Ventricular-Pulmonary Vascular Coupling after the Total Cavopulmonary Anastomosis (Fontan)

6th International Neonatal & Childhood Pulmonary Vascular Disease Conference

Fontan Physiology
- Single ventricular pump
- Series of resistors

Ventricular Morphology
- Dominant LV
- Dominant RV
- Indeterminate
- HLHS
- HRHS
- Balanced ventricles

RV Morphology

Predictors of outcome after the Fontan operation: Is hypoplastic left heart syndrome still a risk factor?

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Objective: This study was undertaken to evaluate factors in early mortality and morbidity after the Fontan procedure between January 1, 1992, and December 31, 1999.
**Systolic function**

- RVEF lower than LVEF  
  *(Every paper ever written)*
- Lower ejection fraction/shape change in TA  
  *(Br Heart J 1988;60:324-31)*
- Abnormal circumferential wall stress over 2 years
- Normalised if Fontan <10 years  

**Force-frequency relationships**

Cheung et al. Heart 2005

**The systemic vessels**

Thicker veins  
Thinner arteries  
*Unrelated to hemodynamics, ACE use, type of Fontan*

Sarkola et al. In press Heart & Vessels 2011
Ventriculo-Vascular coupling

Khammadkone et al. Circulation 2002;106:II-358

Ventricular-Vascular coupling


Ventricular-Vascular coupling


Ventricular-Vascular coupling

ACE inhibition

Enalapril and exercise

- 18 patients (age 14±6 years) 4-19 years post Fontan
- Randomised DB, PC, X-over study of enalapril for 10 weeks
- Graded treadmill exercise

Kouatli et al. Circulation 1997; 96: 1507-12

ACE inhibition

- No change in resting Doppler indices, HR, cardiac index or exercise duration
- Fifty-eight percent of subjects were taking an angiotensin-converting enzyme inhibitor at enrollment. Diastolic function grade was normal in 38%. The median exerciseinduced increase in cardiac index significantly reduced with treatment

Kouatli et al. Circulation 1997

Low ‘preload reserve’

- Diastolic dysfunction
- Pulmonary vascular bed
**FONTAN CIRCULATION**

**Preload reduction - Diastole**
- Acute preload reduction
- Maintained shortening
- Constant LV mass

- Similar geometric change with BCVS


**Acute transition**

Increase

Preoperative  Postoperative

**Wall motion abnormalities**
- Inward motion during IVR
- 10% preop
- 80% postop


**Wall motion abnormalities**

**Incoordinate relaxation**

Incoordinate relaxation refers to an abnormal pattern of relaxation in the heart muscles, where the relaxation of one muscle fiber is not synchronized with the relaxation of another. This can lead to inefficient cardiac function and potentially cardiac dysfunction. The diagram shows a pressure-area loop (intraoperative transesophageal echocardiography (TEE)) which is a graphical representation of the pressure-volume relationship during the cardiac cycle.

**Increased ventricular stiffness**

Increased ventricular stiffness is a condition where the ventricular walls become harder and less elastic, leading to increased workload and reduced cardiac output. This can be assessed using pressure-volume loops. The table below compares characteristics of patients with right ventricular (RV) vs left ventricular (LV) dominance:

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>RV (n=11)</th>
<th>LV (n=5)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years)</td>
<td>3.1 ± 0.8</td>
<td>3.4 ± 1.2</td>
<td>0.6</td>
</tr>
<tr>
<td>BSA (m²)</td>
<td>0.57 ± 0.07</td>
<td>0.61 ± 0.09</td>
<td>0.4</td>
</tr>
<tr>
<td>EDP (mm Hg)</td>
<td>7.7 ± 2.6</td>
<td>9.4 ± 2.5</td>
<td>0.2</td>
</tr>
<tr>
<td>Mean PA (mm Hg)</td>
<td>11.4 ± 1.3</td>
<td>11.2 ± 1.9</td>
<td>0.8</td>
</tr>
<tr>
<td>β Pre-bypass</td>
<td>0.132</td>
<td>0.128</td>
<td>0.6</td>
</tr>
</tbody>
</table>

CL indicates confidence limit. β values were compared using Mixed Models methodology. All other values were compared by t test and are listed as mean ± SD.

**LV Pressure - dimension loops**

LV Pressure - dimension loops are graphical representations of the pressure-volume relationship during the cardiac cycle in the left ventricle. These loops can be used to assess ventricular function and stiffness.
Poor LV compliance

Normal aging

- Approximate 2 mmHg rise in LVEDP every decade after 4th
- Increased if volume load or systemic hypertension

Late changes

- Diastolic performance assessed ~8 years apart
- IVRT flow in 70%
- Shortened IVRT
- Faster e-wave deceleration

Contemporary Outcomes After the Fontan Procedure

A Pediatric Heart Network Multicenter Study

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Objectives
- We characterized a large cohort of children who had a Fontan procedure, with emphasis on functional health status, ventricular size and function, exercise capacity, heart rate variability, and brain natriuretic peptide (BNP).

Background
- The characteristics of contemporary Fontan survivors are well and described.

Methods
- We enrolled 561 children ages 6 to 18 years, mean 11.1 years, and compared them with pre-specified analytic risk factors for postoperative outcomes. All assessments were obtained within a 2-month period.

Results
- Preoperative ventricular morphology was 49% left ventricular (LV), 30% right ventricular (RV), and 21% mixed. Qp/Qs (mixed defects) was higher in 73% of patients, while functional status was worse in 39% of children in mixed defects. Late changes in ventricular size and function were observed in patients who had a Fontan procedure at an earlier age. LV size and function, as well as RV size and function, were greater in children who had a Fontan procedure at an earlier age. BNP was greater in children who had a Fontan procedure at an earlier age. BNP was greater in children who had a Fontan procedure at an earlier age.

Conclusions
- Measures of ventricular wall thickness and functional health status, although lesser in magnitude in the cohort compared with control subjects, were in the majority of subjects, within 2 standard deviations of the mean for control subjects. Right ventricular morphology was associated with poorer functional and cognitive function. RV size and function were more frequently observed in patients who had a Fontan procedure at an earlier age.

Cheung et al. Heart 2000
At peak exercise mean pulmonary arterial pressure increased from 10 to 18 mmHg (max Δ = 8 to 27)

PCWP increased from 6 to 10 mmHg (max Δ = 7 to 19)

**Fontan Physiology**
- Single ventricular pump
- Series of resistors

**Systemic venous-pulmonary vascular bed**

Khammadkone et al. *Circulation* 2002;106:II-358

Redington et al. *Br Heart J* 1990
Systemic venous-pulmonary vascular bed

Hepatic veins

Hsai TY et al. Circulation 2000
Lars Sondegard 2001

Pulmonary endothelial function

- Reduced NO production with low pulsatile stretch/shear

Pre NO Post NO

N=15, p<0.005

Khambadkone et al. Circulation 2003:107; 3204-8

Pulmonary endothelial function

- Greater NO responsiveness if ↑PBF pre-Fontan
- Higher PVR predicted lower functional class
- Role of NO donor/sildenafil?

Kambadkone et al. Circulation 2003

Conclusions

- Intrinsic abnormalities in some forms of ‘Single ventricle’
- Systolic function usually preserved
- Adverse relaxation abnormalities amplified by early and late reduction in compliance
- Abnormal systemic vascular properties
- Abnormal pulmonary vascular bed
Myocardial Fibrosis Identified by Cardiac Magnetic Resonance Late Gadolinium Enhancement Is Associated With Adverse Ventricular Mechanics and Ventricular Tachycardia Late After Fontan Operation

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