“What is the Definition of Small Systemic Ventricle”

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Contents

• Introduction
✓ Aortic valve stenosis
✓ Aortic coarctation ± hypoplastic aortic arch
✓ Hypoplastic left heart complex
✓ RV pressure and/or volume overload
• Conclusion
Hypoplastic left heart physiology

- Inability of LV to sustain adequate CO following birth because of underdevelopment of one or more left heart structure despite surgical or medical intervention
- Physiologic, not morphologic
- Present within a relatively narrow band of broad continuum of hypoplastic lesion of left heart

*Kirklin et al, Cardiac surgery 3rd, 2003*
Pre-operative determination can be extremely difficult, particularly in the presence of a Borderline (small) ventricle!!

- Whether LV is adequate to sustain the systemic circulation
- Whether it may became adequate with the available surgical approaches, and therefore a bi-ventricular type of repair is feasible
1-1. Morphometric parameters

- MV diameter and indexed MV area
- LV inflow dimension and cross-sectional area
- Ratio between LV and RV dimension
- LV long axis to heart long axis ratio
- LVEDV, LV mass index
- Ratio of the right/left ventricular wall thickness
1-2. Morphometric parameters

- Endocardial fibroelastosis (EFE)
- Cardiac apex not formed by LV
- Diameter of LVOT
- Diameter of AV annulus
- Indexed aortic root
2. Functional parameters

• LV ejection fraction
• LVEDP
• mean PAP
• Direction of the blood flow in the ascending Aorta & the level of PDA
Quantitative evaluation allows a scientific approach to the problem.

- Tables with normal left ventricular dimensions
- $z$-values, with SD from the normal values

Inadequate to sustain a biventricular type of repair:
- Indexed MV area < 4.75 cm$^2$/m$^2$
- LV inflow dimension < 25 mm
- Ratio between the apex-to-base LV & RV < 0.8
- Aortic annulus < 6 mm

*Kirklin et al, Cardiac surgery 3rd, 2003*
4 different groups of CHD with VA concordance

- Aortic valve stenosis
- Aortic coarctation, with or without hypoplastic aortic arch
- Hypoplastic left heart complex
- RV pressure and/or volume overload
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Neonate with AS & small systemic ventricle
Risk factors of Aortic valve stenosis correlated with poor results

- LV cross-sectional area <1.6 cm², Small (60–80% of normal) and hypoplastic (<60% of normal) LVEDV, indexed LVEDV <20 mL/m², extensive EFE, LV inflow dimensions <25 mm, diameter of MV <11 mm, small MV orifice <9 mm and diameter of AV <6 mm, LVOT <5 mm, cardiac apex not formed by the left ventricle
- Preop. clinical conditions - severe CHF, cardiomegaly on X-ray and RVH (as possible indication of diminished LV volume or mass)
- LVEDP >20 mmHg and LVEF <40%, Pre-operative mean PAP >50 mmHg
A very extensive retrospective analysis by the Boston group on echo data

- left ventricular long axis to heart long axis ratio < 0.8
- indexed aortic root diameter < 3.5 cm/m²
- indexed mitral valve area < 4.75 cm²/m²
- LV mass index < 35 g/m²

Rhodes, JACC, 1991
Score of Rhodes

- Based on multivariate analysis, the ‘Score of Rhodes’
  \[=14.0 \text{ (BSA)} + 0.943 \text{ (iROOT)} + 4.78 \text{ (LAR)} + 0.157 \text{ (iMVA)} - 12.03\]
- allowed prediction of death after bi-ventricular type of treatment in the presence of a discriminating Score < - 0.35

*Rhodes, JACC, 1991*
A prospective multi-institutional clinical study: risk factors for hospital mortality by the CHSS

• Predict the best chance for survival
• Bi-ventricular type of repair - younger age, presence and degree of EFE, LV length and lower $z$-value of the aortic valve diameter (only aortic valvotomy ? )
• Uni-ventricular type of repair - presence of moderate or severe TV regurgitation, lower diameter of the ascending aorta

http://www.chssdc.org/
Hemodynamic and morphologic predictors, John Kovalchin, 1998, JACC

Table 2. Comparison of Hemodynamic Variables for Survival and Nonsurvival in Patients With Two-Ventri cle Repair

<table>
<thead>
<tr>
<th>Variable</th>
<th>All Pts With Two-Ventri cle Repair</th>
<th>Norwood Group: All Pts</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Survival (n = 14)</td>
<td>Nonsurvival (n = 5)</td>
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<tr>
<td>Ascending aortic flow</td>
<td></td>
<td></td>
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<tr>
<td>Antegrade</td>
<td>13</td>
<td>2</td>
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<tr>
<td>Retrograde</td>
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<td>3</td>
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<tr>
<td>Transverse arch flow</td>
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<td></td>
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<tr>
<td>Antegrade</td>
<td>12</td>
<td>2</td>
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<tr>
<td>Retrograde</td>
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<td>3</td>
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<tr>
<td>Ductus arteriosus flow</td>
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<tr>
<td>Antegrade</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>Retrograde</td>
<td>6*</td>
<td>4</td>
</tr>
<tr>
<td>Aortic valve Doppler gradient (mm Hg)</td>
<td>52 ± 23</td>
<td>27 ± 17</td>
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<tr>
<td>LVEF</td>
<td>0.27 ± 0.12</td>
<td>0.19 ± 0.16</td>
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</tbody>
</table>
Hemodynamic and morphologic predictors
John Kovalchin, JACC, 1998

Table 3. Comparison of Morphometric Variables for Survival and Nonsurvival in Patients With Two-Ventricle Repair

<table>
<thead>
<tr>
<th>Variable</th>
<th>All Pts With Two-Ventricle Repair</th>
<th>Norwood Group: All Pts (n = 9)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Survival (n = 14)</td>
<td>Nonsurvival (n = 5)</td>
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<tr>
<td>Age at initial procedure (days)</td>
<td>15.5 ± 22.1</td>
<td>1.4 ± 0.9</td>
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<td>Body surface area (m²)</td>
<td>0.23 ± 0.03</td>
<td>0.24 ± 0.03</td>
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<tr>
<td>Indexed aortic annulus (cm²/m²)</td>
<td>2.9 ± 0.5</td>
<td>1.9 ± 0.3</td>
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<tr>
<td>Indexed aortic root (cm²/m²)</td>
<td>3.6 ± 0.5</td>
<td>2.6 ± 0.6</td>
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<tr>
<td>Indexed ascending aorta (cm²/m²)</td>
<td>3.5 ± 0.7</td>
<td>2.5 ± 0.3</td>
</tr>
<tr>
<td>Indexed LV long axis (cm²/m²)</td>
<td>13.2 ± 2.5</td>
<td>10.0 ± 0.8</td>
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<tr>
<td>Indexed transverse arch (cm²/m²)</td>
<td>2.3 ± 0.3</td>
<td>2.0 ± 0.3</td>
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<tr>
<td>Indexed aortic isthmus (cm²/m²)</td>
<td>1.9 ± 0.4</td>
<td>1.8 ± 0.3</td>
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<tr>
<td>Relative LV long axis</td>
<td>0.9 ± 0.1</td>
<td>0.9 ± 0.1</td>
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<tr>
<td>Indexed LV EDV (cm³/m²)</td>
<td>42.6 ± 26.8</td>
<td>21.8 ± 7.2</td>
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<tr>
<td>Indexed LV ESV (cm³/m²)</td>
<td>33.1 ± 24.5</td>
<td>18.2 ± 8.6</td>
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<td>Indexed LV mass (g/m²)</td>
<td>85.4 ± 30.1</td>
<td>88.3 ± 41.3</td>
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<tr>
<td>Indexed MV area (cm²/m²)</td>
<td>6.3 ± 2.1</td>
<td>4.8 ± 0.8</td>
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<tr>
<td>Relative MV area</td>
<td>0.6 ± 0.2</td>
<td>0.5 ± 0.1</td>
</tr>
</tbody>
</table>
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• Conclusion
COA ± hypoplastic aortic arch

• Main difference between critical AS and COA (even indexed LVEDV 10 mL/m2 and cardiac apex not formed by LV)

  “Predictable growth of LV”

• Morphometric parameters like the score of Rhodes should be used with caution in lesions with systemic obstructions
Two considerations: hemodynamic parameters and the presence of ASD

- Antegrade flow in the ascending aorta & presence of bi-directional blood flow at the level of PDA characterize LV with potential for growth
  → suitable for bi-ventricular type of repair
- Pre-operatively a significant left-to-right shunting at the atrial level, largely due to diastolic dysfunction secondary to the increased after-load
Follow-up of COA repair in neonate,
Puchalski et al, 2004, JACC

MV

AV

* p < 0.001

2005-11-03 small systemic ventricle 22
LV volume after correction of isolated COA in neonate, Krauser, 2000, Am J Card
**LV volume after correction of isolated COA in neonate, Krauser, 2000, Am J Card**

<table>
<thead>
<tr>
<th>Pt.</th>
<th>Additional Lesions</th>
<th>BSA</th>
<th>ROOT</th>
<th>LAR</th>
<th>MVA</th>
<th>LV Volume (ml [ml/m²])</th>
<th>%Δ</th>
<th>Rhodes</th>
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<td>0.83</td>
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<td>2</td>
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<td>0.23</td>
<td>3.95</td>
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<td>3.98</td>
<td>8.2[35.5]</td>
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<td>0.23</td>
<td>4.78</td>
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<td>3.69</td>
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<td>3.29</td>
<td>0.97</td>
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<td>0.23</td>
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<td>VSD</td>
<td>0.27</td>
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<td>0.27</td>
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<td>3.28</td>
<td>9.9[43.0]</td>
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<td>10</td>
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<td>11</td>
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<td>0.79</td>
<td>1.97</td>
<td>4.5[20.3]</td>
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<tr>
<td>12</td>
<td>PFO</td>
<td>0.26</td>
<td>3.96</td>
<td>0.97</td>
<td>4.58</td>
<td>14.3[54.8]</td>
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<td>13</td>
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<td>0.24</td>
<td>4.46</td>
<td>0.82</td>
<td>3.85</td>
<td>10.1[42.0]</td>
<td>8.9[37.1]</td>
<td>8.8[36.5]</td>
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<tr>
<td>14</td>
<td>VSD, PFO</td>
<td>0.21</td>
<td>4.81</td>
<td>0.95</td>
<td>4.81</td>
<td>8.0[38.1]</td>
<td>7.5[35.7]</td>
<td>7.3[34.8]</td>
</tr>
</tbody>
</table>

**Mean** | 0.24 | 4.1  | 0.89 | 3.7 | 8.9[36.5] | 7.9[32.6] | 9.1[37.5] | 18.4 | 0.06 |

**SD** | 0.02 | 0.6  | 0.07 | 1.1 | 3.0[10.2] | 3.0[10.5] | 3.0[10.0] | 21.0 | 0.95
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  ✓ Hypoplastic left heart complex
  ✓ RV pressure and/or volume overload
• Conclusion
Hypoplastic left heart complex

• Favorable end of the spectrum of HLHS
• Characterized by hypoplasia of the structures of the left heart-aorta complex,
• Aortic and mitral valve hypoplasia without valvular stenosis or atresia, hypoplasia of LV, hypoplasia of LVOT, hypoplasia of the ascending aorta and of the aortic arch ± COA.
Before and after aortic coactectomy in patient with the borderline LV
Hypoplastic left heart complex - pathophysiology

- Constant presence of antegrade blood flow in the ascending aorta and the proximal branches of the aortic arch
- Absence of LV EFE
- Growth of LV, proved after bi-ventricular repair
Controversy for management of the frequently associated inter-atrial communication.

- **Benefit**
  - reduce the risk of low CO
  - improve the left ventricular filling and therefore the subsequent left ventricular growth

- **Risk**
  stormy post-operative course with very elevated left atrial pressure particularly during the first 24–48 hr

- Neonate with hypoplastic left heart complex
- Severe aortic coarctation
- Hypoplastic aortic arch and ascending aorta
- Large ASD (diameter=10 mm)
- Small MV, 6mm, TV, 11 mm)
- Small LVEDV 14.5 mL/m2
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• Neonate with TAPVR
  small LV
• LVEDV=14 mL/m²
RV pressure and/or volume overload

- Total anomalous pulmonary venous connection,
- Unbalanced atrio-ventricular septal defect
- Cor triatriatum
RV pressure and/or volume overload

- LV with normal mass, despite a reduced size of the aortic valve circumference

→ caused by low LV output probably compromised by
- the left to right shunt
- the septal displacement
Hypoplastic LV or simply a squashed ventricle? Collin Phoon, 1997, JACC

- Circumference
- Potential area
- Potential LV volume

\[ r = 0.75, \; p < 0.0001 \]
Hypoplastic LV or simply a squashed ventricle? Collin Phoon, 1997, JACC

- Hypothesized PO LV capacity may be better reflected by Preop. potential LV volume
  - calculated as LV volume if the septal position were normal, without the reverse bowing of the inter-ventricular septum
  - the right-to-left bowing of the inter-ventricular septum does not induce any change of the endocardial circumference nor of the LV length
Hypoplastic LV or simply a squashed ventricle? Collin Phoon, 1997, JACC

- Small pre-operative LV volume is not primarily due to compression by an overloaded right ventricle, but rather to underfilling of LV
- “LV is not ‘hypoplastic’, is simply ‘squashed’”
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Elements to be considered for the decision making process

- Morphometric parameters
- Functional parameters
- Hemodynamic data
- Available surgical options
- Results of the personal and institutional experience
Following considerations may help…

- Fontan after a failed attempt of 2 ventricular repair → increased mortality
  vs. the opposite → better outcome
- the question ‘Is a high-risk bi-ventricular repair always preferable to conversion to a single ventricle repair?’
- the most important consideration ‘every patient is unique’