RVOT Reconstruction Options in Tetralogy of Fallot: a data manager review

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Disclosure

• I have no relevant financial disclosures
Tetrad of Fallot

- First described in 1888 by Louis Arthur Etienne Fallot
- Most common cyanotic CHD in the world - *La Maladie Bleue* – the ‘blue baby’
- Approximately 3 out of every 10,000 live births
- Anatomic features
  - Large anterior malalignment ventricular septal defect (VSD)
  - Overriding of aorta - biventricular connection to the aorta
  - Pulmonary stenosis
  - Right ventricular hypertrophy

https://americanheart.org
Embryology of TOF

• Classic second heart field defect
• Anterior and cranial displacement of conal septum

https://embryology.med.unsw.edu.au/embryology
https://behnace.net
What is "like" TOF

- TOF/PA
  - MAPCA
- TOF/AVC
- TOF/absent pulmonary valve
- DORV
- VSD/PS
What is “like” TOF

• TOF/PA
  • VSD/PA
  • Extreme anterior deviation of septum
  • Ductal dependent pulmonary blood flow
  • Neonatal intervention
    • Palliation – surgical or percutaneous
    • Complete repair
  • 340=Pulmonary atresia, VSD (Including TOF, PA)
What is “like” TOF

• TOF/PA
  • MAPCA
What is “like” TOF

• TOF/PA
  • MAPCA – Major aorto-pulmonary collaterals
  • Diminutive or absent central PA
  • Significantly more complex management paradigms
  • 350=Pulmonary atresia, VSD-MAPCA

Kumar SR. J Thorac Cardiovasc Surg. 2018;156:1205-6
What is “like” TOF

• TOF/PA
  • MAPCA
• TOF/AVC
What is “like” TOF

- TOF/PA
  - MAPCA
- TOF/AVC
  - Combined TOF and AV canal defect – usually complete canal
  - Complete surgical repair includes TOF and canal repair
  - 300=TOF, AVC (AVSD)
What is “like” TOF

• TOF/PA
  • MAPCA
• TOF/AVC
• TOF/absent pulmonary valve
What is “like” TOF

- TOF/PA
  - MAPCA
- TOF/AVC
- TOF/absent pulmonary valve
  - Severe pulmonary insufficiency
  - Dilated branch pulmonary arteries
  - Can have serious respiratory issues in newborn period
  - Usually need to address PA dilation at surgery
  - 310=TOF, Absent pulmonary valve

https://medlineplus.gov
What is “like” TOF

- TOF/PA
  - MAPCA
- TOF/AVC
- TOF/absent pulmonary valve
- DORV
What is “like” TOF

- TOF/PA
  - MAPCA
- TOF/AVC
- TOF/absent pulmonary valve
- DORV
  - Aortic mitral discontinuity
  - Only outlet to LV is VSD
  - Multiple sub-types – TOF-type common
  - 940=DORV, TOF type
What is “like” TOF

- TOF/PA
  - MAPCA
- TOF/AVC
- TOF/absent pulmonary valve
- DORV
- VSD/PS
What is “like” TOF

- TOF/PA
  - MAPCA
- TOF/AVC
- TOF/absent pulmonary valve
- DORV
- VSD/PS
  - Location of VSD – not outlet septum
  - Pulmonary stenosis is flow-related (no anatomic defect in valve or RVOT)
  - Distinction can be difficult in rare cases

https://ctsurgerypatients.org
Why is this distinction important

• Accurate coding
• TOF is a benchmark operation
  • Table 18 of report provides separate results
  • Table 23/24 provide data on TOF palliation and repair respectively
• USNWR specific datapoint

**Question:** What was the total number (cumulative incidence) of patients who underwent Tetralogy of Fallot (TOF) repair surgeries and the total number of these patients the required an unplanned cardiac reoperation following TOF?
Associated defects and anomalies

• Syndromes/Chromosomal anomalies
  • 22q11 microdeletion - DiGeorge syndrome (velocardiofacial syndrome) (conotruncal anomaly face syndrome), especially with VSD/PA
  • Trisomy 21 (Down’s syndrome)
  • Alagille Syndrome
  • CHARGE association – CHD7 genetic anomaly
  • Fetal alcohol exposure
  • VACTERL syndrome (VACTER/VATER/VATERR syndrome)

• Non-cardiac anatomic abnormalities
  • Major abnormality of head, Cleft lip /palate
  • Major abnormality of spinal cord, Myelomeningocele
  • Major abnormality of larynx - trachea - or bronchus, Tracheoesophageal fistula (TEF)
  • Major abnormality of gastrointestinal system, Biliary atresia – Alagille syndrome
TOF - Presentation

- Degree of pulmonary stenosis and right ventricular outflow tract (RVOT) obstruction determines timing and severity of presentation
- Cyanosis
- Ductal dependence in severe RVOT obstruction, pulmonary atresia
- Heart failure is rare
- Medical measures to improve perfusion
- Definitive management is surgery –
  - Palliation
  - Complete repair
TOF - palliation

• Primarily to improve pulmonary blood flow
• Usually in the setting of small PA or factors precluding full repair
• Percutaneous interventions - PDA stent or RVOT stent (remember status post diagnosis)
TOF – palliation

- **Systemic to pulmonary shunting**
  - ☑ 1590= Shunt, Systemic to pulmonary, Modified Blalock-Taussig Shunt (MBTS)
  - ☑ 1600= Shunt, Systemic to pulmonary, Central (shunt from aorta)
  - ☑ 3130= Shunt, Systemic to pulmonary, Central (shunt from aorta) Central shunt with an end-to-side connection between the transected main pulmonary artery and the side of the ascending aorta (i.e. Mee shunt)
  - ☑ 3230= Shunt, Systemic to pulmonary, Potts – Smith type (descending aorta to pulmonary artery)
  - ☑ 1610= Shunt, Systemic to pulmonary, Other
TOF – palliation

- RV-PA Conduit
  - Valved (homograft, xenograft)
  - Non-valved (Sano shunt)
  - 610= Conduit placement, RV to PA
- Transannular patch augmentation
  - Non-valved patch
  - Monocusp patch
  - 510= RVOT procedure

Turrentine M, CTSNET
TOF – palliation

• PA reconstruction
  • Unifocalization

2730 = Unifocalization MAPCA(s), Bilateral pulmonary unifocalization - Complete unifocalization (all usable MAPCA[s] are incorporated)

530 = PA, reconstruction (plasty), Main (trunk)

540 = PA, reconstruction (plasty), Branch, Central (within the hilar bifurcation)

550 = PA, reconstruction (plasty), Branch, Peripheral (at or beyond the first lobar branch)

552 = PA, reconstruction (plasty), Branch, Peripheral (at or beyond the first lobar branch, proximal to first segmental branch)

554 = PA, reconstruction (plasty), Branch, Peripheral (at or beyond the first lobar branch, beyond the first segmental branch)

### TOF-repair

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>350</td>
<td>TOF repair, No Ventriculotomy</td>
</tr>
<tr>
<td>360</td>
<td>TOF repair, Ventriculotomy, Nontransanular patch</td>
</tr>
<tr>
<td>370</td>
<td>TOF repair, Ventriculotomy, Transanular patch</td>
</tr>
<tr>
<td>372</td>
<td>TOF repair, Ventriculotomy, Transanular patch, plus native valve reconstruction</td>
</tr>
<tr>
<td>374</td>
<td>TOF repair, Ventriculotomy, Transanular patch, with monocusp or other surgically fashioned RVOT valve</td>
</tr>
<tr>
<td>380</td>
<td>TOF repair, RV-PA conduit</td>
</tr>
<tr>
<td>390</td>
<td>TOF - AVC (AVSD) repair</td>
</tr>
<tr>
<td>400</td>
<td>TOF - Absent pulmonary valve repair</td>
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- **420**: Pulmonary atresia - VSD (including TOF, PA) repair
- **2700**: Pulmonary atresia - VSD – MAPCA repair, Complete single stage repair (1 stage that includes bilateral pulmonary unifocalization + VSD closure + RV to PA connection [with or without conduit])
- **2710**: Pulmonary atresia - VSD – MAPCA repair, Status post prior complete unifocalization (includes VSD closure + RV to PA connection [with or without conduit])
- **2720**: Pulmonary atresia - VSD – MAPCA repair, Status post prior incomplete unifocalization (includes completion of pulmonary unifocalization + VSD closure + RV to PA connection [with or without conduit])
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TOF-repair

- Includes VSD closure and relief of PS at all levels
- Do not use VSD repair (110) and PS relief (510, 590) codes for TOF repair
- Do not add codes for RVOT relief (510) or pulmonary valvotomy (590) to TOF repair codes
- Remember procedure-specific factors for TOF

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<th>Major coronary crossing RVOT - Coronary anomaly restricting RVOT enlargement</th>
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<tr>
<td>PSFMajCorRVOT(952)</td>
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<th>VSD, Multiple, Repair PSFVSDMultRep (953)</th>
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<th>Restrictive VSD PSFRestrictVSD (954)</th>
<th>□ Yes □ No</th>
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<th>Hypoplastic branch pulmonary arteries (diminished pulmonary vascular bed)</th>
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Questions?