Long-term outcomes after first-onset arrhythmia in Fontan physiology


Royal Children’s Hospital, Melbourne
Disclosures

- Y d’Udekem: consultancy fees MSD
Background

- Arrhythmias reported in 13-54% of patients after Fontan surgery
- Incidence of tachy- vs bradyarrhythmias unclear
- Outcomes after a first onset arrhythmia unknown
- Efficacy of medical strategies (anti-arrhythmic medications, DCR, ablation) unclear
Redefining Expectations of Long-Term Survival After the Fontan Procedure
Twenty-Five Years of Follow-Up From the Entire Population of Australia and New Zealand

Yves d’Udekem, MD, PhD*; Ajay J. Iyengar, MBBS(Hons), BMedSci, GCALL*;
John C. Galati, BSc, PhD; Victoria Forsdick, MBBS;
Robert G. Weintraub, MBBS, FRACP; Gavin R. Wheaton, MBBS, FRACP;
Andrew Bullock, MBBS, FRACP; Robert N. Justo, MBBS, FRACP;
Leeanne E. Grigg, MBBS, FRACP; Gary F. Sholler, MBBS, FRACP;
Sarah Hope, BSc, BMedSci(Hons), MBChB, FRACP, PhD;
Dorothy J. Radford, MBBS, MD, FRACP; Thomas L. Gentles, MBChB, FRACP;
David S. Celermajer, MBBS, PhD, DSc, FRACP; David S. Winlaw, MBBS(Hons), MD, FRACS
ANZ Fontan Registry

Number of Patients Alive with a Fontan Circulation

Year

Number Living

ECC=851
LT=264
AP=157
Patients and methods

• In June 2014: 210 patients identified to have arrhythmias in the Australia and New Zealand Fontan Registry

• Review of every individual patients files with particular focus on onset of first arrhythmia and decreased ventricular function (mostly subjective)
Type of arrhythmias (n=210)

- **Tachy-**
  - SVT: 99
  - Atrial fibrillation: 19
  - Junctional tachycardia: 28
  - Ventricular tachycardia: 28

- **Brady-**
  - Complete heart block: 28
  - Sick sinus syndrome: 28
  - Bradycardia: 24
  - Atrial flutter: 41
  - Complete heart block: 10
Freedom from Arrhythmia

Freedom from tachyarrhythmia (a)

Freedom from bradyarrhythmia (b)

Number at Risk:
962 688 384 161 38

Number at Risk:
968 714 412 209 88

95% CI
Freedom from Arrhythmia

Atriopulmonary vs LT & ECC: p<0.01
Survival after onset of arrhythmia

Survival from onset of tachyarrhythmias (a)

Survival from onset of bradyarrhythmia (b)

72% 81%

Number at Risk:
174 127 91 49 30

Number at Risk:
80 63 37 22 10
Survival after onset of arrhythmia

### Survival from onset of tachyarrhythmia (c)

- **AP**
- **LT**
- **ECC**

<table>
<thead>
<tr>
<th>Number at Risk</th>
<th>0</th>
<th>5</th>
<th>10</th>
<th>15</th>
<th>20</th>
</tr>
</thead>
<tbody>
<tr>
<td>AP</td>
<td>103</td>
<td>83</td>
<td>63</td>
<td>35</td>
<td>24</td>
</tr>
<tr>
<td>LT</td>
<td>36</td>
<td>24</td>
<td>19</td>
<td>12</td>
<td>5</td>
</tr>
<tr>
<td>ECC</td>
<td>35</td>
<td>20</td>
<td>9</td>
<td>2</td>
<td>1</td>
</tr>
</tbody>
</table>

### Survival from onset of bradyarrhythmia (d)

- **AP**
- **LT**
- **ECC**

<table>
<thead>
<tr>
<th>Number at Risk</th>
<th>0</th>
<th>5</th>
<th>10</th>
<th>15</th>
<th>20</th>
</tr>
</thead>
<tbody>
<tr>
<td>AP</td>
<td>37</td>
<td>32</td>
<td>19</td>
<td>16</td>
<td>9</td>
</tr>
<tr>
<td>LT</td>
<td>22</td>
<td>16</td>
<td>12</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>ECC</td>
<td>21</td>
<td>15</td>
<td>6</td>
<td>2</td>
<td>0</td>
</tr>
</tbody>
</table>
Transplant free survival

No arrhythmia vs tachy- & bradyarrhythmia: p<0.01
Freedom from decreased ventricular function

Freedom from decreased ventricular function (a)

Freedom from decreased ventricular function (b)
Freedom from adverse events
(death, transplantation, Fontan conversion, protein losing enteropathy, ventricular dysfunction, thromboembolic event or NYHA III/IV)
Freedom from adverse events
(death, transplantation, Fontan conversion, protein losing enteropathy, ventricular dysfunction, thromboembolic event or NYHA III/IV)

Extracardiac conduit vs AP & LT: p=0.04
Tachyarrhythmias: treatment

- Single episode of tachyarrhythmia in 29 patients (17%)
- More than 5 episodes in 75 patients (43%)

- Anti-arrhythmic medications: 320 in 149 patients (86%)
- Direct current reversion: 275 in 82 patients (47%)
- Ablation: 86 in 57 patients (33%)
Freedom from DCR after onset of tachyarrhythmia

The graph illustrates the percentage freedom from DCR from tachyarrhythmia onset or previous DCR over years since the last DCR. The data is represented for different values of DCR (dcr = 0 to dcr = 5). The number of patients at risk decreases over time as the cumulative percentage freedom from DCR increases. The table below shows the number at risk for each group:

<table>
<thead>
<tr>
<th>Years since last DCR</th>
<th>dcr = 0</th>
<th>dcr = 1</th>
<th>dcr = 2</th>
<th>dcr = 3</th>
<th>dcr = 4</th>
<th>dcr = 5</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>174</td>
<td>102</td>
<td>82</td>
<td>65</td>
<td>53</td>
<td></td>
</tr>
<tr>
<td>2.5</td>
<td>82</td>
<td>41</td>
<td>27</td>
<td>18</td>
<td>15</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>53</td>
<td>22</td>
<td>12</td>
<td>8</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>7.5</td>
<td>28</td>
<td>22</td>
<td>12</td>
<td>8</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>21</td>
<td>7</td>
<td>4</td>
<td>3</td>
<td>0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>15</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td></td>
</tr>
</tbody>
</table>

The graph shows a decreasing trend in the number of patients at risk as time progresses, indicating a higher likelihood of freedom from DCR over time.
Freedom from death, transplant and Fontan conversion after an ablation

57%
Conclusion

• Current treatments provide symptomatic relief for around 60% of patients for 5 years
• Ablation therapy should be recommended after requirement for repeated cardioversion
• Patients with tachy- & bradyarrhythmias are at risk of decreased ventricular function
• Fontan conversion should likely be recommended at the earliest sign of decreased ventricular function.