

**Supplementary Table 1.** Results from spirometry, lung volume, and DLCO<sub>10s</sub> in PFT-I and PFT-II, presented as median percent predicted (%pred). Patients are categorized into a pediatric (under 18 years at PFT-I) and an adult group (18 years or older at PFT-I). All %pred values are calculated with the GLI calculator. The  $\Delta$  value indicates the change from PFT-I to PFT-II.

	Pediatric group (n=32)				Adult group (n=16)			
	PFT-I %pred	PFT-II %pred	$\Delta$	p	PFT-I %pred	PFT-II %pred	$\Delta$	p
<b>FEV1</b>	99.7 (92.4, 104.4)	89.3 (84.9,97.2)	-7.3 (-14.4,-2.7)	<b>&lt;0.001</b>	84.2 (79.1,91.4)	87.1 (78.7,97.3)	0.8 (-8.1,6.8)	0.793
<b>FVC</b>	98.3(87.8,106.1)	96.7(86.7,100.6)	-5.8(-8.9,-0.3)	<b>0.008</b>	87.9 (82.7,92.7)	89.9 (83.3,100.2)	3.8 (-5.6,7.2)	0.298
<b>FEV1/FVC</b>	100.7 (96.7,105.2)	96.9 (93.8,100.7)	-4.8 (-6.7, -1.1)	<b>&lt;0.001</b>	100.4 (93.8,104.6)	101 (93.1,102.3)	-2.3 (-5.5,0.3)	0.065
<b>DLCO<sub>10s</sub></b>	71.4 (62.1,81.6)	69.1 (52.8,77.8)	-5.2 (-13.6,4.6)	0.062	70.2 (52.4,74.5)	68.8 (56.7,76.3)	3.6 (-8.2,13.8)	0.562
<b>DLCO/VA</b>	74.5 (69.6,84.5)	81.9 (66.4,87.9)	2.1 (-5.9,14)	0.477	81 (62.6,89.8)	83.2 (75.1,92.7)	1.5 (-8.9,18.4)	0.274
<b>VA</b>	95.5 (89.5,101.6)	89.5 (79.7,93.2)	-8.7 (-16.3,-3.1)	<b>&lt;0.001</b>	85.1 (78.7,90.1)	78.5 (74.6,92.6)	-3.9 (-9.2,0.2)	0.175

Data are presented as median (IQR)

FEV1 (forced expiratory volume in 1 second; FVC (forced vital capacity); DLCO<sub>10s</sub> (diffusing Capacity for Carbon Monoxide 10 seconds); DLCO/VA (DLCO corrected for alveolar volume); VA (alveolar volume)

**Supplementary Table 2.** Results from the single-breath determination of nitric oxide uptake (DLNO) in PFT-I and PFT-II in the adult group, presented as percent predicted (%pred). All %pred values were calculated using the reference equations published in the ERS task force report by Zavorsky et al. [21]

Adult group n = 16	PFT-I	PFT-II	$\Delta$	p
<b>DLNO</b>	58.4 (53.3, 63.5)	53.7 (44.1, 57.3)	-7.7 (-10.6, 0.1)	<b>0.005</b>
<b>DLCO<sub>5s</sub></b>	63.1 (49.2, 72.6)	54.2 (49.1, 63.3)	-3.9 (-12.2, 0.9)	0.051
<b>DLNO/VA</b>	82.4 (77.3, 99.6)	78.3 (71.6, 82.1)	-9 (-20.6, -2)	<b>0.006</b>
<b>DLCO/VA</b>	81.3 (73.9, 102.0)	75.9 (62.7, 84.2)	-10 (-24.2, 4.3)	0.058
<b>VA</b>	77.1 (70.9, 83.3)	74.0 (70.2, 87)	-2.1 (-6.3, 3.4)	0.597
<b>Dm</b>	49.1 (43.9, 56.3)	39.6 (31.6, 42.1)	-12.6 (-15.1, -7.7)	<b>p &lt; 0.001</b>
<b>Vc</b>	64.4 (54.1, 85.9)	60.7 (57.8, 79.8)	3.6 (-5.5, 7.3)	0.706
<b>DLNO/DLCO ratio</b>	4.6 (4.3, 4.8)	4.3 (4.1, 4.4)	-0.3 (-0.5, 0)	0.083

Data are expressed as median (IQR).

DLNO (diffusing capacity for nitric oxide); DLCO<sub>5s</sub> (diffusing capacity for the lungs of carbon monoxide - 5 seconds measurement; KNO (transfer coefficient of the lung for nitric oxide); KCO (transfer coefficient of the lung for carbon monoxide); VA (alveolar volume); Dm (membrane diffusing capacity); Vc (capillary blood volume).

**Supplementary Table 3.** Simple regression models investigating the relationship between percent predicted (%pred) PFT measurements in PFT-II and clinical characteristics. The %pred values were calculated with the GLI calculator for FEV1, FVC, VA, and DLCO, and with the reference equation provided by Zavorsky et al. for DLNO.

	FEV1 %pred		FVC %pred		VA %pred		DLCO <sub>10s</sub> %pred		DLNO %pred	
	Estimate ± SD	P	Estimate ± SD	P	Estimate ± SD	P	Estimate ± SD	P	Estimate ± SD	P
<b>n = 48</b>										
<b>Sex</b>										
Reference: Male										
Female	6.17 ± 3.11	0.054	1.62 ± 3.40	0.636	0.45 ± 3.34	0.983	4.15 ± 4.15	0.322	4.73 ± 3.04	0.126
<b>Age</b>	0.03 ± 0.25	0.912	-0.05 ± 0.27	0.860	0.02 ± 0.26	0.948	0.43 ± 0.32	0.186	-0.50 ± 0.23	<b>0.035</b>
<b>Ventricular morphology</b>										
Reference: Left ventricle					-1.10 ±					
Right ventricle	5.78 ± 3.12	0.072	1.68 ± 3.40	0.624	3.35	0.744	-3.76 ± 4.16	0.370	-2.67 ± 3.09	0.392
<b>Age at Fontan completion</b>										
Reference: < 5 years					-1.67 ±					
> 5 years	4.88 ± 3.37	0.155	2.75 ± 3.60	0.450	3.56	0.641	7.43 ± 4.33	0.093	-1.07 ± 3.32	0.759
<b>Fontan type</b>										
Reference: Extracardiac tunnel										
Lateral tunnel/Classical Fontan	0.87 ± 3.45	0.802	-0.07 ± 3.62	0.985	4.48 ± 3.44	0.200	-2.32 ± 4.37	0.598	-7.61 ± 3.06	<b>0.016</b>
<b>%pred VO<sub>2peak</sub></b>	0.05 ± 0.16	0.747	0.13 ± 0.16	0.424	0.16 ± 0.16	0.320	0.20 ± 0.20	0.324	0.45 ± 0.13	<b>0.001</b>

FEV1 (forced expiratory volume in 1 second; %pred (percent predicted); FVC (forced vital capacity); VA (alveolar volume); DLCO<sub>10s</sub> (diffusing Capacity for Carbon Monoxide, 10 seconds measurement); DLNO (diffusing capacity for Nitric Oxide)