Pre-operative assessment of Tetralogy of Fallot – Imaging

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Introduction

• Described in 1888 by Louis Etienne Fallot
• ToF accounts for 10% of all antenatally diagnosed CHD
• Live birth incidence of 1/3600
Fig. 1 Graph shows the spectrum of major congenital heart defects in 9,834 foetuses.

AVSD: atrioventricular septal defect; Coarc: coarctation of the aorta; DORV: double outlet right ventricle; HLHS: hypoplastic left heart syndrome; TGA: transposition of the great arteries; TOF: Tetralogy of Fallot; VSD: ventricular septal defect
Fallot’s Tetralogy

- Overiding Aorta
- Ventricular Septal Defect
- Pulmonary Stenosis
- Right ventricular hypertrophy
Fallot’s Tetralogy

• Hallmark is an anterior and superior deviation of the conal septum
• Lies as part of a spectrum ranging between a mal-aligned ventricular septal defect and pulmonary atresia with ventricular septal defect (Tetralogy with pulmonary atresia)
Morphology

- Muscular subpulmonary obstruction
- Right ventricular hypertrophy
- Right ventricular origin of aorta
- Interventricular communication

*Paediatric Cardiology 3rd Edition RH Anderson*
Morphology

Paediatric Cardiology 3rd Edition RH Anderson
Fallot’s Tetralogy

- Increasingly diagnosed prenatally
- Presents with a cardiac murmur within the first few days of life as pulmonary vascular resistance decreases
- Presents with late onset cyanosis
- Can present with tachypnoea (pulmonary over-circulation in mal-aligned ventricular septal defect)
Prenatal Diagnosis
When does the child require Surgery?

- Duct-dependent pulmonary circulation
- Tet spells
- Symptomatic cyanosis
- Pulmonary over-circulation
Pre-operative assessment of Tetralogy of Fallot

• Main imaging modality remains 2D Echocardiography
• Echo is able to demonstrate the key anatomical features of Tetralogy
• “Living Anatomy”
What do we look for with Echo?

- Confirmation of a diagnosis of Tetralogy
- Additional ventricular septal defects
- Size of pulmonary annulus
- Size of main pulmonary artery
- Size of branch pulmonary arteries
- Additional pulmonary arterial supply
What do we look for with Echo?

- Coronary arterial anatomy
- Aortopulmonary collaterals
- Right and left ventricular function
Fallot’s Tetralogy
Right Ventricular Outflow Tract
Right Ventricular Outflow Tract

FR 23Hz
7.0 cm

2D
72%
C 49
P Med
HGen
CF
80%
4.5 MHz
WF High
Med

VSD 10.3 mm

0/09/2016 09:56:55AM TIS1.2 MI 0.9
CARDIAC S8-3/KKHped

94 bpm
Additional Pulmonary Supply - Patent Ductus Arteriosus
Pulmonary Arterial Confluence

Hypoplastic PAs
ECHOCARDIOGRAPHY IN TETRALOGY OF FALLOT
Z Values

- Useful in guiding decision making
- Important to understand the basis by which values are derived
- Important to standardize which reference values are adopted
- Transannular patch at -2.5 Z – 4.0 Z?
  (consider also bicuspid versus trifoliate PV, intraoperative Heger sizing)
But…

- Echo underestimates PA sizes at hilum
- May not be able to delineate coronary arterial supply in patients with limited thoracic windows
- May not be able to visualize/ exclude MAPCAs
Do we need Cardiac Catheterization?

- Less frequently performed for Tetralogy of Fallot
- Distal pulmonary arteries
- Inability to visualize coronary arteries on 2D Echocardiography
- Major aortopulmonary collateral vessels – intervention
Figure Legend:

Graph showing an increase in coronary echocardiography and a concomitant decrease in diagnostic cardiac catheterization in 598 patients with all forms of tetralogy of Fallot examined from 1983 through 1995. Cath = diagnostic cardiac catheterization; Echo = echocardiography.
Distal Pulmonary Arteries in ToF

• In most infants the pulmonary arteries are sufficiently well visualized for surgical planning
• If ECHO windows are limited or if there is poor echogenicity further imaging e.g. cardiac cath may be required
Distal Pulmonary Arteries in ToF

Good-sized distal pulmonary arteries
Assessing the Hilar Pulmonary Arteries

- Mc Goon ratio > 1(2.0 – 2.5)
- Nakata Index > 200 mm²/BSA (>330 mm²/BSA)
Imaging Coronary Arteries in ToF

• Most frequent coronary consideration is exclusion of a coronary artery crossing the right ventricular outflow tract (where the surgeon would like to work)

• Other common variants of concern are the dual left anterior descending arteries and the left descending artery from the right coronary artery
Coronary Arterial Supply in Fallot’s Tetralogy

• Normal coronary artery anatomy in ToF has a spiral appearance due to rotation of the outflow tract

• 5-7% incidence of coronary artery abnormalities in ToF
Coronary Arterial Supply in Fallot’s Tetralogy

• Large conal branch arising from the RCA may preclude typical repair and require RV-PA conduit placement
Patterns of coronary artery anatomy in tetralogy of Fallot as seen from the parasternal short-axis view. The percentage of each pattern seen in 598 patients with all types of tetralogy of Fallot is indicated in the left lower corner. Ant = anterior; br = branch; Cx = left circumflex coronary artery; L = left; LAD = left anterior descending coronary artery; LCA = left coronary artery; Post = posterior; R = right; RCA = right coronary artery.
Figure Legend:

Origin of the LAD from the RCA crossing the right ventricular outflow tract. (A) High parasternal short-axis view. The enlarged proximal RCA supplies the LAD. (B) Leftward angled parasternal long-axis view profiling the right ventricular outflow tract (infundibulum) free wall. The LAD is seen in cross-section approximately 7 mm below the pulmonary valve. A = anterior; AoV = aortic valve; Inf = infundibulum; L = left; LAD = left anterior descending coronary artery; MPA = main pulmonary artery; RCA = right coronary artery; R/S = right/superior.
Collateral Vessels in ToF

- Collateral vessels may be an additional source of pulmonary supply in 5% of ToF patients
- Develop overtime secondary to hypoxia
- More common in late ToF presentation
The problem with MAPCAs

• Gross enlargement with erosion of bronchi resulting massive hemoptysis
• Excessive return to the left heart when the aorta is cross clamped on CPB flooding the operative field
• Can lead to pulmonary edema after operation
getting at MAPCAs

- surgical ligation or transcatheter embolization
- MAPCAs were arising from left subclavian artery and had difficulty in engaging the collaterals so the procedure time was prolonged, however MAPCAs from descending aorta were straight forward with much less procedure and fluoroscopic time.
Coiling of MAPCAs

CARDIAC CATHETERIZATION IN TETRALOGY OF FALLOT
Other Imaging Modalities

- CT Angiography
- Cardiac magnetic resonance imaging
Conclusion

• Echocardiography is sufficient to define the anatomy for surgical planning in most ToF cases (in particular in the younger child/infant)

• Cardiac catheterization is useful in delineating the distal pulmonary arteries, visualizing coronary arterial supply and defining MAPCAs
Conclusion

• Coiling of MAPCAs can also be performed
• CT Angiography and Cardiac MRI may be alternative emerging modalities in defining the anatomy of ToF for corrective surgery
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23 & 24 September 2016
Academia, Singapore
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This congress will be another major step forward in the field of cardiology by encouraging more young professionals to play a leading role in the future cardiology field. There will be state-of-the-art updates and cutting edge technology applied for cardiology practice in this congress.