

TETRALOGY OF FALLOT

AMALDEV

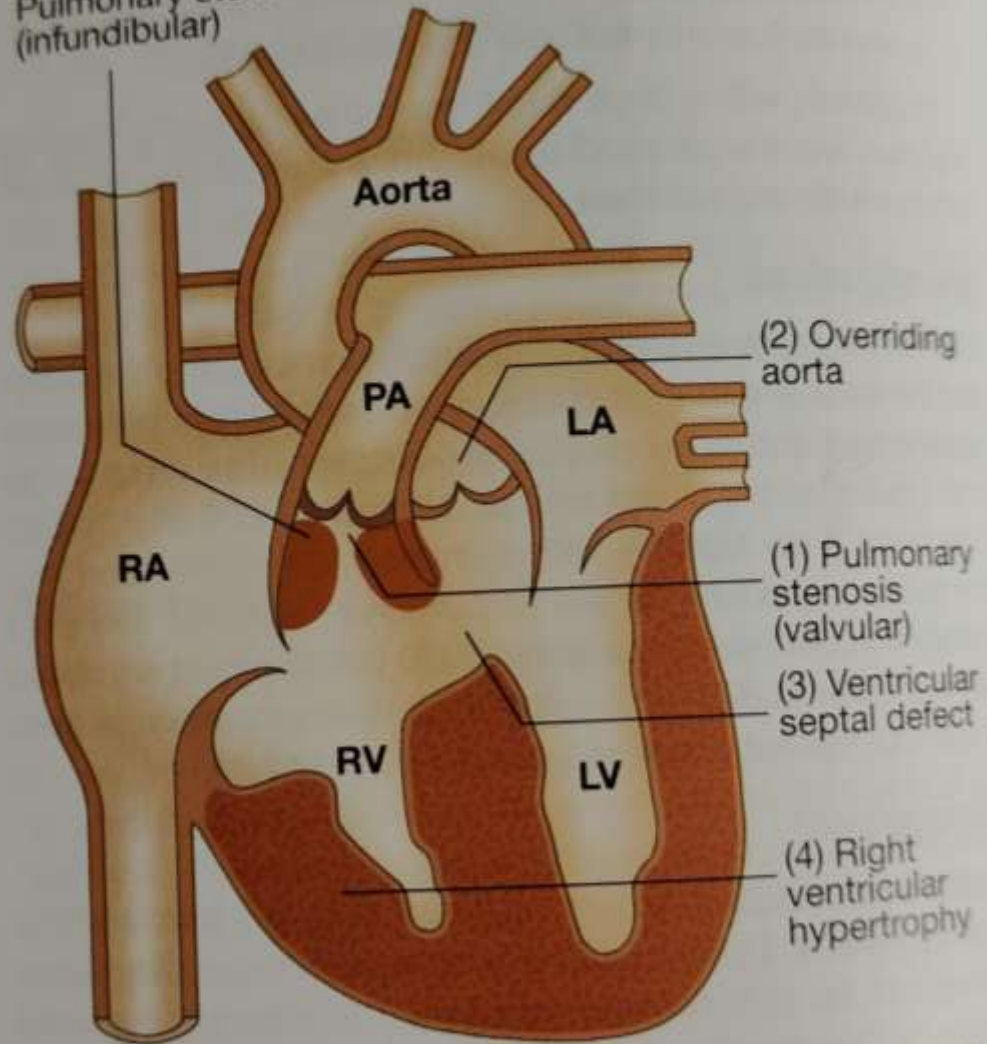
ROLL NO 18

DEFINITION

- ◇ Tetralogy of Fallot is a congenital malformation of the heart involving four cardinal defects:
- ◇ 1. Ventricular Septal Defect (VSD)
- ◇ 2. Pulmonary Stenosis
- ◇ 3. Overriding Aorta
- ◇ 4. Right Ventricular Hypertrophy

It occurs in about 1 in 2000 births

Pulmonary stenosis
(infundibular)



(2) Overriding
aorta

(1) Pulmonary
stenosis
(valvular)

(3) Ventricular
septal defect

(4) Right
ventricular
hypertrophy

PATHOGENESIS

Tetralogy of Fallot occurs as the result of abnormal development of the **bulbar septum** that separates the ascending aorta from the pulmonary artery, and which normally aligns and fuses with the outflow part of the interventricular septum.

- **The right ventricular outflow obstruction is most often subvalvular (infundibular) but may be valvular, supra-valvular or a combination of these**
- **The subvalvular component of the right ventricular outflow obstruction is dynamic and may increase suddenly under adrenergic stimulation.**
- **The ventricular septal defect is usually large and similar in aperture to the aortic orifice.**
- **The combination results in elevated right ventricular pressure and right-to-left shunting of cyanotic blood across the ventricular septal defect into the aorta.**

CLINICAL FEATURES

- ◇ Children are usually cyanosed
- ◇ But this may not be the case in the neonate because it is only when right ventricular pressure rises to equal or exceed left ventricular pressure that a large right-to-left shunt develops

◇ **Fallot's spells**

- ◇ **The affected child may suddenly become increasingly cyanosed, often after feeding or a crying attack, and may become apnoeic and unconscious.**
- ◇ **In older children, Fallot's spells are uncommon but cyanosis becomes increasingly apparent, with stunting of growth, digital clubbing and polycythaemia.**

◇ **Fallot's sign**

- ◇ **Some children characteristically obtain relief by squatting after exertion, which increases the afterload of the left heart and reduces the right-to-left shunting.**
- ◇ **The natural history before the development of surgical correction was variable but most patients died in infancy or childhood.**

On examination

- ◆ The most characteristic feature is the combination of cyanosis with a loud ejection systolic murmur in the pulmonary area (as for pulmonary stenosis).
- ◆ Cyanosis may be absent in the newborn or in patients with only mild right ventricular outflow obstruction, however. This is called acyanotic tetralogy of Fallot.

INVESTIGATIONS

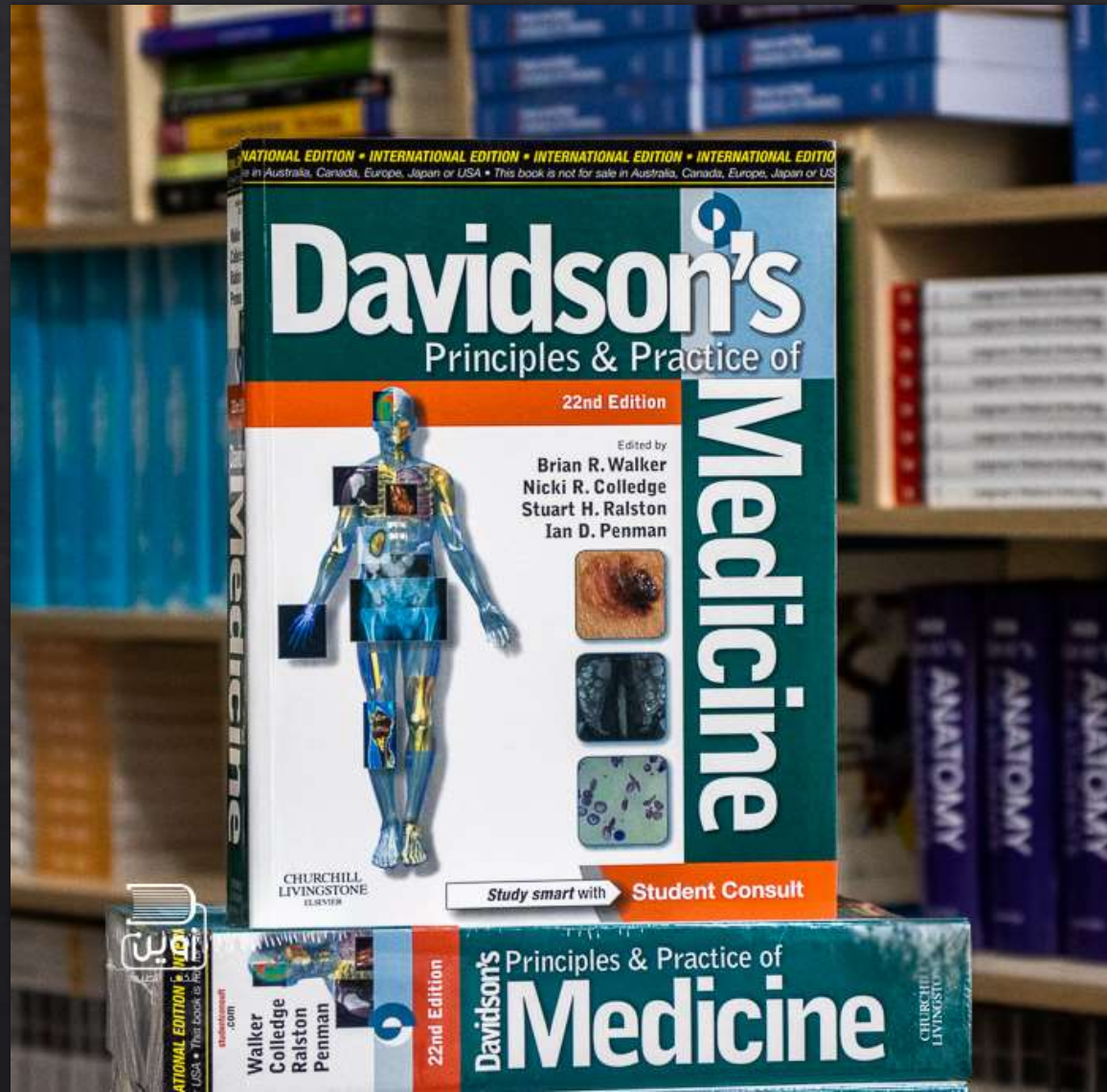
- ◆ Echocardiography is diagnostic and demonstrates that the aorta is not continuous with the anterior ventricular septum.
- ◆ The ECG shows right ventricular hypertrophy and the chest X-ray shows an abnormally small pulmonary artery and a 'boot-shaped' heart.

MANAGEMENT

- ◆ The definitive management is total correction of the defect by surgical relief of the pulmonary stenosis and closure of the ventricular septal defect.
- ◆ Primary surgical correction may be undertaken prior to the age of 5 years.
- ◆ If the pulmonary arteries are too hypoplastic, then palliation in the form of a **Blalock-Taussig shunt** may be performed, with an anastomosis created between the pulmonary artery and subclavian artery.
- ◆ This improves pulmonary blood flow and pulmonary artery development, and may facilitate later definitive correction.

- ◆ **The prognosis after total correction is good, especially if the operation is performed in childhood.**
- ◆ **Follow-up is needed to identify residual shunting, recurrent pulmonary stenosis and arrhythmias.**
- ◆ **An implantable defibrillator is sometimes recommended in adulthood**

REFERENCE



THANKS FOR YOUR

PATIENCE