Techniques for Recruiting the Borderline Left Ventricle

Ram Emani
Boston Children's Hospital
Borderline Left Heart

- Surgery can address:
  - Aortic / mitral Valve if annulus OK
  - LVOT pathology
  - Arch Hypoplasia / Coarctation
  - Endocardial Fibroelastosis
  - VSD
- Surgery MAY be able to influence:
  - Size of LH structures (LV, AoV, MV)
- What we cannot fix:
  - Systolic dysfunction
  - Non compliant LV
  - Pulmonary Vascular Disease
Borderline Left Heart Structures

Single Ventricle Palliation

Initial Biventricular Repair
Borderline Left Heart Structures

- Single Ventricle Palliation
- Initial Biventricular Repair
- Transplantation
- Staged LV Recruitment
- Comfort Care
Staged LV Recruitment

Initial Single Ventricle Palliation

↓

Maneuvers to rehabilitate L heart

↓

Subsequent biventricular conversion
Considerations / Assessment

- Echocardiogram
  - LV
    - EDV < (20 ml/m2) or Z < (-3)
    - “not apex forming” < 2/3
    - Systolic dysfunction (> moderate)
  - Aortic and mitral valve
    - annulus Z < -3
    - MV – single papillary
  - Retrograde flow in transverse arch
  - Presence of EFE
  - Multi-level obstruction
  - Beware of gradients in face of ASD / low CO

Staged LV recruitment

or

Single ventricle palliation
Considerations / Assessment

- Echocardiogram
  - RV Systolic dysfunction (≥ moderate)
  - Tricuspid Regurgitation (≥ moderate)
  - Pulmonary vein concerns (TAPVR, PV stenosis)

- Non cardiac morbidity
  - Genetic Syndrome
  - Significant Airway / Lung disease

Biventricular Repair

or

Staged LV Recruitment
Timeline - Staged LV recruitment

• Neonatal Stage 1 Palliation
• 4-6 mo – Bidirectional Glenn and LV recruitment
  – HLHS - u CAVC
    • Ao V / MV repair septate common AVV ± cleft closure
    • EFE resection VSD closure
    • Fenestrated ASD closure (4 mm)
    • Consider leaving shunt in place

– Patient still has 1V circulation
Mitral valve repair

Divide secondary Chords
Split Papillary muscles
Debride Papillary muscle
Leaflet debridement
Leaflet Augmentation
Mitral valve repair

Divide secondary Chords
Split Papillary muscles
Debride Papillary muscle
Leaflet debridement
Leaflet Augmentation

Take down posterior +/- anterior leaflet
Mitral valve repair
EFE resection

Trans Mitral approach
Septum, Papillary, Apex
Difficult to get LVOT EFE
Difficult as Neonate
Taking down Anterior leaflet improves exposure to septal surface
EFE resection
Aortic Valve repair

- Commissurotomy for stenosis
- Leaflet augmentation for regurgitation
- Tricuspidization of Bicuspid valve
Aortic Valve repair

- Leaflet debridement
- EFE extends from LVOT onto leaflet
- Relieve AS at the risk of AI?
Accessory pulmonary blood flow

Sano or BTS
Increases flow through lungs
Generally done after Glenn
Only minimal increase in Glenn pressures
ASD restriction

Can partially close asd
Fenestrated ASD patch
(4 mm)
Timeline - Staged LV recruitment

- 6 mo – 2 yrs - Close follow up
  - Trans-septal gradient of 5-8 mm Hg ideal
  - Watch for desaturation / SVC syndrome
  - Low threshold for cath to dilate ASD or occlude shunt
- 2 - 4 yrs – Assess adequacy of LV
  - MRI – LV volume / transmitral flow calculation
  - Cath – LVEDP with test occlusion of ASD
  - Echo – residual valve pathology
- Decision for biventricular conversion vs. Fontan - a process in evolution
Biventricular conversion

- Disconnect aorta-pulmonary connection
- Ross or Aortic valve repair
- Reconnect pulmonary artery
- Reconnect Aorta
- Take down Glenn
Valve replacement

Prosthetic Options Limited

If Staged LV Recruitment
  – avoid replacement
  – Influence decision for BiV conversion

If Biventricular circulation
  – Residual disease impacts LV function
  – low threshold for replacing
Staged LV Recruitment

- Staged LV Recruit: 96 Patients
  - SV circulation: 61 Patients (2 pts)
  - Bi V conversion: 35 Patients (7 pts)
  - Death / OHT / VAD: 9 patients
Summary

• Tools to guide BiV vs. SVP imperfect
• When in doubt – Stage 1 palliation
• Staged LV recruitment = SVP + Left heart rehab
• MV repair – focus on subvalvular apparatus
• Aortic Stenosis – leaflet debridement
• EFE – takedown anterior leaflet for superior exposure
Following Bi V conversion

The most recent RV pressure estimated by echocardiography was less than half-systemic in 40%
Expandable Mitral Valve?
Major Risk factors for SVP?

Yes

Tolerate Discontinuation of PGE?
Yes

Subsequent failed BiV circulation?
Ao V / MV repair
EFE resection
LA hypertension?
Transplant vs. VAD early

No

Tolerate Discontinuation of PGE?
No

Staged LV recruitment
Bi V conversion
LA hypertension?

Traditional SVP

Severe TR / RV dysfx
Genetic Syndrome
Airway / pulmonary
Pulm Vein stenosis
TAPVR
## Borderline Left Heart Structures

<table>
<thead>
<tr>
<th>Condition/Outcome</th>
<th>Single Ventricle Palliation</th>
<th>Biventricular Repair</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Initial mortality</strong></td>
<td>~ 10%, independent of LH structures</td>
<td>- Early mortality depends upon LH structures, presence of EFE</td>
</tr>
<tr>
<td><strong>Long term</strong></td>
<td>- Fontan physiology morbidity</td>
<td>- LA hypertension / pulmonary vascular disease</td>
</tr>
<tr>
<td><strong>Bail out options</strong></td>
<td>- Transplantation</td>
<td>Early (No vascular disease) - Crossover to 1 V palliation - Transplantation</td>
</tr>
<tr>
<td><strong>Risk Factors</strong></td>
<td>genetic syndromes, TR, pulmonary / airway dz, pulm vein stenosis</td>
<td>Severe LH hypoplasia, EFE</td>
</tr>
</tbody>
</table>
Staged LV Recruitment - Rationale

- Growth potential of Left Heart
- Lessons from the fetus
  - Growth of cardiac structure stimulated by flow through that structure
- LV size increases following dilation of critical AS

Han et al. 2007 (Toronto)  McElhenney et al. 2005
FIGURE 3. Kaplan–Meier survival curve comparing the Yasui and Norwood procedures.
LVEDV Z score

Aortic Valve Z score

Mitral Valve annular Z score

LV Long axis Z score

Initial (prior to stage 1)

Pre-biational Glenn

Pre- Fontan or BiV conversion

Traditional SVP

LH Rehab
LV “Growth” associated with restriction of ASD

Figure 4A

<table>
<thead>
<tr>
<th></th>
<th>Prior to Stage I</th>
<th>Prior to BDG</th>
<th>Prior to Fontan</th>
</tr>
</thead>
<tbody>
<tr>
<td>ASD Restriction</td>
<td>N = 17</td>
<td>13</td>
<td>11</td>
</tr>
<tr>
<td>No ASD Restriction</td>
<td>N = 13</td>
<td>8</td>
<td>8</td>
</tr>
</tbody>
</table>

P = 0.12
P = 0.002
P = 0.84
Prior to Stage I
Prior to BDG
Prior to Fontan

LVEDV Z-Score
-6
-5
-4
-3
-2
-1
0
Staged LV: N =             30                                        21                                        19
Trad. SVP: N =              28                                        24                                        17

P=0.001
P<0.001
P=0.28

Traditional SVP
Staged LV Recruitment

Figure 3A

LV dimensions LV Recruit vs. SVP
Left Atrial Hypertension

- Occurs commonly with restriction of ASD
- 55% pts require balloon dilation ASD
- Magnified by presence of
  - Residual EFE
  - Residual Aortic Stenosis
  - Aortic or mitral insufficiency
  - 50% require reoperation for aortic / MV
- If Pulmonary vascular disease progresses, could jeopardize “bail out” options
- Early consideration of transplantation / SV conversion