Surgical Options for Congenitally corrected TGA with VSD and PS

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Congenitally corrected TGA is a complex congenital heart defect characterized by the presence of both atrioventricular & ventricular discordance.
Commonly associated with
Tricuspid valve regurgitation
VSD
Pulmonary stenosis or atresia
Complete heart block
Case

- 3 years old boy TGA, VSD, small ASD, subpulmonic PS
- BCPC was done at 6 months old to relieve cyanosis
- Currently well with mild cyanosis (SaO₂ of 82%)
- Echo:
  - good RV and LV function
  - mild/moderate TR;
  - mild MR;
  - mild Ebstein malformation of TV;
  - severe subpulmonic PS without pulmonary regurgitation;
  - VSD of not so large in size and with possible dynamic obstruction;
  - small ASD;
  - mild aortic regurgitation with mild aortic root pathology;
  - good MPA and both pulmonary arteries
Surgical Options

I. Classic Repair (Physiologic Repair)

• Classic Repair (Physiologic Repair)
  – VSD closure
  – LV-PA conduit or direct connection

Hraska et al.
Long-term outcome of surgically treated patients with corrected transposition of the great arteries

*J Thorac Cardiovasc Surg* 2005;1296:182

1963 ~ 1996, 123 patient

Palliative surgery and/or Intracardiac Procedure
Surgical Options

I. Classic Repair (Physiologic Repair)

The probability of survival

One-and-a-half ventricle repair

VSD closure
Pulmonic valvotomy
Conal septum resection
BCPS

Surgical Options

II. Anatomic Repair (Double Switch Operation)

Senning/Mustard procedure
Arterial Switch or Rastelli procedure
Surgical Options

III. Fontan-type Operation

Extension of ASD
Division of MPA
Factors to be considered

• Surgical complexity
• Operative risk
  – Operative mortality
• Late results
  – Late mortality
  – Late complication
  – Re-operation
  – Late functional class
Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

• Atrial Switch
  – Senning / Mustard procedure
• Arterial Switch
• Rastelli Operation
Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

Atrial Switch: Senning procedure
Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

Atrial Switch: Mustard procedure
Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

Arterial Switch

• VSD patch closure
• Resection of Conal septum (subpulmonary obstruction)
Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

Rastelli Operation

- Baffling of LV outflow thorough VSD to Aorta
- RV-PA conduit
Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

Conduction system

Bundle of His runs from the anterior node, through the fibrous trigone, to pass anterior and immediately inferior to the pulmonary valve annulus.

When VSD is present bundle of His runs along the supraanterior border fo the VSD.
Surgical Complexity

II. Anatomic Repair (Double-Switch Operation)

• Ilbawi et al.
  – Mustard procedure for venous switch moderate to deep hypothermia (20~24C)
  – CPB time: 180±23 min
  – ACC time: 147±17 min

• Langley et al.
  – Deep hypothermia (18C) in all cases
  – CPB time: 149 min
  – ACC time: 131 min

• Repair of the tricuspid left atrioventricular valve may be technically difficult, because of the posterior location.
Surgical Complexity

III. Fontan-type Operation

Extracardiac Lateral Tunnel
Extracardiac conduit TCPC
## Operative Risk

### II. Anatomic Repair (Double-Switch Operation)

<table>
<thead>
<tr>
<th>Author</th>
<th>Period</th>
<th>Case</th>
<th>Mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Langley SM</td>
<td>1991~2001</td>
<td>54</td>
<td>3 (5.6 %)</td>
</tr>
<tr>
<td>Devaney EJ</td>
<td>1993~</td>
<td>17</td>
<td>0 (0 %)</td>
</tr>
<tr>
<td>Ilbawi MN</td>
<td>1989~2000</td>
<td>12</td>
<td>1 (9 %)</td>
</tr>
<tr>
<td>Mee RB</td>
<td>1991~2000</td>
<td>22</td>
<td>3 (33 %)</td>
</tr>
<tr>
<td>Imai H</td>
<td>1989~1993</td>
<td>18</td>
<td>2 (11 %)</td>
</tr>
<tr>
<td>Yagihara T</td>
<td>1987 ~ 1990</td>
<td>10</td>
<td>3 (30 %)</td>
</tr>
</tbody>
</table>
## II. Anatomic Repair (Double-Switch Operation)

### Postoperative complete heart block

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Case</th>
<th>Incidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Langley SM</td>
<td>1991~2001</td>
<td>54</td>
<td>7/51 (14%)</td>
</tr>
<tr>
<td>Ilbawi MN</td>
<td>1989~2000</td>
<td>12</td>
<td>1/12 (9%)</td>
</tr>
<tr>
<td>Mee RB</td>
<td>1993~1999</td>
<td>27</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Karl TR</td>
<td>1989~1996</td>
<td>14</td>
<td>3/10 (30%)</td>
</tr>
<tr>
<td>Yagihara T</td>
<td>1987 ~ 1990</td>
<td>10</td>
<td>1/10 (10%)</td>
</tr>
</tbody>
</table>
## III. Fontan-type Operation

<table>
<thead>
<tr>
<th>Author</th>
<th>Years</th>
<th>Case</th>
<th>Mortality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gentles TL</td>
<td>1973~1991</td>
<td>500</td>
<td>16.8% (27.1→7.5)</td>
</tr>
<tr>
<td>Stamm C</td>
<td>1987~1991</td>
<td>220</td>
<td>5.4 %</td>
</tr>
<tr>
<td>Yoshimura N</td>
<td>1988~2000</td>
<td>76</td>
<td>10.5 %</td>
</tr>
<tr>
<td>Azakie A</td>
<td>1994~1998</td>
<td>60</td>
<td>5.6 %</td>
</tr>
<tr>
<td>Woods RK</td>
<td>1995~2001</td>
<td>58</td>
<td>5 %</td>
</tr>
</tbody>
</table>
Operative Risk

III. Fontan-type Operation

Yoshimura et al.
Risk factors influencing early and later mortality after total cavopulmonary connection
*J Thorac Cardiovasc Surg* 1997;114:376

Risk factors for early mortality
- Ventricular morphology (RV)
- Prolonged CPB time (240 min)
- EF < 60%
Operative Risk

III. Fontan-type Operation

Gentles et al.
Fontan operation in five hundred consecutive patients: Factors influencing early and late outcome
J Thorac Cardiovasc Surg 1997;114:376

Risk factors for early failure
- high mean PA pressure
- younger age
- Heterotaxy syndrome
- Tricuspid as only AV valve
- PA distortion
- AP connection
- absence of fenestration
- Longer CPB time
Operative Risk

III. Fontan-type Operation

Stamm et al.
Long-term results of the lateral tunnel Fontan operation

*J Thorac Cardiovasc Surg 2001;121:28*

Predictors of early failure
- Age at Fontan
- Single RV
- Left AVV stenosis or atresia
- PA distortion
- Preop PVR
II. Anatomic Repair (Double-Switch Operation)

Ilbawi et al.
Intermediate Results of the anatomic repair for congenitally corrected transposition

12 patients with ccTGA and VSD, PS (n=10)
Mustard + ASO (n=2), Mustard + Rastelli (10)
Follow up: 0.6 ~ 10 (7.6 ± 3.1) year

Operative mortality  9 %
Surgical complete heart block  9 %
SVC obstruction  9 %
bradyarrhythmia  36 %
conduit replacement in 5.2 year follow up  45%
mild to moderate TR  18%
LV EF = 49 ~ 70%
Early and Late Results

II. Anatomic Repair (Double-Switch Operation)

Langley et al.
Midterm results after restoration of the morphologically left ventricle to the systemic circulation in patients with congenitally corrected transposition of the great arteries

*Ann Thorac Surg 2002;73:594*

1991 ~ 2001
54 patients
ccTGA
AV discordance with DORV
Senning + ASO
Senning + Rastelli
Senning + intraventricular rerouting
Follow-up 4.4 year

Early mortality 5.6%
Heart block 13.7%
Chest reopen within 24 hour 9.3%
Kaplan-Meier Survival
  1 year 94.4%
  9 year 89.7%
Long-term Results

II. Anatomic Repair (Double-Switch Operation)

Kaplan-Meier Survival for Anatomic Repair

II. Anatomic Repair (Double-Switch Operation)

Freedom from reoperation

II. Anatomic Repair (Double-Switch Operation)

• Still, long-term data are lacking
• Long-term problems associated with Senning procedure for concordant TGA
  – Pathway obstruction
  – RV dysfunction
  – Arrhythmia
• Long-term problems associated with Rastelli procedure for concordant TGA
Long-term Results

II. Anatomic Repair (Double-Switch Operation)

Gelatt et al.
Arrhythmia and mortality after the Mustard procedure: A 30-year single-center experience
J Am Coll Cardiol 1997;29:194

534 children who underwent Mustard operation
Follow up: 11.6 ± 7.2 years.
There were 77 late deaths (16.1%)
with sudden death (n=31): the most frequent cause
Sinus rhythm was present in 77% at 5 years and 40% at 20 years.
Reoperation was required in 54 (10%).
  Repair of an obstructed or leaking baffle 25 (5%)
II. Anatomic Repair (Double-Switch Operation)

Kreutzer et al.
Twenty-five-year experience with Rastelli repair for Transposition of the great arteries

1973 ~ 1998, 101 patients with D-transposition, VSD
Pulmonary stenosis (73), Pulmonary atresia (18), LVOTO (10)

Intraventricular Baffling + RV-PA continuity establishment

Early mortality: n=7 (7%)
Median follow-up: 8.5 years (0.4 ~ 22 years)
Long-term Results

II. Anatomic Repair (Double-Switch Operation)

Overall survival after the Rastelli Repair

II. Anatomic Repair (Double-Switch Operation)

Freedom from reintervention, reoperation, or death

Long-term Results

II. Anatomic Repair (Double-Switch Operation)

Causes of late death or failure

- Sudden death 5
- LV dysfunction 7
- Conduit pseudointima rupture 1
- Myocarditis 1
- Unknown 2
- At re-operation 2

Long-term Results

II. Anatomic Repair (Double-Switch Operation)

Freedom from RVOTO and LVOTO reintervention

Long-term Results

III. Fontan-type Operation

Gentles et al.
Fontan operation in five hundred consecutive patients: Factors influencing early and late outcome
*J Thorac Cardiovasc Surg* 1997;114:376

1973 ~ 1991
Various modification of Fontan procedures
500 patients
Early failure 16.8% (27.1 → 7.5%)
Follow-up: 2464 patient-year
Long-term Results

III. Fontan-type Operation

Probability of survival with a Fontan circulation

Long-term Results

III. Fontan-type Operation

Probability of survival with a Fontan circulation

Late Results

III. Fontan-type Operation

Actuarial survival after the modified Fontan procedure

Long-term Results

III. Fontan-type Operation

Stamm et al.

Long-term results of the lateral tunnel Fontan operation

*J Thorac Cardiovasc Surg* 2001;121:28

1987 ~ 1991

Lateral tunnel Fontan procedures

220 patients

Early failure: 5.4%

Follow-up: 10.2 ± 0.6 (3.1~12.3) year
Kaplan-Meier estimated overall survival after the lateral tunnel Fontan operation

Estimates for 5 and 10 years:
- Estimated Survival: 93% and 91%
- Freedom from failure: 90% and 87%

Stamm et al. JTCS 2001;121:28
Long-term Results

III. Fontan-type Operation

Gentles et al.
Fontan operation in five hundred consecutive patients: Factors influencing early and late outcome  
*J Thorac Cardiovasc Surg* 1997;114:376

<table>
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<tr>
<th>Risk factors for early failure</th>
<th>Risk factors for late failure</th>
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<tr>
<td>high mean PA pressure</td>
<td>pacemaker before Fontan operation</td>
</tr>
<tr>
<td>younger age</td>
<td></td>
</tr>
<tr>
<td>Heterotaxy syndrome</td>
<td></td>
</tr>
<tr>
<td>Tricuspid as only AV valve</td>
<td></td>
</tr>
<tr>
<td>PA distortion</td>
<td></td>
</tr>
<tr>
<td>AP connection</td>
<td></td>
</tr>
<tr>
<td>absence of fenestration</td>
<td></td>
</tr>
<tr>
<td>Longer CPB time</td>
<td></td>
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Late Results

III. Fontan-type Operation

Stamm et al.
Long-term results of the lateral tunnel Fontan operation

*J Thorac Cardiovasc Surg* 2001;121:28

**Predictors of early failure**
- Age at Fontan
- Single RV
- Left AVV stenosis or atresia
- PA distortion
- Preop PVR

**Predictors of late failure**
- Prior systemic-pulmonary shunt
- Prior CoA repair
Long-term Results

III. Fontan-type Operation

Yoshimura et al.
Risk factors influencing early and later mortality after total cavopulmonary connection
*J Thorac Cardiovasc Surg* 1997;114:376

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<tr>
<td>Ventricular morphology (RV)</td>
<td>Heterotaxy syndrome</td>
</tr>
<tr>
<td>Prolonged CPB time (240 min)</td>
<td>AV valve regurgitation</td>
</tr>
<tr>
<td>EF &lt; 60%</td>
<td>Prolonged CPB time (240 min)</td>
</tr>
<tr>
<td></td>
<td>Prolonged ACC time (70 min)</td>
</tr>
</tbody>
</table>
### III. Fontan-type Operation

Stamm et al.

Long-term results of the lateral tunnel Fontan operation

*J Thorac Cardiovasc Surg 2001;121:28*

<table>
<thead>
<tr>
<th>Arrhythmia</th>
<th>5 yr</th>
<th>10 yr</th>
</tr>
</thead>
<tbody>
<tr>
<td>Freedom from new supraventricular tachyarrhythmia</td>
<td>96%</td>
<td>91%</td>
</tr>
<tr>
<td>Freedom from new bradyarrhythmia</td>
<td>88%</td>
<td>79%</td>
</tr>
<tr>
<td>Protein Losing Enteropathy</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Thromboembolic event</td>
<td>7</td>
<td></td>
</tr>
</tbody>
</table>
Long-term Results

III. Fontan-type Operation

Stamm et al.
Long-term results of the lateral tunnel Fontan operation
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Functional State

<table>
<thead>
<tr>
<th>NYHA</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>NYHA I</td>
<td>41</td>
</tr>
<tr>
<td>NYHA II</td>
<td>53</td>
</tr>
<tr>
<td>NYHA III</td>
<td>6</td>
</tr>
<tr>
<td>NYHA IV</td>
<td>0</td>
</tr>
</tbody>
</table>
Conclusion

III. Fontan-type Operation

- Less complex surgical procedure
- Comparable operative risk
- Fewer development of complete AV block
- Arrhythmogenic suture lines on atrium can be avoided.
- Favorable candidate for Fontan operation
  - Biventricular morphology
  - no PA distortion, normal PVR
- The patient might not need further operations such as conduit changes and atrial baffle revision
- Proven long-term outcome