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BIVENTRICULAR REPAIR OR  
FONTAN
Introduction

- Aim
- Risks / Benefits
- Anatomy
- Hemodynamics
- Long Term Outcomes
- Families
Fontan

- How to interpret studies
  - Morphology
  - Hemodynamics
  - Pre Fontan Palliation
  - Fontan Modification
  - Time
Anatomic subtypes

- HLHS 53%
- Single RV 8%
- Heterotaxy 7%
- LV-TGA 14%
- LV-NRGA 13%
- Other 5%

Kaplan-Meier estimate of freedom from death or transplantation

5 yr Freedom from Death or Transplant: 94.9% (91.6-97.0%)  Risk Set = 230
10 yr Freedom from Death or Transplant: 93.0% (88.8%-95.7%)  Risk Set = 78

Number of Fontan-related rehospitalizations per patient

<table>
<thead>
<tr>
<th>Health Status</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Excellent</td>
<td>57.1%</td>
</tr>
<tr>
<td>Good</td>
<td>37.5%</td>
</tr>
<tr>
<td>Poor</td>
<td>0.8%</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>School Performance</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Above Average</td>
<td>29.8%</td>
</tr>
<tr>
<td>Average</td>
<td>39.9%</td>
</tr>
<tr>
<td>Below Average</td>
<td>30.2%</td>
</tr>
</tbody>
</table>
Hasaniya N. W. et al.; J Thorac Cardiovasc Surg 2010;140:1076-1083
Biventricular Repair

- Ventricular Size, Position and Relation
- Atrioventricular Valves
- Aortic Valve
- Ventricular Septal Defect
- Coronary Anatomy
cc Transposition of the Great Arteries

- Definition
- Biventricular Repair
- Anatomic Repair
- Double Switch Operation
- ? Fontan
Survivals for the following operative groups: Fontan pathway (dotted line; n = 17), VSD surgery (solid line; n = 76), and TV surgery (dashed line; n = 14)

Freedom from right ventricular dysfunction after 2-ventricle intracardiac procedure (75 patients analyzed)

Survivals for patients with CHB (dotted line; n = 35) and without CHB (solid line; n = 78)

A comparison of freedom from death/transplantation after the double-switch procedure (A); death/transplantation, moderate-to-severe morphologic left ventricle (mLV) dysfunction, or both after the double-switch procedure (B); and the actuarial prevalence of patients free from death/transplantation but with moderate-to-severe mLV dysfunction (C).
A. The probability of survival, in years, for the following operative groups: conventional repair (thin line, n = 36), conventional Rastelli procedure (round dotted line, n = 31), DSO (dotted line, n = 15), Senning/Mustard-Rastelli procedure (solid line, n = 69), Fontan pathway (dashed line, n = 38)

A, The probability of survival, in years, for the following operative groups: TR (-) (thin line, n = 120) and TR (+) (thick solid line, n = 29)


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A, Freedom from reoperation for the following operative groups: conventional repair (thin line, n = 36), conventional Rastelli procedure (round dotted line, n = 31), DSO (dotted line, n = 15), Senning/Mustard-Rastelli procedure (solid line, n = 69), Fontan pathway (dashed line, n = 38)

cc Transposition of the Great Arteries

- Dextrocardia especially Senning/ Rastelli
- Anomalous Coronary
- Inlet VSD
Inadequate Left Ventricle and Aortic Stenosis

- Can be difficult especially in Neonate
- PDA Status
- Hickey 2007
  - Risks for Death in Biventricular Repair
    - Endocardial Fibroelastosis
    - Lower z Score of the Aortic Valve
    - Age
  - Risks for Death in Univentricular Repair
    - TR
    - Ascending Aorta Size
Inadequate Left Ventricle and Aortic Stenosis

- Hickey 2007
  - Overuse of Biventricle Repair
  - CHSS Calculator
Inadequate Atrioventricular Valve

- Z scote less than -3
- Coronary Fistulae
Unbalanced Complete AV Canal

- Difficult
- Valve Anatomy
- Ventricular Size
- Aortic Stenosis/ Aortic Coarctation
- Down Syndrome and Elevated PAP
Unbalanced Complete AV Canal

- Atrioventricular Valve Index (AVVI)
  - Left atrioventricular valve area/total atrioventricular valve area
  - CHSS
  - 356 patients

Jegatheeswaran A et al, Circulation 2010;122S:S209-S215
Unbalanced Complete AV Canal

Jegatheeswaran A et al, Circulation 2010;122S:S209-S215
Unbalanced Complete AV Canal

Probability of a given repair bases on AVVI

Overall Survival

Jegatheeswaran A et al, Circulation 2010;122S:S209-S215
Unbalanced Complete AV Canal

- Left Ventricular Inflow Index
  - As the secondary Color inflow diameter indexed to the left atrioventricular valve annulas diameter

Unbalanced Complete AV Canal

Heterotaxy

- Complex Ventricular Relation
- Venous Abnormalities
- Complex Baffle ad Conduit
- Outcomes of Fontan
(A) Kaplan-Meier estimated survival of patients with heterotaxy syndrome after a modified Fontan procedure

Summary

- Difficult
- Evaluation
  - Anatomy
  - Physiology
  - Reoperation rate
  - Fontan Outcomes
  - Family
  - Cost