Mechanical Support in the Failing Fontan-Kreutzer

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Fontan-Kreutzer Failure

Early vs. Chronic

Mode of Failure:
Pump vs. Non Pump

Fontan-Kreutzer Failure

Mode of Failure:

Pump
Indications for Mechanical Support

- Failed maximal medical therapy with no surgical options
- Inability to wean from cardiopulmonary bypass
- Cardiogenic Shock
- Decreased organ perfusion
- Intractable Arrhythmias

Bridge to Transplant or Destination Therapy
VAD: Surgical Challenges

**Relative Contraindications**

- Patient is NOT a Transplant Candidate
  - Exception – Pulmonary Hypertension: Appropriate for some patients
- Anatomical Diagnosis
- Neurologic Concern
  - Intracranial Hemorrhage, Severe Neurologic Impairment
- Isolated Pulmonary Dysfunction
- Sepsis
- Multisystem Organ Failure
**Mechanical Assist Devices for the Fontan-Kreutzer**

**Cardiac Dysfunction:**
1. Severe systolic dysfunction
   - Less than 35% by echocardiogram
   - Less than 30% by MRI
2. Moderately depressed dysfunction when accompanied by moderate or severe systemic AV valve regurgitation
3. Significant growth derangement or failure to thrive
4. Decreasing exercise tolerance by patient report as measured on sequential exercise testing
5. Significant electrophysiologic abnormalities including arrhythmia or aborted sudden death

**Fontan Pathway Dysfunction:**
1. Symptomatic chronic fluid overload despite increasing diuretic therapy
2. Occurrence of chronic ascites or pleural effusion refractory to therapy
3. Major hemodynamic disturbance resulting in symptoms including diastolic failure or symptomatic cyanosis

**Lymphatic Dysfunction:** PLE or plastic bronchitis requiring multiple admissions in 12 months

**Extracardiac Dysfunction:** Hemoptysis, liver disease or chronic kidney disease
Heart Transplantation in Children after a Fontan Procedure: Better than People Think

Kirk R. Kanter

Figure 1 Kaplan-Meier actuarial freedom from death stratified by patients who had a previous Fontan procedure (closed boxes) and those who did not (open circles). The survival estimates are not statistically different (P = .2622). The oval emphasizes the increased attrition in the first 6 months after transplantation in the Fontan patients.
Is Four Stage Management the Future of Univentricular Hearts? Destination Therapy in the Young

Robert D.B. Jaquiss, and Hamza Aziz


Table 4 Comparison of Transplant Versus Fontan with Durable RVAD

<table>
<thead>
<tr>
<th></th>
<th>Transplant</th>
<th>Fontan + RVAD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Resting cardiac output</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Exercise cardiac output</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Hepatic venous pressure elevated</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>Chronic anticoagulation</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Power cord</td>
<td>No</td>
<td>Probably</td>
</tr>
<tr>
<td>Immunosuppression risks</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Diabetes, hypertension, renal failure</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>Additional surgery needed?</td>
<td>Yes (re-do transplant)</td>
<td>Yes (re-do VAD or transplant)</td>
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Abbreviations: RVAD, right ventricular assist device; VAD, ventricular assist device.
# Mechanical Assist Devices Choices at CHOP

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<th>Position</th>
<th>Pulsatile</th>
<th>Discharge</th>
<th>Destination</th>
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<tr>
<td>ECMO</td>
<td>Paracorporeal</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>Pedimag</td>
<td>Paracorporeal</td>
<td>N</td>
<td>N</td>
<td>N</td>
</tr>
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<td>Berlin</td>
<td>Paracorporeal</td>
<td>Y</td>
<td>N</td>
<td>N</td>
</tr>
<tr>
<td>Heartware</td>
<td>Intracorporeal</td>
<td>N</td>
<td>Y</td>
<td>Y</td>
</tr>
<tr>
<td>HM3 Thoratec</td>
<td>Intracorporeal</td>
<td>N</td>
<td>Y</td>
<td>Y</td>
</tr>
<tr>
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<td>Intracorporeal</td>
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Options for Mechanical Support at CHOP
Patient Size

Selection of pump size

<table>
<thead>
<tr>
<th>Weight [kg]</th>
<th>Pump output [l/min]</th>
</tr>
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<tbody>
<tr>
<td>10</td>
<td>10 ml</td>
</tr>
<tr>
<td>20</td>
<td>20 ml</td>
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<tr>
<td>30</td>
<td>30 ml</td>
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<tr>
<td>40</td>
<td>40 ml</td>
</tr>
<tr>
<td>50</td>
<td>50 ml</td>
</tr>
<tr>
<td>60</td>
<td>60 ml</td>
</tr>
</tbody>
</table>

Adult Devices
Options for Mechanical Support at CHOP
Patient Size

Selection of pump size

Berlin Heart

Heartmate 3 or Heartware

Adult Devices

Adult Devices
VAD: Surgical Challenges

Basic Principles of Mechanical Assistance

– Simple, rapid employment
– Decompression of myocardium
– Maintenance of end-organ perfusion
  • Elimination of edema
  • Improvement in nutrition
– Long-term reliability
– Portability
  • Ability to rehabilitate

\[\text{Acute} \quad \downarrow \quad \text{Chronic}\]
Surgical Challenges – Implantable VAD

- Consideration of chest or abdominal DOMAIN
  - Chest closure challenging. Need for a bioprosthetic membrane to approximate chest or abdomen (fascial closure)
- Consider AV valve resection
- Pre-peritoneal vs Intra-abdominal placement
- Make drive line tunnels LONG
- TEE use after insertion is a must!
  - No residual PFO, Obstruction of inflow cannula by septum
  - Obstruction of Right Atrial Conduit
  - Ventricular position
Table 1
Ventricular assist devices in Fontan patients.

<table>
<thead>
<tr>
<th>Author</th>
<th>Diagnosis</th>
<th>Age at VAD</th>
<th>Type of VAD</th>
<th>Outcomes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fraser et al. 2011</td>
<td>HLHS</td>
<td>15 years</td>
<td>Heartmate II</td>
<td>VAD to TXP at 71 days</td>
</tr>
<tr>
<td>Carderelli et al. 2009</td>
<td>HLHS</td>
<td>18 months</td>
<td>Berlin Heart</td>
<td>Cardiac recovery, VAD explanation at 6 mos</td>
</tr>
<tr>
<td>Russo et al. 2008</td>
<td>Tricuspid atresia</td>
<td>14 years</td>
<td>Centrifugal VAD, Heartmate</td>
<td>VAD to TXP</td>
</tr>
<tr>
<td>Calvaruso et al. 2007</td>
<td>Mitral atresia</td>
<td>10 years</td>
<td>Berlin Heart</td>
<td>VAD to TXP at 7 days</td>
</tr>
<tr>
<td>Newcomb et al. 2007</td>
<td>DILV, TGA, subpulmonary stenosis</td>
<td>25 years</td>
<td>Thoratec</td>
<td>VAD to TXP at 5 mos</td>
</tr>
<tr>
<td>Nathan et al. 2006</td>
<td>HLHS</td>
<td>4 years</td>
<td>Berlin Heart</td>
<td>VAD to TXP at 28 days, died 8 days post-txp</td>
</tr>
<tr>
<td>Frazier et al. 2005</td>
<td>Tricuspid atresia</td>
<td>14 years</td>
<td>Heartmate, IP LVAS</td>
<td>VAD to TXP at 45 days</td>
</tr>
</tbody>
</table>
VAD: Surgical Challenges – Single Ventricle

Successful implantation of a Berlin heart biventricular assist device in a failing single ventricle

Meena Nathan, MD, Christopher Baird, MD, Francis Fynn-Thompson, MD, Christopher Almond, MD, MPH, Ravi Thiagarajan, MD, MPH, Peter Laussen, MBBS, Elizabeth Blume, MD, and Frank Pigula, MD, Boston, Mass

Figure 1. The left and right ventricles were resected leaving a 5mm residual just distal from the AV valve annulus.
VAD: Surgical Challenges – Single Ventricle


**Figure 3.** The inflow cannula from the Berlin-Heart EXCOR system introduced in each atria through the cuffs.
VAD: Surgical Challenges – Single Ventricle


**Figure 4.** Connection of the outflow conduit from the Thoratec system with the pulmonary artery and the ascending aorta.
HeartWare
HeartWare Systems, Framingham, MA

- Long-term device
- Centrifugal
- Continuous flow
- 10L/min flow
- BSA as low as 0.5m²
- Discharge to home
- Small
  - Pericardial space
An overview of mechanical circulatory support in single-ventricle patients

Jacob R. Miller¹, Timothy S. Lancaster¹, Connor Callahan², Aaron M. Abarbanell³, Pirooz Eghtesady³

Figure 3 HeartWare HVAD placement in patients with a failing Fontan. (A) The HVAD inflow cannula is placed within the ventricular apex with the outflow cannula into the neo-aorta; (B) the HVAD inflow cannula is placed into the atrium with the outflow cannula into the neo-aorta.
CAUTION: ALWAYS ensure the inflow cannula position is pointed toward the mitral valve and parallel to the interventricular septum to optimize HVAD Pump operation.
**Table 2** Bioengineering Considerations for the Ideal Cavopulmonary Assist Device

1. 3-way or 4-way flow capability with a single pump/impeller
2. Percutaneous endovascular support which functions in the existing TCPC pathway and maximizes native endothelium exposure (non-synthetic pathways)
3. Optimization of passive TCPC flow patterns - whether functioning or not
4. Partial support in a location where there will be no myocardial recovery to assume function of the pump *(therefore, it is not a VAD)*
5. Pressure in the TCPC is low (10 to 12 mmHg); therefore thrombogenicity potential is high
6. Pressure boost required is very low (optimum, 6 to 8 mmHg; range, 0 to 30 mmHg)
7. Low preload and afterload dependence (a high degree of fluid slip):
   A. Prevent upstream vena caval suction collapse
   B. Prevent downstream perfusion lung injury
8. Ability to address/modify split differential vena caval inflow:
   A. SVC predominant flow in neonates/infants (SVC/IVC: 60/40)
   B. IVC predominant flow in adults (SVC/IVC: 30/70)
9. No fluid reservoir for the pump inlet to draw from
10. No barrier to recirculation should be required
11. The Fontan venous pathway must remain unobstructed during full support, during weaning, in the event of device failure, and after removal of the device

**Figure 2** Von Karman viscous pump. *Left,* Fluid is induced to rotate by disc rotation, resulting in radial outflow. The outgoing fluid is replaced by inflow from the axial field. *Right,* On both sides of the disk, this results in opposed axial (vena caval) inflow and orthogonally opposed (pulmonary arterial) outflow.

**Table 3** Therapeutic Potential of Cavopulmonary Assist

**Existing paradigm:**
- Adult failing Fontan
  - Bridge-to-recovery
  - Bridge-to-transplant
- Stage-2 and -3 repair
- Stabilization after repair
- Destination therapy: the "biventricular Fontan"
- Static percutaneous implant
- Passive flow optimization

**New paradigm:**
- Combined Stage 2-3
  - One-stage Fontan conversion
- Combined Stage 1-2
  - Norwood (no shunt) + Glenn
- Combined Stage 1-3
  - Neonatal Fontan
- Support for other hybrid approaches
  - Percutaneous Fontan completion
THANK YOU