth weight (kg)	3,2 (2 – 4,4)	8
Z-score median birth weight	-0,4	
Median birth height (cm)	50 (40 – 54)	8
Z-score mean birth height	0 ± 1	
Weeks of gestation	39,2 ± 1,7	0
Primary diagnosis		0
Tricuspid atresia / stenosis	16 (25%)	
Double outlet right ventricle	12 (18,7%)	
Double inlet left ventricle	12 (18,7%)	
Unbalanced atrioventricular septal defect	6 (9,4%)	
Hypoplastic left heart syndrome	5 (7,8%)	
Pulmonary atresia with intact septum	5 (7,8%)	
Double discordance	5 (7,8%)	
Other atrioventricular connection	3 (4,7%)	
Ventricular dominance		0
Left ventricle	34	
Right ventricle	22	
Balanced ventricular size	8	

## Interventional details

Type of intervention	Variable	Value
First surgery	Number of patients	57 (89%)
	Median age at surgery (days)	20 (0 – 1456)
	Type of procedure (*)	
	Blalock-Taussig shunt	36
	Pulmonary artery banding	14
	Arch repair + shunt	6
	Arch repair + banding	3
	Damus-Kay-Stansel + shunt	2
	Section MAPCA	1
	Associated procedures	10
BCPA	Number of patients	56 (87%)
	Median age at BCPA (months)	11,9 (5,5 – 76)
	Type of procedure	
	Unilateral BCPA	26
	HemiFontan	23
	Bilateral BCPA	7
	Associated procedures (*)	50
TCPC	Number of patients	64 (100%)
	Median age at TCPC (years)	3,2 (2,2 – 18,3)
	Median interval between BCPA and TCPC (years)	2,3 (0,6 – 14,4)
	Type of procedure	
	Lateral tunnel	50 (78%)
	Extracardiac conduit	13 (20,5%)
	Intracardiac conduit	1 (1,5%)
	Fenestration	54 (84%)
Additional interventions	Number of patients	30 (47%)
	Total number of procedures (*)	46

## Parameters of somatic growth

	Birth	First surgery	BCPA	TCPC	Last FU	P-value
Mean weight ± SD (kg)	3,1 ± 0,55	3,8 ± 1,6	8,5 ± 2,6	16,3 ± 7,6	37,5 ± 18	
Z-score mean weight (kg)	-0,3 ± 1,2	-1,3 ± 1,9	-2,1 ± 1,2	-1,2 ± 1,3	-0,8 ± 1,6	<0,001 <sup>a</sup>
Median weight (range)(kg)	3,2 (2-4,4)	3,4 (2,1-11)	7,8 (5,3-17)	14 (9,8-52)	31,5 (17,2-80)	
Z-score median weight (kg)	-0,4	-0,8	-2,2	-1,1	-0,6	<0,001 <sup>a</sup>
Mean height ± SD (cm)	49,2 ± 2,8	53 ± 7	75 ± 12	103 ± 18	142 ± 24	
Z-score mean height (cm)	0 ± 1	-0,9 ± 1,7	-1,6 ± 1,3	-0,7 ± 1,4	-0,7 ± 1,3	<0,001 <sup>b</sup>
Median height (range)(cm)	50 (40-54)	51 (40-81)	70 (59-114)	98 (79-160)	141 (97-193)	
Z-score median height (cm)	0	-0,4	-1,5	-0,6	-0,5	<0,001 <sup>b</sup>

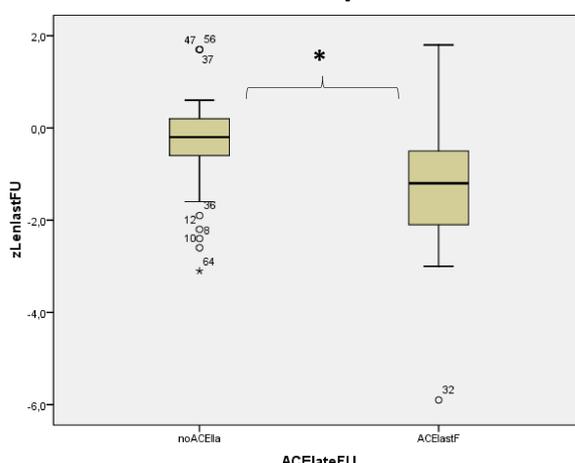


**Results** Median age at BCPA and TCPC was 0.9 (0.4-6.4) and 3.2 (2.2-18.3) years, 8 patients underwent unstaged TCPC after previous neonatal surgery. Median **z-scores for weight** changed significantly after each surgical stage (-0.4 at birth, -0.8 at neonatal surgery, -2.2 at BCPA, -1.1 at TCPC, and -0.6 at latest FU; p<0,001 for change between surgeries), **with the largest decline occurring before BCPA**, and the most marked improvement before and moderate increase after TCPC. **Z-scores for height** showed the same pattern up to the TCPC stage (median 0 at birth, -0.4 at neonatal surgery, -1.5 before BCPA, -0.6 at TCPC; p<0,001 for each interval), **but did not any more improve after TCPC** (median -0.5 at latest FU). Somatic growth at latest FU was **negatively influenced by the need for heart failure treatment** (p<0,05), but not by age at TCPC, ventricular morphology or cyanosis.

## Factors influencing body growth in different stages

Variable	Z-score for weight P-value	Z-score for height P-value
<b>At BCPA</b>		
BCPA before 6 months	0,53	0,37
Previous surgery	0,09	<b>0,04*</b>
<b>At TCPC</b>		
Previous BCPA	0,83	0,51
SaO2 <85%	0,62	0,96
ACE-I at BCPA discharge	0,99	0,48
ACE-I before TCPC	0,46	0,20
TCPC after 4 years of age	0,25	0,34
<b>At last follow-up</b>		
Type of ventricle	0,06	0,06
SaO2 <90% for > 1 year	0,41	0,82
ACE-I at last follow-up	<b>0,04*</b>	<b>0,02*</b>
Additional interventions	0,69	0,39

## Influence of the use of ACE-inhibitors on patient stature at last follow-up



zLenlastFU= median z-score for height at last follow-up  
ACEIlateFU= yes/no use of ACE-inhibitors at last follow-up  
**\*p<0,05**

**Conclusions** Body growth is decreased in patients with univentricular heart treated with a Fontan circulation. The severe growth impairment occurring before BCPA would suggest **advancing this stage** where possible, allowing an earlier start of catch-up growth before TCPC. After TCPC, patients continue to increase z-scores for **weight**, but **stature** remains more impaired than weight, suggesting other influences on bone maturation. **Insidious heart failure** after Fontan completion should be treated promptly, in view of its negative impact on somatic growth.

