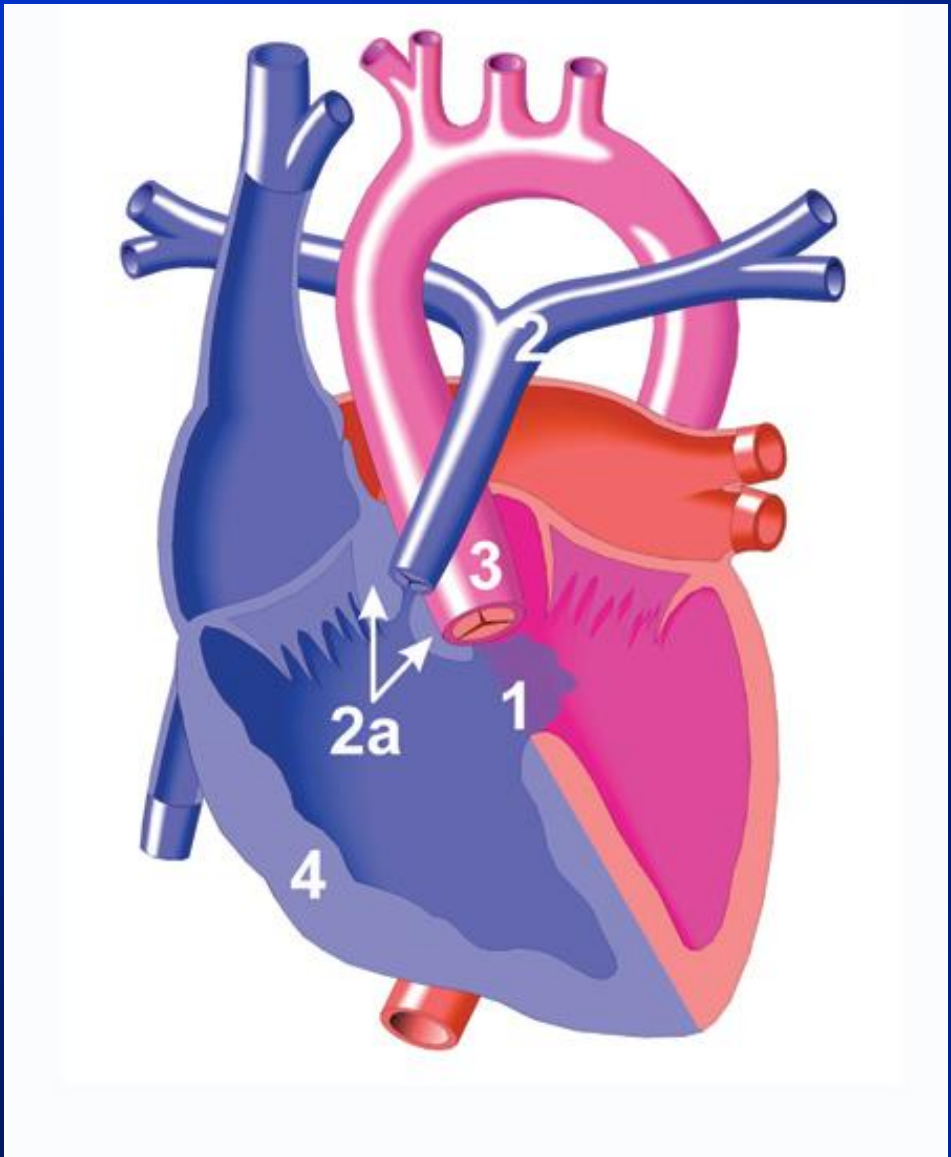
A lifetime of specialist care

Royal Brompton & Harefield NHS
NHS Foundation Trust

Tetralogy of Fallot

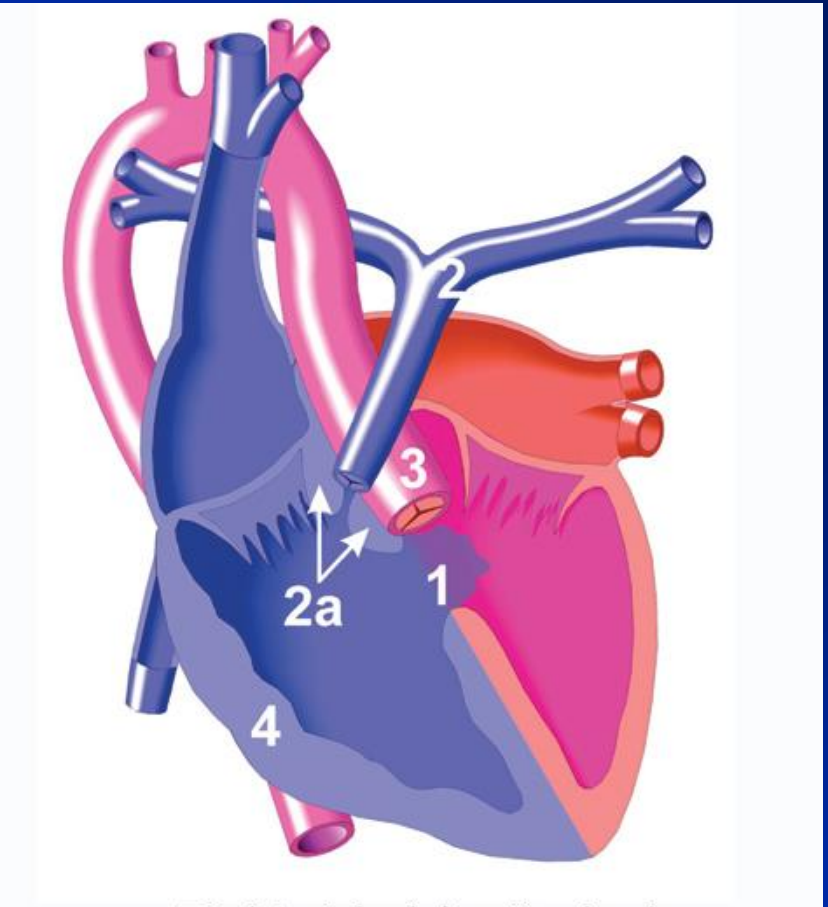
Dr Nitha Naqvi

Tetralogy of Fallot



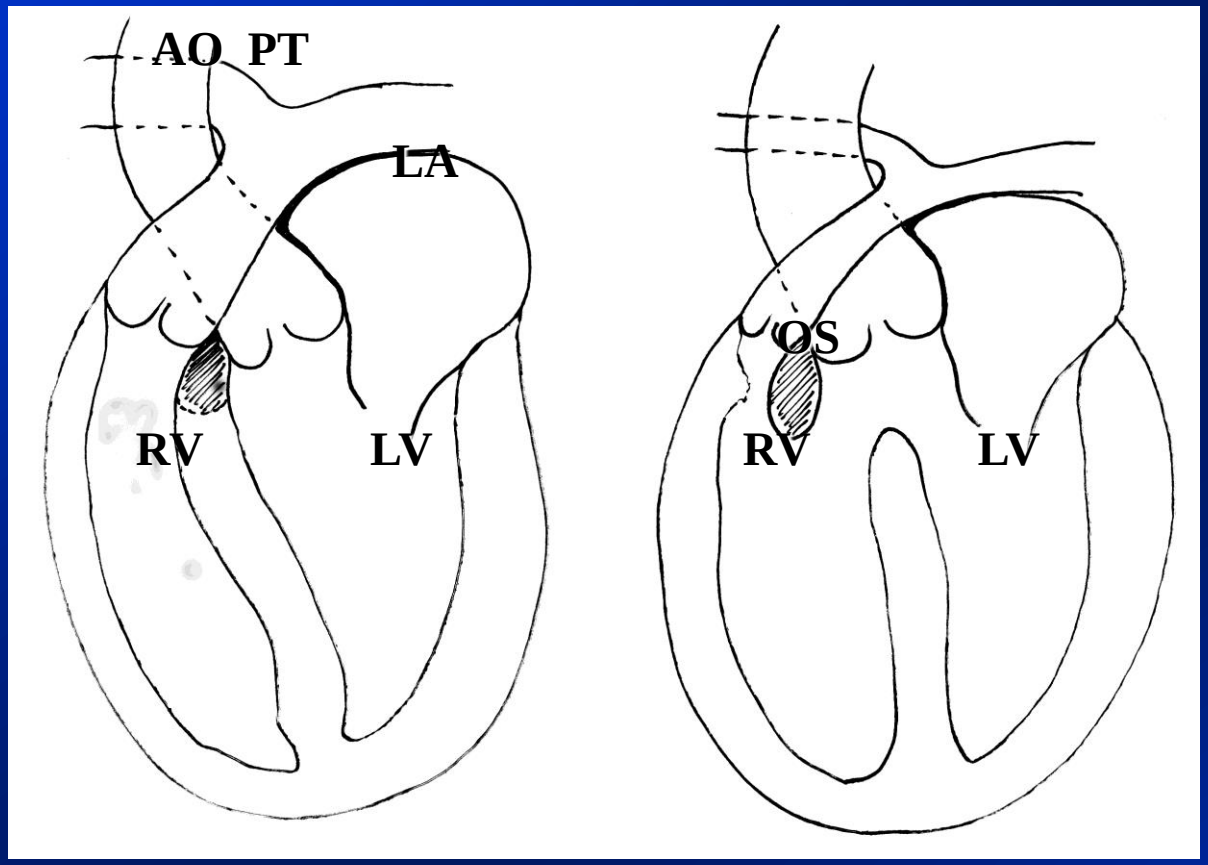
- 1 – VSD
- 2 – pulmonary stenosis
- 2a – infundibular stenosis
- 3 – enlarged aorta overriding VSD
- 4 – right ventricular hypertrophy

Tetralogy of Fallot



Right-sided Aortic Arch

Tetralogy of Fallot



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

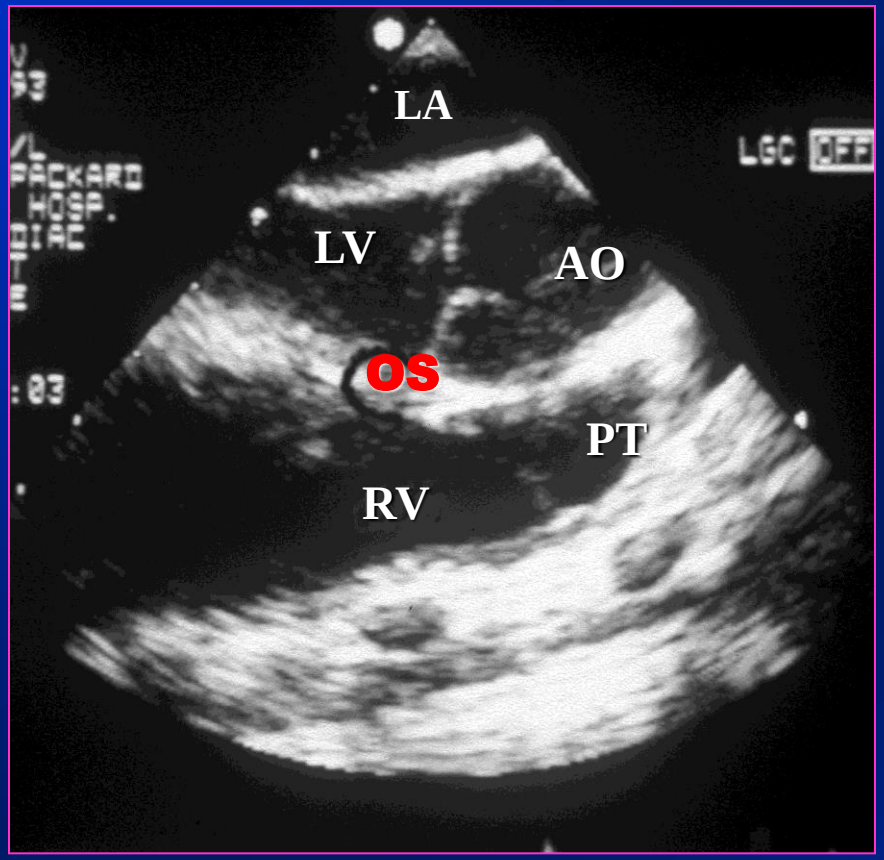
Variants

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

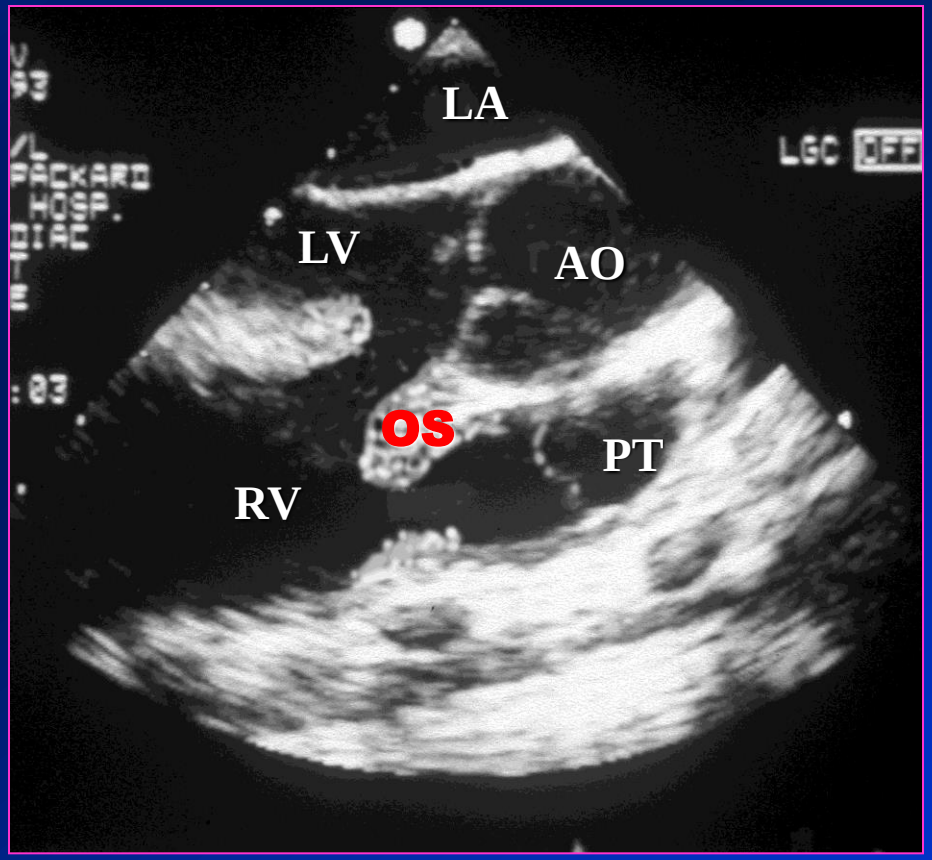
Tetralogy of Fallot

Definition

Normal Outlet Septum



Ant Deviated Outlet Septum



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: ~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**

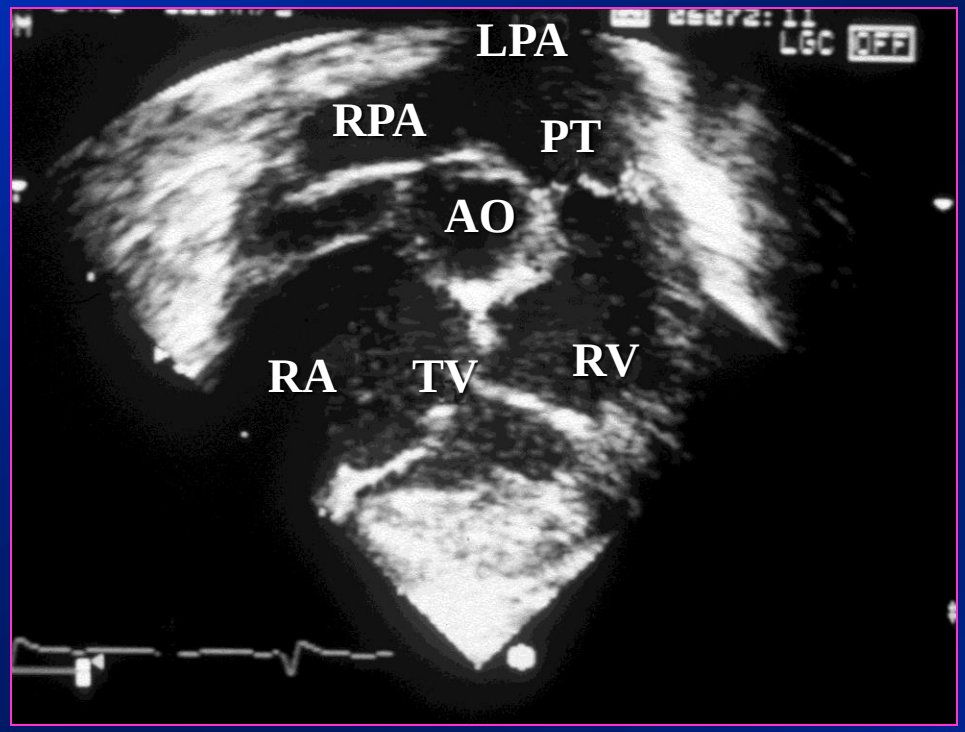
Tetralogy of Fallot
Diagnosis

- **Initially: echocardiography**
- **Pre-definitive repair:**
  - **echocardiography in majority of cases**
  - **Angiography, 64-slice CT scan, MRI in minority**

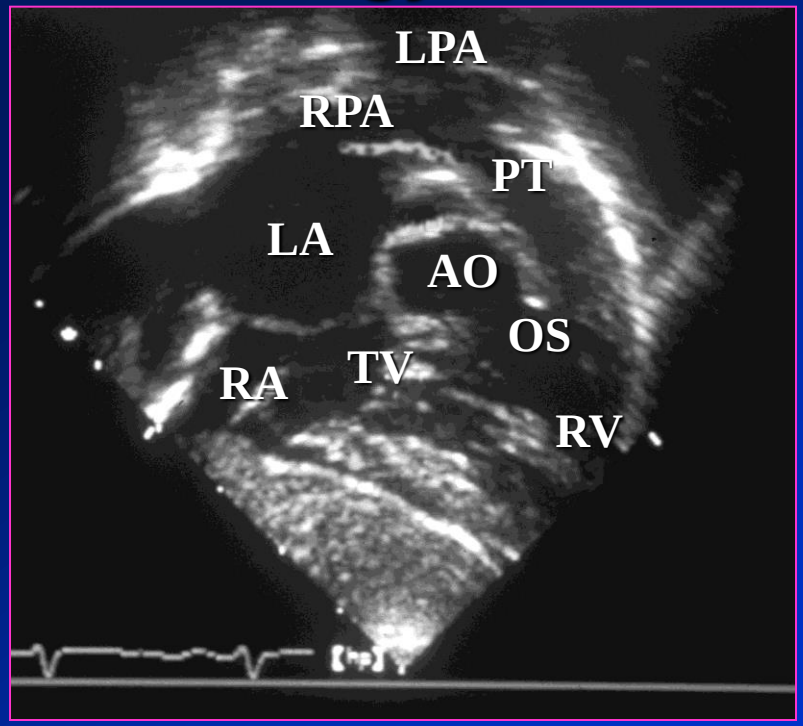
Normal vs Tetralogy of Fallot

Echo: subcostal right anterior oblique

Normal



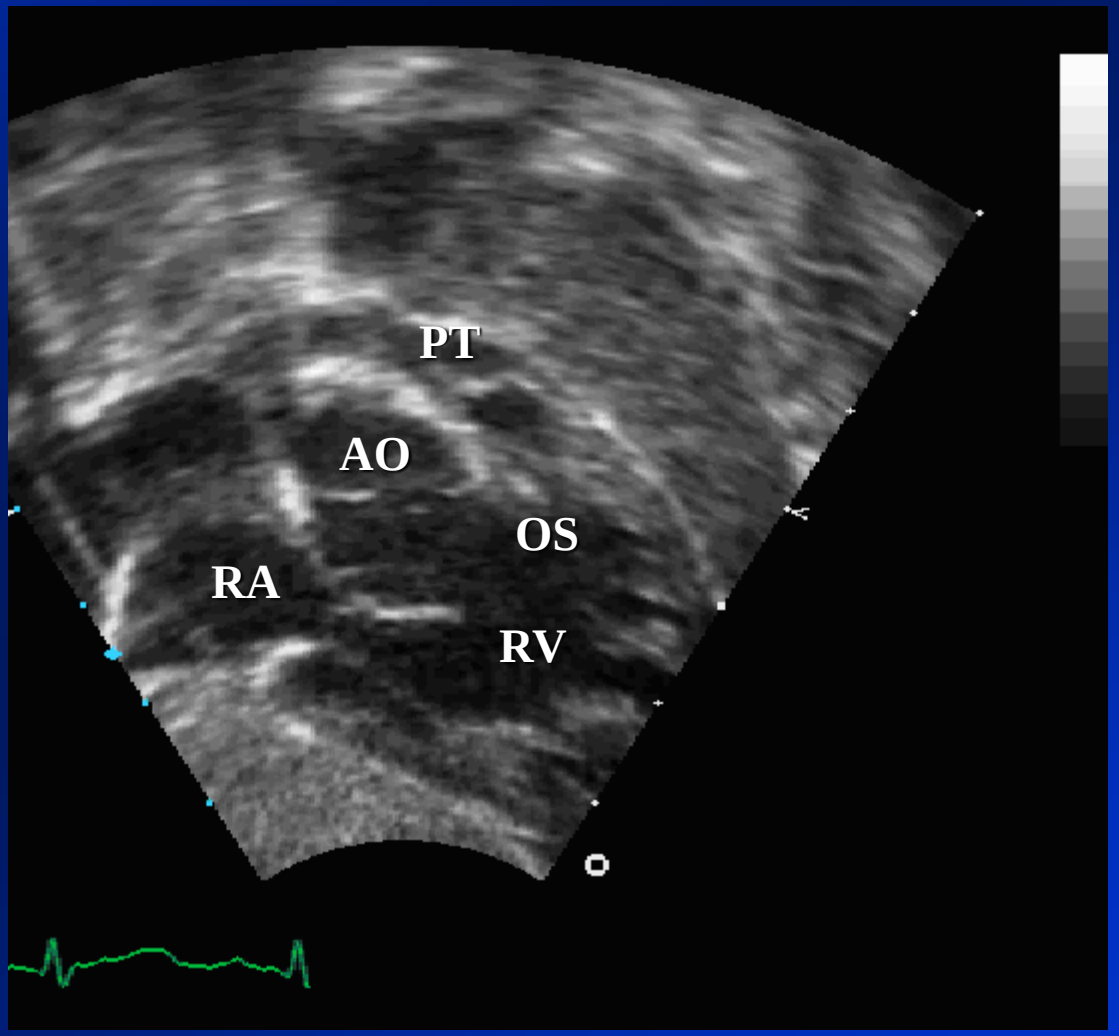
Tetralogy of Fallot



subcostal right anterior oblique

Tetralogy of Fallot

subcostal right oblique



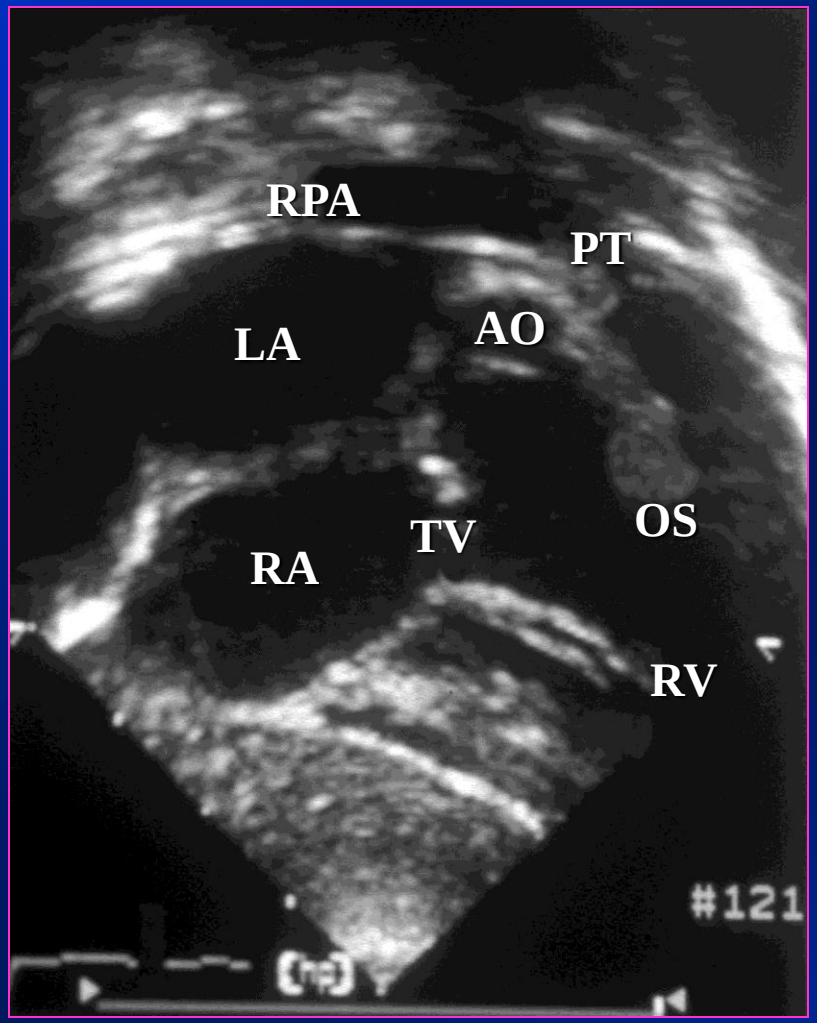
TETRALOGY OF FALLOT
DIAGNOSIS

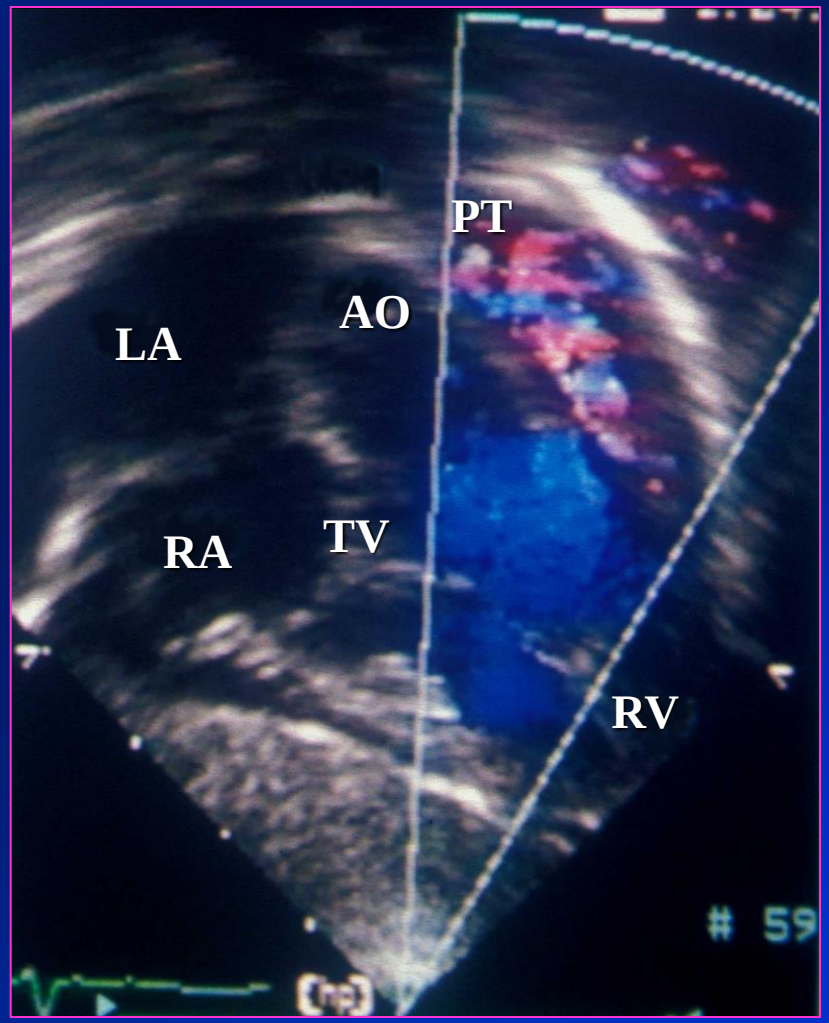
- **ECHOCARDIOGRAPHY alone IN MAJORITY !**
- **CT – coronaries**
- **CARDIAC CATH/ANGIOGRAPHY**
  - **PULMONARY ARTERY ANOMALIES**
  - **CORONARY ANOMALIES (EARLY INFANCY)**
  - **AORTOPULMONARY COLLATERALS**
  - **INADEQUATE ECHO IMAGING**
- **MRI**
  - **AN ALTERNATIVE TO ANGIOGRAPHY**

ESPECIALLY IN OLDER CHILDREN AND ADULTS

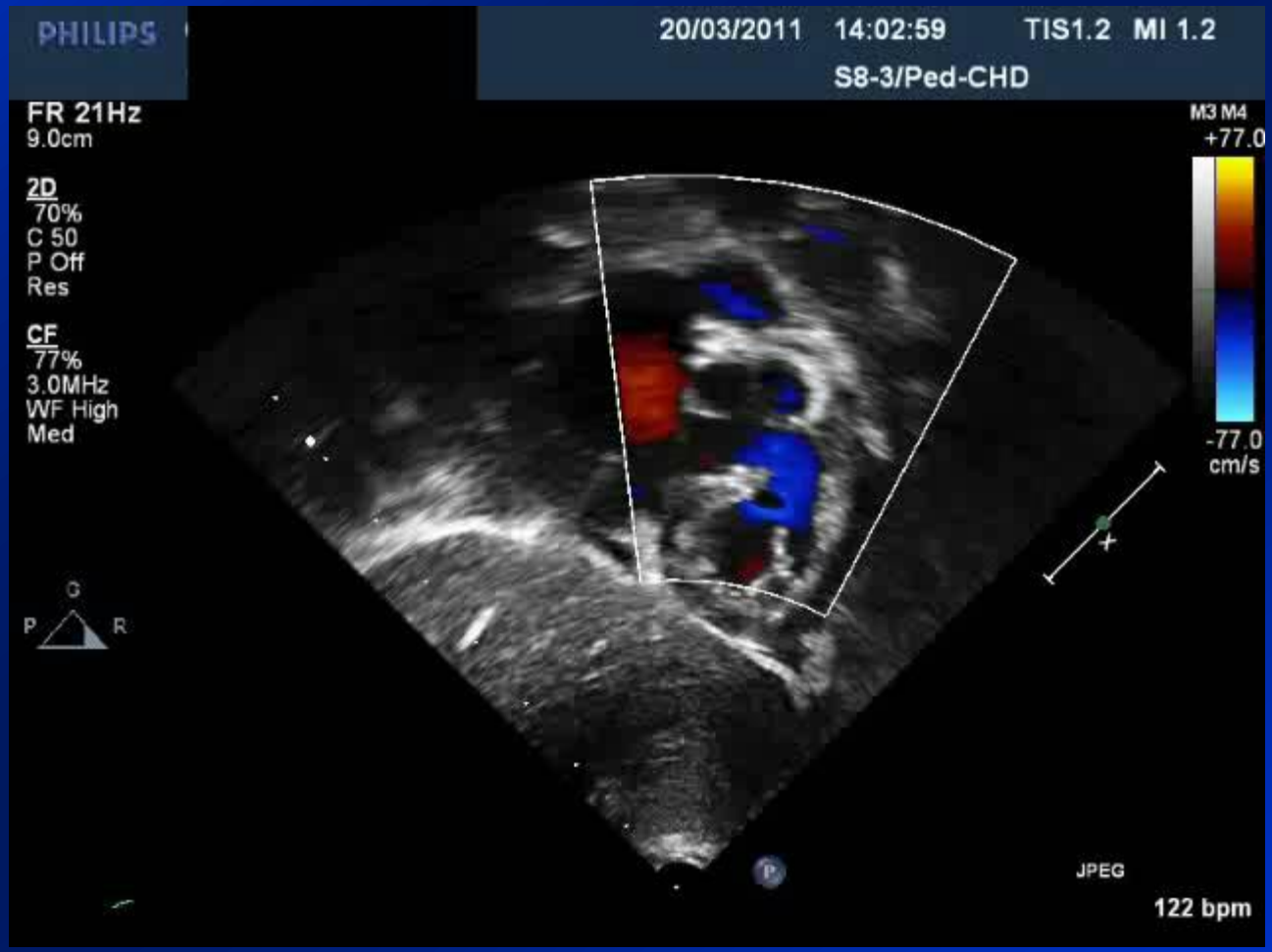
Tetralogy of Fallot

Infundibular PS /Colour Flow



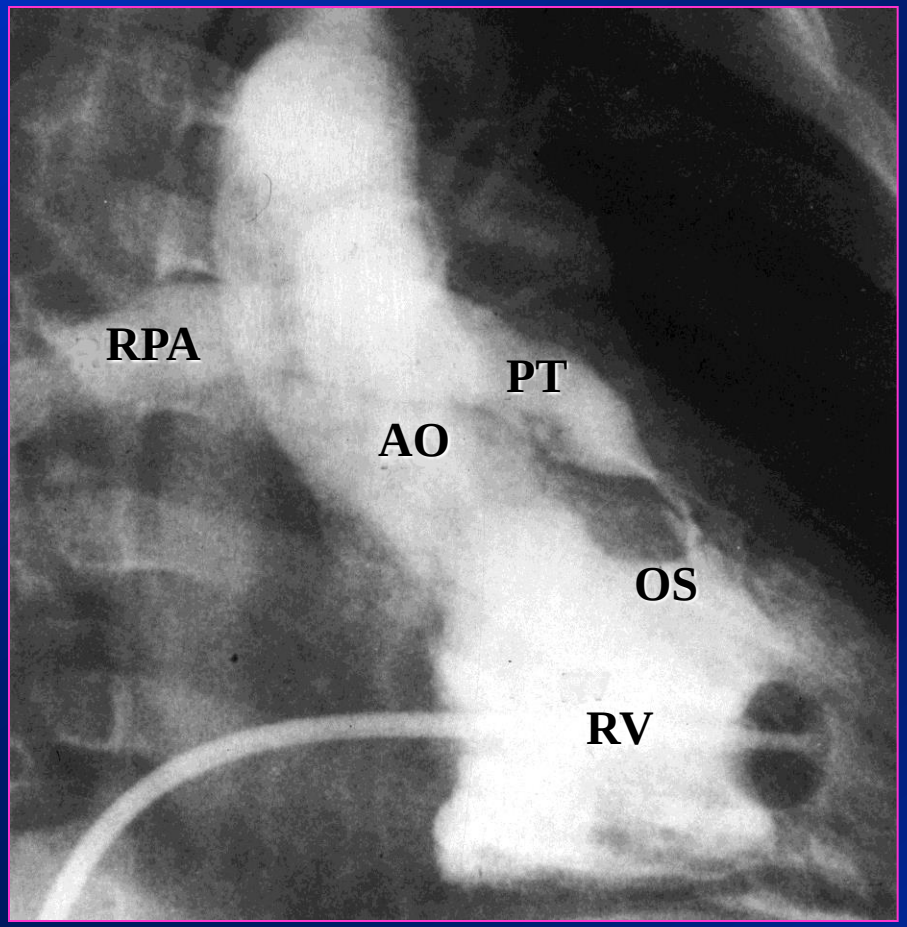


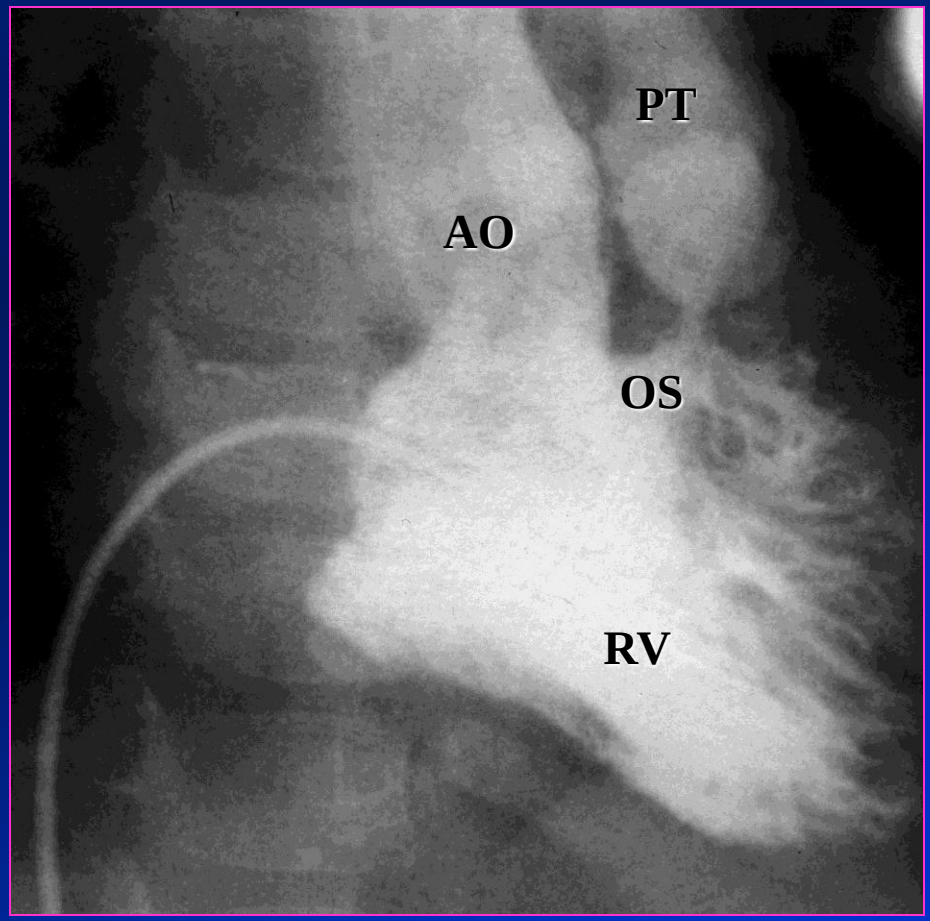
Infundibular PS /Colour Flow



Tetralogy of Fallot

RV angiography (RAO)



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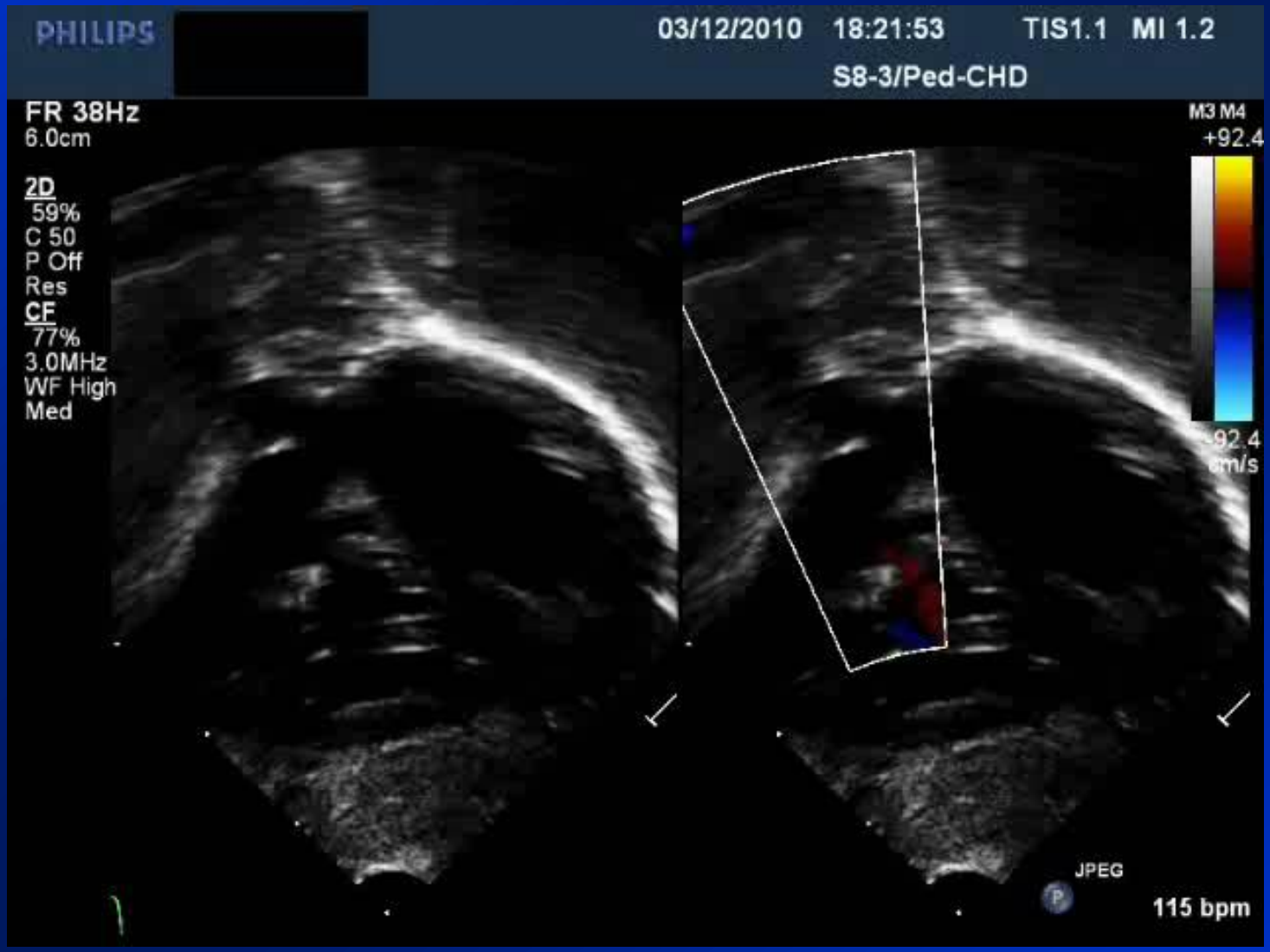
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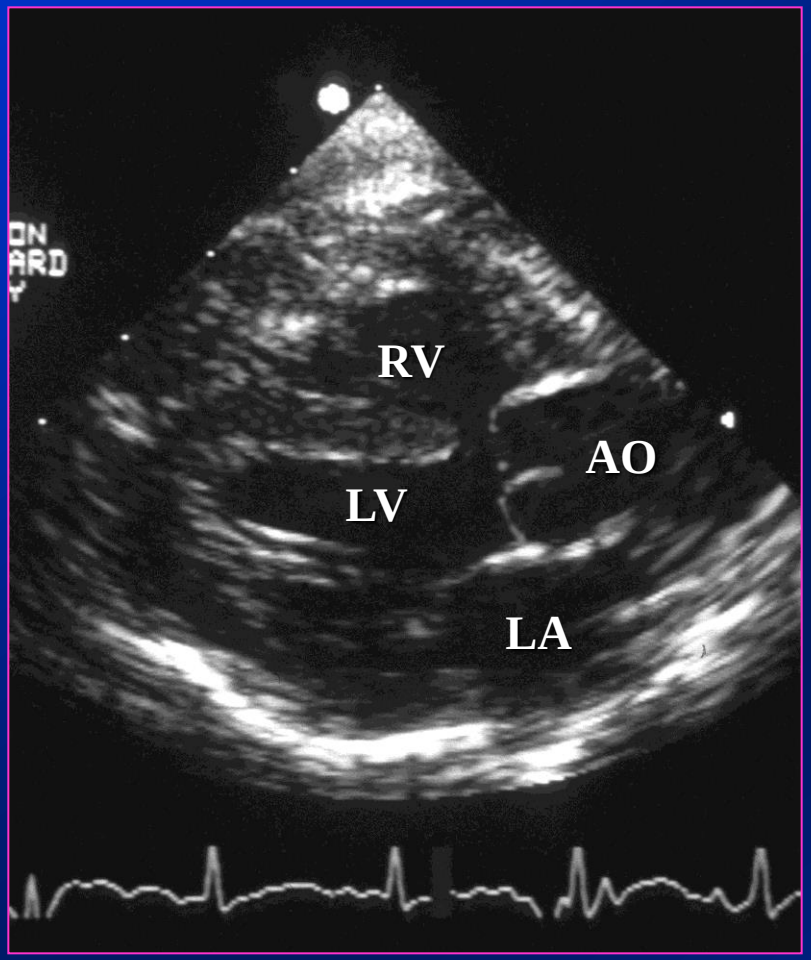
PARASTERNAL LONG AXIS

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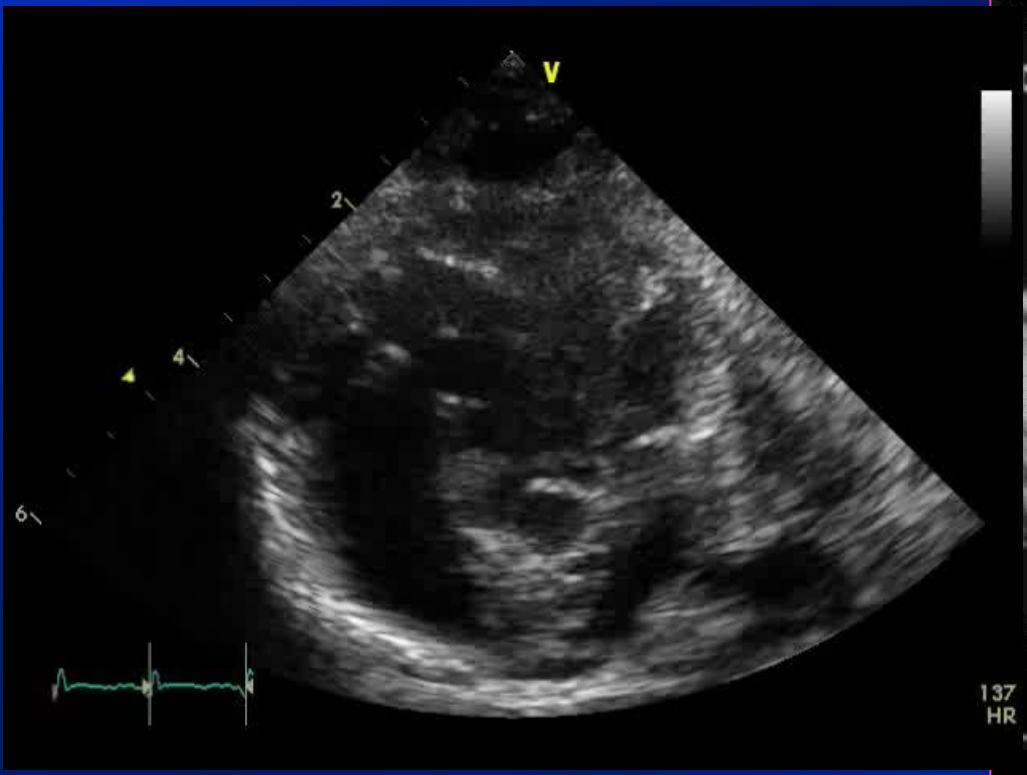
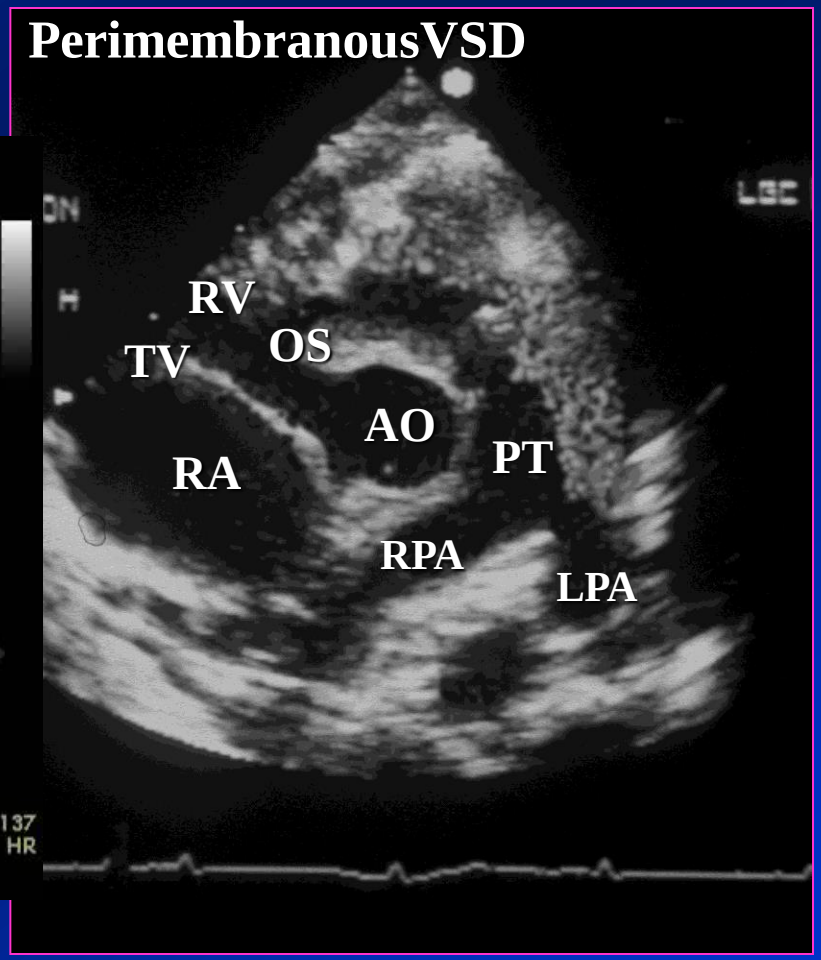
Tetralogy of Fallot

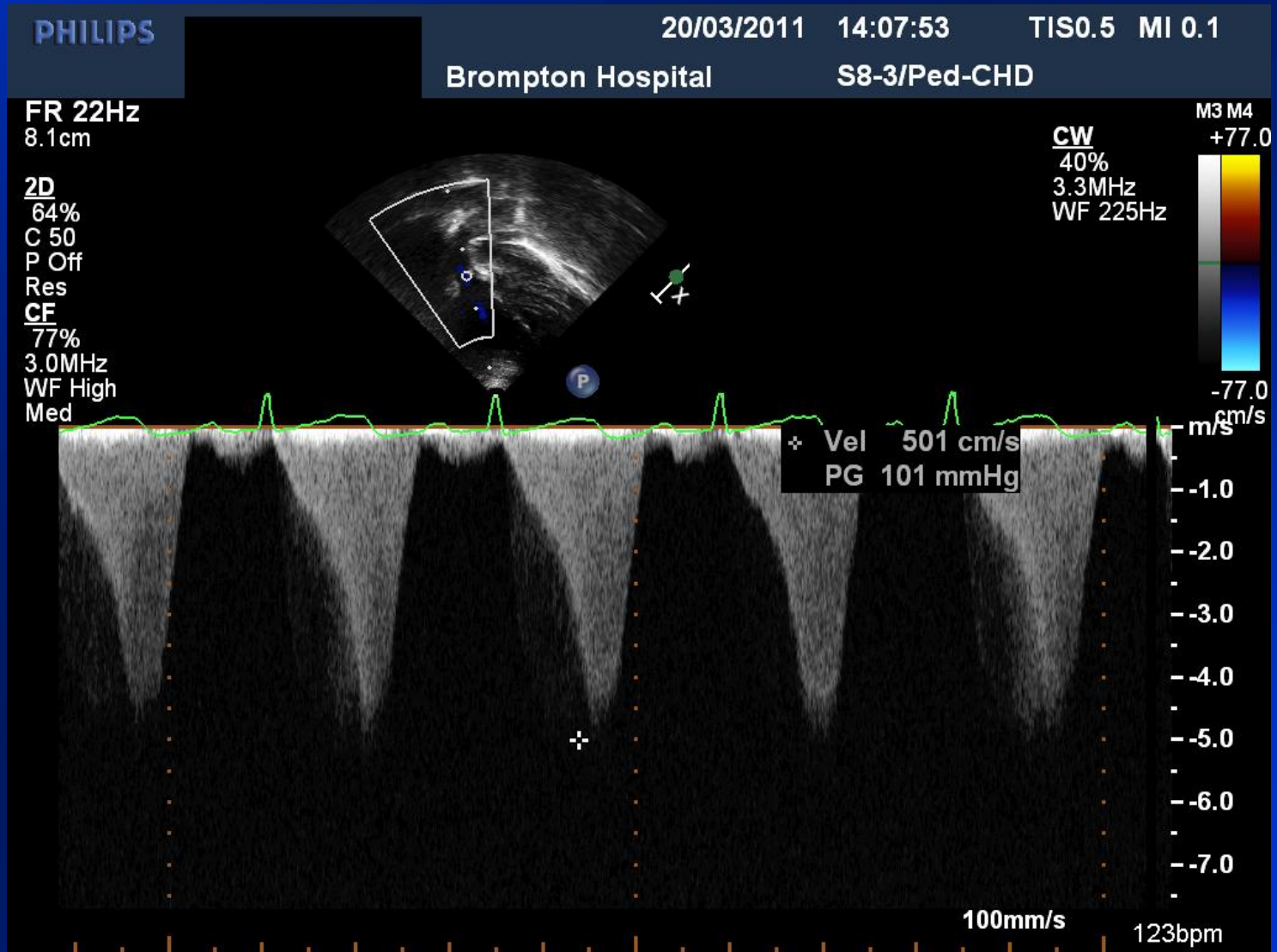


PARASTERNAL SHORT AXIS




IPTON

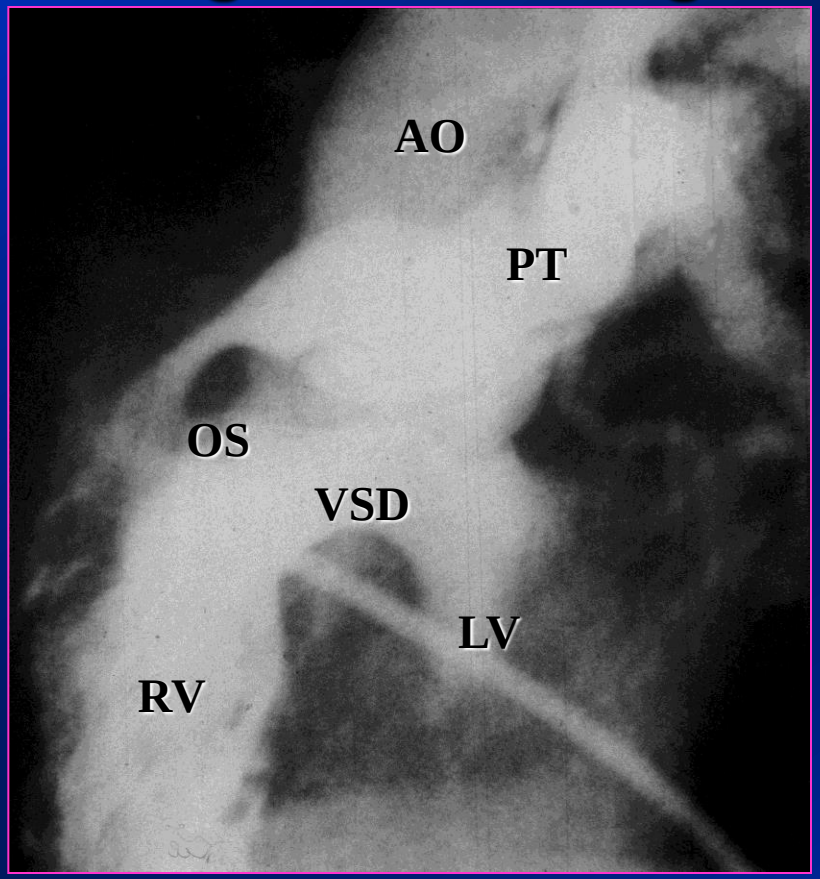
RVOT doppler



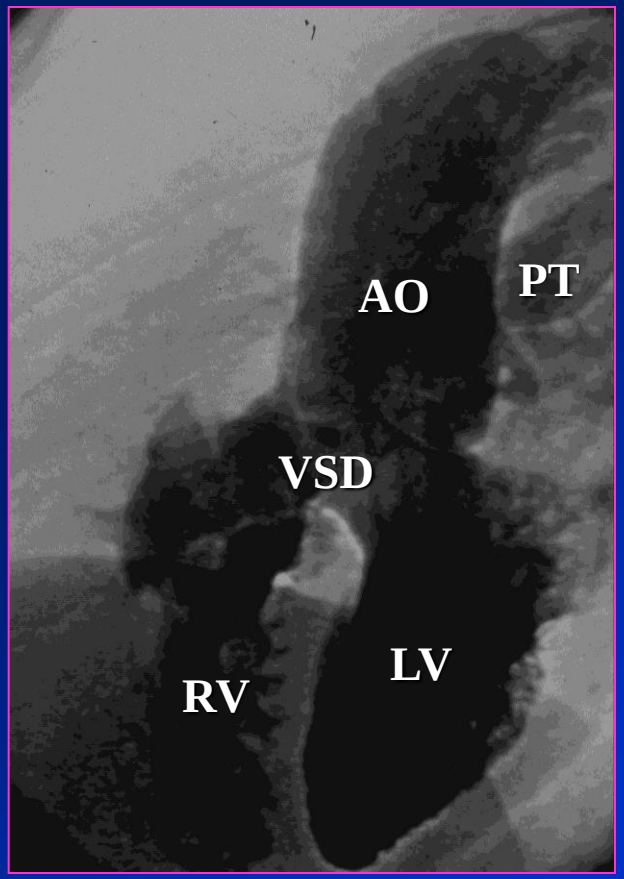
Angiography

aortic override & rule out additional VSD(S)

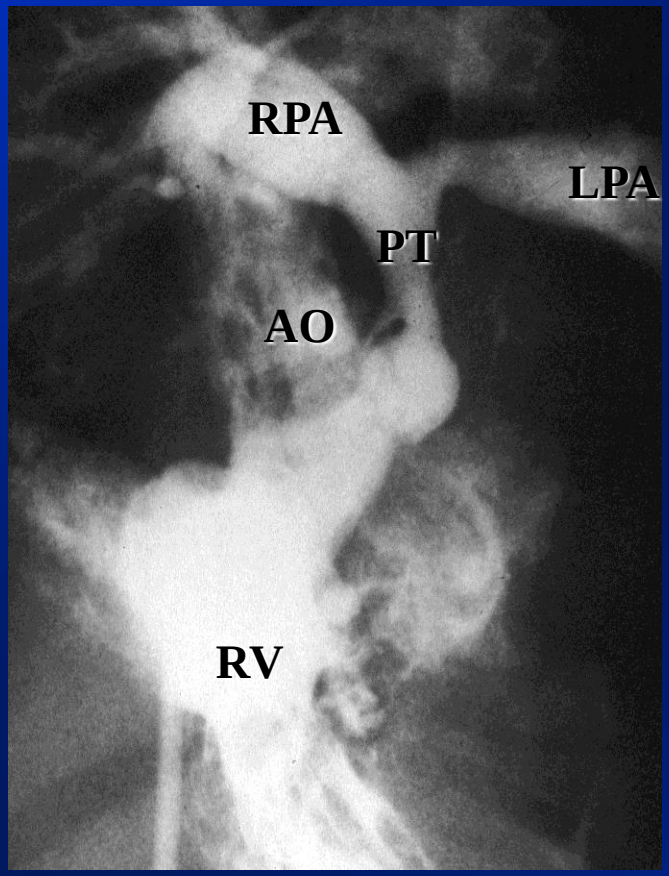
Long Axis RV Angio



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 PAtresia)

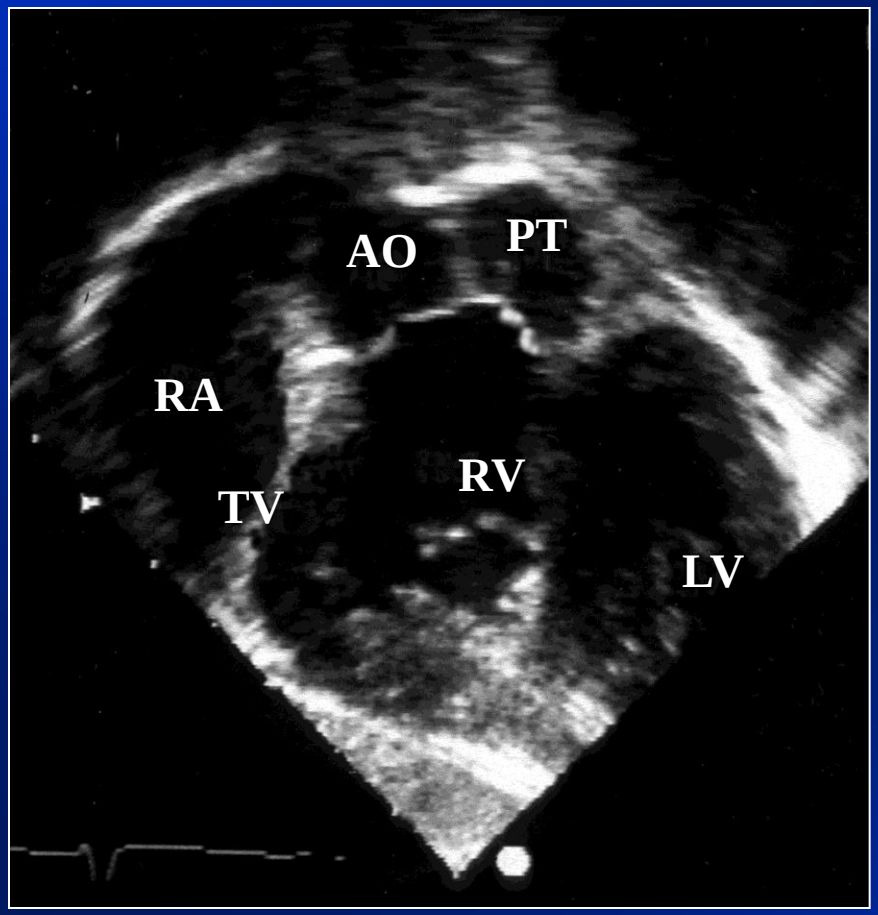
- **Aortic arch - *Right Arch 25%***

- **Coronary Arteries – 6% abnormal**

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

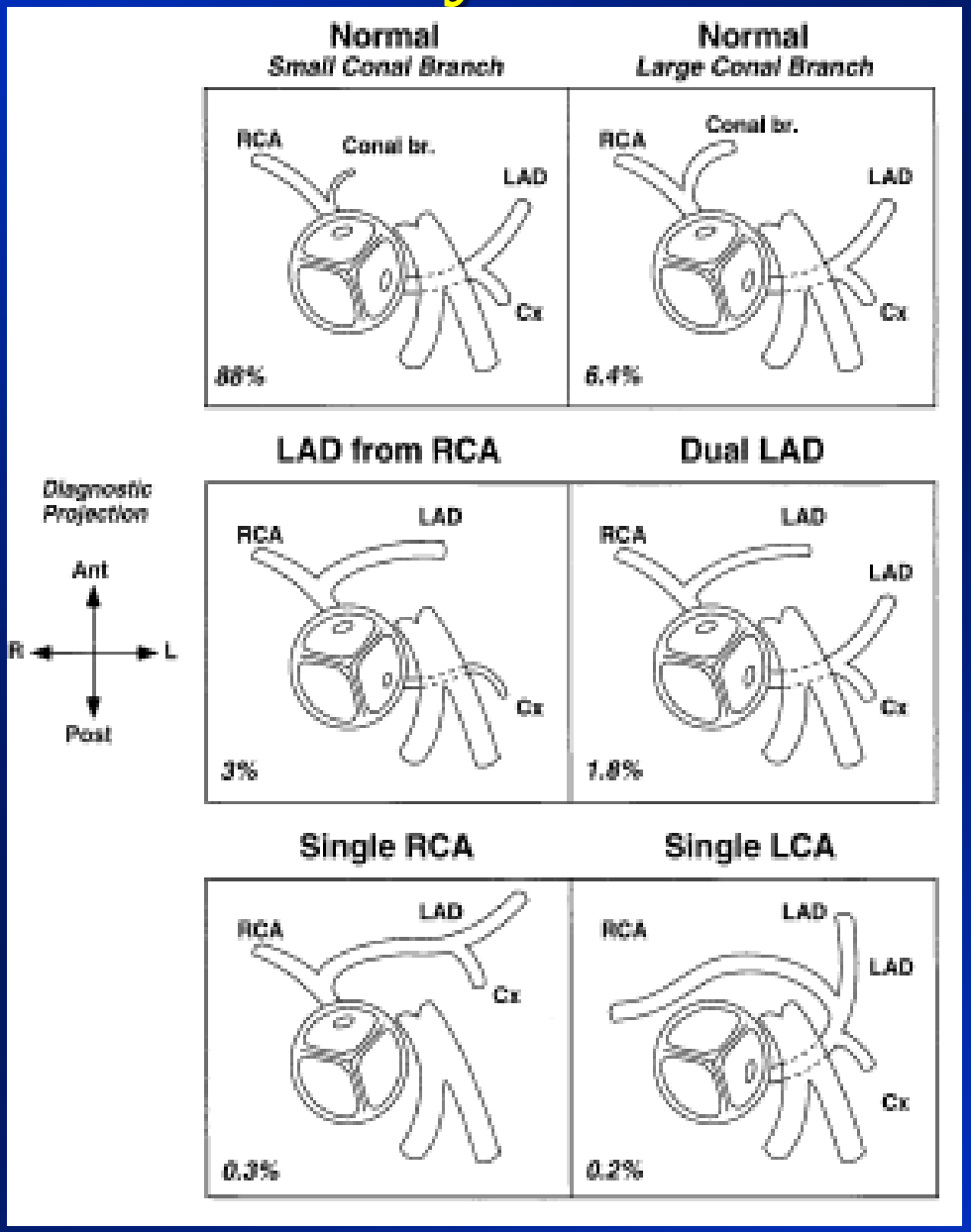
Tetralogy of Fallot

Doubly Committed Subarterial VSD



Fallot: Coronary Arterial Patterns

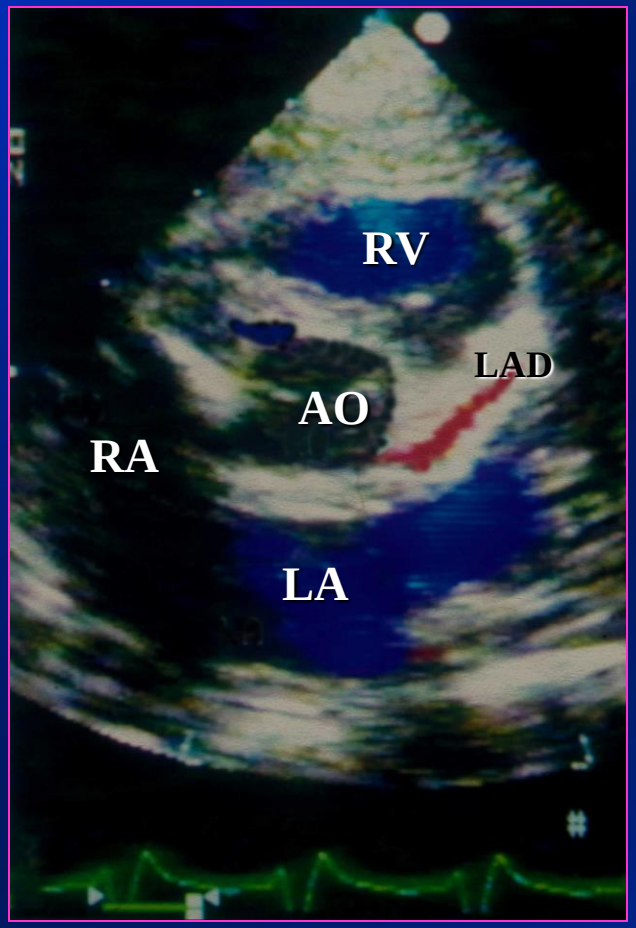
N = 598



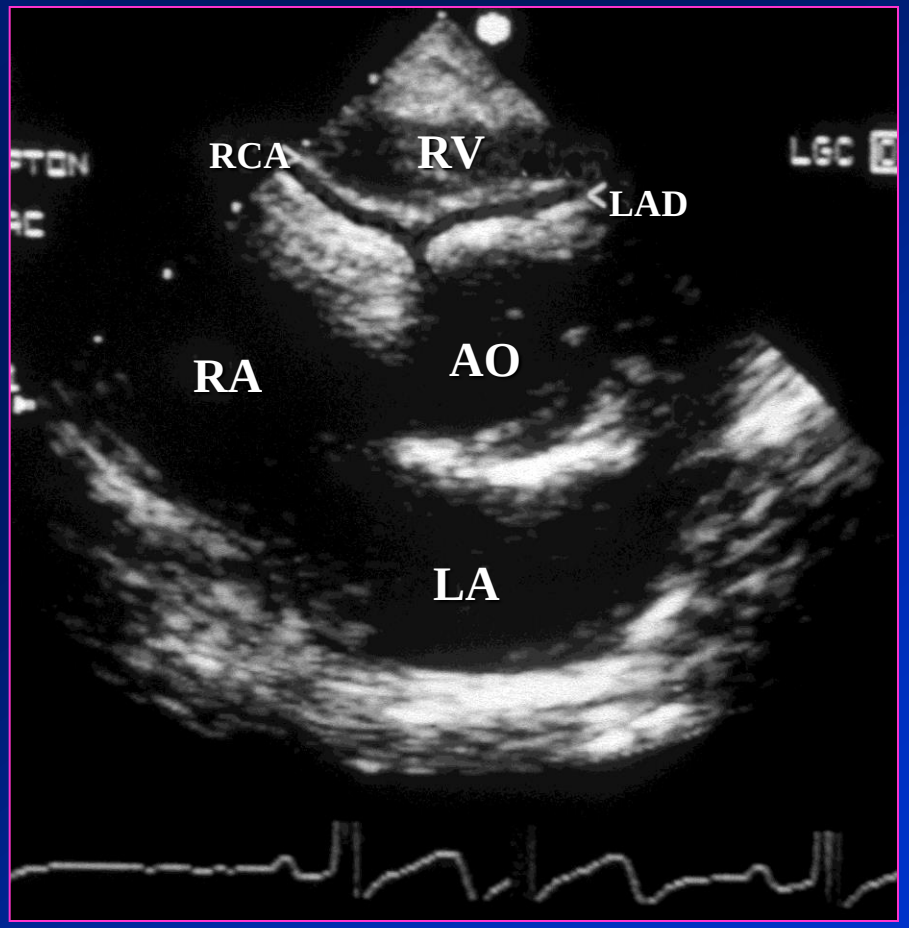
Need et al. JACC 2000

Coronary Arteries

Normal

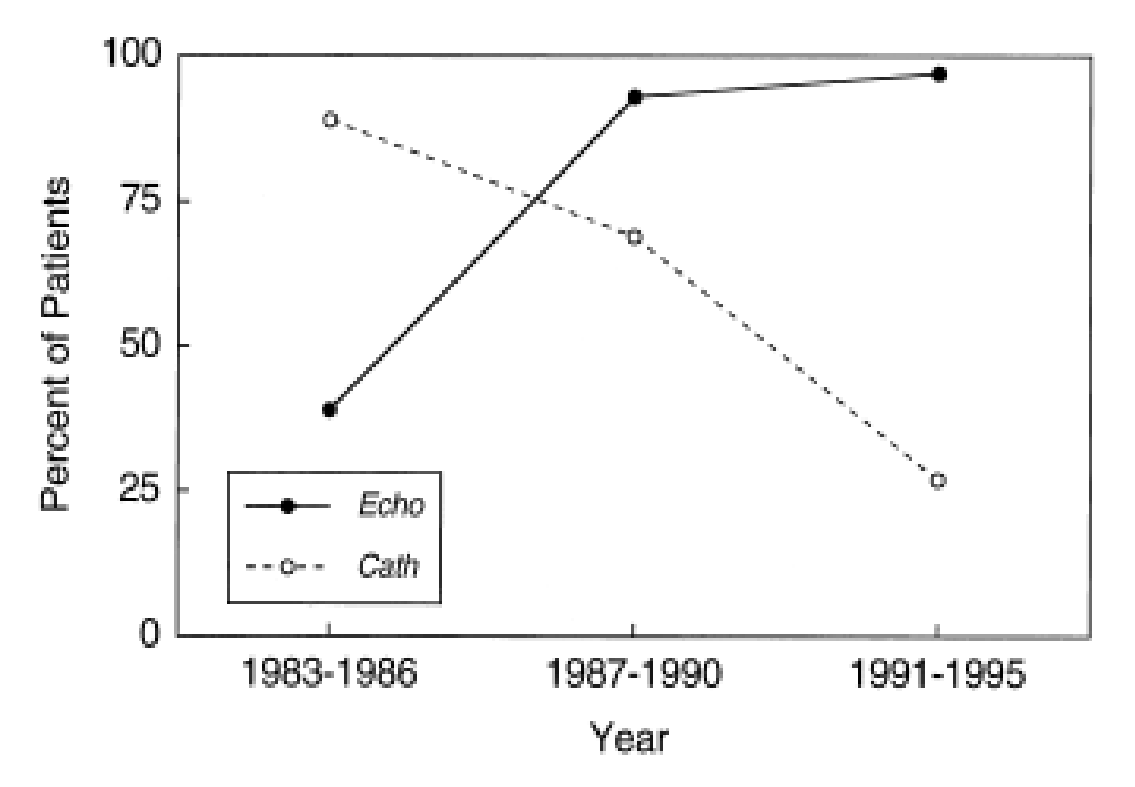


LAD from RCA



Fallot repair: echo vs cath diagnosis

Coronary assessment Boston: N= 598

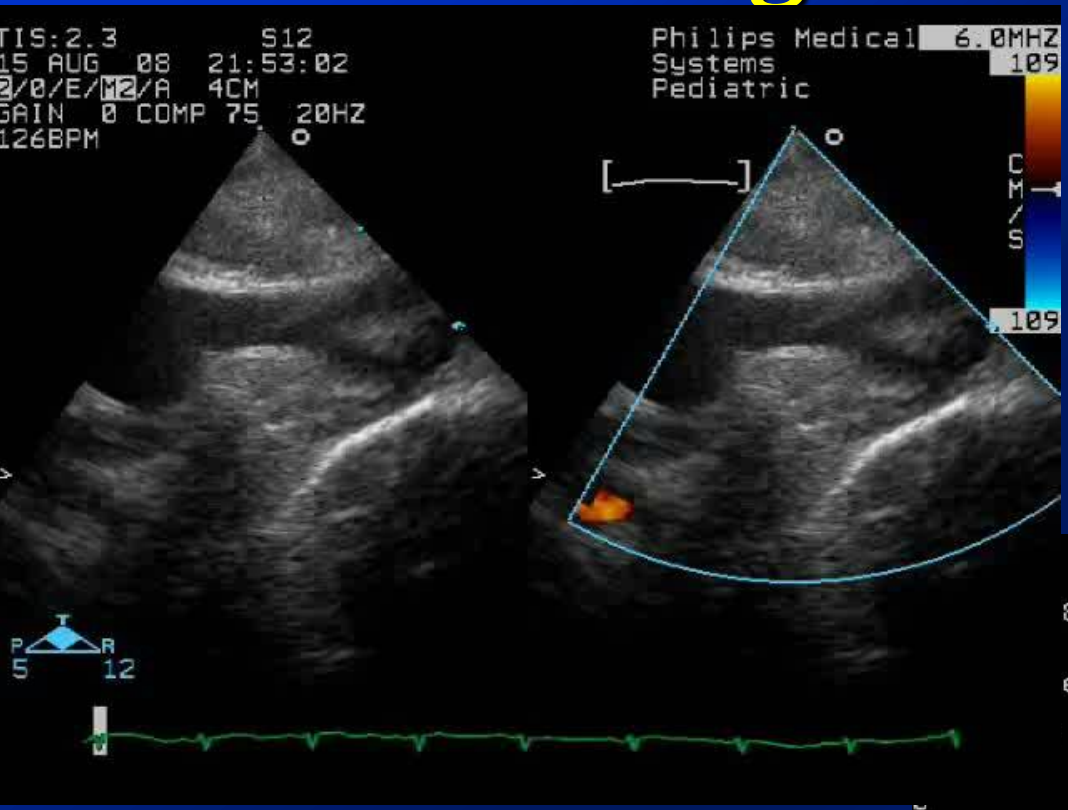


"If the echocardiographic diagnosis is felt to be equivocal, the surgeon is alerted and is prompted to carefully examine the proximal coronary arteries."

Need et al. JACC 2000

Arch sidedness

Right Arch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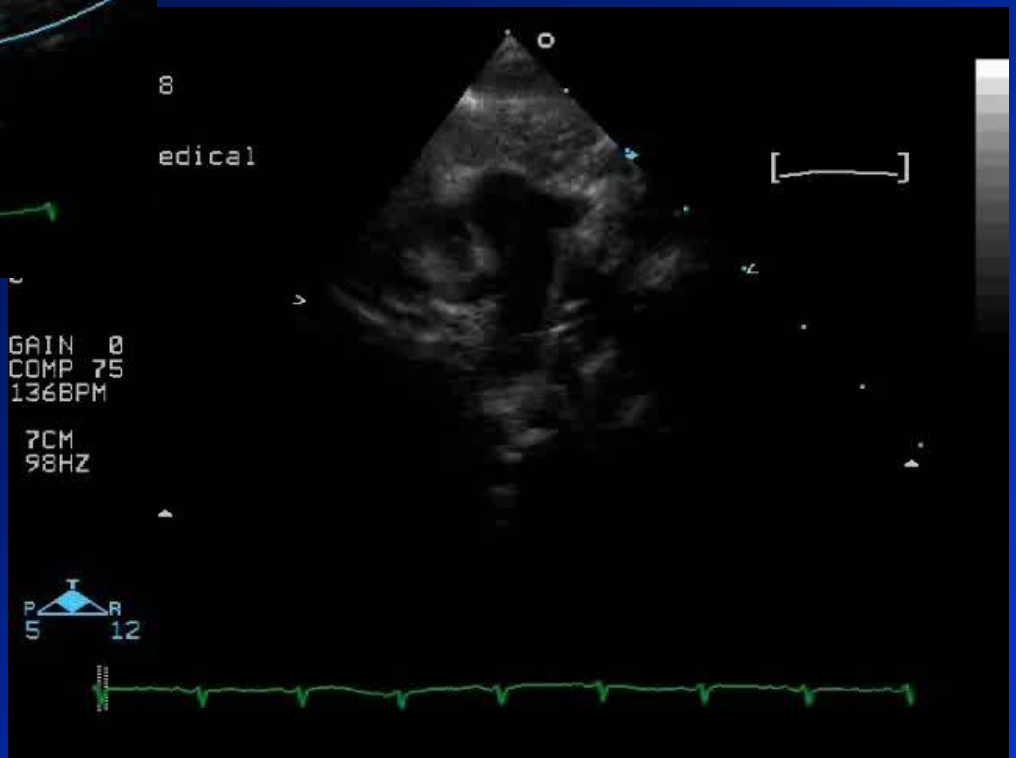
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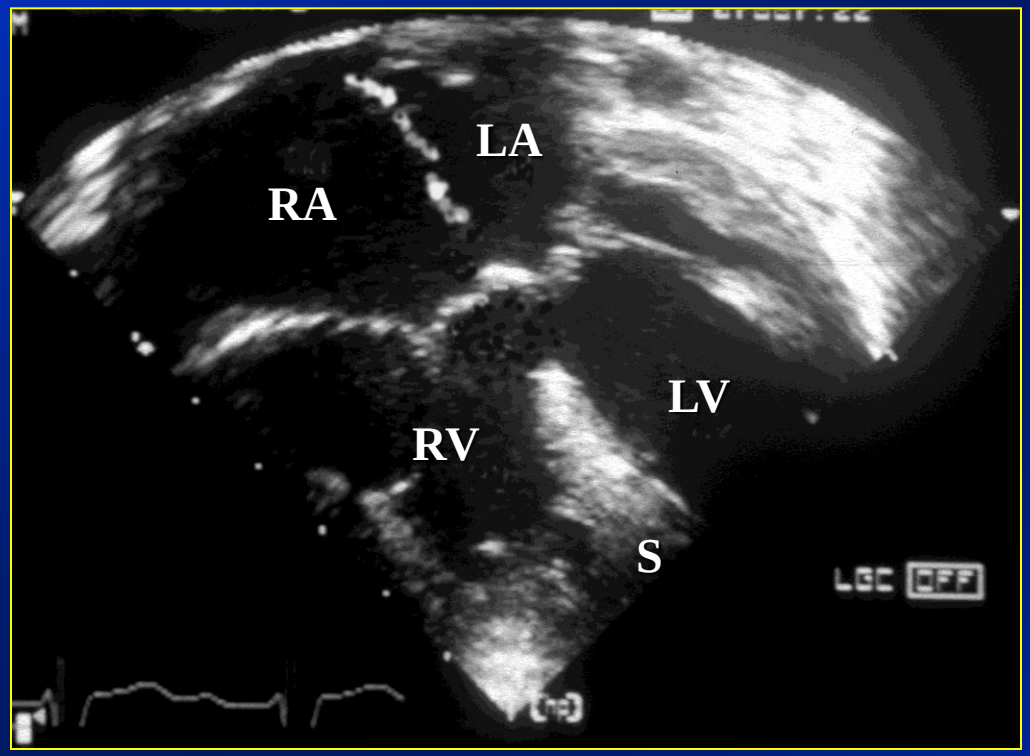
Associated Anomalies

- 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)

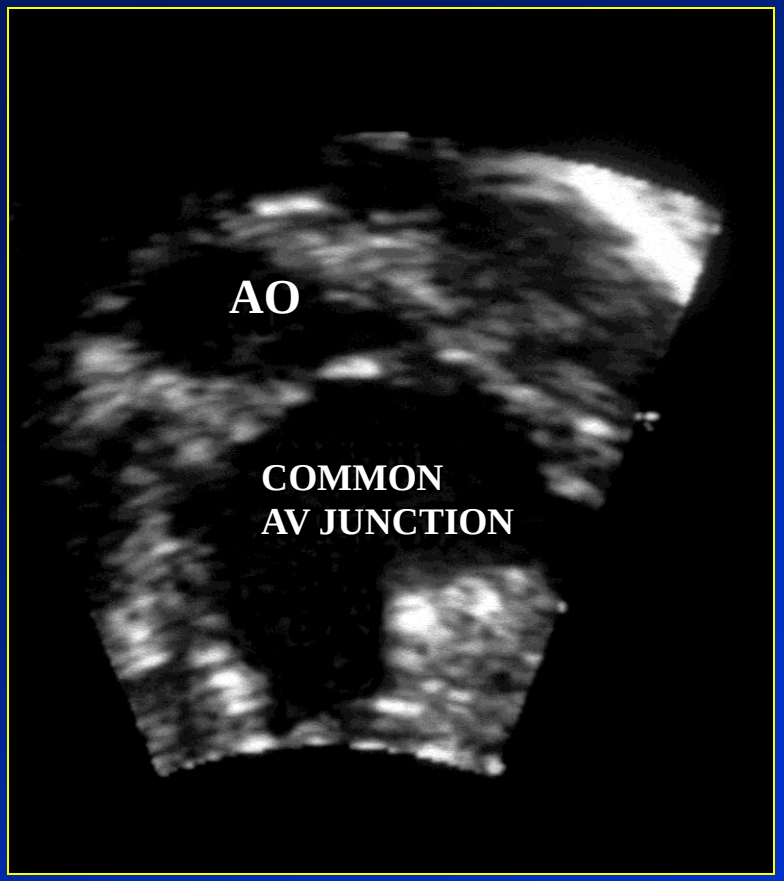
Tetralogy of Fallot

AV Septal Defect

Parasternal 4 Chamber



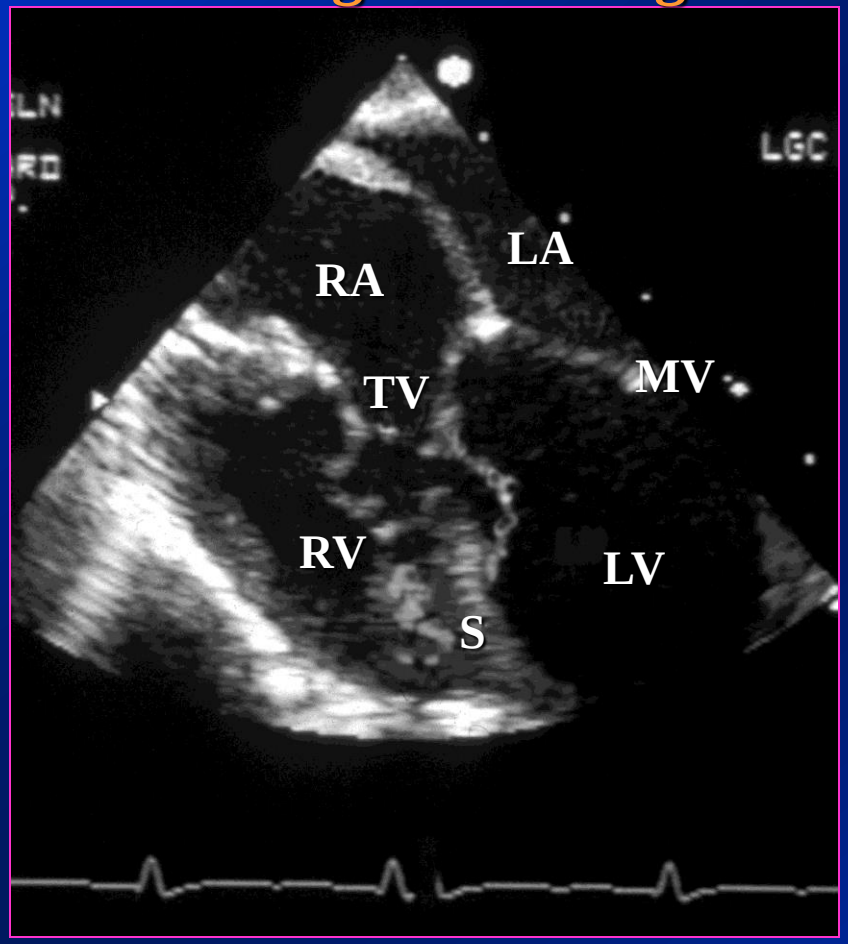
Subcostal Short Axis



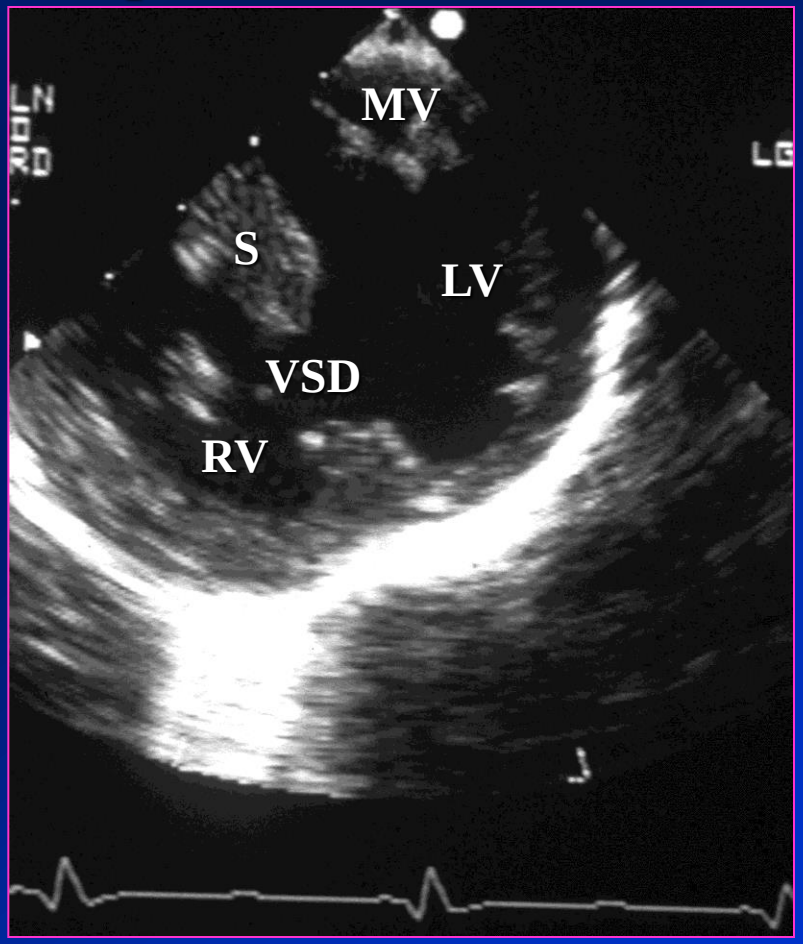
Tetralogy of Fallot

Associated Malformations

Straddling/Overriding TV

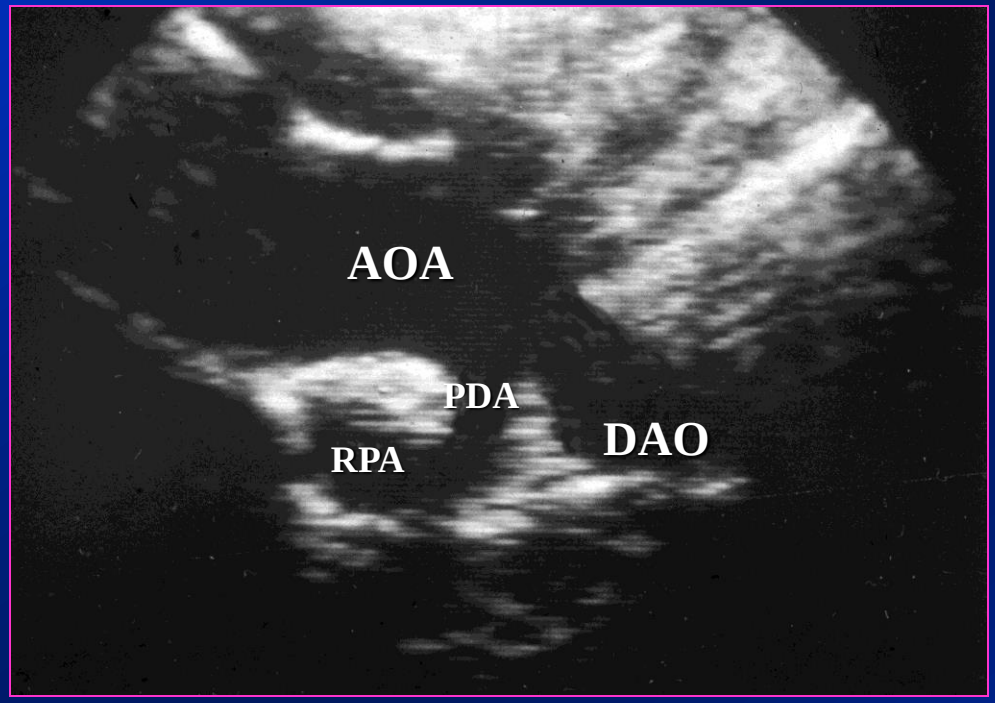


Apical Muscular VSD

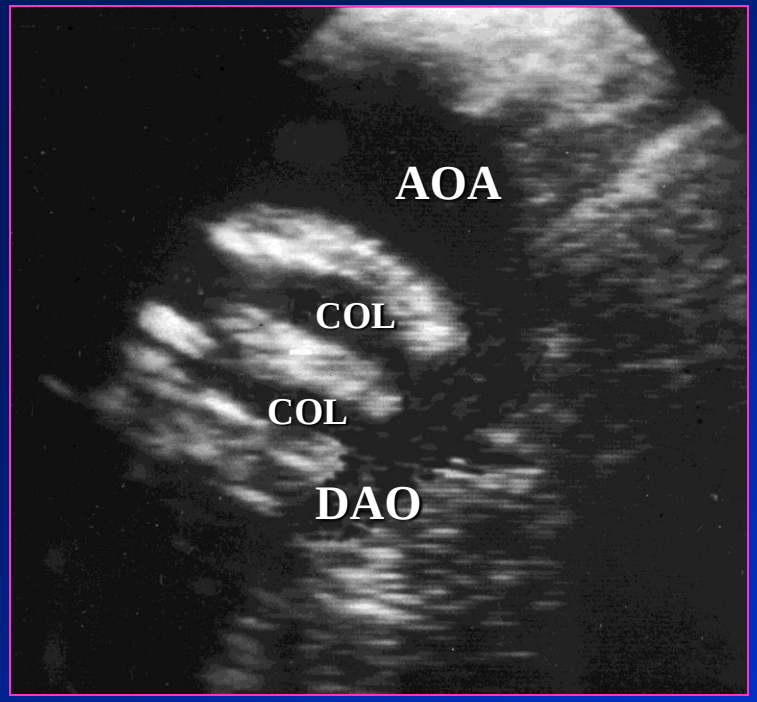


Additional sources of pulmonary flow

Patent Arterial Duct

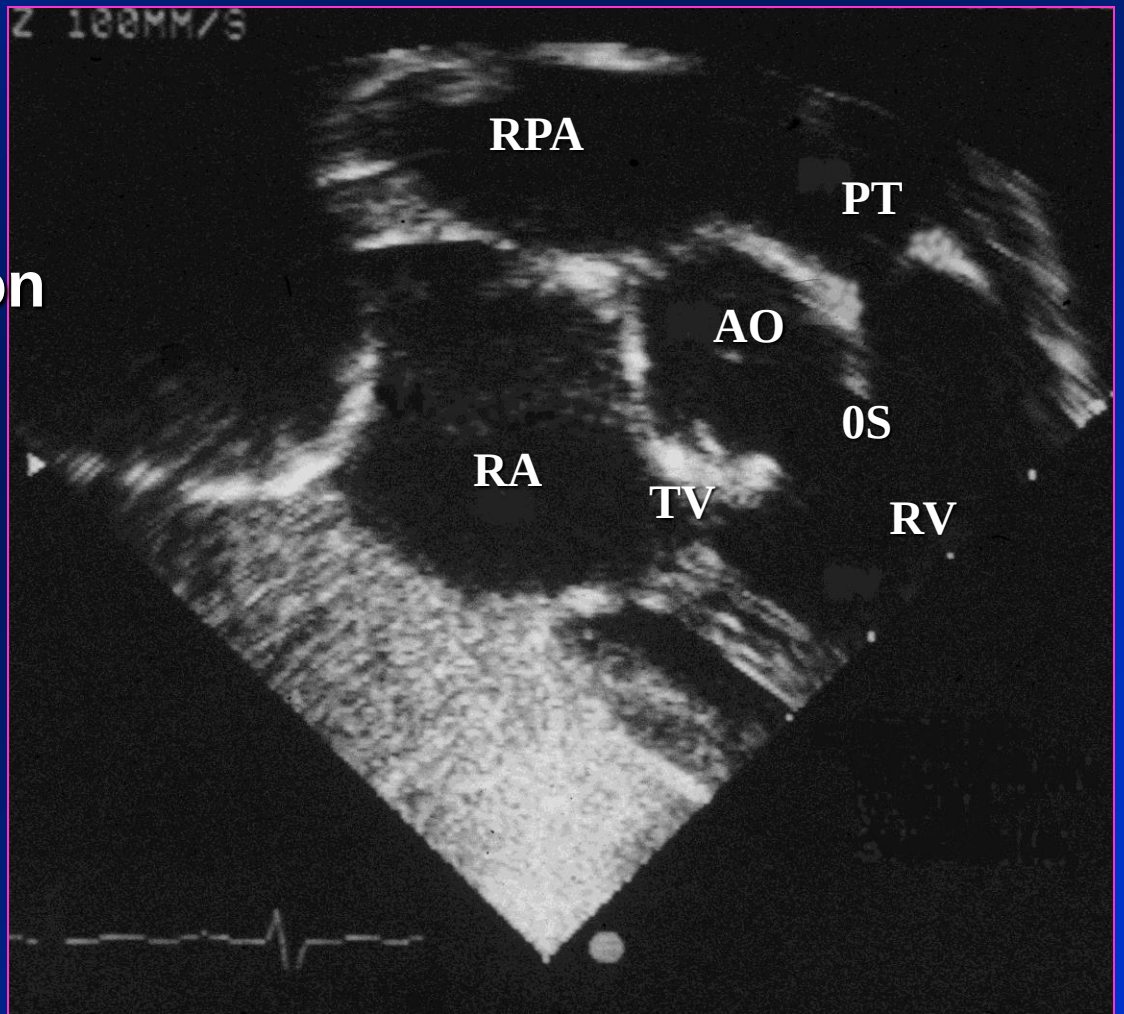


Aortopulmonary Collaterals

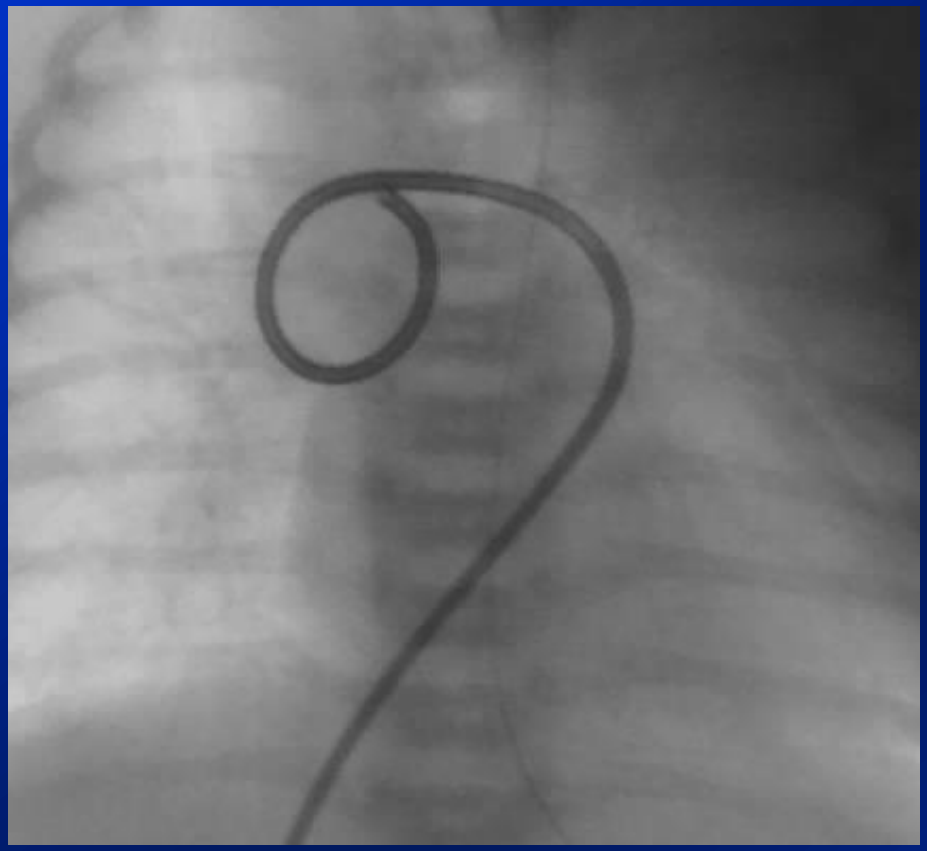


Absent Pulmonary Valve Syndrome

- Dilated Pulm Arteries
- Bronchial Compression
- Chest Infections
- Pulm Regurgitation
- Without Cyanosis



Absent Pulmonary Valve Syndrome



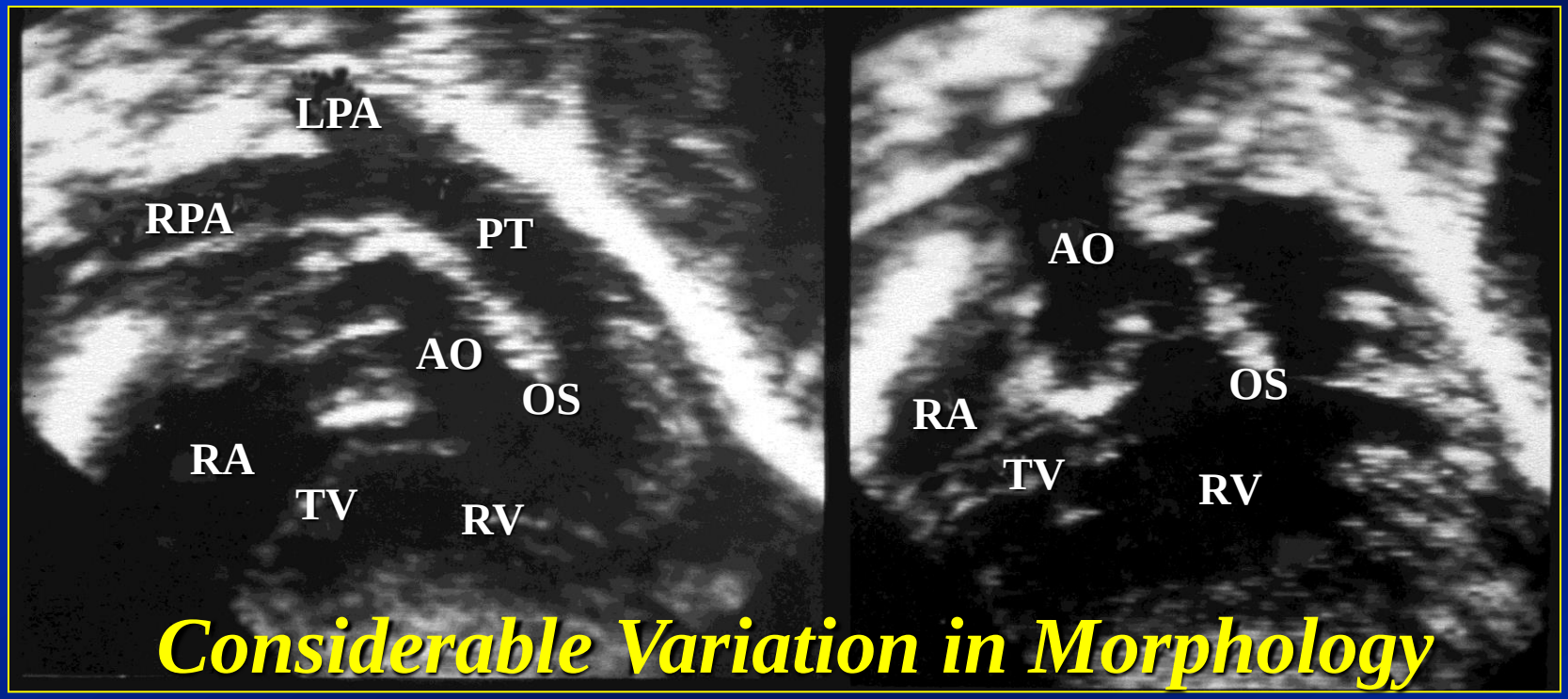
TETRALOGY OF FALLOT

- VARIATION IN MORPHOLOGY
- IMPORTANT to asses 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?

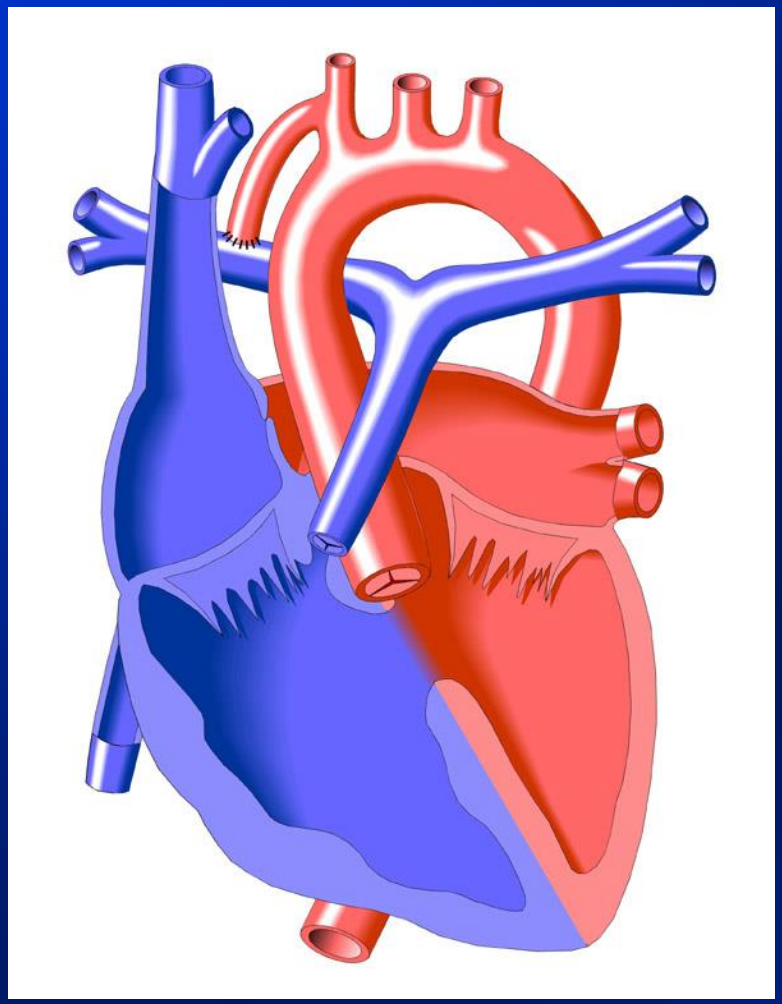
Tetralogy of Fallot
Pre-repair Diagnosis - Conclusions

- 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)