Evaluation of Invasively Measured Pressures and Echocardiographic Measures of Diastolic Function in Children with Fontan Circulation

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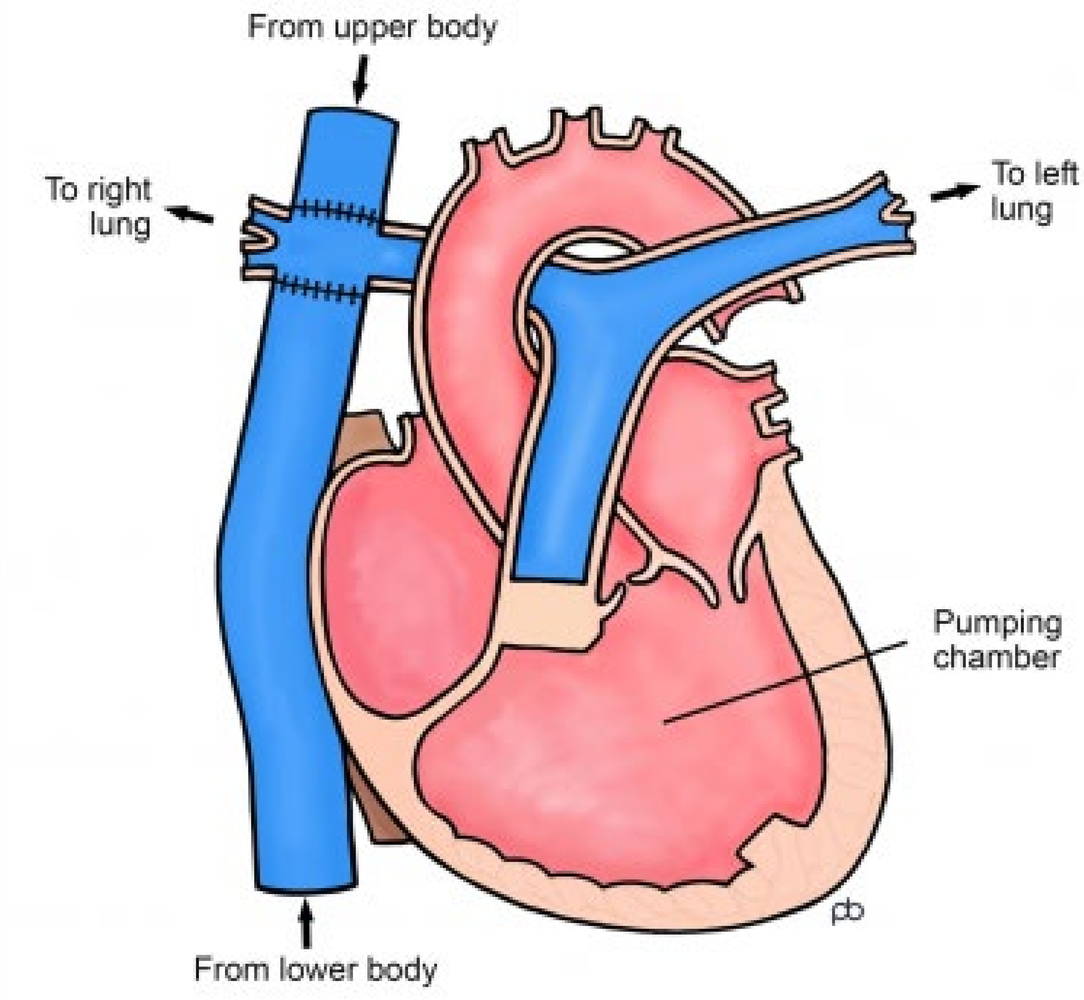
Presenting Author Disclosure Information

- I have no relevant financial relationships with the manufacturer(s) of any commercial product(s) and/or provider(s) of commercial services discussed in this CME activity.

- I do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation.

Introduction

- Fontan procedure is the final step in a series of palliative surgeries for patients with single ventricle physiology
- Ongoing efforts to improve Fontan outcomes have largely focused on pulmonary arteries and systolic dysfunction
  - Many patients have neither overt systolic dysfunction or evidence of reversible pulmonary endothelial dysfunction



Introduction

- Diastolic dysfunction likely remains in under-recognized cause of Fontan failure
  - Pediatric Heart Network Fontan Cross-Sectional Study, 72% of patients had evidence of abnormal DD with normal EF
- Unlike the biventricular heart, established techniques such as echocardiogram are not well validated
- Invasive hemodynamic evaluations are frequently unrevealing
- Improved diagnostic criteria are necessary

Aims

- Primary aim
  - To determine the association of echocardiographic measures of diastolic dysfunction with direct pressure measurements made by right heart catheterization before and after rapid fluid expansion
- Secondary aims
  - Assess pressure changes in the Fontan circuit following volume expansion
  - Identify any association of clinical status with diastolic dysfunction

Methods

- Single center, retrospective study
- Patients < 5 years post Fontan procedure at the time of catheterization were excluded
- Fontan patients with Doppler echocardiogram within 6 months of completing routine right heart catheterization from 8/17/20 to 11/22/21
- Diastolic dysfunction (DD) was defined as a post-bolus ventricular end-diastolic pressure (VEDP) of at least 15mmHg or >20% increase from baseline and greater than 12 mmHg
- Differences between groups was determined using Mann-Whitney U test or Fisher's exact where appropriate, correlation between variables was measured using Spearman's rho.

Demographic Data

	Total	No DD	DD
Demographics	n=20	n=10	n=10
Age	15.1 (9.5-21.1)	13.3 (10-17.9)	15.8 (12.6-22)
Sex (% female)	8 (40%)	5 (50%)	3 (30%)
Weight (kg)	55 (27.5-82)	42.1 (27.3-66.9)	73.1 (36.6-93.9)
Duration of Fontan to catheterization (years)	12.4 (6.5-18.1)	10.6 (7.2-15.1)	12.7 (6.8-18.8)
Systemic ventricle morphology			
Left ventricle	9 (45%)	3 (30%)	6 (60%)
Right ventricle	14 (70%)	10 (100%)	4 (40%)
Type of Fontan			
Extracardiac Fontan	19 (95%)	9 (90%)	10 (100%)
Lateral tunnel	1 (5%)	1 (10%)	
Fenestration present	10 (50%)	7 (70%)	3 (30%)
Protein losing enteropathy	5 (20%)	4 (40%)	1 (10%)
Fontan-associated liver disease	5 (20%)	3 (30%)	2 (20%)

Results

	Total	No DD	DD
E wave (m/s)*	0.63 (0.52-0.78)	0.57 (0.48-0.66)	0.73 (0.56-1.02)
E/A ratio	1.0 (0.70-1.63)	0.91 (0.70-1.4)	1.14 (0.76-1.57)
DT (sec)*	0.18 (0.15-0.22)	0.15 (0.12-0.16)	0.22 (0.19-0.27)
Lateral e' (m/s)	0.08 (0.07-0.09)	0.082 (0.07-0.087)	0.082 (0.074-0.085)
Lateral E:E'*	7.5 (6.6-9.2)	6.7 (5.4-9.0)	8.8 (6.5-11.1)
PV systolic velocity (cm/sec)	40.3 (20.1-47.4)	43.7 (30.6-47.5)	44.8 (37-45.9)
PV diastolic velocity (cm/sec)	67.9 (55.9-79.8)	73.7 (52.7-81.8)	62.1 (55.2-65.5)
PV S/D ratio	0.67 (0.57-1.0)	0.61 (0.53-0.98)	0.75 (0.6-1.6)
PV atrial reversal velocity (cm/sec)	23.3 (15.7-37.0)	25.2 (10.7-36.6)	18 (15-24.6)
PV atrial reversal duration	0.08 (0.07-0.1)	0.08 (0.04-0.11)	0.07 (0.04-0.11)

Results

- Fluid expansion resulted in significant increase in Fontan pressure, PCWP, VEDP.
- There were no significant changes in TPG or PVR
- In multivariable logistics regression, patients with DD were independently associated with a decrease in CI after fluid expansion.

	Total	No DD	DD
Hemodynamics			
Fontan pressure			
Baseline*	14 (11-16)	12 (10.3-13)	15.5 (15-16.8)
After RVE*	18 (14.2-20)	14.5 (12.3-16.8)	19.5 (18-20)
PCWP			
Baseline*	10 (7-12)	6.8 (6.1-7.9)	11.8 (10-12.9)
After RVE*	12.5 (11-15.5)	10.25 (8.5-11.1)	14.75 (13.1-15.9)
TPG			
Baseline	4 (3-5)	4 (3.5-5.6)	3.75 (3.5-4.8)
After RVE	3.5 (3-5)	4 (3-4.8)	3.5 (3-4.8)
VEDP			
Baseline*	8 (5-11)	5 (3.3-7.8)	11 (8-12)
After RVE*	11 (7-14)	8 (7-9)	14 (12-15)
CI			
Baseline*	3.1 (2.6-4.0)	4.0 (3.6-4.8)	2.6 (2.3-3.2)
After RVE*	3.3 (2.8-3.7)	4.5 (3.9-4.9)	2.4 (2-3.1)
PVRi			
Baseline	1.7 (1.2-2.1)	2 (1.3-2.3)	1.6 (1.3-1.9)
After RVE	1.6 (1.1-1.9)	1.7 (1.3-1.9)	1.3 (1.2-1.4)

Limitations

- Although statistically significant results were obtained, the study had a small number of patients.
- There were insufficient numbers to analyze the impact of ventricular type and morphology and other clinical factors on the echocardiographic and catheterization data.
- Patients with different anatomic diagnoses and ventricular morphology may affect Doppler values

Conclusions

- Diastolic dysfunction in patients with a Fontan circulation was associated with changes in lateral E:E’ and deceleration time
- Abnormal diastolic function was independently associated with a lower CI regardless of pulmonary vascular resistance or transpulmonary gradient
- Large multi-center studies are needed to better define invasive and non-invasive measures of DD.

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Questions?