The Role of Fontan Conversion

Tim Hornung

Green Lane Congenital Cardiac Service
Starship & Auckland Hospital
Progressive RA Dilation  →  Atrial Arrhythmias

Atrial Arrhythmias After Atrio-Pulmonary Fontan Operation

Peters, Somerville (London)

Fishberger, Walsh (Boston)
J Th CV Surgery 1997;113:80-86.
Arrhythmia Burden:
ANZ Fontan Registry: AP Fontan Data

- 215 hospital survivors
- 130 developed atrial arrhythmias
  - 101 atrial flutter
  - 50 atrial fibrillation
  - 3 SVT
- Freedom from arrhythmia
  - At 20 years: 45%
  - At 28 years: 23%
- Development of arrhythmia increased the likelihood of death or transplantation: OR 3.1

Medical Treatment
Amiodarone-associated thyroid dysfunction: risk factors in adults with congenital heart disease.

- 92 ACHD patients taking amiodarone for > 6 months
- Mean age 34 years
- 36% developed thyroid dysfunction
- Risk factors included previous Fontan surgery (OR 4.0, p=0.17)

SA Thorne et al (Brompton Hospital); Circulation 1999;100:149-154.
Catheter Ablation
Outcome of Intra-Atrial Re-Entrant Tachycardia Catheter Ablation in Adults with Congenital Heart Disease. S-C Yap et al (Toronto). JACC 2010;56:1589-96.
Fontan Conversion
Australia and NZ Fontan Registry Data

Fontan Conversion Outcomes (Chicago)

- 140 Patients
  - Median age 23 years
  - Median time since Fontan 16 years
- Freedom from recurrent arrhythmia 77% (10 yr F/U)
- Early death or transplant 3
- Late death or transplant 25 (non-cardiac 3)
- Total death or transplant 20%

Fontan Conversion Outcomes (Chicago)

Fontan Conversion Outcomes (Chicago)

Fontan Conversion Outcomes (International)

- Multi-Centre Reviews
  - Early mortality between 5 - 10%
  - Late death or transplant 10%

- Recent single centre reports (since 2010)
  - Early mortality 0 - 15%
The Australia and NZ Experience of Fontan Conversion
“Early” vs “Late” Conversion Groups
Transplant-free Survival

Survival after Fontan Conversion: Deal, Mavroudis

“Early” vs “Late” Conversion Groups
Pre-operative Variables

<table>
<thead>
<tr>
<th></th>
<th>Early Conversion (n=18)</th>
<th>Late Conversion (n=21)</th>
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<tbody>
<tr>
<td>Age at conversion</td>
<td>25.1 yr</td>
<td>23.7 yr</td>
</tr>
<tr>
<td>Time since first arrhythmia</td>
<td>2.9 yr</td>
<td>4.5 yr</td>
</tr>
<tr>
<td>NYHA Pre-op</td>
<td>2</td>
<td>3</td>
</tr>
<tr>
<td>Number of anti-arrhythmias</td>
<td>1</td>
<td>2</td>
</tr>
</tbody>
</table>
“Early” vs “Late” Conversion
Case

- DORV, PS, L-TGA, mitral atresia, large VSD
- Atrio-pulmonary Fontan procedure - aged 21 years
- Atrial arrhythmias from 2008 (age 41 years)
- EPS + RFA 2011 - continued to have arrhythmias
Case

- RA and PA pressure 11-12 mmHg
- Transpulmonary gradient 6 mmHg, PVRi 3.3 U.m²
- Cardiac index 2.1 L/min/m²
- LVEF 41%, Moderate AR
- Normal liver function tests; Normal FVC, FEV₁

- Fontan conversion aged 45 years;
  - Discharged day 12 post-op
  - Remains well 5 years later
“Early” Fontan Conversion

- Not necessarily early in terms of age
- Perhaps early in terms of time since Fontan
  - One of the two patients who died was the furthest out from the Fontan operation (33 years post)
- Early in terms of not waiting for complications
Risk Factors

- PLE
- Severe ventricular dysfunction
- Ascites
- Cirrhosis, Liver dysfunction, HCC
- Severe AV valve regurgitation
- Non-left ventricle morphology
- Plastic bronchitis
- Renal insufficiency
### Table 3. Independent Risk Factors for Cardiac Death or Transplantation After Fontan Conversion

<table>
<thead>
<tr>
<th>Variable</th>
<th>Adjusted Hazard Ratio (95% CI)</th>
<th>p Value</th>
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<tr>
<td>Right or indeterminate ventricular morphology</td>
<td>5.71 (2.37–13.75)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Ascites</td>
<td>3.69 (1.59–8.56)</td>
<td>0.002</td>
</tr>
<tr>
<td>Protein-losing enteropathy</td>
<td>4.93 (1.16–20.98)</td>
<td>0.03</td>
</tr>
</tbody>
</table>

So when should we do Fontan Conversion?

- **Atrial arrhythmias**
  - Unless controlled easily by a single agent (?)
  - Definitely convert if amiodarone required

- **Patients requiring other surgical intervention for haemodynamic indications**
  - Valve surgery, VSD enlargement

- **Progressive symptoms / reduced VO$_2$ etc**
  - Monitor carefully with CPET, Albumin, MRI etc
And when should we **not** do Fontan Conversion?

- Factors to consider include:
  - **Protein losing enteropathy**
  - **Severe ventricular dysfunction** (unless potentially reversible)
  - Other organ system disease
    - **Cirrhosis, renal impairment, pulmonary disease**
  
  …But remember suboptimal transplant outcomes
Conclusions

• Atrial arrhythmias often become progressively harder to control medically or with ablation

• Fontan conversion can be performed with an acceptable mortality risk and with reasonably good medium-term outcomes

• Lower threshold for Fontan conversion is likely to reduce morbidity and mortality
Acknowledgements

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• CAMRI: Anna Lydon and the team

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• Starship Echo team