Introduction
Tetralogy of Fallot (TOF)

- Incidence: 3·5% of infants born with CHD - 0·28/1000 live births
- Among the most common cyanotic heart disease
- Surgical repairs date back decades:
  - 1945- Blalock-Taussig shunt
  - 1954 –Open heart repair (Lillehei)
Introduction

- Tetralogy of Fallot: anterocephalad malalignment of conal septum resulting in:
  - Large unrestricive VSD
  - RVOT obstruction
  - RVH
  - Over-riding aorta

- In this discussion we have excluded patients with:
  - Pulmonary valve atresia
  - Absent pulmonary valve syndrome, and,
  - TOF associated with AV septal defect

Pathology of TOF

Courtesy: Dr W.D Edwards, Mayo Clinic, Rochester, MN
Commonly associated anomalies

- Secundum atrial septal defect
- L-SVC
- PDA
- Coronary artery anomalies:
  - Prominent conal branch (frequent)
  - Anterior descending artery from RCA
  - Dual anterior descending arteries (one each from RCA and LCA)
- Right sided aortic arch:
  - usually with mirror image branching

Indications for Surgery

- Diagnosis alone is indication for surgery
- Type of surgery and optimal timing in asymptomatic patients discussed later
- Earlier surgery for:
  - Hypercyanotic spells
  - Severe systemic desaturation
TOF repair

Palliative
1. AP shunt
2. RVOT stenting

Complete repair

Goals of primary repair

- VSD closure
- Relief of RVOT obstruction
  - While maintaining as competent a pulmonary valve as possible
  - Modest degree of RVOT obstruction with less regurgitation is currently preferred over complete relief of obstruction with severe PR
- Correction of other major associated defects (ASD/PDA)
Age of Repair of TOF

- 1993-1998, 227 consecutive children underwent complete repair
- Shunt palliation ↓ from 38% to 0%
- Mortality 2·6%, ↓ with transition to primary repair (0% in 1996–98)
- Primary repair ≤ 3 mo associated with longer ICU/hospital stay
- Likely optimum age of elective repair is 3–6 mo

Van Arsdell G et al: Circulation 2000

### Age at complete repair

- **Neonatal**
  - Longer post-op ICU/hospital stay
- **Infancy**
  - Lowest morbidity is in patients 3-11 mo age
- **Late (> 1 year)**
  - Risk of death and spells
  - Longer duration of hypoxia

Van Arsdell G et al: Circulation 2000
Candidates for initial palliation with shunt/RVOT stent

- < 2kg weight
- High risk for surgery
- Severe branch PA hypoplasia
- Coronary artery anomalies
  - Coronary crossing RVOT

Types of Complete Repair

- Tetralogy of Fallot with significant RVOT obstruction
- VSD repair with
  - Resection of sub-PS +/- pulmonary valvotomy
  - Infundibular patch +/- valvotomy
  - Transannular patch
Determinants of Surgical Strategy

Morphologic aspects of TOF

- Severe PA hypoplasia
- Coronary crossing RVOT
- Infundibular stenosis & adequate annulus
- Severe annular hypoplasia (Z-score < -3)

- Shunt RVOT stent
- Fenestrated VSD patch
- Shunt/RVOT stent/RV-PA conduit
- RVOT patch or subPS resection and valvotomy
- Transannular patch

Determination of branch PA hypoplasia

- Absolute size and Z-scores
- Nakata index:
  - Angiographic index using the AREA of branch PAs proximal to origin of 1st branch
  - LPA area + RPA area/BSA (m²)
  - Normal 330 mm²/m²
  - In general, in TOF Nakata index of > 100 mm²/m² can have complete repair
Assessment prior to initial repair/palliation

- Echocardiography usually provides a complete diagnosis
- Need for angiography/MRI uncommon
- Elements of complete echo assessment:
  - Complete segmental approach
  - Atrial shunt
  - VSD
  - RVOT, pulmonary annulus and Z-score, MPA/branch PA size, Z-score and Nakata index
  - Aortic arch sidedness and branching
  - Coronary artery anatomy

Late post-operative issues and re-operations
Long term sequelae of TOF repair

- Pulmonary regurgitation
- Residual RVOT obstruction
- Pulmonary artery stenosis
- Residual VSD/ASD
- RVOT aneurysm
- Tricuspid regurgitation
- Myocardial fibrosis
- Ventricular dysfunction
- Arrhythmias

Long term sequelae of PR

- Most asymptomatic in childhood
- By age 40, 29% symptomatic
- Death in 3/72 during follow-up about 3 yrs after onset of symptoms
- PR may result in:
  - Right heart dilation
  - Right heart failure
  - Tricuspid regurgitation
  - Arrhythmia
  - Sudden death

PR may increase with time

Increased PA capacitance → PR → Longer diastole

Increased RV compliance → RV dilation

PR relates to degree of right heart dilation

\[ y = 88.2 + 1.64x \]

\( r = 0.61 \)

\( p < 0.0001 \)
Restrictive Physiology

- Antegrade flow in MPA with atrial systole (throughout respiratory cycle)
- ↓ cardiac output and longer post-op stay in kids
- ? Beneficial in adults (↓ CT ratio, ↑ exercise tolerance, ↓ RV size, narrower QRS) – mixed results

Indications for PVR

- Moderate or more PR PLUS:
  - New onset of arrhythmia (VT)
  - Symptomatic exercise intolerance
  - Right heart failure
  - Asymptomatic with “significant” right heart dilation/dysfunction: more controversial
    - What is threshold for reverse remodeling?
Assessment of RV size

- 2D echo is limited:
  - Complex shape of RV – no simple geometric assumptions unlike LV
  - Poor visualization of endocardial border
  - Near field artifacts
- 3D echo may be better but still limited in patients with poor acoustic windows and needs validation when RV geometry is unusual
- MRI is gold-standard

Advantages of MRI

- Accurate assessment of RV volumes, no geometric assumptions
- Excellent visualization of RV endocardial borders in patients with poor echo windows
- Measure flow volumes
- Visualize extracardiac vasculature
- Free of ionizing radiation
Limitations of MRI

- Excludes patients with metallic implants, most pacemakers and ICDs
- Gd contrast contraindicated in patients with renal dysfunction and in pregnancy
- Long study, need for sedation/anesthesia in young
- Needs expertise and training, not widely available
- Expensive

Indications for PVR Based on MRI

- Moderate or more PR (Regurgitant fraction > 2%) PLUS: 2 or more of the following:
  - $\text{RVEDV} \geq 160 \text{ ml/m2} \ (\text{Z-score} > 5)$
  - $\text{RVESV} \geq 70 \text{ ml/m2}$
  - $\text{LVEDV} \leq 65 \text{ ml/m2}$
  - $\text{RV EF} \leq 45\%$
  - RVOT aneurysm

Landmark papers
Are We Operating Too Late?

- 25 adults post PVR. Radionuclide angiography performed 8.2 mo before PVR & 28.0 mo after
- Age at TOF repair 12 yrs, PVR 33 yrs
- Mean RVEDV, RVESV and RVEF remained unchanged after PVR
- Of 10 patients with RVEF > 0.40 before PVR, 5 maintained RVEF > 0.40 following
- Only 2 of 15 patients (13%) with pre-op RV EF < 0.40 reached an RVEF 0.40 post-op (p = 0.001)

Webb, G et al: J Am Coll Cardiol 2000

Landmark papers
Optimal timing of repair

- 17 adults, repaired TOF followed post PVR (mean age 34 ± 12 yrs), many symptomatic, many had initial BT shunts, all had late repair beyond infancy
- Mean follow-up 21 months
- RV EF 32 ± 7% to 34 ± 10% (p = NS)
- None achieved normal RV end-diastolic volume (EDV < 108 ml/m2) or RV end-systolic volume (ESV < 47 ml/m2) if RV EDV > 170 ml/m2 or RV ESV > 85 ml/m2

Therrien, J et al: Am J Cardiol 2005
Landmark papers
PVR in preadolescents

- 101 pts ≤ 13 yrs had MRI.
- Median age at complete repair 6 mo
- Mean RVEDV at 1st study = 135 ± 39 mL/m²
- In 32 with serial MRI, RVEDV ↑ 9 mL/m²/yr
- RVEF ↓ in 46 (46%)
- 42 had PVR at mean age of 8 ± 3 years
  - Initial surgical criteria: RV:LV > 2:1 with ↓ decreased RV or LV EF, arrhythmias or symptoms
  - Current criteria: Indexed RVEDV > 165 mL/m², ↓ RV EF, or NYHA class III symptoms
- No deaths, no reinterventions


RV remodeling after early PVR in children

Valsangiacomo Buechel ER et al: Eur Heart J 2005
Clinical Evaluation (Repaired TOF)

- **History**: Symptoms, NYHA class, syncope
- **ECG**: Ectopy, QRS duration (>180ms risk factor)
- **Holter**: Asymptomatic ventricular arrhythmia
- **Exercise test**: Objective evidence of deteriorating cardiorespiratory function
- **Echocardiography**: Ventricular size and function, RV pressure, TR, status of RVOT
- **MRI (rarely CT if MRI contraindicated)**: Volumes, flow
- **EP study/cardiac catheterization**: When clinically indicated