Long-term results of a strategy of aortic valve repair in the paediatric population: Should we avoid cusp extension?

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Presenter Disclosure

Yves d’Udekem

The following relationships exist related to this presentation:

No relationships to disclose
Paediatric aortic valve disease

- Favorite approaches

**Stenosis** ►►► Balloon valvuloplasty

**Regurgitation** ►►► Ross procedure
Ross dimensions: 76 pts

- **Annulus***
  - 0.09 cm/y
  - z: 0.31/y

- **Root***
  - 0.18 cm/y
  - z: 0.75/y

- **ST Junction***
  - 0.18 cm/y
  - z: 0.59/y

Years (n):
- D/C (74)
- 0-2 (64)
- 2-4 (45)
- 4-6 (37)
- 6-8 (25)
- 8-10 (7)
The Melbourne experience

• Favour primary aortic valve repair

• We know they will likely fail

• We hope to grow the native annulus during infancy and childhood, to prepare for the best ultimate procedure.
The Melbourne experience

- Favour primary aortic valve repair

- **Aim:**
  - Review outcomes after aortic valve repair for aortic valve disease in terms of survival and need for reoperation
Patients

- **142 consecutive patients** operated between 1996 and 2009 in the Royal Children’s Hospital, Melbourne, Australia

- **Exclusion criteria**
  - International patients
  - Rheumatic heart disease
  - Simple commissurotomy
  - Aortic valve sparing root surgery

- **NB:** During the same time period, 57 primary balloon valvuloplasties were performed
Aortic valve procedures in Melbourne: Balloon valvuloplasty vs surgery
Patients’ Characteristics (n=142)

- **104 males/ 38 females**
- **Median age at surgery: 9 years (0-20)**
  - Neonates (≤ 30 days): 13
  - Infants (31- 365 days): 17
  - 1-18 years: 101
  - ≥ 18 years: 11
- **Aetiology:**
  - Congenital 138
  - Endocarditis 4
- **Previous cardiac interventions: 45 patients**
  - Balloon valvuloplasty: 17 pts
  - Aortic valve commissurotomy: 10 pts
## Stenosis/ insufficiency per age group

<table>
<thead>
<tr>
<th></th>
<th>Neonates &amp; infants (≤ 1yr) (No,%)</th>
<th>Non-infants (&gt; 1yr) (No,%)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>AS</strong></td>
<td>29 (97)</td>
<td>47 (42)</td>
<td>76</td>
</tr>
<tr>
<td><strong>AI</strong></td>
<td>1 (3)</td>
<td>54 (48)</td>
<td>55</td>
</tr>
<tr>
<td><strong>AS/AI</strong></td>
<td>0 (0)</td>
<td>11 (10)</td>
<td>11</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>30</td>
<td>112</td>
<td>142</td>
</tr>
</tbody>
</table>
Patients’ characteristics (n=142)

- 48 tricuspid valves
- 87 bicuspid valves
- 7 unicuspid valves
- 52 bicuspid
- 35 tricuspid
Patients’ characteristics (n=142)

- Patches of glutaraldehyde preserved autologous pericardium: 96 pts
  - Cusp extension: 51 pts
    - 1 cusp: 3 pts
    - 2 cusps: 7 pts
    - 3 cusps: 41 pts
  - Recreation of a new commissure: 28 pts
  - Repair of perforation: 17 pts

- Repair without patches: 46 pts
Aortic valve repair without patches (n= 46)

30 pts: Commissurotomy, leaflets thinning, resection of nodular dysplasia

8 pts: Triangular plication

19 pts: Triangular resection

7 pts: Subcommissural annuloplasty
Tricuspidisation with 3 cusp extensions

Patients
Patients’ characteristics (n=142)

48 concomitant cardiac procedures in 39 pts

- VSD closure: 17
- Relief of LVOVT: 16
- Aortic arch repair: 6
- Mitral valve repair: 4
- Other: 5
Results (n=142)

• 3 early deaths
  – 2 sudden unexpected deaths day 4 and 17
  – 1 neonate: ECMO, stroke, LOS

• 3 early reinterventions
  – 1 Ross procedure
  – 1 reoperation for bleeding
  – 1 pacemaker

• Complications
  – 1 pt: pulmonary hem., stroke without sequellae
Follow-up (n=142)

• Mean follow-up: 3.4 ± 3.5 years (up to 14y)
• 2 pts lost to f-up.
• One late non-cardiac death (MVA).
• No neurologic or thrombo-embolic events
• Reintervention: 18 pts
  – Ross procedure: 11 pts
  – Mechanical valves: 3 pts
  – Re-repair: 3 pts
  – Bentall procedure: 1 pt
Freedom from reintervention (n=142)

Indep. predictors: < 1 year (p=0.005)

cusp extension (p=0.004)
Freedom from reintervention (n=142)

- No Cusp Extension: 66%
- Cusp Extension: 90%

No. at risk
<table>
<thead>
<tr>
<th>No Cusp Extension</th>
<th>89</th>
<th>42</th>
<th>24</th>
<th>14</th>
<th>9</th>
<th>4</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cusp Extension</td>
<td>49</td>
<td>28</td>
<td>19</td>
<td>10</td>
<td>5</td>
<td>1</td>
</tr>
</tbody>
</table>
Freedom from valve replacement

Indep. predictor: cusp extension (p=0.013)
Status at last follow-up

- 119 patients surviving with their native valves
  - Moderate regurgitation: 23 pts
  - Moderate to severe regurgitation: 1 pt
  - Severe regurgitation: 1 pt
  - ≥ moderate stenosis: 12 pts
  - mean peak gradient: 26 ± 18 mmHg

- 82 patients (57%) were event-free at last f-up.
Myocardial ischemic complications

- 16 y old, Ao Stenosis
- Tricuspidisation + 3 cusps extensions
- Arrest on day 1
- ECMO, Thoratec VAD
- Heart transplantation on day 58
Myocardial ischemic complications

8 year-old, bicuspid aortic valve

LCA
Myocardial ischemic complications

- Tricuspidisation and 3 cusps extension
- Removal of cross-clamp, ongoing V fib.
Myocardial ischemic complications

- Second run
- Trimming of left cusp patch
- Pulling of commissure R/Non cor cusp
Myocardial ischemic complications

5/33 pts with bicuspid valves repaired by tricuspidisation and 3 cusps extension

<table>
<thead>
<tr>
<th>History</th>
<th>Resulting anatomy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 14 y old, <strong>sudden death</strong> day 17</td>
<td>Small L sinus</td>
</tr>
<tr>
<td>2 7 y old, <strong>sudden death</strong> day 4</td>
<td>Small R sinus</td>
</tr>
<tr>
<td>3 16 y old, <strong>cardiac arrest</strong>, VAD, Transplant</td>
<td>Small R sinus</td>
</tr>
<tr>
<td>4 3 y old, <strong>recurrent inferior isch changes</strong></td>
<td>Small R sinus</td>
</tr>
<tr>
<td>5 8 y old, <strong>V fib on unclamping</strong>, second run</td>
<td>Small L sinus</td>
</tr>
</tbody>
</table>
Conclusion

• Aortic valve repair is effective in postponing reintervention (2 thirds free of reintervention at 9 years).

• Reintervention occurs earlier if the procedure is required at a young age and if cusp extension technique is used.

• Aortic valve repair restricted to native tissues may have more promising outcomes.
Conclusion

• Aortic valve repair should be offered and attempted as the first operation.

• Patients are likely to need a reoperation …but… we hope that the aortic valve will grow to a size allowing a better Ross procedure …. 
Ross procedure: Inclusion techniques

- Patients are likely to need a reoperation ...but... we hope that the aortic valve will grow to a size allowing a better Ross procedure ....
Conclusion

• Aortic balloon valvuloplasty is associated with a 1 month rate of sudden death of 4%. It seems impossible to prove retrospectively the mechanism of ischemia in suspected cases.

• We suspect that tricuspidisation of bicuspid valves creates asymmetric valves and that, in some instances, cusp extension may cause coronary obstruction.
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