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Is there any role for computed tomography imaging in anticipation functional status of adults late after total cavopulmonary connection? Retrospective evaluation

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Short title: Role of computed tomography after total cavopulmonary connection

ABSTRACT

Background: The Fontan procedure is performed in patients with congenital heart diseases and abnormal anatomy which preclude intracardiac repair separating the systemic and pulmonary circulation. The role of computed tomography (CT) imaging in assessing patients after total cavopulmonary connection (TCPC) is not well defined.

Aim: Aim of the study was to determine potential role and capability of CT imaging in functional assessment of adults after TCPC.

Methods: Data of eighteen patients (10 women, mean age 27.9±6.3 years) after TCPC procedure were retrospectively analyzed. All underwent biochemical evaluation, cardiopulmonary exercise test (CPET), transthoracic echocardiography (TTE) and CT examination. In CT study dimensions of the left and right pulmonary artery (LPA and RPA), superior and inferior venae cavae (SVC and IVC), all pulmonary veins and conduit were measured. CT measurements were correlated with TTE, CPET, and biochemical parameters.

Results: The mean time after TCPC was 18.5±6.5 years. The area and circumference of the IVC significantly correlated with the age of patients ($r=0.503, p<0.05$). A significant positive correlation was found between area and circumference of conduit with predicted VO2peak ($r=0.664, p<0.01$). A significant predictor of predicted VO2peak was the area ($\beta=0.746, R2=0.556, p<0.01$) and the circumference ($\beta=0.757, R2=0.572, p<0.01$) of the conduit.

Conclusion: Our study showed considerable association between conduit measurements with the functional status, time after the procedure and age of adult patients who underwent TCPC in childhood. These findings support more extensive use of CT in patients after TCPC examination with measurements of the SVC, IVC and the conduit.

Key words: cardiopulmonary exercise test (CEPT), computer tomography (CT), total cavopulmonary connection (TCPC).

Introduction

Fontan procedure is performed in patients with congenital heart diseases and abnormal intracardiac anatomy which preclude repair separating systemic and pulmonary circulation. This procedure is reserved for patients who cannot undergo biventricular repair for anatomical reasons. The first procedure was performed by Francis Fontan in 1968 [1,2]. Since then, many technical modifications have been implemented, however, the idea of the treatment remains the same. Nowadays, the superior vena cava (SVC) is directly connected to the right pulmonary artery whereas inferior vena cava (IVC) to the pulmonary artery using an intra-atrial lateral tunnel or extracardiac conduit which creates total cavopulmonary connection (TCPC). In these settings pulmonary blood flow is passive and driven mostly by pressure gradient between the systemic veins and the left atrium [3]. Successful procedure leads to decreased chronic volume overload of the single ventricle, normalisation of arterial saturation and improved survival [4,5,6,7]. Despite these obvious advantages, the Fontan circulation is associated with unfavorable late sequelae such as progressive heart failure, thromboembolism, protein losing enteropathy, plastic bronchitis and Fontan-associated liver disease resulting from chronically increased pressure in the systemic veins, disorders of lymphatic flow and increased pulmonary vascular resistance [8,9,10,11]. Therefore, clinical evaluation of these cohort is very complex and requires blood tests, noninvasive imaging, cardiopulmonary exercise test (CPET), and cardiac catheterization [12]. Transthoracic echocardiography (TTE) is the most widely available imaging modality but its capability of the conduits evaluations is substantially limited. Cardiac magnetic resonance imaging (CMR) is the noninvasive method of choice for postoperative evaluation of cardiac anatomy and function in patients after TCPC. It provides comprehensive information about cardiovascular anatomy, regional and global ventricular function, blood flow velocities and volumes [13,14]. In the case of contraindications to CMR or equivocal morphological information, computed tomography

(CT) may be a valuable alternative method, which is capable of assessing morphological abnormalities and complications such as thrombus, or stenosis of the conduit, pulmonary embolism, pulmonary arteriovenous malformations, arterial and venous collaterals [15,16]. Cardiac CT is a method dedicated to a morphological evaluation. Its role in an overall assessment of the patients with Fontan circulation is not well defined.

Therefore the aim of this retrospective analysis is to answer the question whether is there any role for CT imaging in predicting functional status of adults late after surgical total cavopulmonary connection.

Material and methods

Into this retrospective study patients after TCPC procedure were involved who underwent routine follow-up examinations including biochemical evaluation, TTE, CPET and in whom CT imaging was additionally performed between 2012 and 2018 year in Congenital Heart Disease Department in the Institute of Cardiology in Warsaw.

The protocol was approved by the Ethics Committee of the Institute of Cardiology in Warsaw. All patients provided the written informed consent for the CT examination.

Contraindications for CT examination included renal insufficiency, hypersensitivity to iodine-containing contrast material and uncontrolled hyperthyroidism.

The CT examination was performed using dual source computed tomography (DSCT), 7 patients had CT scanning performed using a second-generation DSCT scanner (Somatom Definition Flash Siemens Healthcare, Forchheim, Germany) and 11 patients had CT scanning performed using a third-generation DSCT scanner (Somatom Force Siemens Healthcare).

CT acquisition parameters for the second-generation DSCT were: gantry rotation time 280ms, tube voltage 100-120kV, slice collimation 128x0.6. CT acquisition parameters for third-generation DSCT were: gantry rotation time 250ms, slice collimation 192 x 0.6mm, tube voltage 70-100kV, tube current 320-500 mAs (depending on the patient body mass). A

retrospective or prospective electrocardiographically gated acquisition protocol was used at the operator's discretion.

The CT protocol included arterial and delayed phase scans (the time of scanning was calculated after administration of a bolus of 10-20 ml of a contrast agent). The contrast material (Ultravist 370 Bayer Pharma AG) was administrated through the antecubital vein at a rate of 4-5 ml/s.

The presence of possible conduit complications was evaluated. Subsequently, dimensions of the left and right pulmonary artery, both venae cavae, pulmonary veins and the conduit were measured.

The SVC was measured 1 cm above the anastomosis with the right pulmonary artery (RPA); the IVC was measured 1 cm below the anastomosis with the conduit or intracardiac tunnel; the RPA was measured 1 cm distal to the anastamosis and the left pulmonary artery (LPA) was measured 1 cm distal to the pulmonary trunk. The conduit was measured 2 cm above the anastomosis with the IVC. All measurements were performed orthogonal to the centerline of the vessel. The diameter (long and short axis), the area and the circumference were measured. Every area and circumference were indexed to the body surface area (BSA).

All analyses were performed by a single expert with experience in cardiac CT and training in interpretation of adult congenital heart diseases.

All patients underwent a complete TTE with a GE Medical System Vivid E95 with a 2.5 MHz transducer. The TTE protocol included a complete two-dimensional and Doppler echocardiographic evaluation of cardiac chambers, atrioventricular valves and semilunar valves, pulmonary and systemic veins, great arteries in multiple imaging planes. Univentricular end-diastolic diameter (UVEDD) and volume (UVEDV), as well as wall thickness were measured in the long axis view at the level of base segments. The wall thickness was the arithmetic mean of posterior wall and intraventricular septum. Systolic

function of single (systemic) ventricle was evaluated by ejection fraction (EF) and global longitudinal strain (GLS). EF was calculated using single-plane Simpson formula. For GLS evaluation the analysis of 2D strain was performed offline by manually tracking of the single (systemic) ventricle. GLS was defined as the average of systolic negative strains of 6 segments in the apical four-chamber view.

TTE was performed by a cardiologist highly experienced in the evaluation of congenital cardiac defects.

Each patient underwent CPET with a modified Bruce protocol (Sheffield protocol) to evaluate exercise tolerance. The following parameters were recorded: the time of exercise, heart rate, blood pressure, peak oxygen uptake (VO2peak), the percentage of VO2peak, normalized to age, sex, and weight-based normative values (predicted VO2peak).

Blood samples were taken after overnight fasting. The parameters of laboratory tests - total protein, albumin, N-terminal pro-brain natriuretic peptide (NT pro-BNP), C-reactive protein (CRP), bilirubin, creatinine, hemoglobin were determined by routine assays.

Statistical analysis

The descriptive statistics of nominal variables were presented as absolute numbers and percentages in relation to the entire study group and continuous variables as means (M) and standard deviations (SD). Non-parametric tests were performed (Spearman's rank correlation test) for variables with the non-normal distribution and linear regression for variables with the normal distribution. Tested regression models were assessed for one predictor. $P < 0.05$ was considered statistically significant. The results of the statistical analysis were obtained using SPSS 20.0.

Results

The analysis involved 18 patients (10 women, mean age 27.9±6.5 years) after TCPC procedure. Nine patients had lateral tunnel (Figure 1) and next nine extracardiac conduit (Figure 2).

The mean time from the TCPC was 18.5±6.5 years. Anatomical characteristics of the patients are shown in Table 1.

The most common indication for a CT examination was assessment of the TCPC anatomy prior to cardiac catheterization (10 patients), a suspicion of pulmonary arteriovenous malformations (5 patients), suspicion of thrombosis in the Fontan circulation (3 patients). Biochemical, CPET, TTE and CT parameters are shown in Table 2.

In table 3, the correlation between CT measurements and TTE, CPET, biochemical parameters are shown. The area and circumference of the IVC significantly correlated both with the age ($r=0.503$, $p<0.05$, $r=0.587$, $p<0.01$, respectively) and time after TCPC ($r=0.673$, $p<0.01$, $r=0.701$, $p<0.01$, respectively). A significant predictor of IVC circumference was the patient's age ($\beta=0.665$, $R2=0.429$, $p<0.01$).

A significant positive correlation was found between area and circumference of the conduit and both VO2peak ($r=0.757$, $p<0.01$, $r=0.710$, $p<0.01$ respectively) and predicted VO2peak ($r=0.735$, $p<0.01$, $r=0.664$, $p<0.01$ respectively). A significant predictor of predicted VO2peak was the area ($\beta=0.746$, $R2=0.556$, $p<0.01$) (Figure 3) and the circumference ($\beta=0.757$, $R2=0.572$, $p<0.01$) of the conduit (Figure 4).

The considerable correlations between parameters of the conduit indexed to the BSA and the results of CPET were observed as well (Table 3).

Except for a moderate correlation between the diameter of RPA and a weak correlation between SVC area with GLS no other correlations were found between CT and TTE

parameters. No correlation was found between CT parameters and concentration of NT pro-BNP.

Discussion

Cardiac CT gained a wide acceptance in studying patients with congenital cardiac diseases, though the method is dedicated to a morphological evaluation. Our retrospective study was aimed to establish possible relation between selected CT parameters and functional status of adult late after TCPC.

The morphology and performance of cavopulmonary connections and pulmonary circulation are major determinants of circulation efficiency late following TCPC [10]. A failure of one segment impacts the function of the rest. In present study, we found correlations between dimensions of the IVC and the age of patient and the time after TCPC. It may be speculated that widening of the IVC results from increase in systemic venous pressure secondary to elevated pulmonary vascular resistance, which is part of the unfavorable natural history of the TCPC circulation.

In patients after the TCPC procedure, CPET is a widely acknowledged tool providing information regarding a prognosis including the need of heart transplantation [17,18,19]. Our study revealed a strong positive correlation between dimensions of the conduit and the results of CPET. Area and circumference of the conduit anticipated predicted VO2peak. This finding emphasizes role of proper selection of the initial size of the conduit with respect to the natural patient's growth before the operation [20].

Previous studies have shown that geometry of TCPC and the size of the pulmonary arteries play an important role in short-term and long-term outcomes [21-25]. We found a significant correlation between the diameter of the right pulmonary artery and GLS of the single (systemic) ventricle. This correlation probably results from an influence of the pulmonary vascular system on the function of the single ventricle. However, no correlation was found

between CT parameters and ejection fraction (EF) of the single ventricle, which confirms a limited value of the single-plane Simpson's method used in this group of patients [26]. The Simpson method for calculating left ventricular EF is not applicable to evaluate systolic function of single ventricular due to its complex geometry. In this group of patients, this method may be useful for longitudinal observation of individual patient.

Eindhoven at al. reported significantly higher concentration of NT pro-BNP in patients with Fontan circulation who had moderate to severe impairment of ventricular functions [27]. However, data on the clinical use of brain natriuretic peptides in the Fontan circulation should be interpreted with caution as its levels depend on the type of the Fontan procedure and the degree of the right atrial tissue involvement into the Fontan circulation [28]. Trojnarska et al. reported higher serum concentrations of NT pro-BNT in patients after the Fontan procedure, which did not correlate with CPET [29]. The increased level of NT pro-BNP was also confirmed in the patients in our study. However, we did not found any significant correlation between NT pro-BNP level CT and TTE parameters.

Conclusion

Our study proved association between tunnel measurements and time after procedure, age of patients and revealed relationship with functional status of patients late after TCPC. These findings support routine measurement of SVC, IVC and tunnel in these group patients while studied by CT.

Limitations

This study has several limitations inherent to its retrospective nature and a small number of patients examined in one center. CT is the second-choice modality and was used only due to selected clinical indications. Therefore, we included two types of TCPC (intacardiac and extracardiac) and two types of univentricular morphology (right ventricle and left ventricle) into one group (for example in GLS evaluation).

The study population is heterogeneous regarding the time elapsed after surgery, and initial diagnoses.

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Table 1. Characteristics of patients

Mean age (year)	27.9±6.3
Female/Male (N; %)	10/8; 55.5/44.5
The mean time after TCPC (year)	18.5±6.5
Specification of included patients:	
1. TA, VSD, TGA, PDA, PS-left ventricle morphology-extracardiac conduit; trace AVVI	
2. TA-left ventricle morhology-extracardiac conduit; trace AVVI	
3. TA, PS, LPAHy-left ventricle morphology-intracardiac lateral tunnel; mild AVVI	
4. DORV, MS, PS-right ventricle morphology-intracardiac lateral tunnel; mild AVVI	
5. PS, TGA, LSVC-left ventricle morphology-extracardiac conduit; mild AVVI	
6. PS, TGA, LSVC-left ventricle morphology-extracardiac conduit; trace AVVI	
7. DILV, TGA-left ventricle morphology-intracardiac lateral tunnel; trace AVVI	
8. DILV, TGA-left ventricle morphology-extracardiac conduit; mild AVVI	
9. TA, PS-left ventricle morphology-intracardiac lateral tunnel; moderate AVVI	
10. TA, ccTGA, VSD-left ventricle morphology-extracardiac conduit; mild AVVI, mild systemic-pulmonary collaterals	

11. DORV, TGA, VSD, ASD, CoA-right ventricle morphology-intracardiac lateral tunnel, fenestration; mild AVVI
12. DORV, PS-right ventricle morphology-intracardiac lateral tunnel; moderate AVVI
13. DILV, PS-left ventricle morphology-extracardiac conduit; mild AVVI
14. DORV, PS, AVSD-right ventricle morphology-extracardiac conduit; trace AVVI
15. PS, TGA-left ventricle morphology-extracardiac conduit; trace AVVI
16. HRHS, TD-left ventricle morphology- intracardiac lateral tunnel, fenestration; mild AVVI,
17. TA, PS, PAHy-left ventricle morphology-intracardiac lateral tunnel; mild AVVI
18. TA, PS-left ventricle morphology-intracardiac lateral tunnel; trace AVVI, mild systemic-pulmonary collaterals.

ASD-atrial septal defect; AVSD-atrio-ventricular septal defect; AVVI- atrio-ventricle valve insufficiency; CoA-coarctation of the aorta; DILV-double inlet left ventricle; DORV-double outlet right ventricle; HRHS-hypoplastic right heart syndrome; LPAHy-left pulmonary artery hypoplasia; LSVC-persistent left superior vena cava; PAHy-pulmonary artery hypoplasia; PS-pulmonary stenosis; TA -tricuspid atresia; TD- tricuspid dysplasia; TGA-transposition of the great arteries; VSD-ventricular septal defect;

Table 2. Results of biochemical evaluation and CPET, TTE and CT examination.

Parameters	Mean±SD	Minimum value	Maximum value
Biochemical parameters			
Total protein (g/dl)	7,5±0,9	3,9	8,5
Albumin (g/dl)	4,6±0,8	2,0	5,7

NT pro-BNP (pg/ml)	271,0 (mediana)	52,9	2603,0
CRP (mg/dl)	0,6±0,7	0,1	2,2
Total bilirubine (mg/dl)	1,5±1,2	0,5	4,5
Creatynine (mg/dl)	0,9±0,2	0,5	4,5
Hemoglobin (mg/dl)	14,7±2,3	8,7	18,0
CPET			
V02peak (ml/kg/min)	20,1±7,7	9,4	38,1
Predicted VO2peak (%)	48,4±16,2	21,0	76,0
RER	1,0±0,1	0,8	1,1
HR max (beats/min)	133±35,7	85,0	195,0
Predicted HR max (%)	67,2±17,5	43,0	96,0
TTE			
UVEDD (mm)	55,0 ±10,0	42,0	75,0
UVEDV (ml)	100,7±52,9	52,9	220,0
Wall thickness (mm)	10,1 ±1,7	7,8	13,0
UVEF (%)	50,3±10,5	25,0	70,0
GLS	-17,2±5,1	-3,5	-23,8
SVC Vmax (m/s)	0,5±0,2	0,3	1,0
IVC Vmax(m/s)	0,5±0,2	0,3	1,1

CT			
SVC area (cm2)	2,2±1,2	1,0	5,6
SVC circumference (cm)	5,3±1,2	3,8	8,6
IVC area (cm2)	7,7±3,5	2,8	15,2
IVC circumference (cm)	9,9±2,1	6,4	14,2
Conduit area (cm2)	3,5±2,2	1,0	8,2
Conduit circumference (cm)	6,6±1,9	3,9	10,3
RPA area (cm2)	2,4±1,0	1,0	5,0
RPA circumference (cm)	5,7±1,0	4,0	8,4
LPA area (cm2)	2,2±0,9	0,9	4,0
LPA circumference (cm)	5,5±1,1	3,7	7,4

CPET-cardiopulmonary exercise test; CRP- C-reactive protein; CT- computed tomography; GLS- global longitudinal strain; IVC- inferior vena cava; LPA- left pulmonary artery; NT pro-BNP- N-terminal pro-brain natriuretic peptide; predicted VO2peak- the percentage predicted, normalized to age, sex, and weight-based normative values; RPA- right pulmonary artery; SD-standard deviation; SVC- superior vena cava; TTE- transthoracic echocardiography; UVEDD- univentricular end-diastolic diameter; UVEDV- univentricular end-diastolic volume; UVEF-univentricular ejection fraction; V02peak- peak oxygen uptake.

Table 3. Correlation between CT parameters and TTE, CPET, biochemical parameters**. Statistically significant p-value is bold and marked '*'.**

	SVC area	SVC circ.	IVC area	IVC circ.	Conduit area	Conduit circ.	Conduit area BSA indexed	Conduit circ. BSA indexed	RPA area	RPA circ.	LPA area
UVEF	-0,094 (p=0,702)	-0,172 (p=0,482)	-0,201 (p=0,409)	-0,091 (p=0,710)	0,384 (p=0,116)	0,344 (p=0,162)	0,380 (p=0,116)	0,460 (p=0,055)	-0,415 (p=0,085)	-0,415 (p=0,077)	-0,159 (p=0,517)
GLS	**0,482 (p<0,05)***	0,342 (p=0,151)	-0,190 (p=0,435)	-0,126 (p=0,606)	-0,247 (p=0,324)	-0,256 (p=0,305)	-0,110 (p=0,654)	-0,010 (p=0,976)	**-0,659 (p<0,01)***	**-0,594 (p<0,01)***	-0,056 (p=0,821)
VO2peak	0,097 (p=0,770)	-0,062 (p=0,820)	0,197 (p=0,464)	0,229 (p=0,393)	**0,757 (p<0,01)***	**0,710 (p<0,01)***	**0,640 (p=0,01)***	**0,550 (p<0,05)***	0,168 (p=0,535)	0,209 (p=0,438)	-0,047 (p=0,863)
predicted VO2peak	0,222 (p=0,408)	0,149 (p=0,582)	0,308 (p=0,246)	0,371 (p=0,157)	**0,735 (p<0,01)***	**0,664 (p<0,01)***	**0,730 (p<0,01)***	**0,650 (p<0,01)***	0,163 (p=0,545)	0,199 (p=0,460)	-0,222 (p=0,408)
Albumin	-0,116 (p=0,636)	-0,197 (p=0,418)	-0,215 (p=0,377)	-0,239 (p=0,325)	0,058 (p=0,819)	0,139 (p=0,583)	0,360 (p=0,156)	**0,530 (p<0,05)***	0,189 (p=0,438)	0,176 (p=0,472)	0,363 (p=0,127)

NT pro-BNP	-0,198 (p=0,416)	-0,063 (p=0,797)	0,065 (p=0,792)	0,004 (p=0,989)	-0,195 (p=0,438)	-0,177 (p=0,483)	-0,390 (p=0,108)	-0,350 (p=0,154)	-0,119 (p=0,627)	-0,057 (p=0,817)	-0,144 (p=0,557)
CRP	-0,161 (p=0,509)	-0,003 (p=0,991)	0,072 (p=0,770)	0,169 (p=0,488)	0,154 (p=0,542)	0,075 (p=0,766)	0,300 (p=0,231)	0,240 (p=0,334)	0,104 (p=0,670)	0,184 (p=0,450)	-0,389 (p=0,100)
Total bilirubin	-0,298 (p=0,229)	-0,366 (p=0,135)	0,321 (p=0,194)	0,189 (p=0,453)	0,135 (p=0,606)	0,217 (p=0,403)	-0,150 (p=0,565)	-0,160 (p=0,539)	0,292 (p=0,240)	0,295 (p=0,234)	0,254 (p=0,309)
Age	0,167 (p=0,494)	0,380 (p=0,108)	**0,503 (p<0,05)***	**0,587 (p<0,01)***	0,436 (p=0,070)	0,346 (p=0,160)	0,350 (p=0,159)	0,160 (p=0,524)	0,245 (p=0,313)	0,246 (p=0,311)	-0,307 (p=0,201)
Time after TCPC	0,059 (p=0,810)	0,112 (p=0,648)	**0,673 (p<0,01)***	**0,701 (p<0,01) ***	0,311 (p=0,209)	0,217 (p=0,387)	0,010 (p=0,954)	0,0,20 (p=0,949)	0,301 (p=0,211)	0,197 (p=0,419)	0,092 (p=0,708)

BSA-body surface area; TCPC- total cavopulmonary connection; Other abbreviations — see Table 2.

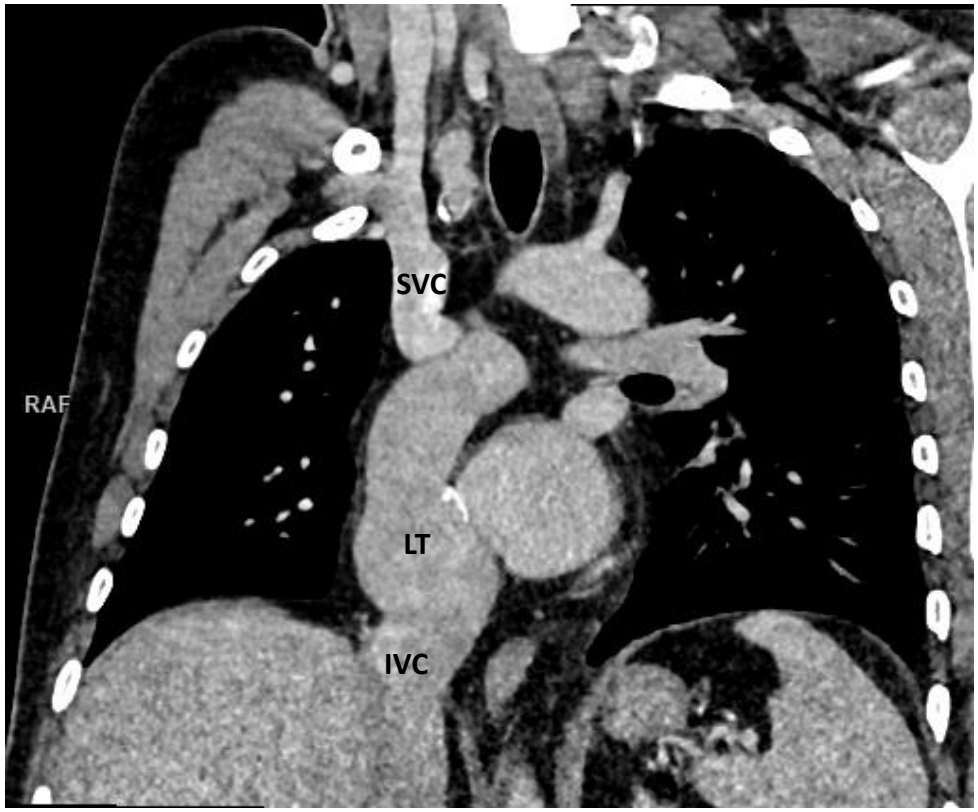

Figure 1. Computed tomography, coronal reformatted image – TCPC lateral tunnel

LT- lateral tunnel, SVC- superior vena cava

IVC – inferior vena cava

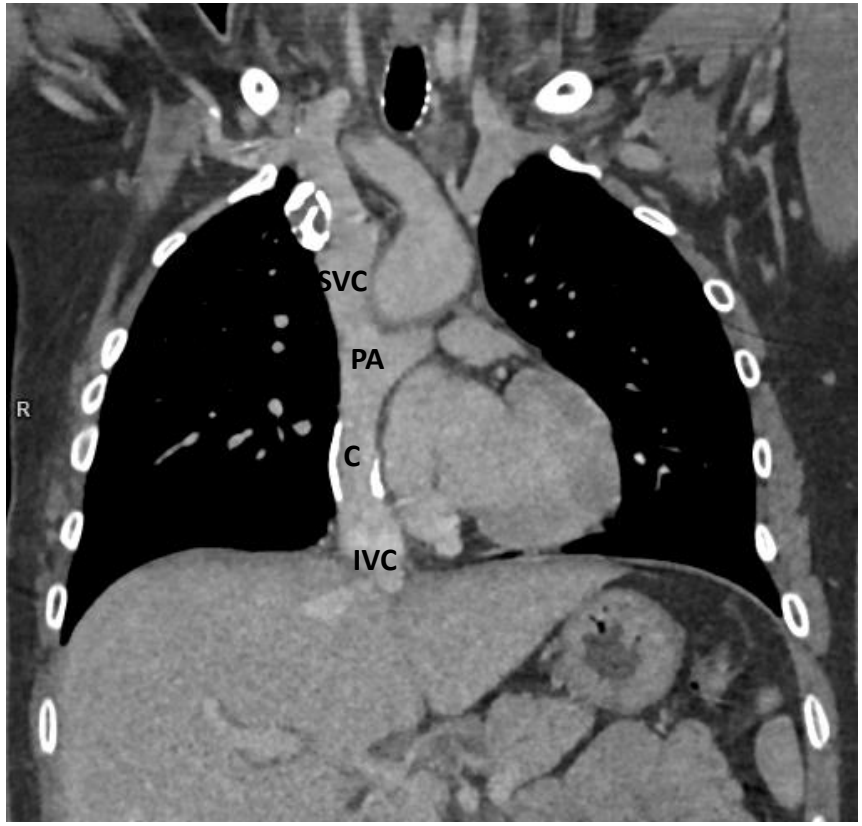

Figure 2 .Computed tomography, coronal reformatted image – TCPC extracardiac conduit

C- conduit, SVC- superior vena cava, IVC – inferior vena cava, PA- pulmonary artery

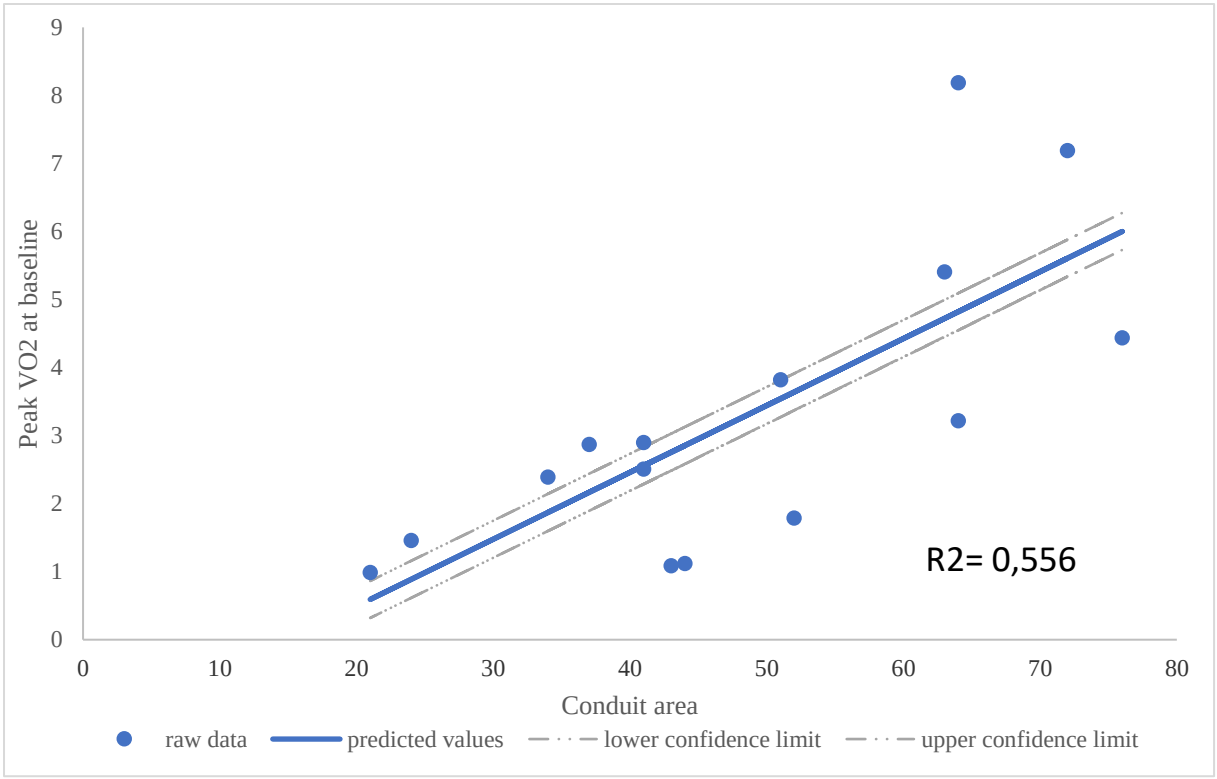

Figure 3. Linear regression predictive model of conduit area with peak V02 at baseline.

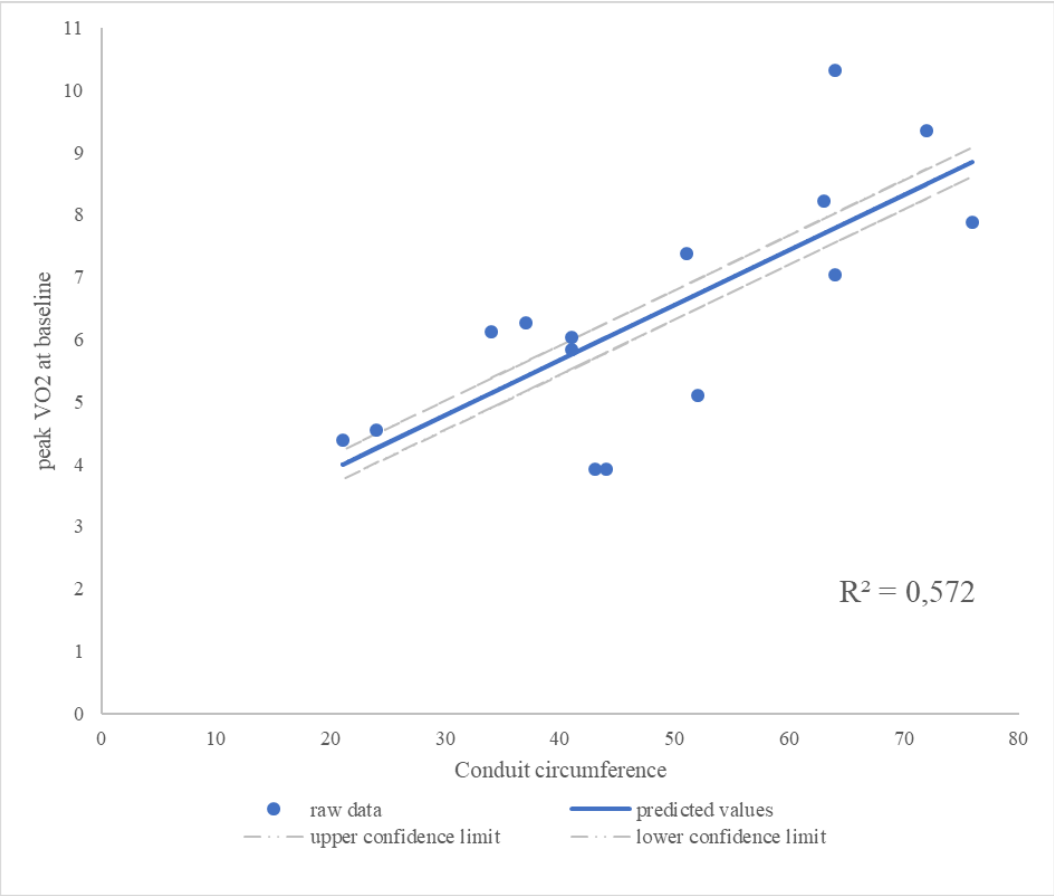

Figure 4. Linear regression predictive model of conduit circumference with peak V02 at baseline.