SUPPLEMENTAL MATERIAL

Echocardiographic Measures and Derived Parameters:

- End-diastolic and end-systolic RV volumes (mL) were obtained using the biplane pyramidal method[1] from a transverse apical 4-chamber view area at the level of the tricuspid annulus and subcostal sagittal length of the RV cavity from RV apex to pulmonary annulus (Appendix Figure 1) and RV % ejection fraction was calculated as:

  End-diastolic volume – End-systolic volume)/End diastolic volume

- The end-diastolic and end-systolic RV area (cm^2) were measured from a transverse apical 4-chamber view and RV % area change calculated as:

  End-diastolic area – End-systolic area/End-diastolic area.

- The RV dP/dt was obtained from the tricuspid regurgitation continuous wave Doppler signal aligned parallel with the jet.[2]
- RV shape was assessed using the eccentricity index, calculated as long axis dimension/short axis dimension from a transverse apical 4-chamber view.[3]
- The tricuspid valve area (cm^2) was calculated from the maximal diastolic annular diameter measured from orthogonal long-axis and apical imaging planes using the formula for an ellipse: $[\pi(\text{diameter}_1/2)(\text{diameter}_2/2)]$.
- Severity of tricuspid regurgitation was assessed by the size of the regurgitant jet vena contracta using color Doppler. Severity of regurgitation was graded as mild for vena contracta width $\leq$2.5 mm and moderate and severe regurgitation for width >2.5 mm. Grading was also performed by qualitative assessment of jet size as none, mild, moderate and severe.
- Peak E and peak A velocities (m/sec), with fused waves measured as a single E wave, and tricuspid closure time (time interval from cessation of diastolic inflow to initiation of diastolic inflow) were obtained from tricuspid inflow Doppler using a transverse apical 4-

chamber view with the pulsed wave Doppler sample volume positioned at the opening of the tricuspid valve leaflets.

- Tricuspid annular Doppler tissue imaging (DTI) velocities and intervals were obtained from the transverse apical 4-chamber view with the Doppler sample volume positioned at the medial (non-septal) annulus. Peak systolic annular early diastolic annular (E'), and peak annular velocity with atrial contraction (A') in cm/sec, systolic ejection time (msec), and tricuspid closure time (time interval from cessation of diastolic annular motion to initiation of diastolic annular motion) were measured and the tricuspid inflow E to annular DTI E' velocity ratio calculated.
- Pulmonary venous inflow was obtained from a transverse apical 4-chamber view with the pulsed wave Doppler sample volume positioned within the mouth of the right upper pulmonary vein and the presence and duration of pulmonary vein reversal with atrial contraction was assessed.

Additional measures were obtained only for the preoperative study:

- The atrial septal defect mean gradient (mmHg) was measured from continuous wave Doppler aligned with flow across the septum, measured from subcostal long or short axis views.
- Native ascending aortic diameter (mm) was obtained from suprasternal notch imaging.
- Native aortic and mitral valve patency were assessed using color Doppler flow.

Additional measures were obtained only at the 3 post-operative stages:

- Myocardial performance index (MPI) assessed using both blood flow pool and annular DTI; MPI is defined as:

Isovolumic relaxation time + Isovolumic contraction time)/Ejection time

and can be derived from the (tricuspid closure time – ejection time)/ejection time from tricuspid and neo-aortic pulsed Doppler and tricuspid annular DTI tracings.[4]

- Neo-aortic valve area (cm^2) from maximal systolic annular diameter measured from orthogonal long axis and apical imaging planes using the formula for an ellipse: [π(diameter$_1$/2)(diameter$_2$/2)].
- Neo-aortic valve regurgitation assessed by the size of the regurgitant jet vena contracta using color Doppler with regurgitation graded as greater than mild when the vena contracta width is greater than 2.5 mm.
- Neo-aortic flow at the annulus using pulsed Doppler, measuring the velocity time integral (VTI) in cm of the systolic flow jet and systolic ejection time (msec) defined as the duration of systolic flow from the Doppler jet.
- Neo-aortic cardiac index estimated as stroke volume [(neoaortic VTI)(neoaortic valve area)] x heart rate in beats per min indexed to body surface area (BSA) in m^2.
- Descending neo-aortic flow patterns using pulsed wave Doppler from the suprasternal notch to characterize antegrade and retrograde flow signals; antegrade flow was defined by the forward systolic VTI and retrograde flow by the reversed diastolic VTI. The percentage of retrograde diastolic flow in the descending aorta was calculated as: 1 - [(systolic VTI – diastolic VTI)/systolic VTI].
- Peak descending aortic velocity (M/sec) measured by continuous wave Doppler from suprasternal imaging, guided by color Doppler through the narrowest portion of the reconstructed aortic arch.
- Pulmonary artery (PA) narrowest diameters (mm) from parasternal/suprasternal notch short axis imaging at 3 locations: a) the anatomic left PA distal to the main PA bifurcation; b) the anatomic right PA distal to the main PA prior to right PA bifurcation;

and c) the anatomic right PA to the right of the superior vena caval anastomosis following stage II procedure (only available at the 14 month study).

Supplement References

1. HelbingWA, Bosch HG, Maliepaard C, Rebergen SA, van der Geest RJ, Hansen B, Ottenkamp J, Reiber JHC, de Roos A. Comparison of echocardiographic methods with magnetic resonance imaging for assessment of right ventricular function in children. Am J Cardiol 1995;76:589-594.
2. Michelfelder EC, Vermilion RP, Ludomirsky A, Beekman RH, Lloyd TR. Comparison of simultaneous Doppler- and catheter-derived right ventricular dP/dt in hypoplastic left heartsyndrome. Am J Cardiol 1996;77(2):212-214.
3. D'Cruz IA, Shroff SG, Janicki JS, Jain A, Reddy HK, Lakier JB. Differences in the shape of the normal, cardiomyopathies and volume overloaded human left ventricle. J Am Soc Echocardiogr 1989;2:408–414.
4. Harada K, Tamura M, Toyono M, Yasuoka K. Comparison of the right ventricular Tei index by tissue Doppler imaging to that obtained by pulsed Doppler in children without heart disease. *Am J Cardiol* 2002;90:566-569.

Online Data Supplement Figure Legend

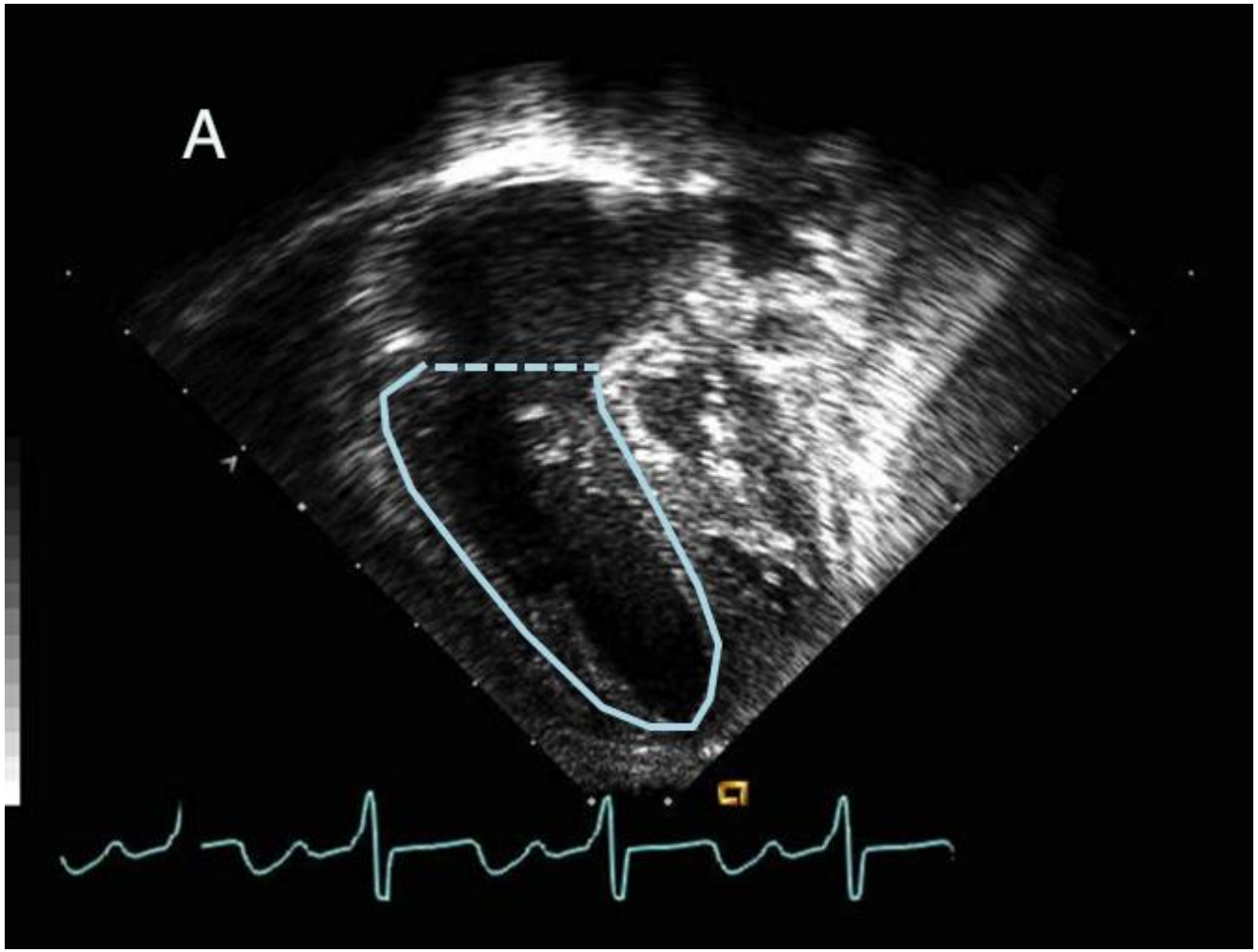
Figure 1. Apical transverse 4-chamber RV area (A) at the level of the tricuspid annulus (dashed line) and subcostal sagittal long-axis length (B) from apex to neo-aortic annulus (arrows) needed to measure RV ventricular volumes using the biplane pyramidal method. End-diastolic and end-systolic volumes are calculated using the equation 2/3 (RV apical area x subcostal long axis length) with the maximal area and length measures at end-diastole and end-systole.

Online Data Supplement Table. Significant echo indices in data limited to 329 subjects alive at 14 months with echo readings deemed acceptable by the echo core lab at all post-operative stages.

	N	MBTS	N	RVPAS	p
Post-Norwood					
RV ejection fraction %	108	45±8	152	49±7	<0.001
RV % area change	142	35±7	178	39±7	<0.001
MPI blood flow Doppler calculation	123	0.37±0.17	151	0.56±0.24	<0.001
Presence of pulmonary vein flow	135	10 (7%)	167	39 (23%)	0.001
LPA diameter, cm	136	0.39±0.10	159	0.43±0.11	0.002
LPA diameter z-score	136	-1.14±1.09	159	-0.71±1.29	0.002
Neoaortic valve annular area (cm^2)/BSA	139	4.70±1.18	172	4.15±1.11	<0.001
Neoaortic valve annular area, z-score	139	7.23±3.09	172	5.79±2.90	<0.001
Neoaortic AP valve annulus diameter (cm)/√(BSA)	143	2.39±0.37	177	2.25±0.35	<0.001
Neoaortic AP valve annulus diameter, z-score	143	4.81±2.15	177	3.95±2.05	<0.001
Neoaortic cardiac index, L/min/m^2	122	8.20±2.67	150	4.40±1.95	<0.001
Neoaortic retrograde fraction, %	123	42 (33,53)	146	0 (0,0)	<0.001
Neoaortic ejection time, msec	130	197±22	158	170±25	<0.001
Neoaortic peak distal arch gradient, m/sec	106	1.86±0.50	141	1.37±0.49	<0.001
Pre-stage II					
MPI blood flow Doppler calculation	125	0.39±0.16	155	0.49±0.19	<0.001
Neoaortic valve annuluar area (cm^2)/BSA	139	4.93±1.22	172	4.05±1.07	<0.001
Neoaortic valve annular area, z-score	139	8.08±3.27	172	5.72±2.87	<0.001
Neoaortic AP valve annulus diameter (cm)/√(BSA)	142	2.44±0.35	177	2.21±0.30	<0.001
Neoaortic AP valve annulus diameter, z-score	142	5.59±2.24	177	4.16±1.96	<0.001
Neoaortic cardiac index, L/min/m^2	128	9.44±3.11	153	6.11±2.24	<0.001
Neoaortic retrograde fraction, %	108	39 (32,47)	138	0 (0,0)	<0.001
Neoaortic ejection time, msec	132	232±28	164	217±28	<0.001
Neoaortic peak distal arch velocity, m/sec	113	2.21±0.69	139	1.89±0.70	<0.001

Data presented as mean±SD or median (inter-quartile range). Data at 14 months is not shown because none of the echocardiographic indices were significantly different between shunt types at 14 months. MBTS=modified Blalock-Taussig shunt; RVPAS=right ventricular-to-pulmonary artery shunt; RV=right ventricle; MPI=myocardial performance index; AP=anteroposterior; LPA=left pulmonary artery

Online Data Supplement Figure 1A.



Online Data Supplement Figure 1B.