

SUPPLEMENTAL MATERIAL

Supplemental Table 1. Patient, Medical and Laboratory Characteristics of 511 Patients.

Variable	n	Value
Age at enrollment (years)	511	11.8±3.4
Gender, male	310	61%
Ethnicity		
White	412	81%
Black	48	9%
Asian	11	2%
Other	39	8%
Predominant ventricular morphology		
Left	251	49%
Right	170	33%
Indeterminant or mixed	90	18%
Staging cavopulmonary connection performed	380	74%
Age at Fontan procedure (years)	511	3.4±2.0
Type of Fontan connection		
Atriopulmonary	69	14%
Intracardiac lateral tunnel	305	60%
Extracardiac lateral tunnel	63	12%
Extracardiac conduit	64	13%
Other	10	2%
Fenestration performed	342	67%
Interval from Fontan procedure (years)	511	8.5±3.4
Current pacemaker	64	13%

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Variable	n	Value
Number of current medications		2 (1, 3 IQR)
0	54	11%
1	129	25%
2	135	26%
3	97	19%
4	37	7%
5	29	6%
6	12	2%
7	7	1%
8	4	1%
9	2	0%
10	3	0%
11	1	0%
12	1	0%
Current cardiac medication(s)	323	63%
Current morbidities		
Stroke	10	3%
Thrombosis	38	7%
Protein-losing enteropathy	20	4%
Arrhythmia	103	20%
Ventricular dysfunction	86	17%
Parent Report Child Health Questionnaire (CHQ)		
CHQ Summary Scores		
Physical functioning	511	45.3±11.9
Psychosocial functioning	511	47.2±10.8
Laboratory Testing		
Brain-Natrietic Peptide (BNP; pg/ml)	483	24±40 (IQR 7.8, 25.6)

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Variable	n	Value
Cardiopulmonary Exercise Testing		
Percent predicted maximum work rate (%)	157	66.2±16.1
Percent predicted VO ₂ at peak (%)	157	66.8±14.8
Percent predicted VO ₂ at anaerobic threshold (%)	153	77.4±21.8
Percent predicted maximum oxygen pulse (%)	157	88.3±22.4
Resting oxygen saturation (%)	156	94.1±4.2
Chronotropic index	134	0.7±0.1
Echocardiography		
End-systolic volume Z score	389	0.2±2.4
End-diastolic volume Z score	389	-0.6±1.9
Total stroke volume Z score	389	-1.0±1.8
Total ejection fraction Z score	389	-0.8±2.0
Total ventricular mass Z score	383	1.0±2.3
Total mass:end-diastolic volume ratio (gm/mL)	383	1.2±0.4
Tei index	345	0.6±0.2
dP/dt _{ic} (mm Hg/sec)	337	1441±965
AVV peak late diastolic velocity (m/sec)	261	0.5±0.1
Early:late AVV diastolic velocity (E/A)	261	1.6±0.5
Duration of AVV late diastolic inflow (msec)	261	139±27
Absolute systemic ventricular flow propagation rate (cm/sec)	113	64±20
Ratio of AV to tissue Doppler early diastolic flow velocity	229	8.5±4.1
Tissue Doppler peak early diastolic velocity (cm/sec)	338	9.3±3.2
Overall AV valve regurgitation	385	
None	97	25%
Mild	224	58%
Moderate/severe	64	17%
Semilunar valve regurgitation	239	
None	122	51%
Mild	97	41%
Moderate	20	8%

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Variable	n	Value
Magnetic Resonance Imaging		
Total end-systolic volume indexed to BSA ^{1.3} (mL/m ²)	155	37±15
Total end-diastolic volume indexed to BSA ^{1.3} (mL/m ²)	155	86±25
Total stroke volume (mL)	155	65±25
Total ejection fraction (%)	155	57±9
Cardiac output (L/min)	153	5.1±1.7
Total ventricular mass indexed to BSA ^{1.3} (gm/m ²)	155	71±20
Total mass/end-diastolic volume ratio (gm/mL)	155	0.9±0.3

AV, atrioventricular; AVV, atrioventricular valve; BSA, body surface area; cm, centimeters; dP, change in pressure; dt, change in time; gm, grams; IQR, interquartile range; L, liters; m, meters; min, minute; mL, milliliters; mm Hg, millimeters Mercury; PLE, protein-losing enteropathy; sec, second; VO₂, oxygen consumption

*values represent frequency, median with interquartile range, or mean and standard deviation

**after logarithmic transformation

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