CARDIOVASCULAR DISEASES

Jonathan

1. Rheumatic Heart Disease

- Also called **Rheumatic Endocarditis**
- Is an acute, recurrent inflammatory disease that causes damage to the heart particularly the valves as a sequelae to group A beta-hemolytic streptococcal infection resulting in valve leakage (insufficiency) and/or obstruction (narrowing or stenosis).
- There are associated compensatory changes in the size of the heart's chambers and the thickness of chamber walls in response to the disease

Pathophysiology

- Rheumatic fever is a sequelae to group A streptococcal pharyngitis
- Heart damage and the joint lesions of rheumatic endocarditis are not infectious in the sense that these tissues are not invaded and directly damaged by destructive organisms rather, they represent a sensitivity phenomenon or reaction occurring in response to hemolytic streptococci.

- Leukocytes accumulate in the affected tissues and form nodules, which eventually are replaced by scar tissue.
- Heart valves are affected, resulting in valve leakage (regurgitation) and narrowing (stenosis)
- Compensatory changes lead to changes in chamber sizes and wall thickness
- Heart involvement (carditis) also includes pericarditis, myocarditis, and endocarditis
- Rheumatic endocarditis is much more severe and symptomatic than pericarditis and myocarditis

Clinical Manifestations

- Symptoms of streptococcal pharyngitis may precede rheumatic symptoms
  - Sudden onset of sore throat; throat reddened with exudate
  - Swollen, tender lymph nodes at angle of jaw
  - Headache and fever(38.9°C - 40° C)
  - Abdominal pain (children)
  - Some cases of streptococcal throat infection are relatively asymptomatic
- Warm and swollen joints (polyarthritis)
- Chorea (irregular, jerky, involuntary, unpredictable muscular movements)

Clinical manifestations-Cont'd

- Erythema marginatum (transient mesh-like macular rash on trunk and extremities in about 10% of patients)
- Subcutaneous nodules (hard, painless nodules over extensor surfaces of extremities; rare)
- Fever
- Prolonged PR interval demonstrated by ECG
- Heart murmurs; pleural and pericardial friction rubs

Diagnosis

- Throat culture - to determine presence of streptococcal organisms
- ESR, WBC count and differential, and CRP increased during acute phase of infection
- Elevated antistreptolysin-O (ASO) titer
- ECG-prolonged PR interval or heart block

Management

- **Rest-**to maintain optimal cardiac function
- **Salicylates or NSAIDs -** to control fever and pain
- **Corticosteroids -** administered to prevent cardiac damage and to treat arthritis and other inflammatory symptoms
- **Antimicrobial therapy -** penicillin is the drug of choice if streptococcal infection is still present
- **Prevention** of recurrent episodes through:
  - Long-term penicillin therapy for 5 years after initial attack in most adults
  - Periodic prophylaxis throughout life if valvular damage is present

Complications

- Valvular heart disease
- Cardiomyopathy
- Heart failure

2. Congestive Cardiac Failure

- Is a clinical condition characterized by laboured breathing because the heart fails to meet circulatory and metabolic oxygen demand of the body
- Almost all predisposed children develop this problem by the end of 6 months if treatment is not initiated

Causes

- **Volume overload** - from left to right shunts where the right ventricle hypertrophies in order to compensate the increased blood volume e.g. in congenital heart defects
- **Pressure overload** - due to obstructive lesions e.g. valvular stenosis, coarctation of the aorta
- **Reduced myocardial contractility** by severe anemia or asphyxia
- **High cardiac output demands** e.g in sepsis, hormonal problems such as hypothyrodism
- **Cor pulmonale** – which is caused by chronic obstructive lung diseases e.g. cystic fibrosis,

Clinical Manifestations

- **Left-Sided Heart Failure**
  - Congestion occurs mainly in the lungs from blood backing up into pulmonary veins and capillaries.
    - Shortness of breath, dyspnea on exertion, paroxysmal nocturnal dyspnea (due to reabsorption of dependent edema that has developed during day), pulmonary edema (manifested by rales, rhonchi, wheezes, and orthopnea)
    - Cough may be dry, unproductive; usually occurs at night
    - Signs and symptoms of pulmonary edema, e.g.
  - Fatigability from low Cardiac Output, nocturia, insomnia, dyspnea, catabolic effect of chronic failure.
  - Insomnia, restlessness.
  - Tachycardia, third heart sound
  - Finger clubbing

- **Right-Sided Heart Failure**
  - Signs and symptoms are due to elevated pressures and congestion in systemic veins and capillaries:
  - Edema of ankles →unexplained weight gain (pitting edema)
  - Liver congestion→hepatomegally - may produce upper abdominal pain
  - Distended jugular veins
  - Abnormal fluid in body cavities (pleural space, abdominal cavity)
  - Anorexia and nausea - from hepatic and visceral engorgement
  - Nocturia - diuresis occurs at night with rest and improved CO
  - Weakness

Signs occurring in both Right and Left-sided failure

- Cardiomegaly (enlargement of the heart) - detected by physical examination and chest X-ray
- Rapid heart rate
- Development of pulsus alternans (alternation in strength of beat)

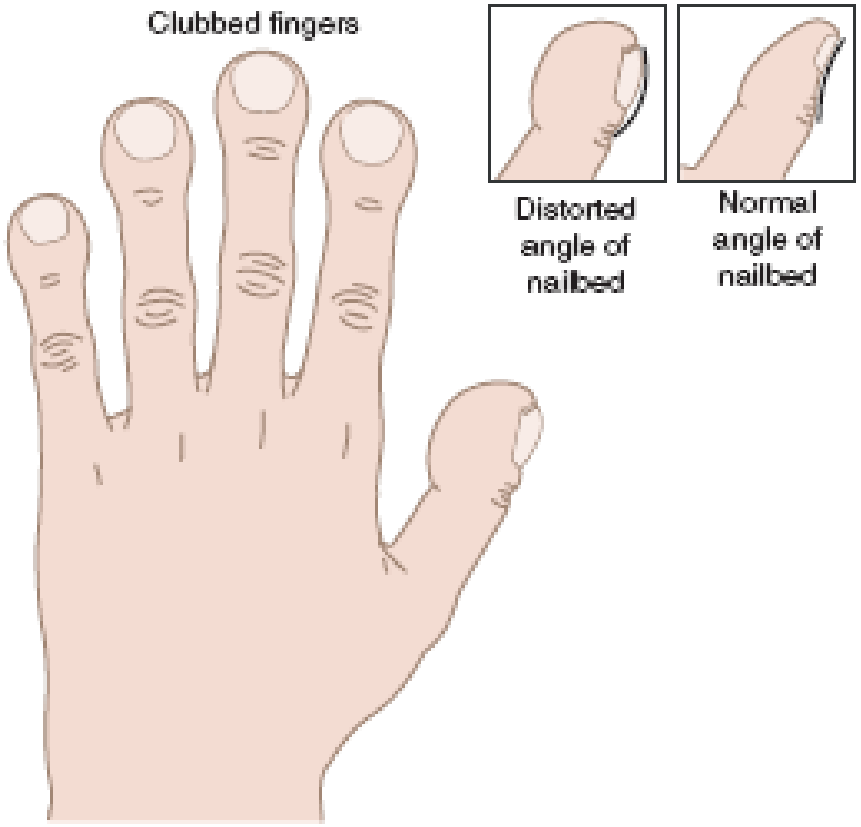


Fig 17.2 Clubbed fingers.

Diagnostic investigations

- Echocardiography
- Electrocardiography
- Chest X-ray - may show cardiomegaly, pleural effusion, and vascular congestion
- Arterial Blood Gases(ABG) studies
- Liver function tests- altered because of hepatic congestion

Management

- The goal is to improve cardiac function
  - Medications used
    - Cardiac glycosides – digoxin
    - Angiotensin converting enzyme (ACE) inhibitors – Captopril
    - Diuretics e.g. furosemide (lasix)
  - General care
    - Restrict and salt intake
    - Decrease cardiac demands-minimal activity, maintaining normal body temperature
    - Reduce breathing effort by keeping patient in semi-prone position or high Fowler's position by proping up the patient's bed
    - Avoid environmental stimuli
    - Improve tissue perfusion by providing supplemental oxygen
    - Daily weighing
    - Nutritional support
    - Alleviating anxiety to both patient and family
    - Advice on importance of keeping appointment schedules, medication adherence

Congenital Heart Problems

- The heart develops very early in the fetal period.
- Initially, it is a single tube through which single cells pass.
- Rapidly, two sets of chambers form—atria and ventricles.
- By the end of the 8th week of fetal development, the atrial and ventricular septa have formed and the pulmonary artery, aorta, and vena cava are all in place.
- Unfortunately, many different cardiac defects can develop within the first 8 weeks of pregnancy because many women remain unaware that they are pregnant through this period and may take medications, drink alcohol, or even develop an infectious disease, all of which can cause defects.
- The most common heart defect seen in neonates is the **ventricular septal defect**.

Fetal Circulatory system

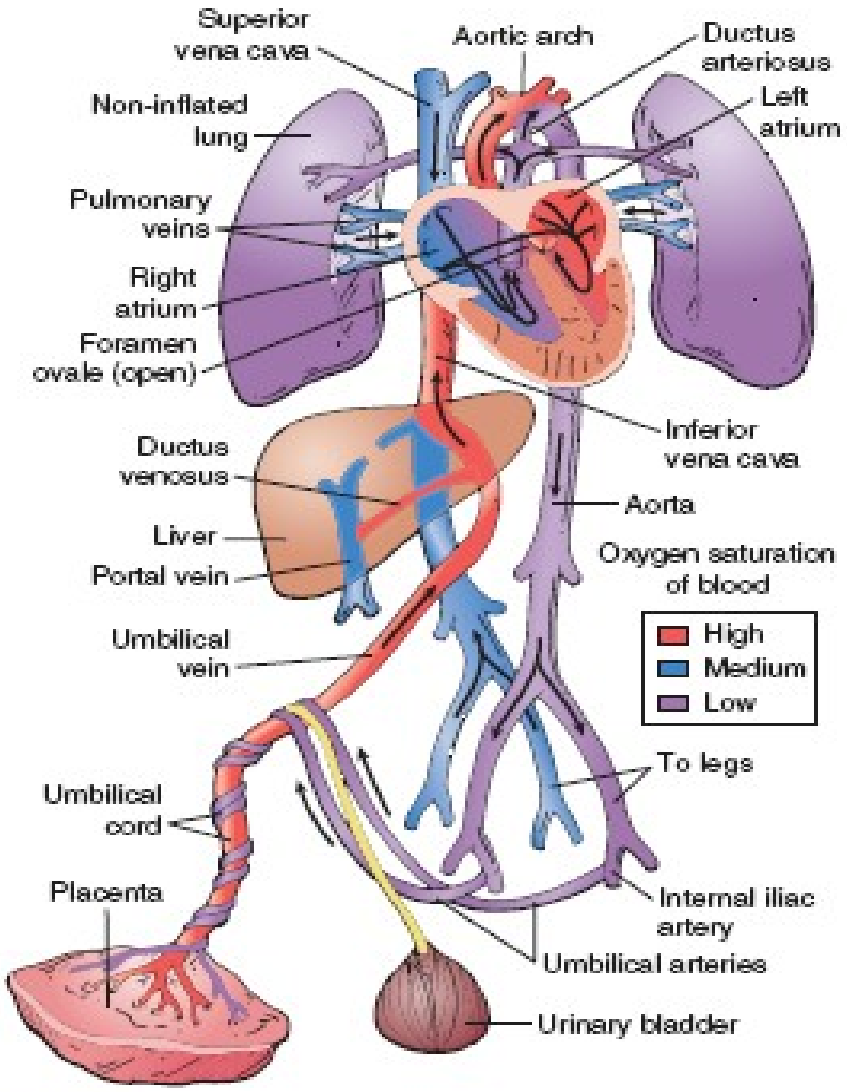


Fig 17.1 The fetal circulatory system.

- Cardiac defects are seen in 4 to 10 out of every 1,000 live births
- **Etiology.**
  - The cause of majority of defects is unknown but may include:
    - Prenatal rubella infection
    - Maternal alcohol consumption
    - Advanced maternal age
    - Maternal diabetes
    - Genetic diseases e.g. Down's syndrome, Klinefelter's syndrome, and Turner's syndrome

Diagnosis of CHDs

- **Echocardiogram** - ultrasound of the heart that is performed to assess the structures of the heart and the blood flow through the heart
- **Cardiac catheterization -** An invasive diagnostic procedure during which a radiopaque catheter is threaded through a peripheral vessel to the heart

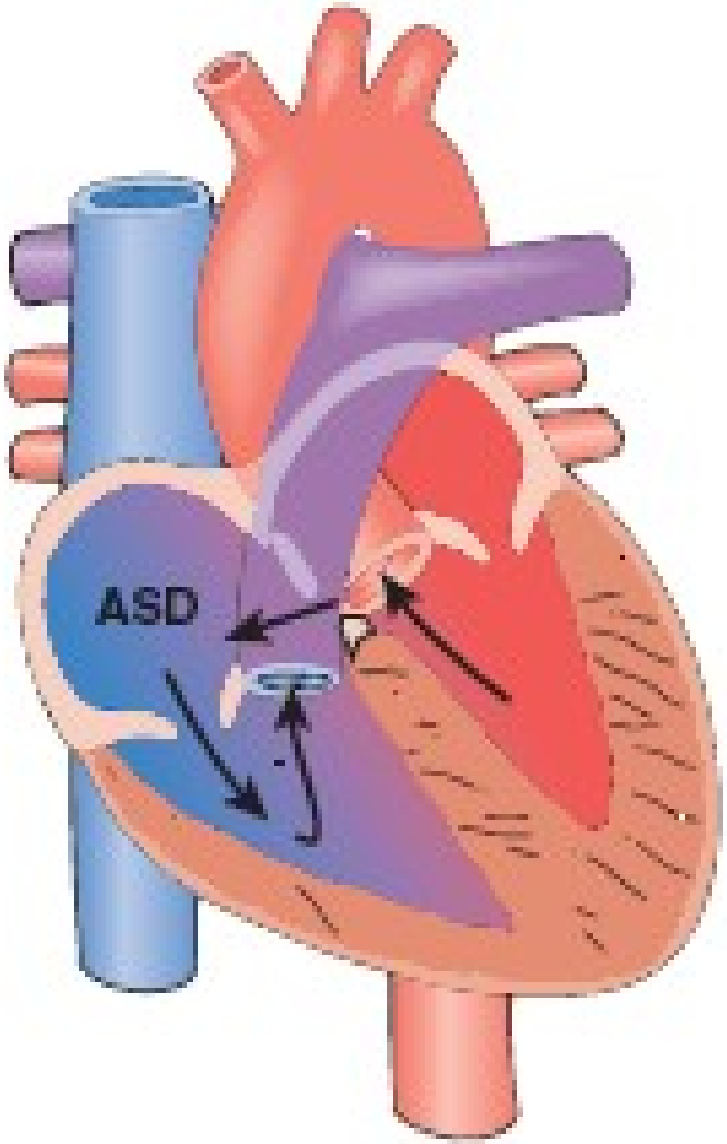
Types of Congenital Heart Defects

1. Acyanotic Defects

– Characterized by defects that result in blood being shunted from the left to the right side of the heart. As a result, the blood enters and re-enters the pulmonary system

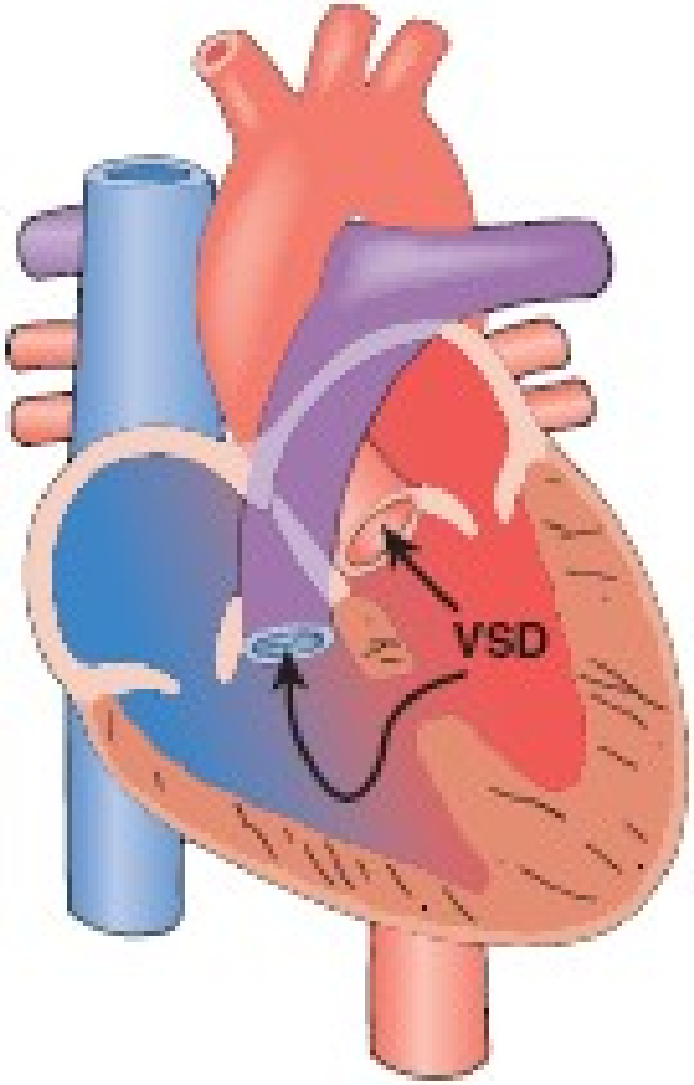
a. Atrial septal defect (ASD)

- Hole between the atria
- May be a foramen ovale that has not closed or a defect unrelated to the fetal duct
- **S & S -** Most have no symptoms, but the child may develop CHF, if the ASD is large. A murmur may be present
- **Mx -** Many ASDs close spontaneously. If not, surgery or interventional cardiology may be performed


ASD

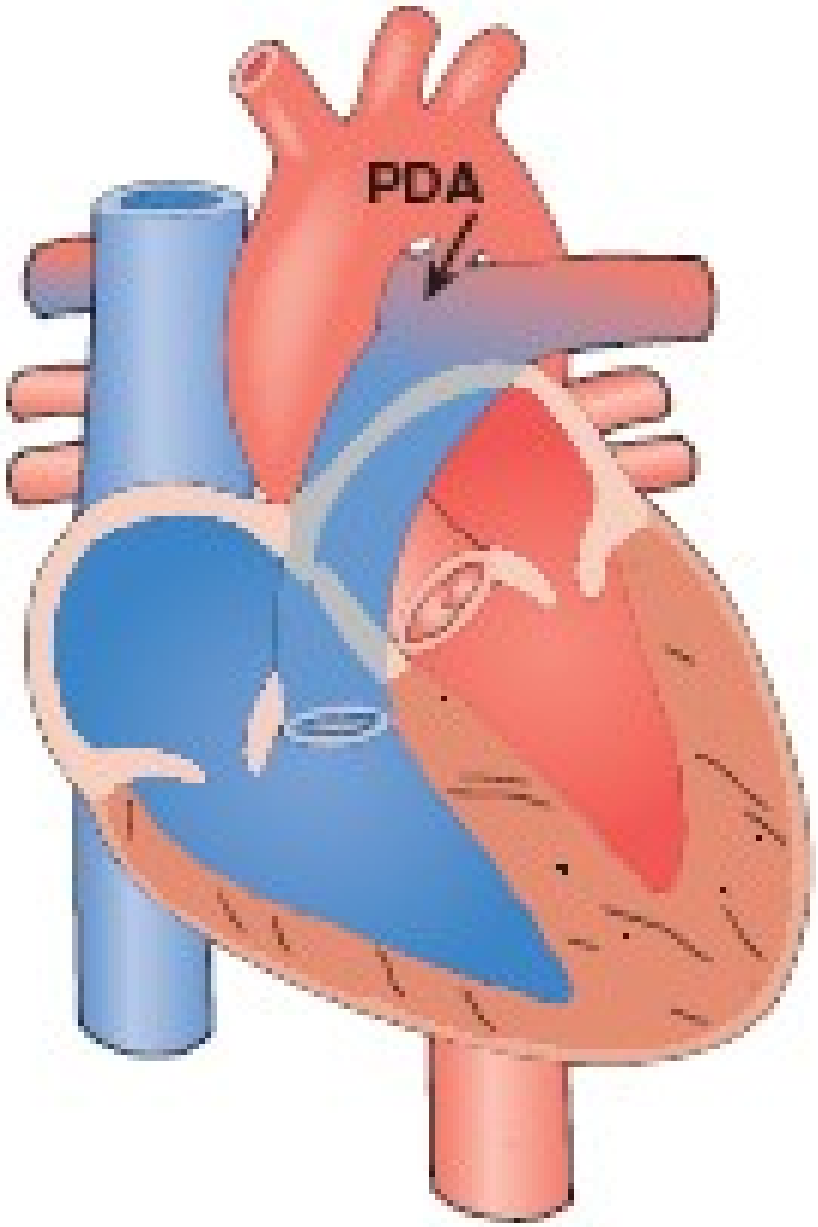
b. Ventricular septal defect (VSD)

- A hole between the ventricles; the most common cardiac defect.
- **S & S -** Most have no symptoms, but the child may develop CHF if the VSD is large. A murmur may be present.
- **Mx -** Small VSDs close spontaneously. If not, surgical repair will be needed


VSD

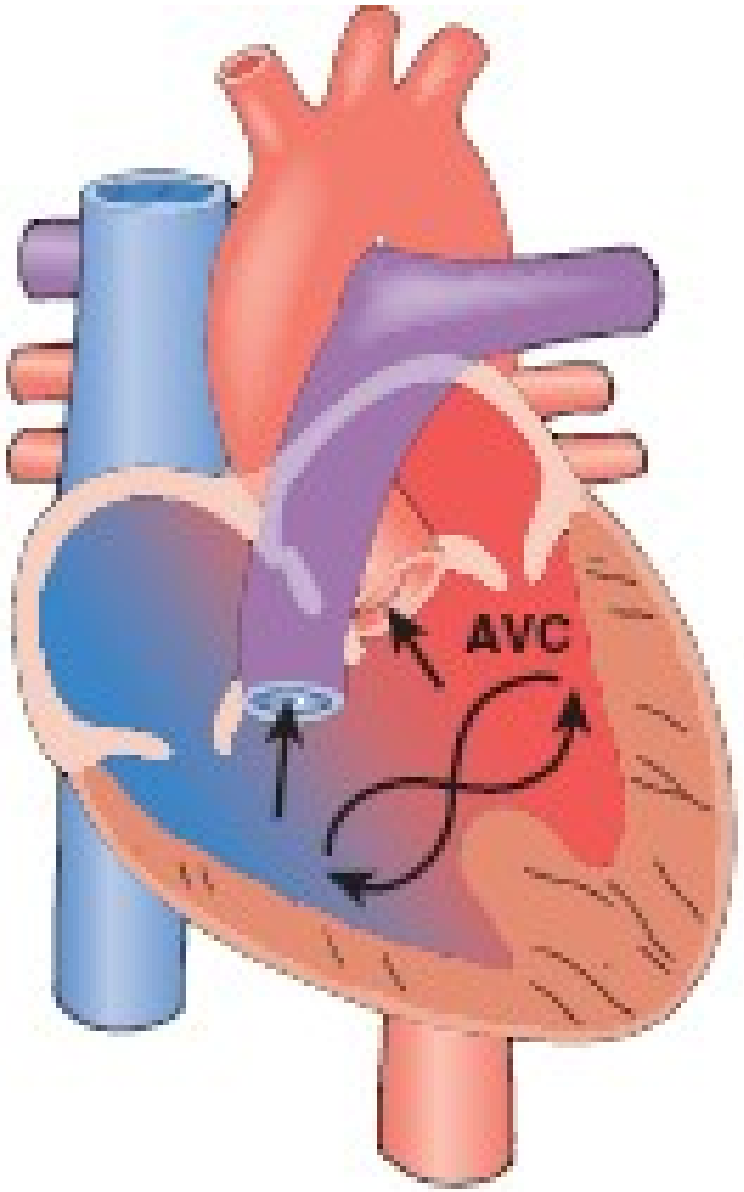
c. Patent ductus arteriosus (PDA)

- Most commonly seen in premature infants, especially when they weigh less ≤ 1,500 g at birth.
- The fetal duct between the pulmonary artery and the aorta fails to close.
- **S & S -** May have no symptoms, but a murmur may be heard, and the child may develop CHF.
- **Mx -** The defect may close spontaneously. If not, it may be closed medically with the administration of indomethacin, a prostaglandin inhibitor. If the medication is unsuccessful, surgery may be needed.


PDA

d. Atrioventricular canal (AVC)

- A large hole in the middle of the heart.
- **S & S -** progressively worsening CHF.
- **Mx -** Surgical repair is required.


AVC

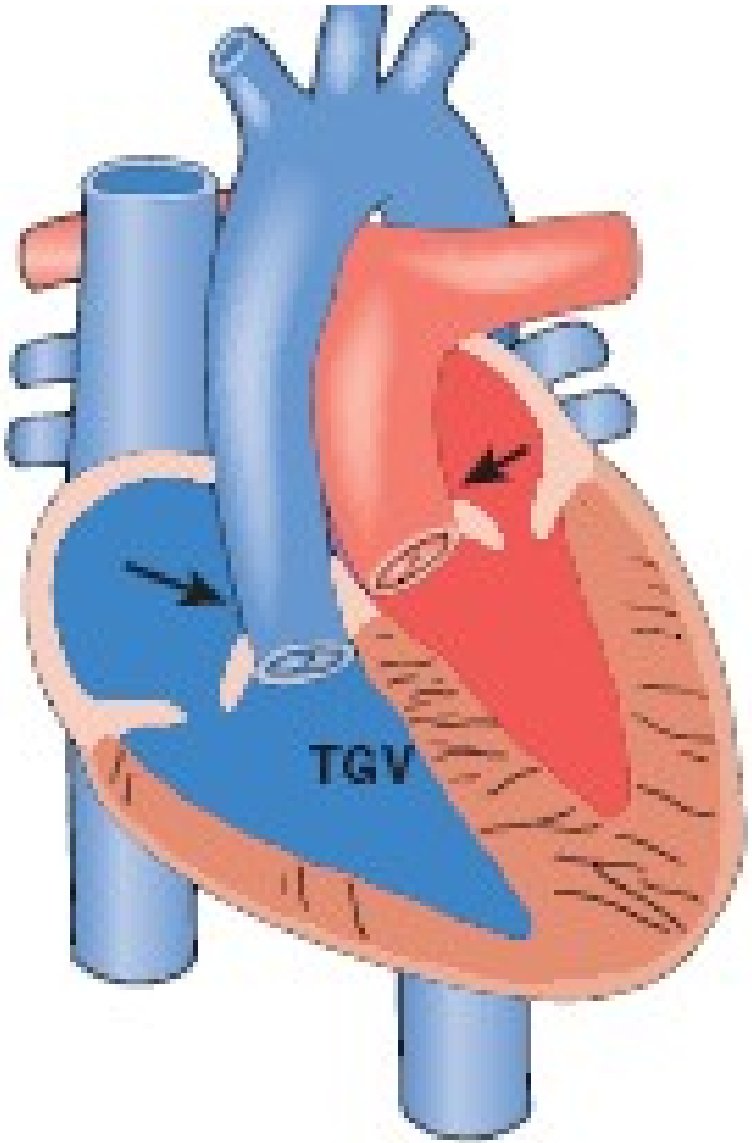
2. Cyanotic Defects

- Some result in the blood being shunted from the right to the left side of the heart.
- As a result, the blood bypasses the pulmonary system. In other cyanotic defects, deoxygenated blood never reaches the pulmonary system.

a. Transposition of the great vessels (TGV)

- The aorta exits off the right ventricle and the pulmonary artery off the left ventricle.
- This defect is incompatible with life unless another defect is present that allows the mixing of blood.

- **S & S -** Rapid and sustained cyanosis
- **Mx -** Surgery to create an intact vascular system

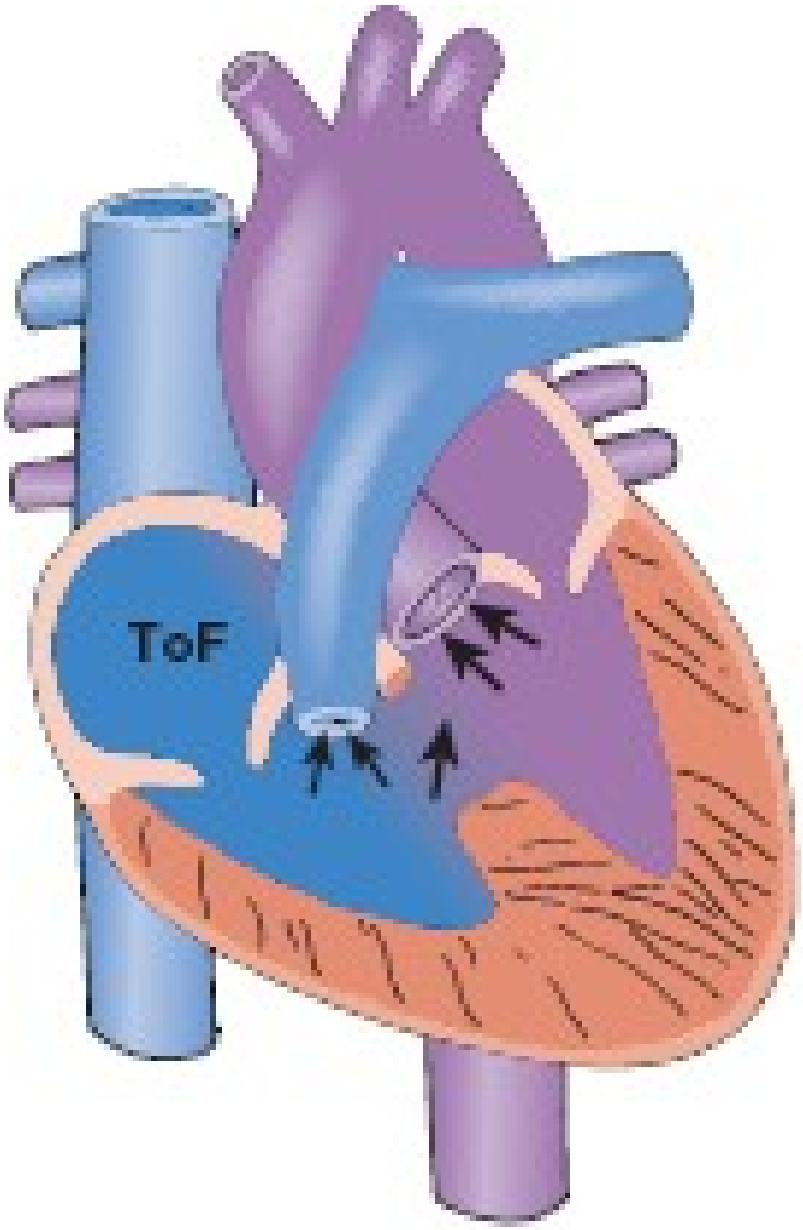

TGV

b. Tetralogy of Fallot (ToF)

- Is the most common cyanotic defect, consisting of four defects:
  - VSD
  - Overriding aorta
  - Pulmonary stenosis
  - Right ventricular hypertrophy.
- The right ventricular hypertrophy develops over time because the ventricle is working extra hard to circulate the blood.

- **S & S-** TET spells, in which the child becomes cyanotic, especially when crying and while eating (infancy) and during play (in older children)
- **Other S & S** – polycythemia(↑ RBC count) and clubbing of the fingers.
- These signs develop as a result of chronic hypoxic.

- **Mx**
  - Surgical repair – if the definitive treatment
  - Supportive management
    - The cyanosis that develops during a TET spell can be relieved when the legs and knees are bent, resulting in reduced blood flow to the lower body and improved blood flow to the vital organs.
    - Infants should be placed in a knee-chest position. If the defect has not been repaired, older children usually squat instinctively.

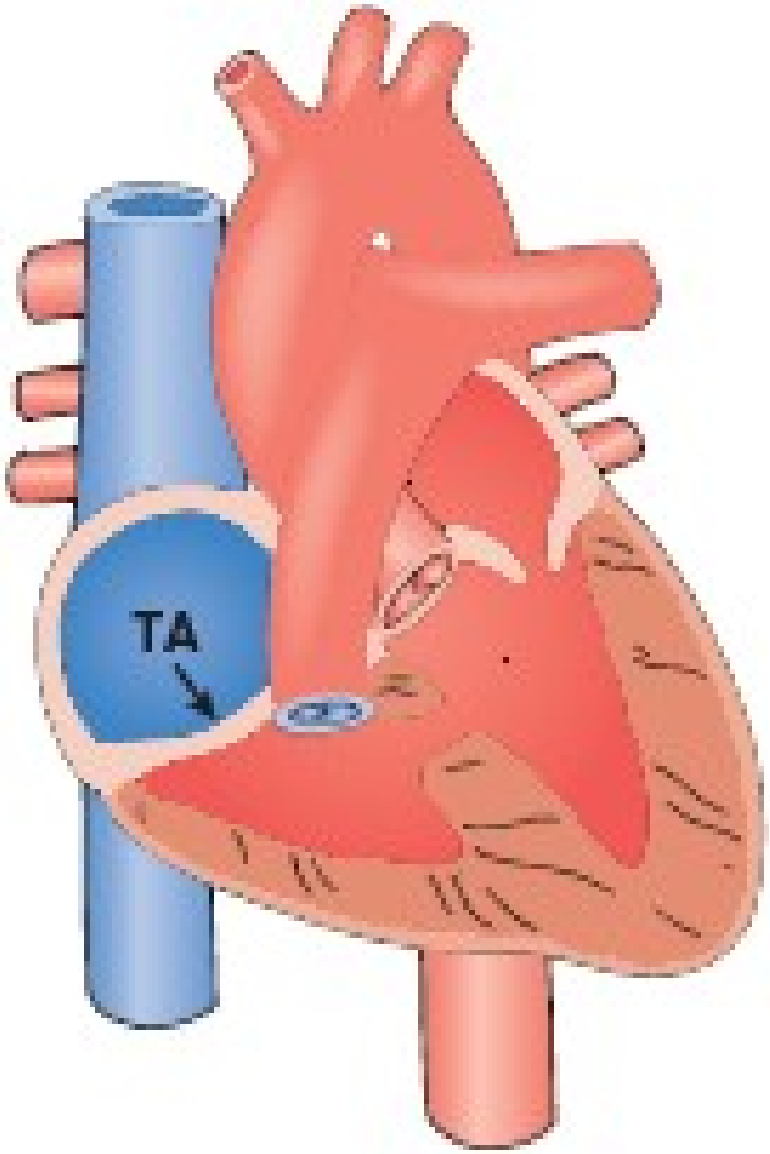

ToF

3. Obstructive Defects

- These are characterized by an intact vascular system but with an obstruction preventing the free flow of blood through the heart

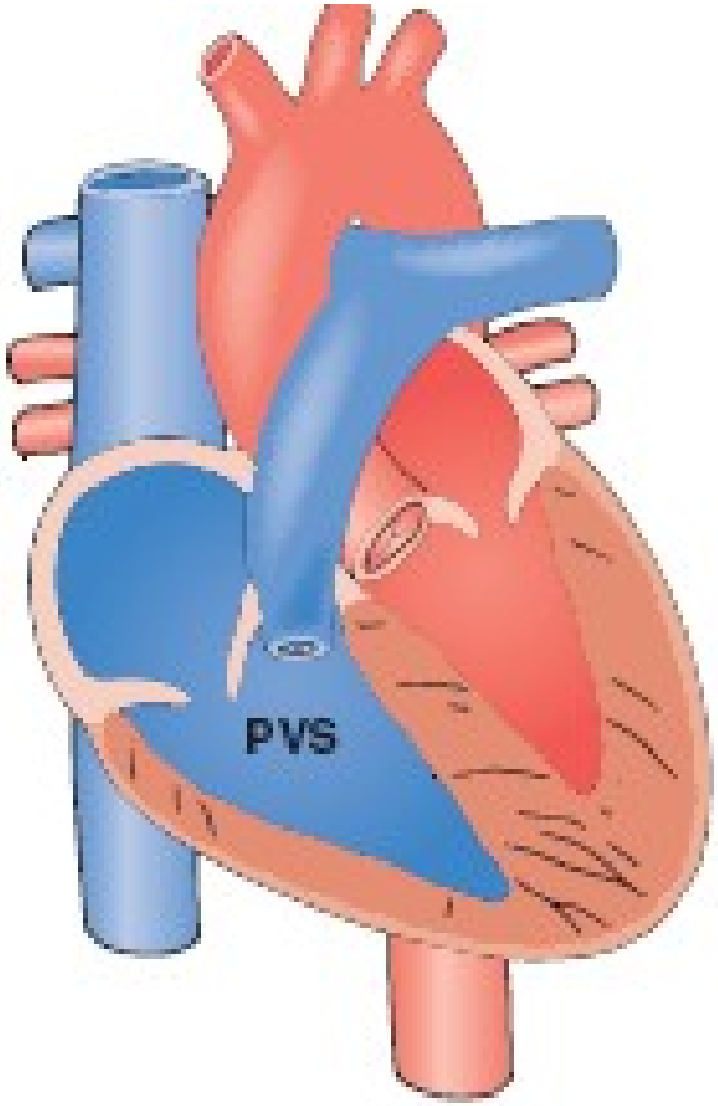
a. Tricuspid atresia (TA)

- Is characterized by a closed tricuspid valve, resulting in no movement of blood from the right atrium to the right ventricle.
- This defect is incompatible with life unless another defect is present that allows mixing of the blood.
- **S & S -** Rapid and sustained cyanosis.
- **Mx -** Surgical repair

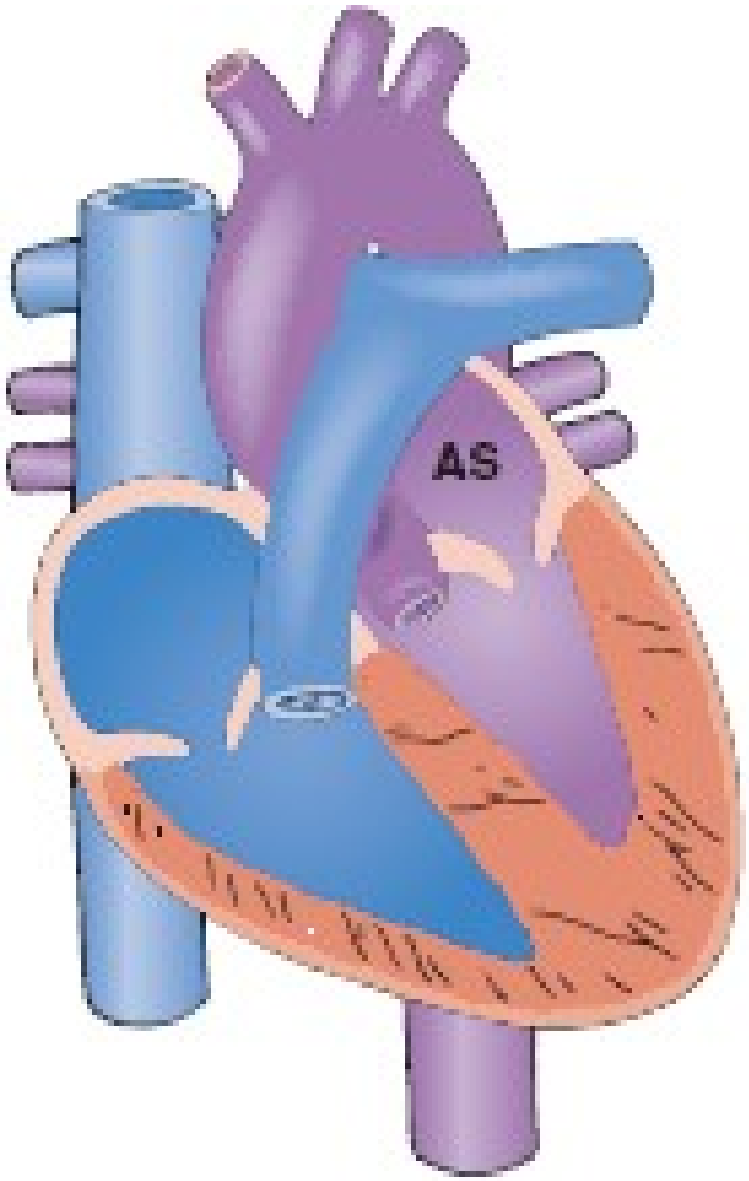

TA

b. Pulmonic stenosis (PVS)

- Is a narrowing of the pulmonary artery or valve.
- **S & S -** Cyanosis during times of activity to severe CHF.
- **Mx -** Balloon angioplasty or surgical repair.

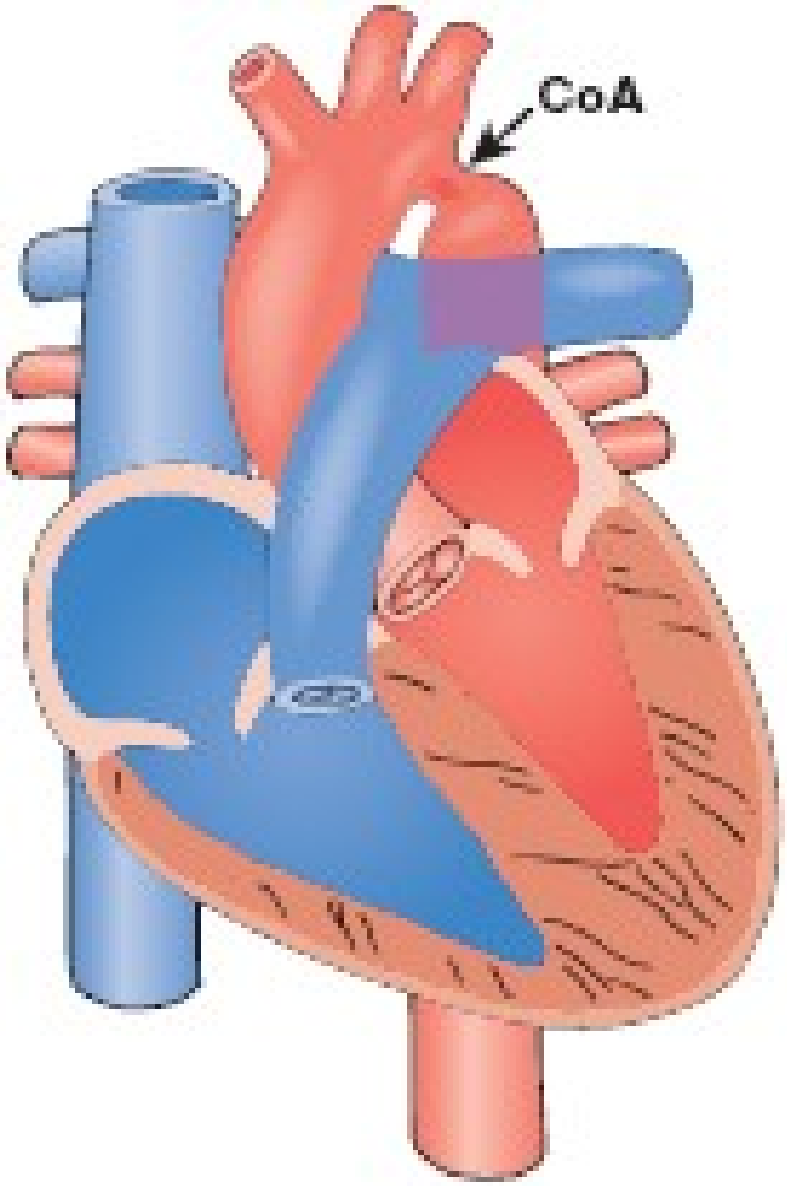

PVS

c. **Aortic stenosis (AS)**
   - Is a narrowing of the aorta or aortic valve.
   - **S & S -** Murmur to CHF.
   - **Mx -** Balloon angioplasty or surgical repair.


AS

d. Coarctation of the aorta (CoA)

- A narrowing of the aorta, usually distal to the ascending vessels.
- **S & S -** Higher blood pressures and pulses in the upper extremities as compared to those in the lower extremities.
- If left uncorrected, older children suffer from recurrent episodes of epistaxis and complaints of leg cramps or leg pain, especially during periods of activity.
- **Mx -** Surgical repair.


CoA