Tetralogy of Fallot

Dr Nitha Naqvi
Tetralogy of Fallot

- 1 – VSD
- 2 – pulmonary stenosis
- 2a – infundibular stenosis
- 3 – enlarged aorta overriding VSD
- 4 – right ventricular hypertrophy
Tetralogy of Fallot

Right-sided Aortic Arch
Tetralogy of Fallot

- Outlet VSD
- overriding aorta
- RV/ pulmonary outflow obstruction
- RVH
Variants

- With pulmonary atresia and patent duct
- With absent pulmonary valve syndrome
- With pulmonary atresia & systemic-pulmonary collaterals
- With double outlet RV
Tetralogy of Fallot

Definition

Normal Outlet Septum

Ant Deviated Outlet Septum

LA  RV  AO  LV  PT  OS

LA  RV  AO  LV  PT  OS
Tetralogy of Fallot - classic

Pathophysiology of presentation

- Large unrestrictive VSD - equal ventricular pressures

- RV-PA outflow obstruction – high RV-PA gradient
  - the more severe, the earlier the clinical presentation

- RVH – secondary to RV outflow obstruction

- Cyanosis - R to L shunt across VSD
  - Often progressive during infancy

- Acyanotic - balanced or L to R shunt across VSD
  - VSD: ~10%
Tetralogy of Fallot - classic
Pathophysiology of presentation -2

- Single S2 – low PA diastolic pressure
- Ejection systolic murmur – RV-PA obstruction
  - the more severe the shorter the murmur
- Spells (40%) – infundibular shutdown
- Heart failure - unusual: systemic-PA collaterals → Continuous murmurs
- ECG: RAD, RVH classically
  - Superior axis suggests additional AV septal defect
Tetralogy of Fallot

Diagnosis

- Initially: echocardiography
- Pre-definitive repair:
  - echocardiography in majority of cases
  - Angiography, 64-slice CT scan, MRI in minority
Normal vs Tetralogy of Fallot

Echo: subcostal right anterior oblique

Normal

Tetralogy of Fallot

AO
PT
RPA
LPA
RV
RA
TV

LA
AO
OS
RA
TV
RV
subcostal right anterior oblique
Tetralogy of Fallot

subcostal right oblique

PT
AO
OS
RA
RV
TETRALOGY OF FALLOT

**DIAGNOSIS**

- ECHOCARDIOGRAPHY alone IN MAJORITY!
- CT – coronaries
- CARDIAC CATH/ANGIOGRAPHY

**AN ALTERNATIVE TO ANGIOGRAPHY**

- MRI

PULMONARY ARTERY ANOMALIES
CORONARY ANOMALIES (EARLY INFANCY)
AORTOPULMONARY COLLATERALS
INADEQUATE ECHO IMAGING

ESPECIALLY IN OLDER CHILDREN AND ADULTS
Tetralogy of Fallot
Infundibular PS /Colour Flow
Infundibular PS /Colour Flow
Tetralogy of Fallot

RV angiography (RAO)
PARASTERNAL LONG AXIS
Tetralogy of Fallot

PARASTERNAL SHORT AXIS
Perimembranous VSD

AO
PT
RPA
LPA

RV
TV
OS
RA
AO
PT
RPA
LPA
RVOT doppler
Angiography
aortic override & rule out additional VSD(S)

Long Axis RV Angio

Long Axis LV Angio
Pulmonary Arteries
Morphological Variables

- Outlet VSD with aortic override:
  - Perimembranous 80% / Muscular Inferior Rim 15%
  - Doubly committed 5% / Restrictive - rare

- Ventriculo-arterial connections:
  - Concordant / Double outlet RV 5-10%

- Pulmonary Stenosis:
  - Infundibular / Valvar / Supravalvar
Morphological Variables

- **Pulmonary Arteries:**
  
  *Hypoplasia / Stenoses / Absent Rt Or Lt* 10-12% *(not PAAtresia)*

- **Aortic arch - Right Arch 25%**

- **Coronary Arteries – 6% abnormal**

- **Systemic to pulmonary collaterals <5% of classic TOF**
Tetralogy of Fallot

Doubly Committed Subarterial VSD
Fallot: Coronary Arterial Patterns

N = 598

Need et al. JACC 2000
Coronary Arteries

Normal

LAD from RCA
If the echocardiographic diagnosis is felt to be equivocal, the surgeon is alerted and is prompted to carefully examine the proximal coronary arteries.

Need et al. JACC 2000
Arch sidedness
Right Arch
Associated Anomalies

- Secundum ASD **10%**
- Additional Muscular VSD(s) – **3%**
- AV Septal Defect - **2%**
- Straddling Tricuspid Valve - **< 0.5%**
- PAPVD – **1%**
- Others (v rare): AS, AR, hypoplastic RV...
- Absent Pulmonary Valve Syndrome
- Additional Sources of Pulmonary Blood Supply:
  - Systemic to pulmonary collaterals (MAPCAs)
  - PDA (Common)
Tetralogy of Fallot

AV Septal Defect

Parasternal 4 Chamber

Subcostal Short Axis

RA

LA

RV

LV

S

AO

COMMON AV JUNCTION
Tetralogy of Fallot
Associated Malformations

Straddling/Overriding TV

Apical Muscular VSD

RA
LA
MV
S
TV
RV
LV
S
MV
LV
RV
VSD
Additional sources of pulmonary flow

Patent Arterial Duct

Aortopulmonary Collaterals
Absent Pulmonary Valve Syndrome

Dilated Pulm Arteries
Bronchial Compression
Chest Infections
Pulm Regurgitation
Without Cyanosis
Absent Pulmonary Valve Syndrome
TETRALOGY OF FALLOT

apsulation in morphology

important to assess on echo

1. Degree of Pulmonary Artery Hypoplasia
2. Left Anterior Descending Coronary Artery?
3. Double Outlet Right Ventricle?
4. Straddling Tricuspid Valve?
5. Atrioventricular Septal Defect?
6. Additional Sources of Pulmonary Blood Supply?
Tetralogy of Fallot
Pre-repair Diagnosis - Conclusions

- Echocardiography in majority of cases
- Angiography, CT, MRI
  - Unclear anatomy, particularly distal PAs
  - Delineation of additional sources of PBF:
    - systemic-to-pulmonary collateral arteries
  - Coronary arterial anatomy - rarely
  - Discrepancy of clinical vs echo findings
  - Interventional procedures
Considerable Variation in Morphology
Repair of Tetralogy of Fallot

Subclavian Artery to Pulmonary Artery Anastomosis (Blalock-Taussig Shunt)