“Failing” FONTAN
Conversion or Transplant?

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Fellow, Pulmonary Vascular Research Institute.
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• Dr. Billie-Jean Martin
• Dr. David Ross
• Dr. Isabelle Vondermuhll
• Dr. Simon Urschel
• Dr. Holger Buchholz
• Dr. Ivan Rebeyka
Surgical repair of tricuspid atresia

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FIG. 7. Case 1. Tricuspid atresia type I B. Drawing illustrates the repair: anastomosis between right atrium and proximal end of right pulmonary artery was made without interposition of an aortic valve homograft.
Figure 2. A, Preoperative angiogram of our first patient showing absent right pulmonary artery. B, Postoperative angiogram. The APA through a homograft is patent and also a fenestration (F) at the atrial septum. C, Schematic representation of APA published by ...  

Guillermo O. Kreutzer, Andrés J. Schlichter, Christian Kreutzer

The Fontan/Kreutzer Procedure At 40: An Operation for the Correction of Tricuspid Atresia

Seminars in Thoracic and Cardiovascular Surgery: Pediatric Cardiac Surgery Annual, Volume 13, Issue 1, 2010, 84–90

http://dx.doi.org/10.1053/j.pcsu.2010.01.002
Experimental Circulatory Bypass of the Right Side of the Heart

- Superior Vena Cava to Right Pulmonary Artery
  - Pleural Effusions
- Inferior Vena Cava to Right Pulmonary Artery
  - Ascites
  - Hypoproteinemia

Nuland SB, Glenn WW, Guilfoil PH: Circulatory bypass of the right heart. III. Some observations on long-term survivors Surgery: 1958 Feb;43(2):184-201
Causes of Fontan “Failure”

Unique to Fontan/Atriopulmonary Connection/TCPC
- Protein Losing Enteropathy
- Plastic Bronchitis
- Hepatic Cirrhosis
- Sequelae of Atrial/Cavo Pulmonary Surgery
  - Pulmonary vein compression
  - Pulmonary artery distortion
  - Enlarged systemic venous reservoir
  - Obstructed cavopulmonary connection
  - Arrhythmias
  - Pulmonary vascular disease

Associated with long term survival of congenital heart disease.....but poorly tolerated after Fontan/APC/TCPC
- Ventricular failure
  - Systolic
  - Diastolic
- Pulmonary vascular disease
- Arrhythmias
- Valve stenosis/regurgitation
- Aortic to Pulmonary Collateral Vessels
Figure 7. A, Patient, aged 53 years, with TA Ib after 34 years of radical FK palliation. B, Brief communication published in the Journal of Thoracic and Cardiovascular Surgery. C, Pre-reconversion Rx showing huge right atrium and severe hydrothorax. D, Rx 1 ...

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http://dx.doi.org/10.1053/j.pcsu.2010.01.002
Conduit stenosis with thrombus formation

G.O. Kreutzer: 32 years after total right heart bypass
Figure 8. A, Pre-reconversion ECG showing atrial fibrilation. B, ECG 2 years after conversion, C, Ergometric test 2 years after conversion.

Guillermo O. Kreutzer, Andrés J. Schlichter, Christian Kreutzer

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http://dx.doi.org/10.1053/j.pcsu.2010.01.002
Evolution of Single Ventricle Palliative Surgery
From Fontan to Total Cavopulmonary Connection

Figure 1 The different types of Fontan circulation. (A) Atiopulmonary connection. (B) Intracardiac total cavopulmonary connection (lateral tunnel). (C) Extracardiac total cavopulmonary connection.

Marc de Leval: Nature Clinical Practice Cardiovascular Medicine
2005:2:4:202
Courtesy of Isabelle Vondermuhl: ACHD University of Alberta Hospital and Mazankowski Alberta Heart Institute, Edmonton, Canada
Courtesy of Isabelle Vondermuhll: ACHD University of Alberta Hospital and Mazankowski Alberta Heart Institute, Edmonton, Canada
• Atriopulmonary connection
• Functional Class 4

• Conversion to extra cardiac TCPC
• Functional Class 1

Courtesy of Isabelle Vondermuhl: ACHD University of Alberta Hospital and Mazankowski Alberta Heart Institute, Edmonton, Canada
Kaplan–Meier survival curve of freedom of death and transplantation in the centre with a low threshold for Fontan conversion versus other centres.

**Graph:**
- **X-axis:** Years since Fontan conversion
- **Y-axis:** Freedom from death/or transplantation (%)
- **Legend:**
  - Other Centres
  - Early Conversion Centre

**Table:**

<table>
<thead>
<tr>
<th>Years</th>
<th>Other Centres</th>
<th>Early Conversion Centre</th>
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<tbody>
<tr>
<td>0</td>
<td>20 (6)</td>
<td>19 (1)</td>
</tr>
<tr>
<td>2</td>
<td>6 (1)</td>
<td>12 (0)</td>
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<tr>
<td>4</td>
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</tr>
<tr>
<td>6</td>
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<tr>
<td>10</td>
<td>1 (1)</td>
<td>5 (0)</td>
</tr>
</tbody>
</table>

**Note:**
- # at risk (#Fail)
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500 TCPC without fenestration


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500 TCPC without fenestration

Freedom from late-occurring morbidities.

<table>
<thead>
<tr>
<th>Freedom from new onset arrhythmias</th>
<th>Freedom from late complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tachyarrhythmia n=13</td>
<td>PLE n=8</td>
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<tr>
<td>Bradyarrhythmia n=19</td>
<td>Systemic thromboembolism n=5</td>
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<tr>
<td></td>
<td>Liver cirrhosis n=1</td>
</tr>
</tbody>
</table>

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500 TCPC without fenestration

Trends in percent predicted value of peak VO2.

Edmonton Fontan Data – WCCHN 1996-2015

- Total 315
  - HLHS 102 (32.4%)
  - Deaths < 30 days 6 (1.9%)
  - Transplants 10 (3.12%)

Martin, Ross, Rebeyka 2015
Edmonton Transplant Free Fontan Survival – HLHS vs Others

Transplant free survival

Time (years)

Number at risk
hlhs = 0 211 185 152 115 95 78 60 34 23 5 1
hlhs = 1 102 89 75 58 39 28 20 7 5 1

hlhs = 0
hlhs = 1

Martin, Ross, Rebeyka 2015
Heart transplantation
No difference in survival Fontan versus non Fontan

Fig 1. Kaplan-Meier actuarial freedom from death stratified by patients who had a previous Fontan procedure (•) and those who did not (◦). The survival estimates are not statistically different (p = 0.4975).

Kirk R. Kanter, William T. Mahle, Robert N. Vincent, Alexandria M. Berg, Brian E. Kogon, Paul M. Kirshbom
Heart Transplantation in Children With a Fontan Procedure
Figure 5. Kaplan Meier survival plot with 95% confidence limits, depicting OHT outcome for failing single ventricles, stratified by palliation staging to OHT. BDG transition to OHT showed 100% survival at 6.5 years follow-up.

Kaplan–Meier freedom from death with patients at risk in the overall cohort.
Kaplan–Meier 5-year freedom from death after orthotopic heart transplantation in late and early Fontan failure (FF) with the number of patients at risk.

Kaplan–Meier 5-year freedom from death after orthotopic heart transplantation (OHT) in early and late Fontan failure (FF) stratified by pre-OHT ventricular function (VF) with the number of patients at risk.


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Kaplan–Meier 5-year freedom from death after orthotopic heart transplantation in protein-losing enteropathy (PLE) and non-PLE with the number of patients at risk.
Conclusion

• For the “Failing Fontan”
• Optimize rhythm, trial of ERA, PDE 5 inhibitor, valve function repair/replacement
• Conversion for
  – For Fontan surgery sequelae and “earlier” Fontan variants
• Transplant
  – for complications unique to Fontan and ventricular failure
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