

Congenital heart disease

- It is the abnormality of the heart present right from the birth.
- 0.8% of live births and higher in premature babies.

Classification

- 1) Malposition (Dextrocardia)
 - Apex points towards the right side.
- 2) Shunts (Cyanotic Congenital heart disease).
 - a) Right to left shunts (Cyanotic group).
 - i) Tetralogy of Fallot.
 - b) Left to Right shunts (Acyanotic or late cyanotic)
 - 1) Ventricular septal defects.
 - 2) Atrial septal defects.
 - 3) Patent ductus arteriosus.
- 3) Obstructive Congenital heart disease
 - 1) Aortic stenosis.
 - 2) Pulmonary stenosis.
 - 3) Coarctation of aorta.

Causes:

- 1) Maternal infection or drugs.
- 2) Maternal rubella.
- 3) Maternal alcohol misuse.
- 4) Chromosomal → Down syndrome
- 5) Genetic → Di George syndrome, Marfan syndrome.

C/F: Central cyanosis, Clubbing, Syncope.

Finnegan syndrome, Pulmonary BP, Growth

Tetralogy of Fallot - (Cyanotic).

- ① Right ^{Ventricular} outflow obstruction (Pulmonary valve or infundibulum stenosis)
- ② Right Ventricular hypertrophy (Boat shaped heart)
- ③ Ventricular Septal Defect
- ④ Overriding of the aorta.

Pathophysiology

Right Ventricular outflow obstruction
(Valvular or subvalvular)

↓
Right Ventricle hypertrophy

↳ Response to adrenergic receptor

↓
VSD → overriding aorta → Deoxygenated Blood in circulation

↓
Central Cyanosis

Clinical Features:-

- Most common congenital defect Cyanotic - 50-70%.
- 10% of congenital heart disease associated with Di George syndrome.
- Cyanosis in lips & Fingertips, Clubbing in fingers & toes.

→ ↓ O₂ causes → low weight, difficult feeding, growth.

Cyathotic spell "tet spell"

During crying or feeding. Normal ← Shunt reversal

↓
Increased O₂ demand

↓
Heart pumps more blood (Deoxygenated)

↓
Cyanosis

↑
↑ Left Pressure

↑
Increased systemic pressure

↓
Squats down → Kinks femoral → ↑ Vascular resistance

↳ Fallot's artery

Murmur $\rightarrow$ Ejection systolic murmur. (PS)

- Cyanosis is absent if mild right ventricular outflow obstruction. called as acyanotic Tetralogy of Fallot.

Investigations:

Echocardiography $\rightarrow$ Diagnostic.

ECG $\rightarrow$ Right ventricular hypertrophy

Chest Xray $\rightarrow$ Boot shaped heart

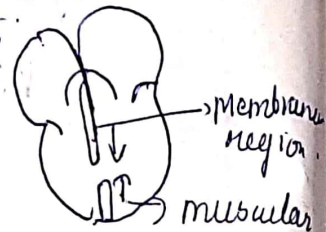
Management $\rightarrow$ Surgical.

Acyanotic (Ventricular Septal defect)

- Most common congenital defect.

But closes spontaneously in 30-50% of people.

- Associated with Down syndrome and Fetal alcohol syndrome.



Pathogenesis

Membranous part $\rightarrow$ Muscular part fuses to form the septum.

Defect in "perimembranous part"

High Pressure in LV

$\downarrow$
RV $\rightarrow$ Pulmonary hypertension

$\downarrow$
After sometime T Pressure in RV

$\downarrow$
Reversed shunt (Eisenmenger syndrome)

$\downarrow$
RV $\rightarrow$ LV $\rightarrow$ Deoxygenated blood

$\downarrow$
Cyanosis

Clinical Features:

- Cyanosis & clubbing.
- Polycytotic Murmur? however, left sternal Bond.
- Cardiac failure may occur if there is large septal defect.