Anomalous Aortic Origin of a Coronary Artery

is always a surgical disease

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risk/benefit ratio of surgical treatment

risk of sudden death
« anatomical » repair
« anatomical » repair : result (left AAOCA)

preop

postop
« anatomic » repair: results in 19 patients

- 5-37 years (mean: 14 yrs)
- ALCA: 37% - ARCA: 63%
- sudden death: 27% - symptoms: 42% - no symptoms: 31%
- 3/12 pts with ARCA had aborted sudden death

- no mortality
- 100% patency
- no residual myocardial ischemia
- one adverse event
  (reoperation for aneurysm)
### surgical treatment: results

<table>
<thead>
<tr>
<th>Authors</th>
<th>No. of pts</th>
<th>Age</th>
<th>Surgical technique</th>
<th>Early mortality</th>
<th>Late mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Romp (2003)</td>
<td>9</td>
<td>7–65</td>
<td>Unroofing</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Davies (2009)</td>
<td>36</td>
<td>13–82</td>
<td>Coronary bypass graft (14) Unroofing (22)</td>
<td>0</td>
<td>1 (non cardiac)</td>
</tr>
<tr>
<td>Mainwaring (2011)</td>
<td>50</td>
<td>0.1–47</td>
<td>Unroofing (35) Coronary reimplantation (6) Pulmonary translocation (9)</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Mumtaz (2011)</td>
<td>22</td>
<td>5–54</td>
<td>Unroofing</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Frommelt (2011)</td>
<td>27</td>
<td>4–20</td>
<td>Unroofing</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Brothers (2007)</td>
<td>24</td>
<td>5–18</td>
<td>Unroofing</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Karl (2008)</td>
<td>7</td>
<td>2–52</td>
<td>‘Physiological’ repair</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td></td>
<td><strong>202 pts</strong></td>
<td></td>
<td></td>
<td><strong>0</strong></td>
<td><strong>1 (0.5%)</strong></td>
</tr>
</tbody>
</table>
risk of sudden death (1)

- 0.1%-0.3% of population
- RACA > LACA (x 6-10)
- both can cause sudden death
- 15%-20% of sudden deaths in athletes

Chetilin MD et al. Circulation 1974;50:780
Taylor AJ et al. Am Heart J 1997;133:428
Frescura C et al. Hum Pathol 1998;29:689
risk of sudden death (2)

- actual risk is difficult to evaluate
- high in LACA: 2%-5%
- much lower in RACA: 0.1%-0.2%
- risk factors:
  - intramural course
  - ostial slit-like deformation
  - young athletes (< 35 yrs)

Corrado D et al. JAMA 2006;296:1593
Harris KM et al. JAMA 2010;303:1255
Eckart R et al. Ann Int Med 2004;141:829
Harmon KG et al. Circulation 2011;123:1594
Redelmeier DA et al. BMJ 2007;335:1275
ACC/AHA Guidelines for the Management of Adults with Congenital Heart Disease  
*(Circulation 2008;118:e714-e833)*

- **Surgical revascularization** should be performed in patients with:
  - anomalous origin of left main coronary artery
  - anomalous origin of right artery with evidence of ischemia (symptoms ± silent ischemia)  
    (level of evidence: B)

- **Surgery** may be beneficial in patients with:
  - evidence of anatomical obstruction
  - evidence of flow restriction (intravascular ultrasound)  
    (level of evidence: C)
ACC/AHA Guidelines for the Management of Adults with Congenital Heart Disease
*(Circulation 2008;118:e714-e833)*

- In patients with anomalous right coronary artery and no evidence of ischemia, a conservative approach may be reasonable.

- This must be argued:
  - this represents the majority of patients
  - there is a risk of sudden death (even if very low)
  - physical activity must be restricted (quality of life)
  - psychological and behavioural consequences
conservative management
  - risk of sudden death
  - restriction in activity
  - impaired quality of life

surgical treatment
  - very low risk
  - high efficiency
  - normal life

for all patients with AAOCA
although long-term follow-up of operated patients is still needed
Corentin M., 14 years old
admitted into training center for professional soccer players
routine echocardiography: anomalous right coronary
no symptom, no evidence of myocardial ischemia
sports licence refused

options:
- licence authorization (despite risk of death)
- accept major change in life project
- surgery

surgery on 01/12/2014 – training center on 01/04/2015