Biventricular Repair of Complete Atrioventricular Septal Defect with Double Outlet Right Ventricle

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What’s the problem?

- Atrioventricular septal defect with common atrioventricular valve and double outlet right ventricle (CAVSD/DORV)
  - Remote nature of VSD
  - Associated defects
    - PS, TAPVC
Background

• Prevalence
  – 6.7% of CAVSD
• Surgical repair
  – High mortality
  – Univentricular palliation
  – Biventricular repair
Repair Techniques

- Two-patch transatrial repair of CAVSD
- Enlargement of the VSD
- Tunnel repair
- RVOT reconstruction
- Correction of venous anomalies
Surgical Anatomy
Surgical Technique
Surgical Technique
Surgical Technique
Surgical Technique
Biventricular Repair of CAVSD/DORV with Heterotaxy and PS or PA

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Study Purpose

• Review a group of patients who had undergone biventricular repair of CAVSD/DORV

• Assess patient outcome with respect to anatomy and surgical technique

• Composite primary endpoint
  – Death or significant complication

• IRB approval obtained
# Patient Demographics (N=16) March 1991 to Present

<table>
<thead>
<tr>
<th>Category</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>Male / Female</td>
<td>6/10</td>
</tr>
<tr>
<td>Median age at biventricular repair</td>
<td>16 months</td>
</tr>
<tr>
<td>Anatomic features</td>
<td></td>
</tr>
<tr>
<td>Unbalanced CAVSD</td>
<td>3</td>
</tr>
<tr>
<td>Rastelli C</td>
<td>15 (1 indeterminate)</td>
</tr>
<tr>
<td>VSD noncommitted or remote</td>
<td>all</td>
</tr>
<tr>
<td>AV valve insufficiency</td>
<td></td>
</tr>
<tr>
<td>Mod-severe</td>
<td>2</td>
</tr>
<tr>
<td>Mild or less</td>
<td>14</td>
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</tbody>
</table>
## Anatomic Features

### Associated Defects (all but 2 patients)

- Heterotaxy syndrome: 12
  - Right isomerism: 9
  - Left isomerism: 3
- TAPVC: 6
- Pulmonary stenosis: 13
- Systemic venous anomalies: 8
- Dextrocardia: 6
- Situs inversus: 2

### Prior operations in 10

(Shunt, PA banding, TAPVC repair, Fontan)
Operative Features of Repair

• Biventricular repair
  – Primary complete repair 6
  – Staged repair 10
• CPB 242 min, XC 158
• VSD enlargement 11
• RV-PA Conduit 13
Early Results

- **Mortality**
  - 1 hospital death

- **Morbidity**
  - Postop ECMO  1
  - Prolonged ICU  1
  - SVT        8
  - Heart block 2

- **AV valve insufficiency**
  - Moderate  5
Late Results

- Median follow-up 66 months (1 mo – 17 yrs)
- Mortality
  - 1 late death
- NYHA I or II
- Reoperation in 6
  - Conduit replacement 2
  - Valve repair or replacement 2
  - Other 2
- AV valve insufficiency
  - More than mild 3
Results of Biventricular Repair

- No cases of left ventricular outflow tract obstruction.
- No atrioventricular valvar stenosis,
- No significant residual VSDs.
Risk Factors for Poor Outcome

Univariate Analysis

Heterotaxy with TAPVC  
P=0.008

Nonsignificant variables
  Age at repair
  Heterotaxy w/o TAPVC
  Form of isomerism
  Dextrocardia
  Staged repair
Conclusions

- Survival for biventricular repair 88%
- Excellent functional results
  - No left ventricular outflow tract obstruction
  - Preserved AV valve function
- Identified risk factor
  - Heterotaxy with TAPVC
Inferences

• Biventricular repair of CAVSD/DORV
  – Alternative to univentricular palliation
  – High risk of reoperation (e.g., conduit replacement)
  – May offer long term benefits