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ORIGINAL PAPER

Body plethysmography – additional information on exercise capacity in patients with congenital heart disease?

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Abstract

Aims In each cardiopulmonary exercise test (CPET), resting spirometry is performed in advance. In patients with a congenital heart defect (CHD), lung volumes are often impaired. This study investigates correlations between lung volumes and CPET parameters and determines, whether body plethysmography provides substantial additional information for these patients.

Methods Data from 102 patients (23.8 ± 10.4 years of age, 39 female) with various forms of CHD were examined from April 2018 to October 2022. All patients underwent spirometry (measuring forced vital capacity, FVC and forced expiratory volume in 1 s, FEV1), body plethysmography (measuring total lung capacity, TLC) and an exhausting CPET. Data is presented as the median and interquartile range (z-scores) and correlated with Spearman's rho.

Results Fifty-five% of all patients had normal results in lung function and 45% had normal peak oxygen uptake (≥ 80% predicted in peak VO_2). Patients with impaired lung function were significantly more likely to have low exercise capacity (Fisher's exact test: $p = 0.028$). FVC z-values and %predicted peak VO_2 ($r = 0.365$, $p < 0.001$) correlated significantly as well as FEV1_z and %predicted peak VO_2 ($r = 0.320$, $p = 0.001$), and TLC z-values and %predicted peak VO_2 ($r = 0.249$, $p = 0.012$). No correlation was found between FEV1/FVC z-values and %predicted peak VO_2 ($r = -0.043$, $p = 0.670$).

Conclusion Spirometry and exercise capacity positively correlate, also in CHD patients. However, body plethysmography does not provide additional or improved prediction and is therefore only recommended in noteworthy results in spirometry to exclude further lung co-morbidities.

Keywords Lung function · Exercise capacity · Body plethysmography · Congenital heart defect

Introduction

Nowadays, patients with a congenital heart defect (CHD) reach not only adolescence but also adulthood [1] and, therefore, research is facing "new" questions. Survival is no longer the main goal; patients' quality of life, exercise capacity and participation in daily life are now of high interest [2–14]. Additionally, co-morbidities or further notable results in patients with CHD occur more often [8].

In this prospective study, lung function and volumes were studied. Previous studies have already shown not only in COPD [15], but also in CHD, that, depending on the type of CHD, one-third to half of all patients have noteworthy results in spirometry [16, 17].

However, next to spirometry, body plethysmography (investigating total lung capacity, TLC and residual volume, RV) may explain restrictive results in spirometry and may be advisable.

This study therefore aims to (I) describe lung volumes in CHD patients, measured by spirometry and body plethysmography with the latest reference; (II) correlate these results with parameter of cardiopulmonary exercise test. Since body plethysmography is performed fewer in this cohort, the (III) whether body plethysmography even provide (better) information on CHD patients' exercise capacity?

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Materials and methods

The study was registered at the "Deutsches Register für Klinische Studien" (DRKS00021120) and approved by the ethical committee of the Technical University of Munich (project number: 110/19 S).

Study subjects

From April 2018 to October 2022 a total of 102 children and adults (23.8 ± 10.4 years of age, 39 female) with various types of CHD underwent spirometry, body plethysmography and a symptom-limited cardiopulmonary exercise test (CPET) at the German Heart Center of Munich. CPET was performed due to regular follow-up. For lung function test (body plethysmography), since it is not part of the routine, all participants signed informed consent. Table 1 shows the anthropometric data and underlying CHD classification.

Lung function and volumes

All tests were performed with the HDpft 2000 and were assessed using the software KoKo Px v7, following the latest ATS/ERS guidelines [18] in both, spirometry and body plethysmography.

Patients first underwent spirometry, and after at least two similar and valid tests were obtained, body plethysmography was performed as described in the manual. This involved having the patient breathe at the functional residual volume for at least six breaths, followed by closing a shutter to assess the volume of thoracic gas (VTG). After three breaths, the shutter automatically opens and the patient performs one normal breath before taking maximal inspiration and expiration (to assess inspiratory and expiratory capacity). Total lung capacity was calculated using the expiratory vital capacity method: average functional residual capacity (FRC) + average inspiratory capacity (IC), which is equal to residual volume (RV) + maximum ventilatory capacity (VC). At least two similar and valid tests were required.

Classification of abnormal results was done following Stanojevic et al. [18]:

- FVC and FEV1 z-scores < -1.645 with normal FEV1/FVC defined as a preserved ratio impaired spirometry (PRISm)

  If TLC in body plethysmography z-score < -1.645 the pattern is restrictive.
  If TLC is normal, it is a non-specific pattern.

- Normal FVC and ≥ FEV1 z-scores but FEV1/FVC z-score < -1.645 is an obstructive pattern.

Table 1 Study Subjects

CHD	*Female*	*Children*	*Age**	*BMI**	*Surgical intervention [n/%]*	*no surgical or transcatheter intervention***	*comorbidity***	*lung comorbidity****
All (n = 102)	39	34	25.3 [15.8;31.3]	21.7 [19.5;24.7]	83 (81%)	14 (14%)	33 (32%)	6 (6%)
Left heart obstruction (n = 11)	5	2	31.4 [18.8;41.6]	21.7 [18.9;24.5]	5 (46%)	4 (46%)	4 (37%)	-
Right heart obstruction (n = 52)	18	21	19.0 [15.7;24.1]	21.7 [18.9;24.5]	51 (98%)	-	15 (29%)	4 (8%)
Ebstein's anomaly (n = 13)	7	4	20.1 [13.2;36.6]	22.0 [19.6;25.6]	6 (46%)	7 (54%)	2 (15%)	-
Isolated shunt (n = 3)	1	1	22.8 [14.3;-]	20.4 [16.0;-]	2 (67%)	-	2 (67%)	1 (33%)
TGA arterial switch (n = 8)	1	5	15.2 [11.7;25.6]	20.3 [17.6;22.2]	8 (100%)	-	1 (13%)	1 (13%)
TGA others§ (n = 6)	2	-	29.0 [25.7;36.1]	22.1 [21.4;23.7]	6 (100%)	-	4 (67%)	-
TCPC (n = 4)	2	1	30.5 [17.3;38.5]	23.3 [20.0;28.0]	4 (100%)	-	3 (75%)	-
Miscellaneous (n = 5)	3	-	31.2 [21.3;36.3]	20.5 [19.4;23.8]	1 (20%)	3 (60%)	2 (40%)	-

*expressed as median [IQR25;IQR75], **e.g.: orthopedic, chronic headache, allergies etc., ***asthma, paralyses of diaphragma, severe skoliosis

§TGA others: Transposition of the great arteries after Senning/Mustard/Rastelli procedure and congenital corrected transposition of the great arteries

CHD: congenital heart defect, BMI: body mass index,, TCPC: Fontan circulation after total cavopulmonary connection

left heart obstruction includes aortic stenosis and coarctation of the aorta; right heart obstruction includes tetralogy of Fallot and pulmonary stenosis and common arterial trunk; isolated shunt includes atrial/ventricular septal defect, atrioventricular septal defect, partial or total anomalous pulmonary venous return, patent ductus arteriosus or foramen ovale

- FEV1/FVC z-score < -1.645 and a TLC z-score < -1.645 is a combination of restrictive and obstructive patterns.
- FVC z-score OR FEV1 z-score < -1.645 but other parameters are normal as un-classified spirometry.

This last group was analyzed separately and, where needed, included in the group with markedly results (Table 2).

Exercise capacity

As part of their routine follow-up, all patients underwent a standardized [19] symptom-limited cardiopulmonary exercise test, as previously described [20, 21]: All patients performed an upright sitting bicycle CPET with a ramp-wise increase of load (5 to 30 Watts per minute). After a rest of three minutes for baseline and unloaded paddling for further three minutes, exhaustion should be reached after 8–12 min (proved by a respiratory exchange ratio ≥ 1.05 in children and ≥ 1.10 in adults). Five minutes after maximum exhaustion, patients were continuously observed. All patients were motivated to cycle to their maximum.

The highest running 30 s average of oxygen uptake was used as peak oxygen uptake (peak VO_2). The percentage of predicted (%peak VO_2) was calculated for children and adults below the age of 25 years by Cooper et al. [22] and adults 25 years and older, Gläser et al. [23] was used. Peak VO_2 results ≥ 80% predicted are defined as normal, 80–60% as slightly impaired and < 60% as severely impaired.

All patients were under permanent 12-lead electro-cardiogram and pulse oximetry surveillance, blood pressure was measured every two minutes.

Data analyses

Due to the low number of participants, anthropometric measures were expressed as median and interquartile ranges [IQR25; IQR75]. Correlations between lung volumes and CPET were calculated using Spearman's rho correlation or Fisher's exact test, if adequate.

All data were analyzed using SPSS 28.0 software (IBM Inc., Armonk, NY, USA) with a two-tailed level of significance at p-value ≤ 0.05.

Results

Detailed results are seen in Table 3 and Table 4.Fifty-six patients (55%) had normal results in FVC and FEV1. While 28 patients (28%) showed a preserved ratio impaired spirometry (PRISm). Four patients had an obstructive pattern, and another four had both patterns. Of the 28 patients with PRISm, 16 (57%) were proven to have a small lung (additional TLC_z < -1.645), while the other 10 were defined as "non-specific-pattern" (normal TLC_z) and required further investigations. Table 4 shows six patients with only FEV1_z below the threshold, who can be classified as having additional obstructive results. Another four patients only had reduced FVC_z, which could be originated from insufficient effort during the test.

Overall, 45% of the patients ($n = 46$) had normal peak oxygen uptake (%predicted ≥ 80), 41% ($n = 42$) showed mild impairments with %predicted below 80 but above or equal to 60 and the others (14% or $n = 14$) showed severe reduction in peak oxygen uptake below 60% of peers.

Patients with impaired lung function more often showed low exercise capacity (Fisher's exact test: $p = 0.028$). However, subgroups showed no significant cluster – a CHD itself does not accumulate impaired results.

There were significant correlations between FVC_z, FEV1_z, and TLC_z with %predicted peak VO2 ($r = 0.365$, $p < 0.001$; $r = 0.320$, $p = 0.001$; $r = 0.249$, $p = 0.012$ respectively), with FVC being the strongest predictor followed by FEV1 and TLC. However, a correlation was only seen in z-score values, not in raw data (measurements in litre). Therefore, it is important to calculate the adjusted data in

Table 2 Classification of ventilatory impairments (mod. from Stanojevic, 2021)

	FVC	FEV_1	FVC/FEV_1	TLC	
Obstruction	normal	(normal)/↓	↓	normal	
Restriction	↓	↓	normal	↓	
Mixed	↓	↓	↓	↓	
Non-specific pattern	↓	↓	normal	normal	
Muscle weakness suboptimal effort	↓	↓	normal	normal/↓	lack of sharp PEF
Dysanapsis	high normal	normal	↓	normal	maybe normal variant

FVC: forced vital capacity, FEV1: forced expiratory volume in 1 s, TLC: total lung capacity, PEF: peak expiratory flow, ↓: z value < -1.645

Table 3 Results in spirometry, body plethysmography and CPET

CHD	FVC_z*	FEV1_z*	FEV1/FVC_z*	TLC_z*	RV/TLC_z*	peak VO_2*	%peak VO_2*
Overall ($n=102$)	-1.31 [-1.91;-0.40]	-1.23 [-2.16;-0.39]	-0.17 [-0.93;0.56]	-0.79 [-1.39;0.02]	0.97 [0.40;1.42]	28.6 [24.7;34.1]	78.6 [65.6;91.7]
Left heart obstruction ($n=11$)	-0.54 [-1.40;0.52]	-0.67 [-1.35;0.49]	-0.62 [-1.29;0.55]	-0.31 [-0.90;0.62]	0.82 [0.25;1.40]	29.1 [26.1;41.3]	93.1 [79.0;102.7]
Right heart obstruction ($n=52$)	-1.63 [-2.44;-1.01]	-1.65 [-2.28;-1.10]	-0.18[-0.89;0.69]	-1.07 [-1.65;-0.41]	0.98 [0.32;1.40]	29.8 [24.6;35.8]	78.3 [62.6;86.6]
Ebstein's anomaly ($n=13$)	-0.34 [-1.46;0.44]	-0.71 [-1.59;0.19]	-0.32 [-1.80;0.97]	-0.01 [-0.63;0.84]	0.97; [0.55;1.70]	25.7 [19.3;30.5]	69.2 [53.2;91.8]
Isolated shunt ($n=3$)	-1.68 [-1.83;-]	-1.17 [-2.14;-]	-0.70 [-1.14;-]	-1.21 [-1.71;-]	0.75 [0.76;-]	33.2 [24.9;-]	80.7 [58.6;-]
TGA arterial switch ($n=8$)	-0.37 [-0.57;0.73]	-0.04 [-0.43;0.38]	-0.04 [-0.28;1.15]	0.26 [-0.45;1.28]	0.63 [0.33;1.28]	36.8 [27.8;46.3]	90.5 [69.3;100.5]
TGA others[$] ($n=6$)	1.30 [-1.84;-0.26]	-0.82 [-2.13;-0.32]	0.39 [-0.55;1.07]	-0.83 [-1.53;-0.39]	1.12 [0.69;1.39]	26.3 [22.9;28.6]	72.0 [69.9;77.2]
TCPC ($n=4$)	-1.42 [-1.90;-0.62]	-1.52 [-2.27;-0.40]	-0.29 [-0.98;0.31]	-0.91 [-1.10;-0.46]	1.06 [-0.30;1.63]	22.4 [20.3;30.8]	71.4 [65.3;77.5]
miscellaneous ($n=5$)	-1.07 [-2.11;0.11]	-0.89 [-2.52;0.16]	-0.44 [-0.98;0.46]	0.49 [-1.78;1.29]	1.86 [1.09;1.93]	31.9 [21.6;40.4]	93.5 [73:6;102.0]

*expressed as median [IQR25;IQR75]

[$]TGA others: Transposition of the great arteries after Senning/Mustard/Rastelli procedure and congenital corrected transposition of the great arteries

CPET: cardiopulmonary exercise test, CHD: congenital heart defect, FVC: forced vital capacity, FEV1: forced expiratory volume in 1 s, TLC: total lung capacity, VO_2: oxygen uptake, TGA: transposition of the great arteries, TCPC: total cavopulmonary connection

lung function and exercise capacity to exclude potential bias.

Discussion

In this cohort, about half of the patients with various CHD exhibit impaired results in spirometry that are associated with a limited exercise capacity.

Published data from several studies support this study's findings: right heart obstruction patients (mostly tetralogy of Fallot) often suffer from low FVC and FEV1 [24, 25], as well as in TCPC patients, PRISm often occur [26]. However, only limited data are available for left heart obstruction or isolated shunts, showing more or less normal results in lung function tests [16, 26–28]. Nevertheless, patients with CHD have lower z-scores than the reference (Table 3). Furthermore, lung function correlates with exercise capacity – which was already studied in previous publications [28–30]. The present study adds that there is also a correlation between exercise capacity with TLC.

However, results show that a significant proportion of patients with a PRISm have significantly smaller lungs in body plethysmography (16 from 28 with a restrictive pattern). Even of more interest are those with a non-specific pattern (restrictive in spirometry with normal TLC). Formerly, these patients were classified as airflow occlusion or air trapping etc. [31, 32] which was revised with the latest task force report in 2021 [18]. In our cohort, twelve patients are grouped as "non-specific pattern". Adding patients with only FEV1_z or FVC_z below the threshold, a total of 22 patients, representing 22% of all patients, have abnormal results that need to undergo further investigations (e.g. bronchodilator test, muscle test) [18, 30]. Co-working with lung specialists is recommended and advisable in patients with CHD.

Limitations

All included patients are under regular tertiary care. Furthermore, patients who consider their lung function test results may more likely refuse to participate. Patients with cognitive restrictions, language barriers or not being able to perform a CPET could not be included. Furthermore, if the patient did the CPET, not until full exhaustion ($n=26$) were excluded. Insufficient quality in spirometry and/or body plethysmography also excluded patients ($n=76$). All of these bias the results ($n=204$ eligible, $n=102$ for analyses).

Table 4 Frequencies in striking results in underlying CHD

CHD	Normal	PRISm*	Restriction**	Obstructive pattern***	Combined pattern	↓FVC but normal FEV1	Normal FVC but ↓FEV1	Normal[£] %peak VO_2	↓%peak[¥] VO_2	↓↓%peak[¥] VO_2
All (*n* = 102)	56 (55%)	28 (28%)	16 (57%)	4 (4%)	4 (4%)	4 (4%)	6 (6%)	46 (45%)	42 (41%)	14 (14%)
Left heart obstruction (*n* = 11)	9 (82%)	-	-	-	1 (9%)	-	1 (9%)	8 (73%)	3 (27%)	
Right heart obstruction (*n* = 52)	23 (44%)	21 (40%)	12 (57% of restr.)	1 (2%)	2 (4%)	2 (4%)	3 (6%)	22 (42%)	23 (44%)	7 (14%)
Ebstein's anomaly (*n* = 13)	7 (54%)	1 (8%)	0 (0% of restr.)	3 (23%)	1 (6%)	1 (8%)		5 (39%)	3 (23%)	5 (38%)
Isolated shunt (*n* = 3)	1 (33%)	1 (33%)	1 (100% of restr.)	-	-	1 (33%)		2 (67%)	-	1 (33%)
TGA arterial switch (*n* = 8)	7 (88%)	1 (13%)	1 (100% of restr.)	-	-			5 (62%)	3 (38%)	-
TGA others[§] (*n* = 6)	4 (67%)	1 (17%)	1 (100% of restr.)	-	-		1 (17%)		6 (100%)	
TCPC (*n* = 4)	2 (50%)	2 (50%)	0 (0% of restr.)	-	-			-	4 (100%)	-
Miscellaneous (*n* = 5)	3 (60%)	1 (20%)	1 (100% of restr.)		-	-	1 (20%)	4 (80%)	-	1 (20%)

CHD: congenital heart defect, VO_2: oxygen uptake, TGA: transposition of the great arteries, TCPC: total cavopulmonary connection, peak VO_2: peak oxygen uptake, FVC: forced vital capacity, FEV1: forced expiratory volume in 1 s, TLC: total lung capacity, ↓: z value < -1.645 (there is no abnormal high) or respectively < 80% and ↓↓ < 60%

[§]TGA others: Transposition of the great arteries after Senning/Mustard/Rastelli procedures and congenital corrected transposition of the great arteries

*FVC and FEV1 z-score < -1.645

**restrictive pattern with approved TLC z-score < -1,645

***normal FVC and FEV1, but FEV1/FVC z-score < -1.645

[£]ranges: > 80% predicted normal; 79–60% predicted mild impaired; < 60% severe impaired

[¥]References for children up to 25 years of age from Cooper et al. [22]; adults above 25 years of age from Gläser et al. [23]

Conclusions

Lung function and exercise capacity correlate significantly. Following the guidelines, performing a CPET in patients with CHD includes a lung function test. Not only since there is a significant correlation, but also since a valid lung function (examine FVC and FEV1) gives further information during the test (see pulmonary limitation etc.). This is why it must always be included.

However, total lung capacity and therefore body plethysmography may not add any extra or better value, except in cases where simple spirometry reveals noteworthy results. Especially if a restrictive pattern is seen, body plethysmography should be performed to ensure a restriction and warrant further examinations by a pulmonary specialist.

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Declarations

Conflict of interest No conflicts of interest.

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