

# Exercise performance in fontan patients – a single-center Bulgarian study

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## Abstract

**Aim:** Exercise capacity in Fontan patients is reduced compared to healthy peers and progressive decline in cardiovascular function is expected over time. The purpose of this study is to present, analyze and discuss the initial Bulgarian experience with exercise performance of Fontan patients and to compare them to age-matched healthy peers. Additionally, we focused on understanding whether peak oxygen consumption can be predicted based on multiple independent variables.

**Materials and methods:** The research is a retrospective observational study of 11 patients aged 8y-18y, who had undergone the Fontan procedure and underwent cardiopulmonary exercise testing in National Heart Hospital in Sofia, Bulgaria. Data were collected for a 1-year period. Simultaneously, 11 age-matched healthy patients were selected for comparison.

**Results:** Patients with Fontan palliation had lower peak oxygen consumption (23.1 ml/kg/min vs. 30.1 ml/kg/min,  $P=0.046$ ), percent-predicted peak oxygen consumption (52% vs. 76%,  $P<0.001$ ), percent-predicted peak work (56% vs. 82%,  $P<0.001$ ), percent-predicted peak oxygen consumption at ventilatory anaerobic threshold (37% vs. 48%,  $P=0.04$ ), resting saturation (93% vs. 97%,  $P<0.001$ ) and saturation at peak work (84% vs. 95%,  $P<0.001$ ). Male subjects and patients with left ventricular dominance exhibited a greater peak oxygen consumption.

**Conclusion:** Exercise capacity in the Bulgarian Fontan cohort is reduced relative to healthy controls. Male gender and subjects with left ventricular dominance have a greater peak oxygen consumption.

## Keywords

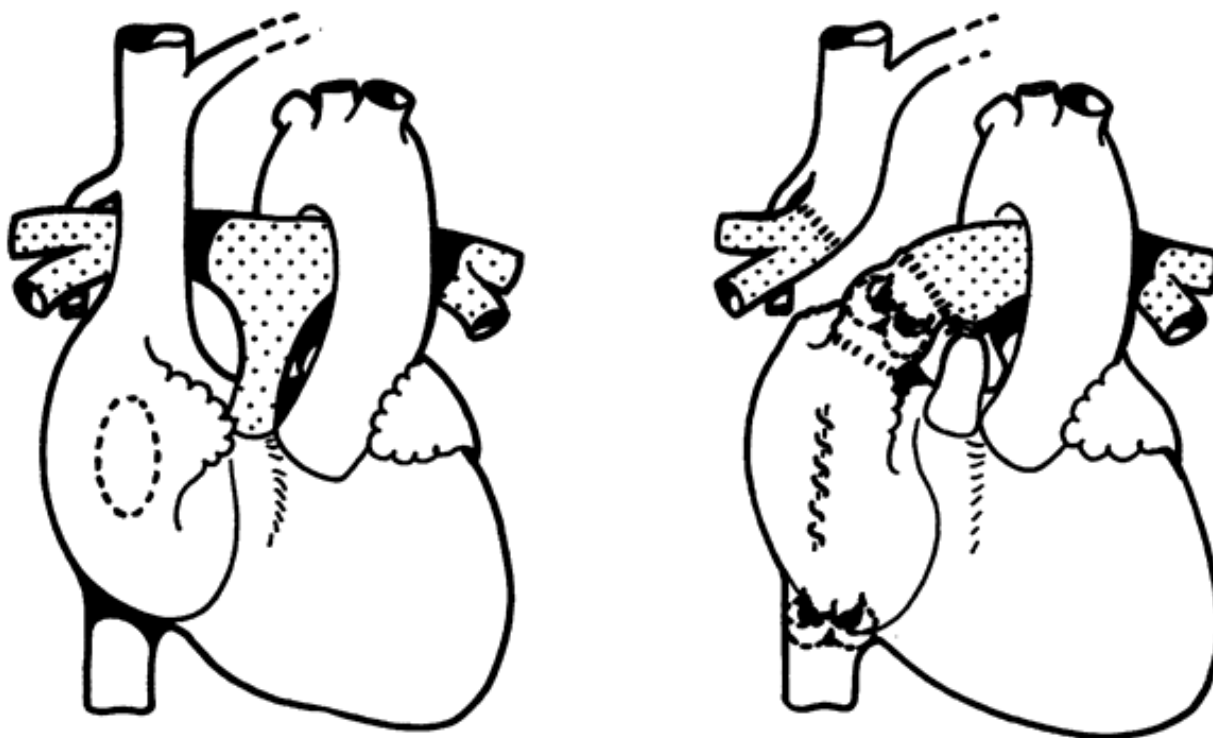
Cardiopulmonary exercise testing, exercise performance, fontan procedure

## Introduction

The Fontan operation is the last of series of operations for a group of congenital cardiovascular anomalies in which it is not possible to maintain two-ventricle physiology [1–2]. The procedure results in separating the systemic and pulmonary circulation by creating a total cavo-pulmonary connection [3–4]. Pulmonary blood flow in Fontan circulation is passive and its adequacy depends on low impedance to passively forward flow and to lower pulmonary vascular resistance

(PVR). This newly constructed system is characterized by elevated central venous pressure and chronically low cardiac output (Fig. 1) [3]. Long term sequelae of the Fontan physiology is progressive decline in cardiovascular function [5].

Deterioration of cardiovascular efficiency leads to reduced exercise capacity. Contributing factors to exercise impairment include failure to increase stroke volume, chronotropic incompetence and reduced arterial blood saturation. Cardiopulmonary exercise testing (CPET) has an



**Figure 1.** Schematic image of Fontane Procedure [3].

important role in the management of patients after Fontan operation. Peak oxygen uptake (peak  $\text{VO}_2$ ) is a significant index of exercise capacity and has a prognostic value in patients with congenital heart disease [6]. Patients after Fontan procedure have percent predicted peak  $\text{VO}_2$  from 60–65% [7–8]. Fernandes et al. reports that the hazard for death for patients with peak  $\text{VO}_2$  less than 16.6 ml/kg/min was 7.5 times higher, compared to patients with higher peak  $\text{VO}_2$  [9]. Worsening of peak  $\text{VO}_2$  over time predicts poorer outcomes including death, transplant, and unscheduled cardiac-related rehospitalization [10–11].

The purpose of this study was to present the initial Bulgarian experience with exercise performance of Fontan patients and compare them to their healthy peers. This paper also focuses on understanding whether peak  $\text{VO}_2$  can be predicted based on multiple independent variables.

## Materials and methods

### Patient characteristics

A retrospective observational study was conducted in Fontan patients referred to the Pediatric Cardiology Department in National Heart Hospital in Sofia, Bulgaria. This is the only center in the country involved in the surgical treatment of congenital heart defects. Patients aged 8y-18y with functionally univentricular heart who had undergone the Fontan procedure and had CPET were investigated. Exclusion criteria from the study were patients less than 8 or above 18 years of age and a submaximal effort test. Data were collected over

a one-year period, from August 2021 to August 2022, involving the patients that met the inclusion criteria (11 out of 19 patients). Simultaneously, 11 age-matched healthy patients from outpatient visits were selected for comparison. Additional factors assessed included ventricular morphology (right, left), ventricular dysfunction and more than moderate atrioventricular valve regurgitation, evaluated by echocardiography. The Ethics Committee in our institution approved the study protocol and waived the need for patient consent due to the study's retrospective nature.

### Primary and secondary outcomes

The authors reviewed patients' medical files for baseline characteristics and anatomic details. We defined the primary outcome of interest: peak  $\text{VO}_2$ , a direct measurement of cardiorespiratory fitness [12]. Secondary outcomes included peak work rate, peak heart rate, peak oxygen pulse, oxygen consumption ( $\text{VO}_2$ ) at ventilatory anaerobic threshold (VAT), minute ventilation/carbon dioxide production slope ( $\text{VE}/\text{VCO}_2$  slope) at VAT, oxygen saturation at rest and at peak exercise. Peak  $\text{VO}_2$ ,  $\text{VO}_2$  at VAT, work rate and heart rate are also expressed as a percentage of predicted normal values in order to eliminate sex, height and weight differences [13–14].

### Echocardiography

Transthoracic echocardiography was performed using Philips Epic 7c ultrasound system with X5-1 transducer. We graded atrioventricular valve regurgitation (AVVR) according to the American Society of Echocardiography recommended crite-

ria: none to trivial, 0; mild, 1; mild to moderate, 2; moderate to severe, 3; and severe, 4 [15]. Ventricular dysfunction was assessed by the modified Simpson method using 4 chamber frames at end systole and end diastole, and was classified as follows: mildly abnormal, 41–51%; moderately abnormal, 30–40%; severely abnormal, <30% [16–17].

### Cardiopulmonary exercise testing

All patients underwent CPET on an electrically braked cycle ergometer (Lode Corival; Lode BV, Groningen, The Netherlands). Meanwhile, VO<sub>2</sub> was measured using a commercial, breath-by-breath gas analysis system (Ultima-CPX, MGC Diagnostics Corp., St. Paul, Minnesota). A RAMP protocol was used, in which the patient should maintain a pedaling speed of about 60/minute. After an initial warm-up period of 2–3 minutes, the pedal resistance is gradually increased at a rate, based on the patient’s age, weight, gender, and level of fitness so that they reach peak exertion in 8–12 minutes. Anaerobic threshold was calculated using the V-slope method (170029). Oxygen pulse - a surrogate for stroke volume - was measured by dividing absolute oxygen uptake by heart rate. Also, VE/VCO<sub>2</sub> slope was assessed at VAT. Blood pressure was measured every 2 minutes with an automatic cuff and a 12-lead ECG was obtained throughout the whole exercise. A maximal effort test was defined as peak respiratory exchange ratio (RER)<sup>3</sup> 1.05.

### Statistical analysis

The authors used descriptive statistics to provide frequencies and percentages for categorical variables and medians, and interquartile ranges for continuous variables. We used the students t-test to compare variables between Fontan patients and healthy patients. Normality was checked based on the normal Q-Q plot and histogram. Gender, age at Fontan, age at CPET, ventricular dominance, pulmonary vasodilator therapy, more than moderate AVVR and ventricular dysfunction were assessed for linearity with peak VO<sub>2</sub>. Out of the 7 independent variables only gender and ventricular dominance were significantly linearly related. A multiple regression was run to predict peak VO<sub>2</sub> from gender and ventricular dominance. A value of P<0.05 indicated statistical significance. We used IBM SPSS Statistics 24 (IBM Corp., Armonk, N.Y., USA) for statistical analysis.

## Results

### Study group characteristics

We had 19 Fontan patients who were referred to the Pediatric Cardiology Department in National Heart Hospital in Sofia, Bulgaria between August, 2021 and August, 2022. A total of 11 patients were included in the study, regarding the including criteria. Fontan patients’ characteristics are summarized in Table 1.

**Table 1.** Fontan patients’ characteristics.

Variable	Fontan (N=11)
Male (n, %)	9 (82%)
Body mass index (BMI), median (range)	19.8 (15.1–22.6)
Age at CPET (years), median (range)	17 (11–17.9)
Diagnosis (n, %):	
• Tricuspid atresia	2 (18.2%)
• Pulmonary atresia/intact ventricular septum	3 (27.3%)
• Double outlet right ventricle	1 (9.1%)
• Complex transposition of the great arteries	2 (18.2%)
• Double inlet left ventricle	3 (27.3%)
Dominant left ventricle (n, %)	7 (63.6%)
More than moderate AVVR (n, %)	1 (9.1%)
Ventricular dysfunction (n, %)	
• Mild	4 (36.4%)
• Moderate	0
• Severe	0
Age at Fontan completion (years), median (range)	3.3 (2.9–4.8)
Conduit size (n, %):	
• 16 mm	3 (37.3%)
• 18 mm	6 (54.5%)
• 19 mm	1 (9.1%)
• 20 mm	1 (9.1%)
Fenestration (n, %)	3 (27.3%)
Pulmonary vasodilator (n, %)	4 (36.4%)

\* Abbreviations: CPET – cardiopulmonary exercise testing; AVVR - atri-ventricular valve regurgitation.

It is accurate to note that Only 1 patient (9.1%) had more than moderate AVVR and none of the observed patients had neither moderate, nor severe ventricular dysfunction and this may explain the fairly preserved numbers compared to larger registries. Hereby, we should mention that low burden of ventricular dysfunction in this group might bias results toward better exercise outcomes.

All of the single ventricle patients had an extracardiac Fontan procedure. Comparison between Fontan patients and healthy subjects is presented in Table 2. Age at CPET, body mass index and gender distribution were similar between the two groups for the purposes of objectivity.

**Table 2.** Comparison between Fontan and healthy patients.

Variable	Healthy patients (N = 11)	Fontan patients (N = 11)	P
Male (%)	7 (63.6)	9 (82)	0.35
Body mass index (BMI), median (range)	19.3 (14.7–22.9)	19.8 (15.1–22.6)	0.89
Age at CPET (years), median (range)	14.1 (9.4–17.2)	17 (11–17.9)	0.13

\* Abbreviations: CPET - cardiopulmonary exercise testing.

**Table 3.** Cardiopulmonary exercise testing parameters between the two groups.

Variables	Healthy patients (N = 11)	Fontan patients (N = 11)	p	Mean difference	95% CI	
					LL	UL
Ramp protocol (W), median (range)	15 (10–20)	15 (10–20)	0.71	0.45	-2.326	3.235
Peak work (W), median (range)	126 (77–186)	125 (45–189)	0.51	18.20	-16.35	52.71
Percent-predicted peak work (%), median (range)	82 (60–100)	56 (27–75)	<0.001	23.73	11.632	35.822
Peak oxygen consumption (ml/kg/min), median (range)	30.1 (19.4–42.3)	23.1 (15.2–40.3)	0.046	5.73	-0.589	12.043
Percent-predicted peak oxygen consumption (%), median (range)	76 (65–100)	52 (31–91)	<0.001	27.20	13.690	40.672
Peak RER, median (range)	1.16 (1.05–1.51)	1.24 (1.06–1.30)	0.53	-0.12	-0.103	0.101
Peak oxygen pulse (%), median (range)	86 (48–98)	63 (53–108)	0.21	7.73	-8.328	23.783
Peak heart rate (bpm), median (range)	176 (149–195)	162 (103–196)	0.09	16.10	-2.625	34.807
Percent-predicted peak heart rate (%), median (range)	84 (71–96)	80 (49–96)	0.12	7.10	-2.456	16.638
Percent-predicted peak oxygen consumption at VAT (%), median (range)	48 (28–65)	37 (25–60)	0.04	10.23	0.385	20.069
VE/VCO <sub>2</sub> at VAT, median (range)	28 (25–31)	32 (21–41)	0.14	-3.10	-7.083	0.901
Resting saturation (%), median (range)	97 (96–99)	93 (78–96)	<0.0001	6.45	2.800	10.108
Saturation at peak exercise (%), median (range)	95 (93–98)	84 (70–94)	<0.0001	10	5.433	14.566
ECG abnormalities	0	0	0.98	0	-0.872	0.145
Abnormal blood pressure response	0	1	0.32	0	-0.090	0.281

\* Abbreviations: RER - respiratory exchange ratio; VAT - ventilatory anaerobic threshold; VE/VECO<sub>2</sub> - ratio of minute ventilation to carbon dioxide production; LL - lower limit; UL - upper limit; p – statistical significance.

### Cardiopulmonary exercise testing parameters between the two groups

All of the study subjects achieved maximal effort test. Exercise capacity in our Fontan cohort was reduced compared to healthy peers - patients after Fontan palliation had lower peak VO<sub>2</sub> (23.1 ml/kg/min vs. 30.1 ml/kg/min, P=0.046), percent-predicted peak VO<sub>2</sub> (52% vs. 76%, P<0.001), percent-predicted peak work (56% vs. 82%, P<0.001), percent-predicted peak VO<sub>2</sub> at VAT (37% vs. 48%, P=0.04), resting saturation (93% vs. 97%, P<0.001) and saturation at peak work (84% vs. 95%, P<0.001). None of the patients exhibited ECG abnormalities and one Fontan patients had an abnormal blood pressure response. Comparison of CPET parameters between the two groups is demonstrated in Table 3.

### Predicting peak oxygen consumption

A multiple regression was run to predict peak VO<sub>2</sub> from gender and ventricular dominance. The multiple regression model statistically significantly predicted peak VO<sub>2</sub>, F (2, 8) = 12.998, P = 0.003, adj. R<sup>2</sup> = .71. Both gender and

**Table 4.** Multiple regression results for peak oxygen consumption.

Variables	B	SE B	P	95% CI for B	
				LL	UL
Constant	8.712	3.394	0.033	0.886	16.539
Gender	11.925	2.936	0.004	4.925	18.925
Ventricular dominance	7.738	2.036	0.005	3.041	12.434

\* Abbreviations: B – unstandardized regression coefficient; SE B – standard error of the coefficient; CI – confidence interval; LL – lower limit; UL – upper limit; p – statistical significance.

ventricular dominance added statistically significantly to the prediction, P=0.003. Male Fontan patients exhibited higher peak VO<sub>2</sub> of 11.925 ml/kg/min. Peak VO<sub>2</sub> for patients with left ventricular dominance was 7.738 ml/kg/min, greater than patients with right ventricular dominance. Regression coefficients and standard errors can be found in Table 4.

### Discussion

Patients with functionally univentricular heart survive satisfactory into adulthood after Fontan completion [18–19]. The creation of such an unconventional circulation results in improved exercise capacity compared to preoperative state, but it is still reduced in comparison with healthy subjects. Patients after the Fontan procedure are expected to have a decline in cardiovascular function over time with subsequent decrease in exercise performance [5]. This leads to poorer outcomes, including death, transplant, and unscheduled cardiac-related rehospitalization. [10–11].

Cardiopulmonary exercise testing (CPET) is a crucial tool to objectively assess exercise performance and functional capacity in patients with congenital heart disease. Access to CPET in Bulgaria is not routine and scarce experience has been gained in recent years. This method provides objective data for assessing functional limitations, tracking disease progression, and guiding personalized treatment strategies. By monitoring objective metrics over time, CPET helps tailor exercise prescriptions, optimize timing for interventions, identify early signs of complications, and guides the creation of personalized, safe exercise programs to improve long-term outcomes.

This study documents that in practice, exercise capacity in Bulgarian Fontan patients is reduced compared to healthy peers. The performed multiple regression analysis reveals that male patients and patients with left ventricular dominance have a greater peak  $VO_2$ .

We demonstrated that peak  $VO_2$ , as well as  $VO_2$  at VAT and percent-predicted peak work vary significantly between Fontan patients and healthy subjects. These findings extend those of others [7–8, 20–22], confirming that exercise capacity is impaired in Fontan patients. Known causes of depressed exercise function may include: inability to increase stroke volume normally in response to exercise; chronotropic incompetence; arterial desaturation; pulmonary abnormalities; deconditioning [23]. Interestingly, peak oxygen pulse, percent-predicted peak heart rate and  $VE/VCO_2$  slope were similar between the two studied groups.

In accordance with previous findings [24–28], anatomic morphology may be associated with difference in exercise performance. Fontan patients with left ventricular dominance had greater peak  $VO_2$  than patients with right ventricular dominance. It would seem reasonable that a systemic right ventricle would less likely adapt to hemodynamic changes, leading to progressive decline in cardiovascular function and exercise intolerance. Dominant right ventricle morphology is associated with poorer outcomes on a long-term follow up [29].

There are limited data on patients with Fontan physiology regarding exercise parameter difference between males and females. Seckler and colleagues previously reported that peak  $VO_2$  decreased by 32% through adolescence in females and did not have the typical increase for adolescence for males [30]. Our study reveals that male patients have a higher peak  $VO_2$ .

## Study limitations

Some limitations are worth noting. A RAMP protocol was used via cycle ergometer, which means that obtained peak  $VO_2$  is lower than uphill treadmill running. Cycle ergometry was used mainly because of the relatively young patients age. Another limitation is the single-center, retrospective, non-randomized design of the study. Also, the small sample size in this research may limit the ability to observe significant differences.

## Conclusion

Cardiopulmonary exercise testing has an important role in management of patients after Fontan operation. Our current data on the topic provide the first description of exercise capacity in Bulgarian Fontan patients. Our initial experience revealed that patients after the Fontan procedure have impaired exercise performance compared to healthy peers. Male gender and left ventricular dominance are associated with greater peak  $VO_2$ . These findings address the need for serial cardiopulmonary exercise testing in Fontan patients to monitor for declining peak oxygen

consumption. Future multicenter collaborations with larger Fontan cohorts are needed for better understanding exercise capacity trajectories and identifying factors associated with outcomes over time.

## Additional information

### Conflict of interest

The authors have declared that no competing interests exist.

### Ethical statements

The authors declared that no clinical trials were used in the present study.

The authors declared that no experiments on humans or human tissues were performed for the present study.

The authors declared that no informed consent was obtained from the humans, donors or donors' representatives participating in the study.

The authors declared that no experiments on animals were performed for the present study.

The authors declared that no commercially available immortalised human and animal cell lines were used in the present study.

### Use of AI

No use of AI was reported.

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No funding was reported.

### Author contributions

All authors have contributed equally.

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### Data availability

All of the data that support the findings of this study are available in the main text or Supplementary Information.

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