Surgical Treatment for Double Outlet Right Ventricle

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History

1949: First surgical description of DORV (Kirklin)
1957: First repair of Taussig-Bing anomaly (Kirklin)
1967: Intracardiac repair of Taussig-Bing anomaly (McGoon)
1968: Intraventricular tunnel (Kawashima)
1971: Contemporary concept of DORV (Lev)

1960s: Detail understanding of anatomy (Newfeld)

Today

Taussig-Bing anomaly (Taussig & Bing)
Indication

• Congestive heart failure
  The patients with no pulmonary stenosis

• Cyanosis
  Patients with pulmonary stenosis
  Streaming due to VSD location
Surgical Treatment - Overview

- Able to commit VSD to aorta?
- If not – able to commit VSD to pulmonary artery, then switch?
- If not – Fontan path
- Is the patient big enough for surgery of choice?
Intracardiac repair feasible

Yes

Commit VSD to aorta

Yes

PS

Yes

TOF type repair

Small VSD

No

VSD closure / Intracardiac tunnel

Small VSD

Yes

Enlargement of VSD

Small VSD

No

Rastelli, REV, Nikaidoh Operation

Small VSD

Yes

VSD closure & Arterial switch

Small VSD

No

Kawashima Operation

Small VSD

No

DKS

PA band or BT shunt / RV-PA shunt

Yes

Commit VSD to PA

No

Fontan Operation

Small VSD

Yes

Enlargement of VSD

Small VSD

No

Yes

No

No

No

Small VSD

Yes

No

No

No

Small VSD

Yes

No

No

No

Small VSD

Yes

No

No

No

Small VSD

Yes

No

No

No

Small VSD

Yes

No

No

No

Small VSD
Surgical Treatment

- Commit VSD to aorta
  - Yes
    - PS
      - Yes: TOF type repair
      - No: VSD closure / Intracardiac tunnel
  - No
    - Commit VSD to PA
      - Yes
        - PS
          - Yes: Rastelli, REV, Nikaidoh Operation
          - No: VSD closure & Arterial switch
      - No
        - Fontan Operation
          - Small VSD
        - Kawashima Operation
          - DKS
        - Enlargement of VSD
          - Small VSD
          - Yes: Enlargement of VSD
Surgical Treatment
– TOF type

SKIP!
Surgical Treatment

Commit VSD to aorta

Yes

PS

- TOF type repair
- VSD closure / Intracardiac tunnel

No

- Small VSD
- Enlargement of VSD

Commit VSD to PA

Yes

PS

- VSD closure & Arterial switch
- Rastelli, REV, Nikaidoh Operation

No

- Small VSD
- Kawashima Operation

No

- DKS
- Enlargement of VSD

Yes

- No

No

- Small VSD
- Enlargement of VSD
Surgical Treatment
- VSD Closure / Intracardiac Tunnel

1. Approach can be transatrial or transventricular
2. Conduction pathway
3. Dacron or ePTFE graft
4. Possible leak – easier with continuous suture
5. Can secure from epicardium

Surgical Treatment
- Enlargement of VSD

1. VSD size ≥ aorta
2. Away from conduction, away from mitral valve
   - stay anterosperiorly

Surgical Treatment

Commit VSD to aorta

Yes
PS

- TOF type repair
  - Yes
  - Small VSD
    - Yes
    - Enlargement of VSD
    - No
    - VSD closure / Intracardiac tunnel
  - No
  - Small VSD
    - Yes
    - Enlargement of VSD
    - No

No
Commit VSD to PA

Yes
PS

- VSD closure & Arterial switch
  - Yes
  - Kawashima Operation
  - No
  - DKS
- Fontan Operation

No
Enlargement of VSD

Yes
No
No

Surgical Treatment – Rastelli Operation

Consists of
a. Rerouting or VSD to aorta
b. RV-PA conduit

1. Tunnel size ≥ aorta
2. Redundant patch
3. Conduit curves to left – redo is inevitable

Surgical Treatment – REV Operation

Consists of
a. Rerouting from VSD to aorta
b. Locompte maneuver
c. Posterior wall reconstruction
d. Anterior patch

1. Tunnel size ≥ aorta
2. Stretch of LPA
3. PR

Surgical Treatment – Nikaidoh Operation

Consists of
a. Harvesting aorta
b. Creating common orifice
c. Aortic translocation
d. Rerouting VSD to neo-aorta
e. Reconstruction of PA

## Surgical Treatment – Rastelli vs REV vs Nikaidoh

<table>
<thead>
<tr>
<th></th>
<th>Rastelli</th>
<th>REV</th>
<th>Nikaidoh</th>
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<tbody>
<tr>
<td>Surgical mortality</td>
<td>5~30%</td>
<td>12~18%</td>
<td>0~7%</td>
</tr>
<tr>
<td>Long-term mortality</td>
<td>20~30% at 15 years</td>
<td>15% at 25 years</td>
<td>NA</td>
</tr>
<tr>
<td>Technical challenge</td>
<td>Normal</td>
<td>Complex</td>
<td>More complex</td>
</tr>
<tr>
<td>Conduit stenosis / pulmonary stenosis</td>
<td>++</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>AR</td>
<td>-</td>
<td>-</td>
<td>+</td>
</tr>
<tr>
<td>PR</td>
<td>++</td>
<td>++</td>
<td>++</td>
</tr>
<tr>
<td>Reoperation</td>
<td>Inevitable</td>
<td>55% at 25 years</td>
<td>NA</td>
</tr>
</tbody>
</table>
Surgical Treatment

Commit VSD to aorta

Yes
- PS
  - Yes
    - TOF type repair
  - No
    - VSD closure / Intracardiac tunnel

No
- Commit VSD to PA
  - Yes
    - PS
  - No
    - Enlargement of VSD

Small VSD

Yes
- Rastelli, REV, Nikaidoh Operation
- Kawanishima Operation

No
- Fontan Operation
- DKS
- Enlargement of VSD
Surgical Treatment
- VSD Closure & Arterial Switch

1. The location varies - conduction pathway is a key
2. Chordae can be re-implanted
3. Tunnel / VSD size ≥ PA size

1. Unusual coronary pattern is common
2. Side-by-side great arteries stretch LPA
   - Lecompte maneuver vs classic switch
   - keep ex-aorta long

Surgical Treatment

Commit VSD to aorta

Yes
PS

No

Commit VSD to PA

Yes

No

TOF type repair

Small VSD

VSD closure / Intracardiac tunnel

Yes
Enlargement of VSD

No

Rastelli, REV, Nikaidoh Operation

Yes

No

VSD closure & Arterial switch

Kawashima Operation

No

Fontan Operation

Small VSD

DKS

Enlargement of VSD

Yes

No

Small VSD

Enlargement of VSD
Surgical Treatment
- Kawashima Operation

1. Resect infundibulum
2. Tricuspid chordae can be detached and re-implanted - not easy
3. Need enough space between tricuspid and pulmonary valve


Surgical Treatment

- Commit VSD to aorta
  - Yes
    - PS
      - Yes
        - TOF type repair
        - Small VSD
        - Enlargement of VSD
      - No
        - VSD closure / Intracardiac tunnel
        - Rastelli, REV, Nikaidoh Operation
        - Enlargement of VSD
  - No
    - Commit VSD to PA
      - Yes
        - PS
      - No
        - Fontan Operation
        - Small VSD
        - Enlargement of VSD
        - DKS
        - Small VSD
        - Enlargement of VSD
Surgical Treatment
- Fontan Operation & DKS / Enlargement of VSD

Enlarged VSD tends to become smaller


Short-Term Mortality

• 1970s – 50%, 1980s – 20%, 1990s - <6%
• Those with high pulmonary flow carries higher risk
• Heterogeneous group of patients
Long-Term Mortality

Due to intra-tunnel stenosis at levels

Short-Term Morbidities

- Residual leak
- Haemolysis due to a large patch
- Heart block
- LAD injury during enlargement of VSD
- Subaortic stenosis
- Aortic regurgitation
- Pulmonary regurgitation
- Tricuspid regurgitation
Long-Term Morbidities

Common morbidities for all types

• Subaortic stenosis
• Pulmonary regurgitation
• Aortic regurgitation
• Leak from long intracardiac tunnel
Conclusion

• DORV has wide spectrum of anomalies

• Surgical techniques include VSD closure to Nikaidoh operation

• High probability of reoperation in the future
Further Reading


Thank you!