Transplantation for Failed Fontan

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488 patients transplanted for CHD (CTRD & PHTS)
Fontan Failure

- Bi-modal
  - Early
    - Unsuspected risk factors
    - Intraoperative myocardial injury
- Late

Systemic-Ventricular Dysfunction

Elevated PVR

Lack of **pulsatility**

Pulmonary recurrent **microemboli**

Elevated pulmonary **lymphatic** pressure

Disordered **NO synthesis/endothelial** dysfunction

Valve Dysfunction and Subvalvar Stenosis

Lymphatic Derangement: PLE (3-15% incidence)

Plastic Bronchitis

50% mortality

Pulmonary AV Malformation

Thrombotic Circuit Occlusion (20% incidence, 25% mortality)

Intractable Arrhythmias
Outcome following “perfect” Fontan
Circulation 1990; 81:1520-36
Indications for Heart Transplantation

- **Failing Fontan Physiology**
  - No obvious remediable lesions
  - Poor ventricular Function

- **Timing**
  - Early
  - Difficult due to disparate modes of failure
  - Evidence of systemic mal-perfusion
  - Lack of reliable mechanical assist methods
- Analysis of anatomy
  - Cardiac Situs
  - Great vessels
  - Location of pulmonary and systemic venous returns
- Fontan type
- Aortopulmonary collateral vessels
- Patent vessels for both hemodynamic monitoring and peripheral cannulation site.
Pulmonary Vascular Resistance in Failed Fontan

- Difficult to assess and often “unreliable”
  - Systemic - to - pulmonary collateral arteries
  - Low cardiac output
  - Non pulsatile flow
  - Insidious Pulmonary Emboli
  - (Presence of pulmonary AV malformation)
Knowledge of recipient anatomy is critical

Vast majority of recipients require vascular reconstruction (PA, systemic or pulmonary veins)

Retrieval of:
- Aortic arch and head vessels
- Branch pulmonary arteries to the hilum
- SVC & innominate vein (for bilateral SVC)
- IVC
Recipient Procedures

- Safe re- sternotomy
  - Multiple reoperation
  - Anterior aorta
- Peripheral CPB may be necessary (> 40% of patients)
- Bi-caval venous anastomosis (less TR)
Bilateral SVC
IVC interruption with azygous drainage

Left sided - Hepatic drainage

Rotated 30-45° in a counter-clockwise
Post - Transplant Survival

J Am Coll Cardiol 2009; 54:160-5 (data from CTRD & PHTS)
Patient Survival (%)

Time Post-Transplant (Years)

Glenn (n=11)

Fontan (n=24)

p = 0.06

J Am Coll Cardiol 2004; 44:2065-72 (data from Philadelphia)
### Causes of death after Transplantation

<table>
<thead>
<tr>
<th>Cause</th>
<th>Fontan Patients (n=23)</th>
<th>CHD (n=47)</th>
<th>Non - CHD (n=85)</th>
</tr>
</thead>
<tbody>
<tr>
<td>*Infection</td>
<td>7 (30)</td>
<td>10 (21)</td>
<td>11 (13)</td>
</tr>
<tr>
<td>*Early graft failure</td>
<td>4 (17)</td>
<td>5 (11)</td>
<td>7 (8)</td>
</tr>
<tr>
<td>Rejection</td>
<td>3 (13)</td>
<td>6 (13)</td>
<td>15 (18)</td>
</tr>
<tr>
<td>Sudden death</td>
<td>3 (13)</td>
<td>10 (21)</td>
<td>13 (15)</td>
</tr>
<tr>
<td>Graft CAD</td>
<td>2 (9)</td>
<td>0</td>
<td>10 (12)</td>
</tr>
<tr>
<td>*Hemorrhage/tech. /Op.</td>
<td>2 (9)</td>
<td>1 (2)</td>
<td>1 (1)</td>
</tr>
</tbody>
</table>

*Circulation 2006; 114:273-280 (data from PHTS)*
Risk factors of Death

Failing Fontan with “Impaired” ventricular function

Failing Fontan with “Preserved” ventricular function

Ann Thorac Surg 2009; 88:558-64 (data from Boston)
Circulation 2006; 114:273-280 (data from PHTS)
Outcomes are acceptable.

Transplantation for “failed Fontan” is technically challenging.

Knowledge of recipient’s cardiac morphology is very important.

Measurement of pulmonary vascular resistance is unreliable.

Clear understanding of its risks and pitfalls is necessary to maximize survival and quality of life among Fontan patients.