MRI (AND CT) FOR REPAIRED TETRALOGY OF FALLOT

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OUTLINE

• Pathogenesis
• Variants
• Initial surgical treatments
• Basic MR protocols
• MR and CT for post-repair treatment planning (PVR)
  • Ventricular size and function, pulmonary insufficiency, pulmonary artery anatomy & flow, coronary anomalies, lungs
INTRODUCTION

- Tetralogy of Fallot (TOF): 6.8% of all congenital heart disease
- Described in 1888 by Fallot - autopsy series cyanotic heart disease
- Malalignment of the conal septum in relation to the ventricular endocardial cushion → lack of stimulus for membranous septum development
- Malaligned VSD, overriding aorta & RVOT obstruction
- Disease severity and age of presentation depends on degree of RVOT obstruction
• TOF represents a spectrum of disease with three major variants:
  • TOF with pulmonary valve or RVOT stenosis
  • TOF with pulmonary atresia
  • TOF with absent pulmonary valve
  • TOF with pulmonary atresia is the most severe form, present in 20%
• 1950s: initial surgical repair
• VSD closure and relief of RVOT obstruction
• Since then increasing population lives with TOF
• Unique problems related to longstanding postoperative physiology & delayed consequences of disease
• Most common problem & most frequent indication for imaging is prolonged post-op pulmonary insufficiency with consequent RV dilation
# Surgery for tetralogy of Fallot

<table>
<thead>
<tr>
<th>Procedure</th>
<th>Description</th>
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<tbody>
<tr>
<td>Transannular patch</td>
<td>Marked obstruction of PV annulus Relieves RVOT obstruction- patch placed across opened RVOT → free PI</td>
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<td>Infundibulectomy</td>
<td>Less severe PS Relieves RVOT obstruction- excision of infundibular muscle</td>
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<td>RV-to-PA conduit</td>
<td>Marked obstruction of PV annulus Valved conduit into the anterior wall of RV, connected to PAs.</td>
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<td>Blalock-Taussig shunt</td>
<td>Palliation or staged for inadequate PA anatomy Subclavian to PA shunt via Gore-Tex tube (modified) or direct anastomosis (classic)</td>
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<tr>
<td>Unifocalization</td>
<td>Pulmonary atresia with extensive systemic collaterals Systemic collaterals detached from aorta, redirected into single vessel to establish blood supply to lung</td>
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IMAGING MODALITIES

• **Echocardiography** – mainstay

• **MR** for specific indications anatomic and quantitative
  - Central and peripheral PA anatomy
  - *Quantitate* ventricular volumes, function, regurgitant fraction, differential PA flow
  - Exploratory role for ventricular fibrosis

• **CT** when MR is contraindicated, coronary artery anatomy and pulmonary parenchymal assessment
MR PROTOCOL - INFORMATION GLEANED

- ECG-gated black double IR axial - anatomic overview
- SSFP cine - short axis, long axis, RVOT - RV LV volumes, EF & wall motion
- Phase contrast MPA (RPA, LPA), aorta - flow volume, velocity, quantification
- Contrast enhanced MRA coronal - central and peripheral PA anatomy
- Delayed enhancement - fibrosis
12 year-old girl s/p TOF repair in infancy

Echo shows free PI and moderate to severe RV dilation

MR requested to check RV volumes and regurgitant fraction
CARIDAC MRI CALCULATIONS & MEASUREMENTS

LEDV = 84ml; LV ESV = 34ml, LV EF = 59 %
RV EDV = 221 ml; RV ESV = 137 ml, RV EF = 38%
Pulmonary Regurgitant Fraction = 45%
MPA = 1.9 cm; RPA = 1 cm; LPA = 1.2 cm
WHAT DOES THIS MEAN?

- Pulmonary insufficiency initially well-tolerated post repair
- Over decades, morbidity & mortality from chronic PI → RV dilation, biventricular dysfunction, heart failure & arrhythmia
- PVR standard practice in symptomatic pts, improved PI & sx, but not RVEF or ? mortality
- Optimal timing of PVR is still being explored, should precede development of sx
- RV ESV indexed to BSA <90 ml/m2, QRS <140 ms associated optimal postoperative outcome (nl RV size, fxn)

Geva et al Circulation. 2010;122:S201–S208 Randomized Trial of Pulmonary Valve Replacement Withand Without Right Ventricular Remodeling Surgery
21-year-old woman s/p TOF repair in infancy who developed shortness of breath on exertion

Echo showed TR and free PI with RV dilation

Underwent PVR and tricuspid valve annuloplasty

MR performed preoperatively & 1 and 2 years postop
RVEF

2/20/08

Mid

D

S

Apical

7/7/09

D

S

9/10/10
Phase contrast orthogonal to direction of flow for PR fraction
<table>
<thead>
<tr>
<th></th>
<th>Indexed RVEDV</th>
<th>Indexed RVESV</th>
<th>RVEF (%)</th>
<th>Regurg. Fraction (%) at PV</th>
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</thead>
<tbody>
<tr>
<td>2/20/08</td>
<td>156</td>
<td>106</td>
<td>32</td>
<td>53</td>
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<td>9/10/10</td>
<td>114</td>
<td>80</td>
<td>30</td>
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LATE GADOLINIUM ENHANCEMENT IN TOF

- More common in patients repaired after infancy
- Indicative of postoperative fibrosis
  - RVOT, VSD patch, RV insertion point & LV apical vent site
- Worse prognosis
- Vascular distribution indicative MI
CT IN TOF

- Ventricular volumes, wall motion and ejection fractions - retrospective ECG-gating
- Inferentially calculate regurgitant volume if isolated PI (no TI or VSD) based on equal right and left heart cardiac outputs
  - RV SV-LV SV = pulmonary regurgitant volume
- Imaging modality of choice for coronary anomalies, pulmonary parenchyma
20 YEAR OLD MALE S/P TOF REPAIR WITH RV-PA CONDUIT AND SINGLE CORONARY ARTERY S/P PACEMAKER
RV EDV = 214 mL
LV EDV = 160 mL

RV ESV = 110 mL
LV ESV = 70 mL

RV EF = 49 %
LV EF = 56%
3D images - useful in surgical planning
TOF WITH PULMONARY ATRESIA-SPECTRUM

- Large confluent central PAs discontinuous but near RVOT
- Absent central PAs with lungs supplied solely by systemic collaterals (pseudotruncus)
- Numerous intermediate forms
- Primary repair for milder forms
- Absent central PA’s difficult to repair (unifocalization)
- MR and CT add value to echo in surgical planning, depict central PAs, systemic collaterals
21-YEAR-OLD MAN
PULMONARY ARTERY ANEURYSM

- Aneurysmal central PAs in a subset of TOF - usually absence of the pulmonary valve
- Cause unknown
- May be at Blalock-Taussig shunt insertion
- Collaterals
52-YEAR-OLD MAN
**TRICUSPID INSUFFICIENCY**

- Tricuspid insufficiency 
  ~10%
  - RV & tricuspid annulus dilation
  - Valve injury during repair
- CT and MR demonstrate enlarged RA

**RESIDUAL VSD**

- Residual postop VSD rare
- Echo adequate
- CT & MR also reliable
- MR- Qp:QS & direction of flow
20-YEAR-OLD WOMAN
PULMONARY PARENCHYMAL & VASCULAR ABNORMALITIES IN TOF

- Lung function often abnormal after repair
- Due to diminished perfusion
- Central and peripheral PA stenoses
- PA stenosis exacerbates pulmonary regurg, increases RV ESP results in decreased exercise tolerance
- TOF with PA, lung segments supplied exclusively by nonstenotic collaterals develop pulmonary HTN
• CT appearance of lung depends local vasculature
• Perfusion: adequate, under or over perfused
• Mosaic perfusion pattern reflects variability in regional lung perfusion & pulmonary HTN
• Systemic collaterals visible directly or inferred by serrated pleural thickening & subpleural parenchymal bands
CONCLUSION

• TOF repair invariably leads to pulmonary insufficiency
• Well-tolerated early, but associated with morbidity & mortality over time
• MR primary tool for evaluating ventricular volumes & function, pulmonary insufficiency- guide to PVR
• MR useful for PA anatomy, differential flow thru PAs, fibrosis
• CT when MR contraindicated & primary tool for coronary anomalies & pulmonary parenchymal imaging
THANK YOU