TETRALOGY OF FALLOT – EVALUATION WITH CT AND MRI IN “KIDS FROM 1 TO 92”

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EDUCATIONAL OBJECTIVES

- I. To show that Tetralogy of Fallot (TOF) is a wide spectrum of abnormalities beyond the classic four findings of RVH, PS, VSD and Overriding Aorta and can lead to additional cardiac, vascular and airway malformations.

- II. Describe the variations in postoperative / post-intervention appearance.

- III. Describe quantitative parameters and imaging findings that need to be reported to cardiologists and cardiothoracic surgeons for management decisions.
BACKGROUND:
WHAT IS FALLOT’S TETRALOGY?

- The classic tetrad of cardiac abnormalities all stemming from anterior/superior malalignment of the outflow septum:
  - Overriding Aorta
  - Ventricular Septal Defect (VSD)
  - Pulmonic stenosis (PS)
  - Right ventricular hypertrophy (RVH)
- These features may manifest in varying degrees of severity
- Additional consequences include: subvalvar RVOT stenosis, branch pulmonary artery (PA) stenoses and aneurysms, aortopulmonary collaterals and airway complications.
WHY TOF?

- TOF is 10% of all congenital heart defects, but it is the most common defect in humans beyond 1 year of age (Rao 2013).
- Incidence (0.31-0.42 per 1000 live births) (Hoffmann et al. JACC 2002, Van der Linde et al. JACC 2011)
- 86% of patients survive up to 32 years post intervention, and the average mortality is beyond 18 years of age (Kalra, Klewer et al. 2010).
- As number of adults with TOF increases, the likelihood of encountering these patients in the daily hospital setting increases.
- Radiologists need to learn about the complexities of TOF in both the child and adult patient.
I. THE SPECTRUM OF TOF

- Common findings in all forms: large subvalvar VSD due to anterior malalignment of the outflow septum.

- MILD: mild PS, normal PAs (VSD physiology)

- MODERATE: subvalvar and/or valvar PS, normal PAs (classic TOF)

- SEVERE: Severe subpulmonic stenosis or pulmonic atresia, hypoplastic PAs with PDA (duct-dependent)

- VERY SEVERE: Pulmonic atresia, absent pulmonary arteries, MAPCA

- RARE: TOF with absent pulmonary valve: Airway obstruction, severe pulmonic insufficiency (PI,) large main pulmonary artery (MPA) and branch PAs.
MILD TOF

- 3 day-old F
- Mild Pulmonic Stenosis (PS)
- VSD Physiology
- Well developed, “criss-crossed” pulmonary arteries (PA) with only mild LPA stenosis
- Pulmonary Edema
MILD-MODERATE TOF

- 11w M, Mild Pulmonic Stenosis (PS)
- VSD Physiology
- Mildly hypoplastic PAs
- Pulmonary Edema
MODERATE TOF

- 4mo F
- Aorta overriding large subvalvar VSD
- Subvalvar PS, RV hypertrophy
- Normal MPA and RPA, moderate LPA stenosis
- Right aortic arch, which is common in TOF
SEVERE (DUCTAL DEPENDENT)
- 3-day-old M
- Pulmonary atresia – hypoplastic MPA
- Preserved contiguous PAs
- PDA dependent.

Pulmonary atresia – patient is dependent on PDA to maintain blood supply to lungs
SEVERE (DUCTAL DEPENDENT)

- 6-week-old with
- pulmonary atresia – hypoplastic MPA
- discontiguous PAs (LPA supplied by PDA)
- Bilateral PDAs (Left PDA from brachiocephalic artery and right PDA from distal arch)
VERY SEVERE

- Newborn with pulmonary atresia
- Absent PAs (Absent RVOT or MPA on images below)
- MAPCAs = major aortopulmonary collateral arteries
RARE

- 5-week-old with absent pulmonic valve
- Markedly engorged PAs causing airway compression
- Note also right arch.
II. EARLY PALLIATION

- Balloon Angioplasty/Stenting
- Blalock-Taussig (BT) shunt / modified BT shunt (mBT) / Central Shunt
- Transannular patch, VSD closure
- Valve sparing RVOT augmentation, PA augmentation
- Homograft or RV to PA conduit
- Unifocalization, RV to neoPA (MAPCA) conduit
Balloons Angioplasty/Stenting

- Stenting of RPA and LPA, Marked RV hypertrophy
SHUNTS TO PROVIDE PULMONARY FLOW

Central Shunt
4 mo F

mBT Shunt, 14-day-old
VSD PATCH
TRANSANNULAR PATCHES (HOOD)
Surgical patches calcify with time (66 y.o. M !!!)
VALVE SPARING RVOT AUGMENTATION
PA AUGMENTATION
5 yo M, RV to PA conduit in pulmonary atresia
18 MO, M, Unifocalization: converting MAPCA into pulmonary arteries by dividing them from the aorta, and creating an RV to neo-PA conduit.
III. ADULT EVALUATION

- Crucial to evaluate and quantify right ventricular function
- RVEF
- RVEDV – RVEDV Index (RVEDV/BSA) an important factor in making the surgical decision regarding valve placement (cutoff approx 165 mL/m2)
- Pulmonic Regurgitant fraction
- Qp:Qs
- RVOT (subpulmonic stenosis)
- MPA and branch PAs
- Retrosternal anatomy
- Coronary artery origins/course
MRI OF 53 YO M

- History of VSD closure and transannular patch.
- SSFP 4ch Cine: mildly dilated RV (indexed RVEDV 116 ml/m2) due to restrictive physiology, which “protected” against otherwise expected severe RV dilatation.

  Phase contrast velocity mapping of MPA: regurgitant fraction of 34%. Note restrictive physiology with early opening of the pulmonic valve and late diastolic antegrade flow.

- Aortic and PA flows yielded a Qp:Qs of 1:1 and balanced flow to the lungs in spite of mild RPA stenosis.
Free breathing coronary MRA for retrosternal anatomy. Note the rotated aortic root, typical in TOF, and RCA coursing immediately behind the sternotomy wire.

- 3D rendering of a time-resolved MRA, showing normal “hood” at the site of patching, but also mild right pulmonic stenosis/hypoplasia and aneurysmal dilatation of the proximal left pulmonary artery.
Axial black blood slices in a 30 yo M with repaired TOF and a trans-annular patch. The indexed RV volume was 185ml/m2
RESIDUAL SUBVALVAR STENOSIS

- SSFP cine MRI in 26yo F subpulmonic stenosis due to subvalvar web and hypertrophied trabeculation in the RVOT.
MRA TO EVALUATE RVOT, PULMONARY ARTERIES AND RETROSTERNAL ANATOMY

- 26 yo F, Free-breathing respiratory navigated non-contrast MRA showing subvalvar trabeculations and retrosternal anatomy, including RCA/acute marginals

For comparison: another patient's MRA showing tortuosity, but no subvalvar stenosis
CT IN ADULT TOF

- CT evaluation in patients with severe claustrophobia or pacemakers/AICDs.
- To obtain RV function contrast is timed to achieve sufficient opacification in the RV.
- Rotated aortic root (RCA leftward and LCA rightward)
- RCA / acute marginals / right atrium (RA) very close to the sternum
IV. LATE INTERVENTION/COMPLICATIONS

- Valve placement surgical/transcatheter
- Homograft replacement
- Pulmonary artery patching
- MAZE procedure (for atrial arrhythmias)
- Pacemaker / AICD
- Tricuspid valve surgery (annuloplasty, valve replacement)
23 y.o. male with TOF and pulmonary atresia, status post RV to PA conduit with bioprosthetic valve (A) LPA atresia and aortopulmonary collaterals and bioprosthetic tricuspid valve (B).

AICD leads in the RV (B), precluding MRI evaluation.

3D VR is helpful for surgical planning: NOTE sub-valvar ridge in the conduit, causing stenosis, warranting this evaluation (C).

Appropriate bolus timing to optimize RV opacification allows quantitative assessment of RV volumes and function(D).
RESIDUAL RVOT OBSTRUCTION

- 35 yo F
- Surgery at age 2
  - VSD patch, pulm valvotomy
- Residual subvalvar PS
- Valvar PS and insufficiency
- Pacemaker
MELODY AND MAZE

- 53yo F, TOF status post transannular patch and VSD closure with severe pulmonic regurgitation and RVEDV index of 168 mL/m²; with severe right atrial dilation as well, and atrial fibrillation.
- MRA also helped visualize coronary artery course

She underwent transcatheter Melody valve placement as well as MAZE procedure.
46 yo M, TOF status post transannular patch and VSD closure with residual VSD (jet shown by arrow).

- Aortic root also dilated (4.9cm), with associated aortic regurgitation. This lead to mild LV dilation with an indexed LVEDV of 94 mL/m2.
- Indexed RVEDV was 165 mL/m2.
SUMMARY / TAKE HOME POINTS

• TOF is more than just the classic four findings.

• TOF is a disease spectrum from mild to very severe forms

• Many patients now live to their 50s and 60s, therefore all cardiac imagers need to be informed about the postsurgical appearance

• CTA and MRI play an important role in the pre and postoperative evaluation in adults

• Important to look for vascular complications

• Important to assess and report quantitative parameters such as RVEDV and RVEF as well as pulmonic regurgitant fraction
REFERENCES


