PATENT DUCTUS ARTERIOSUS (PDA)

It is a channel that connect the pulmonary artery with the descending aorta (isthmus part). It results from the persistence of patency of the fetal ductus arteriosus after birth. It is the most common lesion in infant of mothers with congenital rubella, PDA more common in females.

**CLINICAL FEATURES:**
Patients with small PDA usually are asymptomatic but large PDA will result in heart failure & pulmonary hypertension along with growth failure.

**O/E:** Collapsing pulse with side pulse pressure; on auscultation -- > classical continuous machinery murmur (systolic & diastolic) at the pulmonary area with a thrill when there is severe pulmonary hypertension, only systolic murmur can be heard with loud S2.

**DIAGNOSIS:**
ECG: In small PDA, it is normal, but large PDA, there is normal axis deviation, left ventricular hypertrophy (LVH) or biventricular hypertrophy.

**Chest X-ray:** Cardiomegaly, plethoric lung, and prominent pulmonary conus.

**RX:** Closure should be done soon after the Dx (at 9 months - 1 year) whatever the size due to possible complications. Most PDA closures can be done by transcatheter device and some need transthoracic surgery, usually done by occluder (safe and
Localized, discrete narrowing of the thoracic aorta occurs in 6-8% of congenital heart disease. More common in males with M: F = 2:1. More common cardiac lesion in Turner syndrome.

C/F: In infants, usually presented with congestive heart failure, dyspnoea, poor feeding.

MRI: Used in Dx due to difficulty in infant patients.

RX:
1. Balloon coarctation angioplasty → native or recurrent (previously).
2. Surgery by end to end anastomosis.

PROGNOSIS: The mean age for untreated coarctation is about 34 years and the most common cause of death are: heart failure, aortic rapture, infective endocarditis, intracranial hemorrhage (due to t B.P. or aneurysm).

PULMONARY STENOSIS

Anatomical classification:
1. Valvular 90%
2. subvalvular
3. Supravalvular

Classification according to severity:
1. Mild: when the pressure gradient < 50 mmHg
2. Moderate: when the pressure gradient 50-80 mmHg
3. Severe: when the pressure gradient > 80 mmHg

**C/F:**
Most patients are asymptomatic and discovered accidentally.

**Symptoms:** exertional dypnoea and sometimes chest pain.

**O/E:** The asymptomatic patient has normal growth unless it is a severe case.

**In symptomatic patients:**
1. S1 is normal.
2. Non-fixing splitting S2.
3. Multiple systolic ejection click at pulmonary areas.
4. Ejection systolic murmur with thrill (i.e. > grade 4), maximally at pulmonary area.

**INVESTIGATIONS:**

**CXR:**
1. Prominent pulmonary conus caused by post-stenotic pulmonary dilatation.
2. Normal pulmonary vascularity, i.e. there is no oligemia of the lung as expected after pulmonary stenosis because no intracardiac shunt (i.e. all the blood in the heart from the systemic circulation goes to the lung).

**ECG:** Rt. Axis deviation & peaked P-wave.

**Echo:** For Dx, severity determination, and to Dx the level of stenosis (supra, valvular or sub).

**Catheterization:** Diagnostic and therapeutic.

**RX:**
Mild → just reassurance.
Moderate to severe → balloon valvuloplasty; if failed → surgical valvotomy.

TETRALOGY OF FALLOT
(ToF)

It is the commonest cyanotic congenital heart disease in children and adults. It is a combination of:
1. VSD
2. Pulmonary stenosis (PS).
3. Overriding of aorta.
4. Right ventricular hypertrophy (RVH).

HEMODYNAMICS: The degree of right ventricular outflow obstruction (i.e. P.S.) determines the degree of pulmonary blood flow and the severity of cyanosis.

C/F:
1. Cyanosis: most patients develop cyanosis in the first 6 month - 1 year of life.
2. Dyspnoea: occurs in exertion, so they have a limited exercise tolerance.
3. Squatting: it is the characteristic posture of children with TOF after any physical effort.

Q/E:
1. Cyanosis (central and peripheral).
2. Clubbing (usually after 3 months of age).
3. Single S2 & ejection systolic murmur at the left sternal border.

**INVESTIGATION:**

**ECG:** Right axis deviation (RAD) & right ventricular hypertrophy  
**CXR:**
1. Normal heart size.
2. Rounded apex situated above the diaphragm (hypertrophied RV).
3. Concavity at the main pulmonary artery area.
4. Reduced pulmonary vascularity (oligemic lung).
   The classical radiological picture of TOF is called (boot shape).

**Echo:** It is essential for Dx.

**Catheterization:** Gives the definitive Dx and as a preoperative step.

**COMPLICATIONS:**
1. Hypercyanotic spells.
2. Cerebrovascular accidents (CVA).
3. Cerebral abscess.
4. Infective endocarditis.
5. Haematological complications include bleeding and thrombosis. All complications are essentially due to cyanosis & polycythemia.
   Polycythemia is due to hypoxia which results from right to left
shunt (as the right pressure grows higher than the left one).

**RX:**

1. **Medical:**
   a. Rx of anemia by iron, sometimes considered as relative anemia if we find normal Hb level; also nutritional support.
   b. Phlebotomy: in symptomatic patients with hematocrit more than 65% (as there is a risk for CVA).

2. **Surgical:**
   a. Palliative to ↑ pulmonary blood flow e.g.: Blalock-Taussig shunt (BT shunt).
   b. Total surgical repair.

**Indications for surgery:**

1. Cyanosis
2. Hypecyanotic spells
3. Polycythemia
4. ↓ exercise tolerance
5. Appropriate age and weight (usually between 2-3 years, if there is cyanosis).

**HYPER.CYANOTIC SPELLS:**

Attacks of increasing cyanosis associated with abnormal respiration and altered level of consciousness and it is an important cause of death in TOF patients (especially in mild cyanosis).
Spells are particular problems during the 1st 2 years of life and is more common in the morning and can be precipitated by any activity. Most spells are self-limited, but should be considered as an indication for surgery.

The cause is infundibular spasm.

**RX:**

2. O₂.
3. Sodium bicarbonate to correct metabolic acidosis.
4. Morphine: 0.2 mg/Kg subcutaneously (s.c.) or i.v. can be repeated 4 hourly.