Staged LV recruitment in patients with HLHS

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Conflict of interest disclosure: none to disclose
Case presentation

• Postnatal diagnosis of HLHS
• Arch hypoplasia and coarctation
• Echo Z scores
  – LV EDV = -2.27
  – Aortic Valve = -3.41
  – Mitral V = -2.13
  – Severe LV dysfunction
  – Endocardial Fibroelastosis
• “Borderline LV”
• Single ventricle palliation or biventricular repair?
### Borderline Left Heart Structures

**Single Ventricle Palliation**
- Initial (stage 1) mortality ~ 10%, independent of LH structures
- Long term morbidity related to Fontan physiology
- Generally considered irreversible

**Biventricular Repair**
- Early mortality depends upon LH structures, presence of EFE
- Long term outcome better if survival past early phase
- Crossover to 1 V palliation has high mortality

### Advantages / Disadvantages

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<tr>
<th>Benefits</th>
<th>Drawbacks</th>
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Borderline LV – strategy

- Decision regarding single vs. biventricular repair
- Risk factors after BiV repair – small left structures, EFE, VSD, ventricular dysfunction
- Risk factors for single ventricle repair – Tricuspid regurgitation, small RV
- Tools to weigh competing risks and help choose initial strategy
  - CHSS calculator
  - Rhodes et al.
Borderline LV – Growth potential

- Data from patients with critical aortic stenosis
- Following aortic valve balloon dilation
- LVEDV can normalize
- Is it possible to take advantage of this growth potential?

McElhenney et al. 2005 (CHB)
Han et al. 2007 (Toronto)
Staged LV recruitment

Initial Single Ventricle Palliation

Maneuvers to rehabilitate left heart

LV growth

Subsequent biventricular conversion
Biventricular Conversion - Overview

- **Patient Selection**
- Operative strategies
- Outcomes
- Postoperative care
- Future directions
Borderline LV - Definition

• Left Ventricle characteristics
  – Small size
  – Restrictive LV (EFE)
  – LV dysfunction
• Associated lesions
  – Small mitral valve or congenital mitral stenosis
  – Small aortic valve or LVOT obstruction
  – Coarctation
Patient Selection - Diagnosis

• Hypoplastic Left Heart Syndrome
  – Mitral / Aortic Stenosis variant only
  – Atresia is incompatible even if LV OK.
• Shones Syndrome
• Unbalanced AV canal
• Deemed unsuitable for immediate biventricular repair
Biventricular Conversion - Overview

• Patient Selection
• **Operative strategies**
• Outcomes
• Postoperative care
• Future directions
Operative Strategies

• LV Rehabilitation
  – EFE resection
  – Accessory pulmonary blood flow
  – Restriction of ASD
  – Aortic valve repair
  – Mitral valve repair

• Biventricular conversion procedure
  – Ross procedure
  – Direct reanastamosis
Accessory pulmonary blood flow

RV PA conduit or BTS

Increases pulmonary blood flow and LV throughput

Generally done after Glenn

Only minimal increase in Glenn pressures
Biventricular repair

- Disconnect aorta-pulmonary connection
- Ross or Aortic valve repair
- Reconnect pulmonary artery
- Reconnect Aorta
- Take down Glenn
Biventricular Conversion - Overview

• Patient Selection
• Operative strategies
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Study design

• Retrospective review of patients who
  – HLHS or Shones with borderline LV
  – Underwent single ventricle palliation
  – Surgical attempts to rehabilitate LV with a goal of eventual biventricular repair
    • All underwent EFE resection
    • +/- other rehabilitation procedures

• Case - Matched Controls
  – Patients with borderline LV who underwent traditional single ventricle palliation
### Preoperative patient characteristics and size of LH structures

<table>
<thead>
<tr>
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<th>Left heart rehabilitation (n = 34)</th>
<th>Traditional SVP (n = 34)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Male</strong></td>
<td>25 (73%)</td>
<td>27 (79%)</td>
<td>NS</td>
</tr>
<tr>
<td><strong>LVEDV Z-score</strong></td>
<td>-2.5 ± 1.2</td>
<td>-2.9 ± 0.2</td>
<td>NS</td>
</tr>
<tr>
<td><strong>LV Long Z-score</strong></td>
<td>-2.9 ± 1.8</td>
<td>-3.2 ± 1.9</td>
<td>NS</td>
</tr>
<tr>
<td><strong>Aortic annulus Z-score</strong></td>
<td>-3.1 ± 1.0</td>
<td>-2.9 ± 1.0</td>
<td>NS</td>
</tr>
<tr>
<td><strong>Mitral valve Z-score</strong></td>
<td>-2.0 ± 1.3</td>
<td>-2.2 ± 1.6</td>
<td>NS</td>
</tr>
<tr>
<td><strong>Fetal balloon aortic valvuloplasty</strong></td>
<td>14 (41%)</td>
<td>2 (6%)</td>
<td>( P&lt;0.01 )</td>
</tr>
<tr>
<td><strong>Postnatal balloon aortic valvuloplasty</strong></td>
<td>19 (56%)</td>
<td>5 (15%)</td>
<td>( P&lt;0.01 )</td>
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LV Recruitment Strategy

Stage 1 / LH rehab
34 Patients

BDG / LH rehab
3 patients

Death
3 (9%)

Fontan / LH rehab
15 Patients

Biventricular conversion
12 Patients

Cardiac Transplantation
1 patient
LH dimensions over time in LV rehab vs. control

- LVEDV
- Aortic valve
- LV long axis
- Mitral valve

Time (months)
Ejection Fraction by Echo with Staged LV Recruitment

Ejection Fraction %

Age (months)

SE and 95% CI for regression estimate
Biventricular conversion follow up

- 13 patients have undergone biventricular conversion
- No operative or long term mortality
- One patient underwent cardiac transplantation early following conversion
- Median follow up of 2.9 yrs (range 1-6 yrs)
- 3 reoperations in 2 pts for MR (2 repairs, 1 replacement)
- Mean AoV gradient at recent follow up 12±4 mmHg
Biventricular Conversion - Overview

- Patient Selection
- Operative strategies
- Outcomes
- Postoperative care
- Future directions
Left Atrial Hypertension

- Compliance of ventricle may worsen immediately after EFE resection
- Occurs commonly with restriction of ASD
- Magnified by presence of residual LH disease (valvular stenosis or regurgitation)
Left Atrial Hypertension

• Long Term effects
  – Pulmonary edema
  – Pulmonary hypertension
  – Airway compression
• Cath Lab dilation of restrictive ASD
  – ± stent
  – 9/14 patients in our series required ASD dilation
Conclusions

• LV salvage is possible after single ventricle palliation
• This strategy requires multiple operations and interventions
• Left sided structures grow over time
• Growth may be associated with ASD restriction
• Results of Biventricular repair are acceptable at short term follow-up
Future Directions

- Optimize patient selection - who are most likely to reach conversion
- Define optimal size of ASD for LV growth
  - Compromise between LV growth and LA HTN
- Study effect of EFE resection on ventricular compliance
- Effect of LV recruitment upon single ventricle function
- Multi Site pacing for dyssynchrony
- Increase use of MRI and 3D echo to assess LV capabilities and valve repair