Surgical Treatment for Tetralogy of Fallot

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History

1944 Blalock-Taussig shunt (Blalock & Taussig)
1946 Brock procedure (Brock)
1948 Waterston shunt (Waterston)
1954 Intracardiac repair of TOF with cross-circulation (Lillehei)
1955 Intracardiac repair of TOF with CPB (Kirklin)
1956 Modified BT shunt (de Leval)
1980 Today

Potts shunt (Potts)
Indications

“Blue” Fallot: \( \text{SaO}_2 < 75\% \)
- Hypoxaemic spell
- Otherwise, operate electively around 12 months

“Pink” Fallot: Same as VSD
Surgical Treatment - Overview

- Palliative –
  - Systemic-to-pulmonary shunt
  - Right ventricular outflow tract patch
  - Right ventricle-to-pulmonary shunt
  - Right ventricle-to-pulmonary stent

- Corrective – VSD closure +
  - Infundibulectomy only
  - Transannular patch with/without valve
  - Pulmonary valve preservation

When to choose palliative, when to choose corrective?
Surgical Treatment - Palliative vs Corrective

1. TOF
   - Symptomatic
     - > 3 month-old
       - Yes: Branch PA Z-score >2
         - Yes: Corrective repair
         - No: Palliation
       - No: Palliation
     - No: Palliation
   - Asymptomatic
     - Corrective repair

Aim of palliation
1. Increase pulmonary blood flow
2. Let branch PAs grow
3. Let the patient grow
PA Index

McGoon ratio  \[ \frac{\text{Pre-branching RPA diameter} + \text{LPA diameter}}{\text{Descending aorta diameter at the diaphragm}} \]

The risks of failure of the corrective repair is higher for less than 1.0 (normal range: 2.0-2.5)

Nakata index  \[ \frac{\text{Pre-branching RPA area} + \text{LPA area}}{\text{Body surface area}} \]

Lower Nakata index (< 150-200) has been identified as a risk factor (normal range: 330±30mm²/BSA)

The usefulness of these indices is controversial

They don’t consider
- the compliance of the pulmonary vascular bed
- distortion of the pulmonary arteries
Neonatal Repair vs Late Repair

Benefits of neonatal repair
1. Promote normal growth and development of organs
2. Eliminate hypoxaemia
3. Operate before extensive right ventricular hypertrophy
4. Better late left ventricular function
5. Decreased incidence of late dysrhythmias
6. No multiple procedures

Benefits of late repair (>3~6 months)
1. Lower mortality
2. Better chance of avoiding transannular patch
3. Shorter ICU stay
4. Lower incidence of re-intervention
Surgical Treatment – Palliative

• Palliative –

  Systemic-to-pulmonary shunt
  Right ventricular outflow tract patch
  Right ventricle-to-pulmonary shunt
  Right ventricle-to-pulmonary stent

Which technique to be used?
Surgical Treatment
- Systemic-to-pulmonary shunt

- Modified BT shunt
- Potts shunt
- Waterston shunt
Surgical Treatment
- RVOT Patch & RV-PA Shunt

Surgical Treatment
- RVOT Stent

## BT Shunt vs RA-PA Stent/Patch/Shunt

<table>
<thead>
<tr>
<th></th>
<th>BT shunt</th>
<th>RV-PA stent/patch/shunt</th>
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</thead>
<tbody>
<tr>
<td>Mortality</td>
<td>7~8% in CCAD, STS Some reported &gt;15%</td>
<td>Small cohort reports only (2~3%)</td>
</tr>
<tr>
<td>Diastolic control</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Landmark for coronary artery</td>
<td>No</td>
<td>No \ Yes – if patch or shunt</td>
</tr>
<tr>
<td>during the second stage</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Stress on RV</td>
<td>No change</td>
<td>Reduced</td>
</tr>
<tr>
<td>Shear stress</td>
<td>Arterial pressure</td>
<td>Pulsatile flow</td>
</tr>
<tr>
<td>Flow limiting factor (inflow</td>
<td>Inflow – bracheocephalic</td>
<td>Inflow – size of stent / patch / shunt</td>
</tr>
<tr>
<td>and outflow)</td>
<td>Outflow - RPA</td>
<td>Outflow – MPA / LPA</td>
</tr>
<tr>
<td>CPB</td>
<td>No / sometimes Yes</td>
<td>No / Yes</td>
</tr>
</tbody>
</table>
Concerns in Postoperative TOF & TOF/PA

- Right ventricular hypertrophy
- RV fibrosis
- Restrictive RV physiology
- Severe third spacing
- Haemodynamic instability
- Prolonged chest tube

Is RV-PA patch / shunt better?
Inconsistent Choices

Birmingham Children’s Hospital
- BT shunt for PA
- RV-PA stent for TOF
- RV-PA shunt for Norwood stage I

Great Ormond Street Hospital for Children
- RV-PA shunt for PA
- RV-PA patch for TOF
- BT shunt for Norwood stage I
Birmingham Criteria for RVOT Stent

- Low birth-weight neonates
- < 4 kg
- Branch PAs Z-score < −2
- Complex anatomy or comorbidity
  - AVSD / Fallot
  - anomalous LAD,
  - unroofed coronary sinus,
  - chronic lung disease

PA growth (Z-scores) at the time of RVOT stenting and prior to surgery.

Choice of Size

- **Restrictive patch**
  - Small enough to restrict flow from VSD

- **Should have L→R shunt > R→L shunt**

- **PA pressure < ½ systemic**

- **Some suggested 5, others 6mm for shunt**
Surgical Treatment – Corrective

- Corrective – VSD closure +
  - Infundibulectomy only
  - Transannular patch with/without valve
  - Pulmonary valve sparing surgery

Surgery consists of:
1. Infundibulectomy
2. VSD closure
3. ±RVOT reconstruction
Surgical Treatment
- Infundibulectomy

1. Identify trabecular septomarginalis
2. Resect large septoprietal band
3. No more band before pulmonary valve
4. Divide smaller ones
   - important to understand septum shape

Surgical Treatment
VSD Closure

1. Pay attention to valleys
   1: between tricuspid and aortic valve
   1’: behind aortic valve
   1’’: between superior and inferior rim of TSM

2. Conduction

Surgical Treatment
- RVOT Reconstruction

1. Determine the size you aim for
2. Estimate the patch size
3. Ensure the size is enough in every dimension
4. ± monocusp

Monocusp


Surgical Treatment
- Pulmonary Valve Sparing

1. Only PV thinning and commissurotomy
2. Add intraoperative balloon dilation
3. Incision of anterior leaflet and patch augmentation

Short-Term Mortality

- 1950s – 20%
- 1960 ~ 1980 – 5%
- Contemporary data – 0~3%

EACTS congenital database

<table>
<thead>
<tr>
<th>Mortality, Stratified by Age and Anatomy</th>
<th>Neonates</th>
<th>Infants</th>
<th>All Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tetralogy of Fallot (VSD closure, ventriculotomy, transannular</td>
<td>9.7%, n = 103</td>
<td>2.06%, n = 3157</td>
<td>2.5%, n = 5241</td>
</tr>
<tr>
<td>patch)</td>
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<tr>
<td>Pulmonary atresia VSD (VSD closure, right ventricle-pulmonary</td>
<td>4.69%, n = 64</td>
<td>2.7%, n = 296</td>
<td>4.52%, n = 663</td>
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<td>artery conduit)</td>
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<tr>
<td>Absent pulmonary valve syndrome (all operative strategies)</td>
<td>42.9%, n = 42</td>
<td>6.5%, n = 154</td>
<td>11.33%, n = 256</td>
</tr>
</tbody>
</table>

VSD = ventricular septal defect.

The indications and timing (urgent vs elective) are not accounted for. The five “best centers,” with a total of 261 “classical” tetralogy of Fallot repairs, reported a combined mortality of 0%.

Long-Term Mortality

Short-Term Morbidities

- Residual shunt
- Complete heart block
- Tricuspid valve regurgitation
- Aortic regurgitation
- Pulmonary regurgitation
- Restrictive right ventricular physiology
Long-Term Morbidities

• Arrhythmias
• Right ventricular outflow tract obstruction
• Pulmonary regurgitation
• RV dilatation → PV replacement
• Reduced right ventricular function
Other Variations

• Absent pulmonary valve syndrome

• TOF + AVSD

• Strategy for small branch PAs

• Pulmonary atresia
Conclusion

• Surgical repair has excellent outcome

• While there is general consensus that repair should be performed between 3 and 12 months, controversy persists regarding neonatal repair and staged repair
Further Reading


Thank you!