CONGENITAL HEART DISEASES

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CHD

- Group of abnormalities in the structure of the heart or great vessels.
- CHD are among the most common birth defect, occurring in 1% of live births.
- The recurrence risk in offspring is 3-5%.

ETIOLOGY

	Cardiac abnormalities
Maternal disorders	
Rubella infection	Peripheral pulmonary stenosis, PDA
Systemic lupus erythematosus	Complete heart block (anti-Ro and anti-La antibody)
Diabetes mellitus	Incidence increased overall
Maternal drugs	
Warfarin therapy	Pulmonary valve stenosis, PDA
Fetal alcohol syndrome	ASD, VSD, tetralogy of Fallot
Chromosomal abnormality	
Down syndrome (trisomy 21)	Atrioventricular septal defect, VSD
Edwards syndrome (trisomy 18)	Complex
Patau syndrome (trisomy 13)	Complex
Turner syndrome (45XO)	Aortic valve stenosis, coarctation of the aorta
Chromosome 22q11.2 deletion	Aortic arch anomalies, tetralogy of Fallot, common arterial trunk
Williams syndrome (7q11.23 microdeletion)	Supravalvular aortic stenosis, peripheral pulmonary artery stenosis
Noonan syndrome (PTPN11 mutation and others)	Hypertrophic cardiomyopathy, atrial septal defect, pulmonary valve stenosis

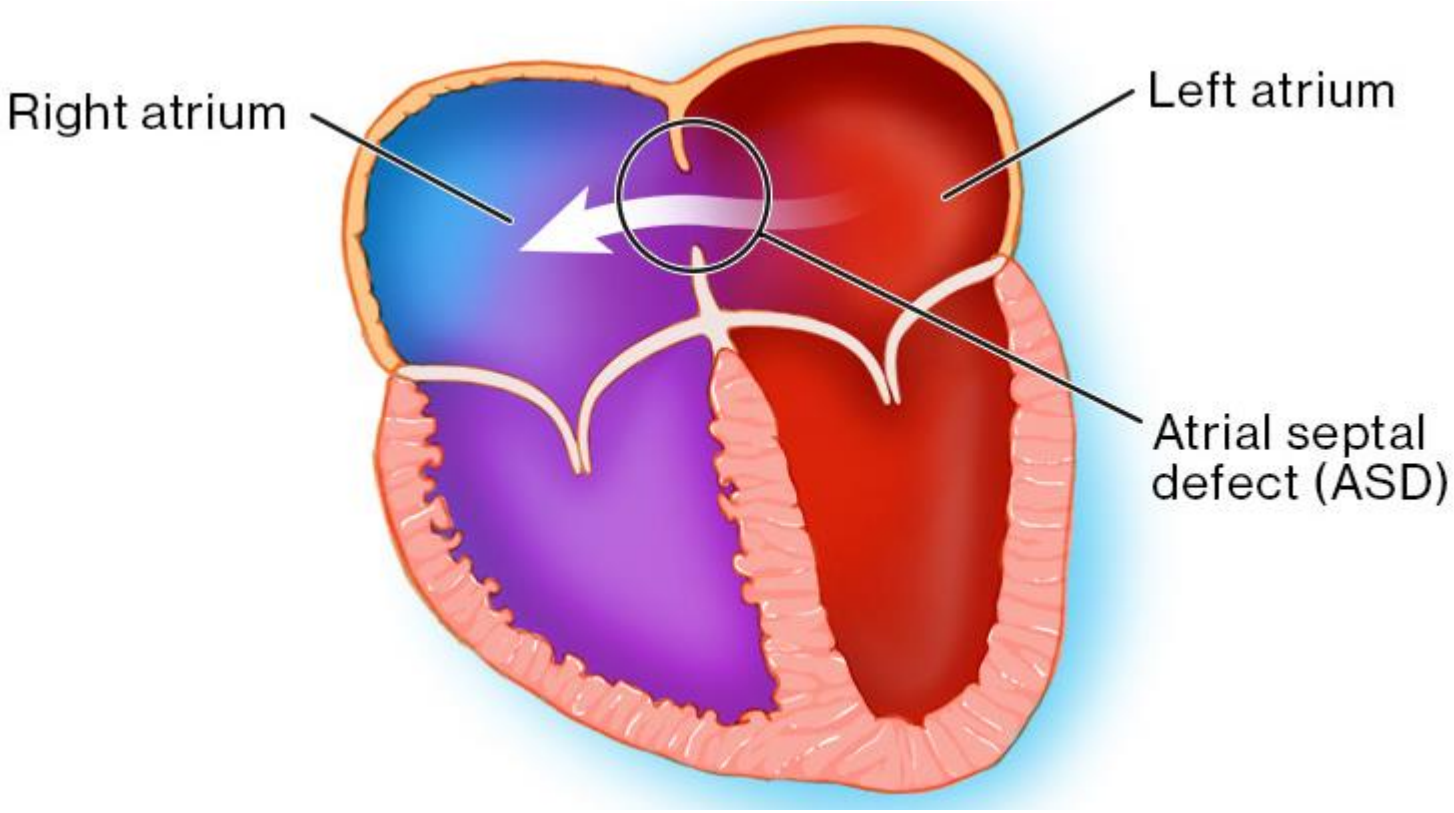
CHD classified into three major groups:

- **Acyanotic CHD:** LR shunt: VSD, ASD, PDA
- **Cyanotic CHD:** RL shunt: TOF, TGA, TA, Tr. Art., TAPVR
- **Obstructive lesions**: AS, PS, COA.

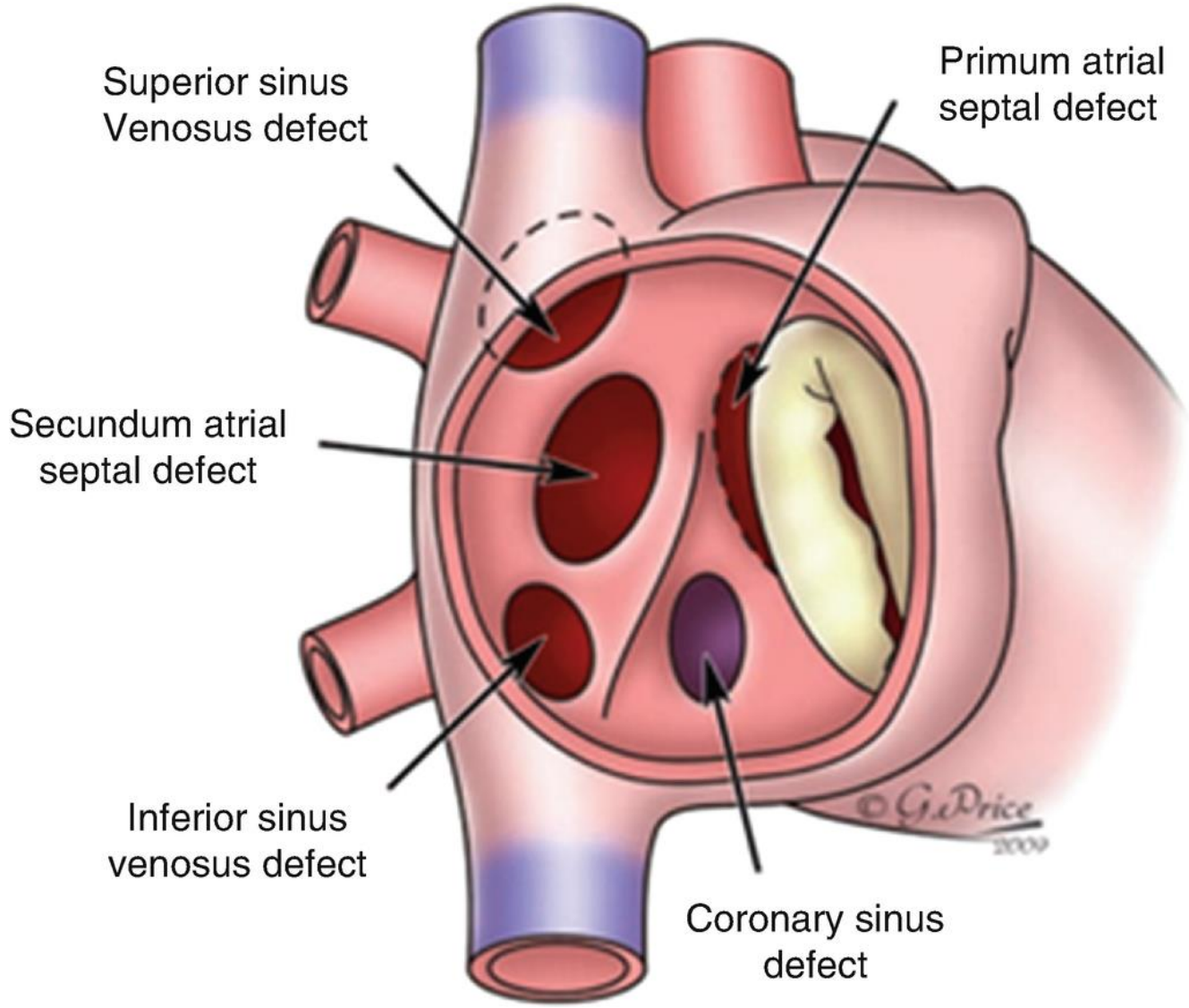
ACYANOTIC CHD

ATRIAL SEPTAL DEFECT (ASD):

- ASDs represent approximately 10% of all CHD.
- Female preponderance (male to female ratio of 1:2)

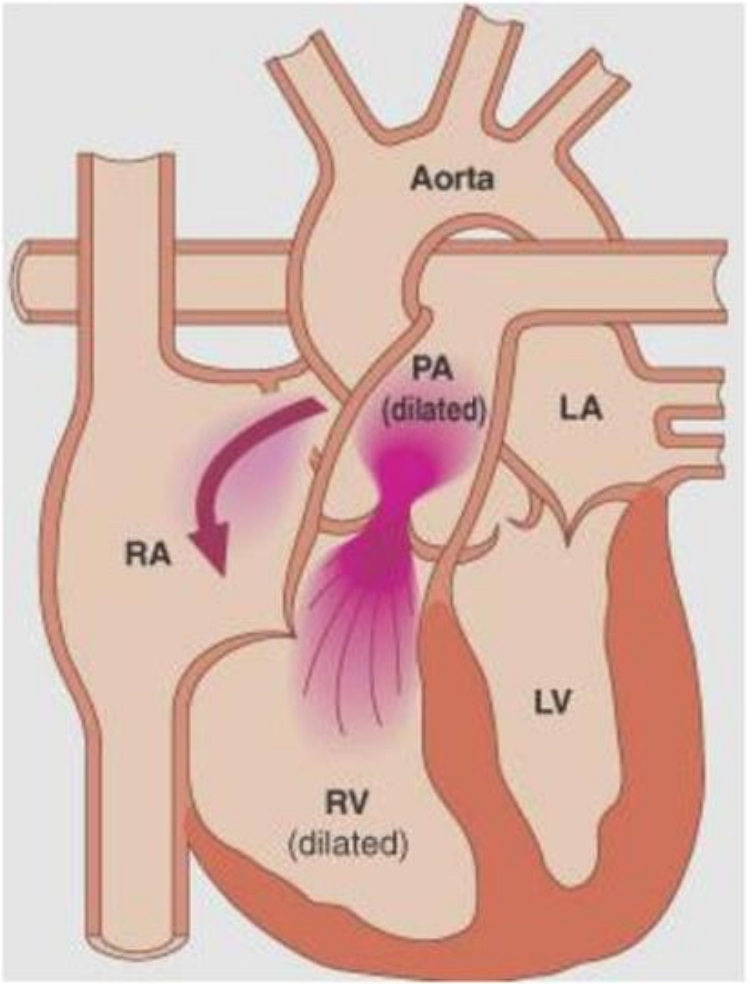


- Four types of ASD:
  - **Secundum ASD:** (75%).
  - **Primum ASD:** (20%).
  - **Sinus venosus ASD:** (5%).
  - **Coronary sinus ASD:** (<1%).



PATHOPHYSIOLOGY

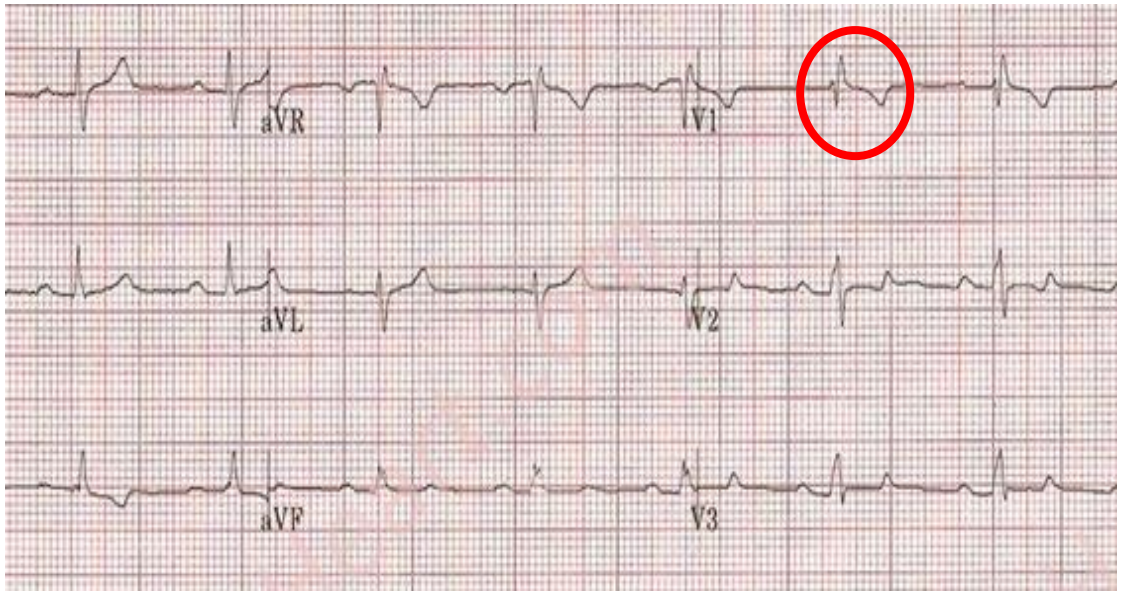
- An L-R shunt occurs through the defect, with a volume overload to the RA and RV and an increase in pulmonary blood flow.



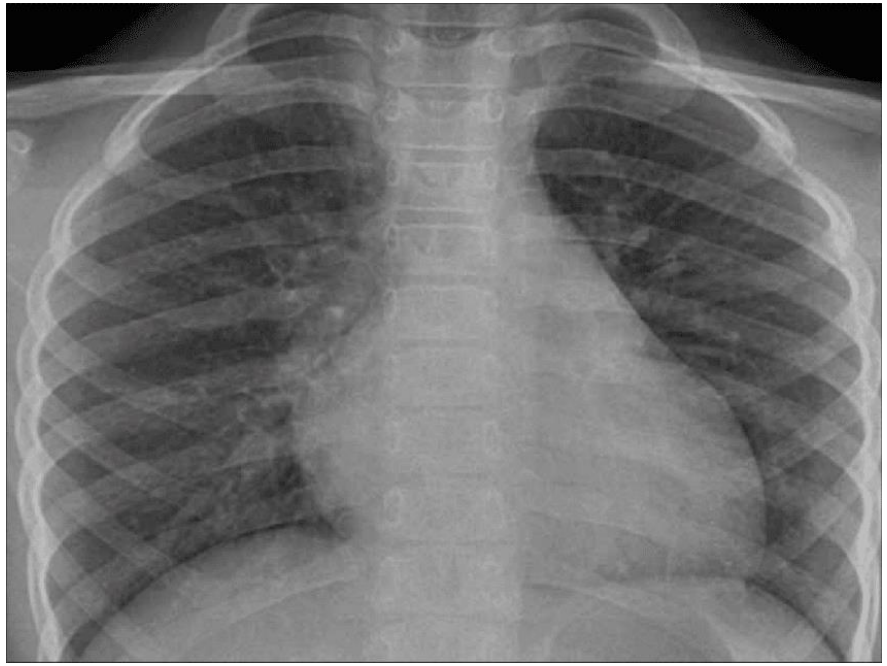
CLINICAL MANIFESTATIONS

- The patients are usually asymptomatic.
- A widely split and fixed S2.
- a grade 2 to 3/6 systolic ejection murmur at the ULSB are characteristic of moderate-size ASD.

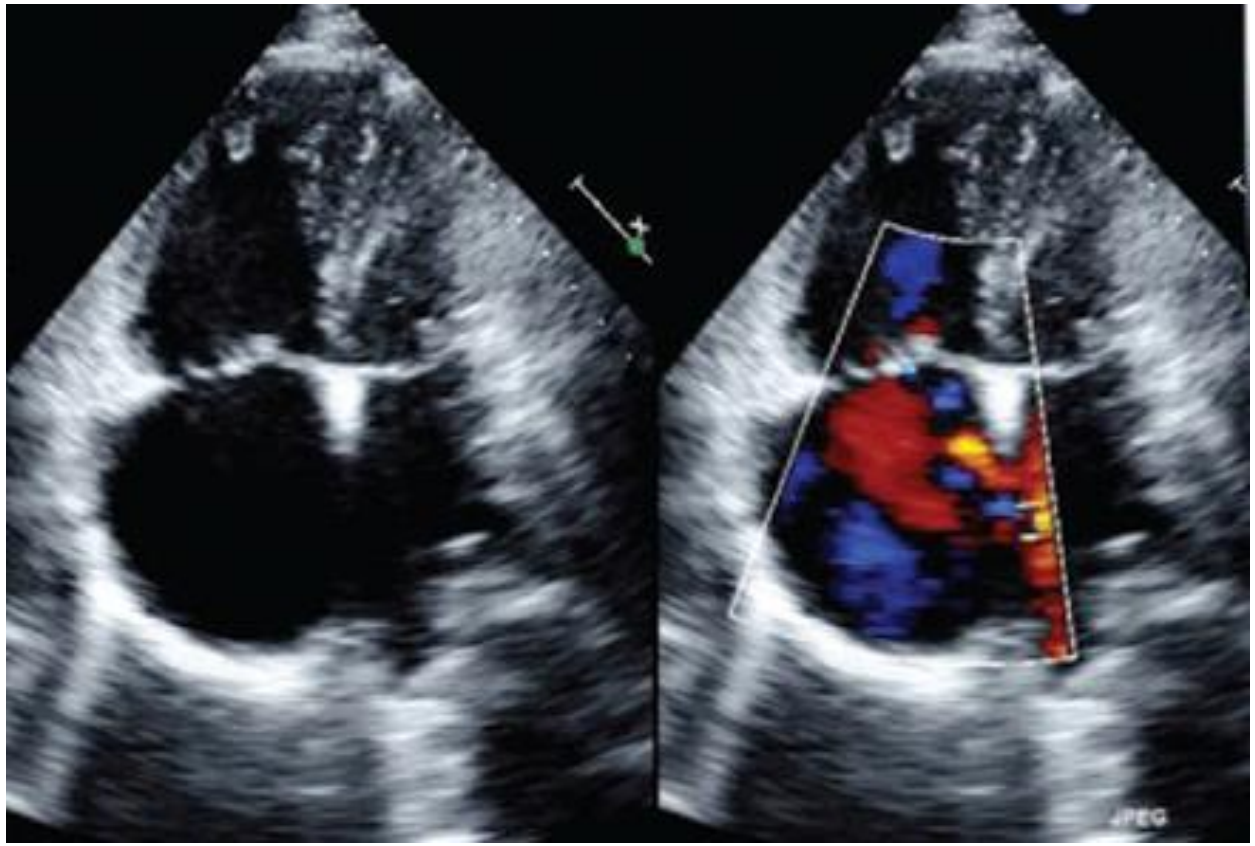
- **ECG** → RAD, RBBB (secundum ASD)
  → LAD, RBBB (primum ASD)



- **CXR** → cardiomegaly, increased PVM, and a prominent MPA segment when the shunt is moderate or large.



- ECHO → the position and the size of the defect.



Natural history

- Spontaneous closure of the defect occurs in more than 80% of patients with defects of 3 to 8 mm (diagnosed by echo) before 1½ years of age.

- An ASD with a diameter >8 mm rarely closes spontaneously. Spontaneous closure is not likely to occur after 4 years of age.

- If the defect is large and left untreated, arrhythmias and PHT develops in the third and fourth decades of life.

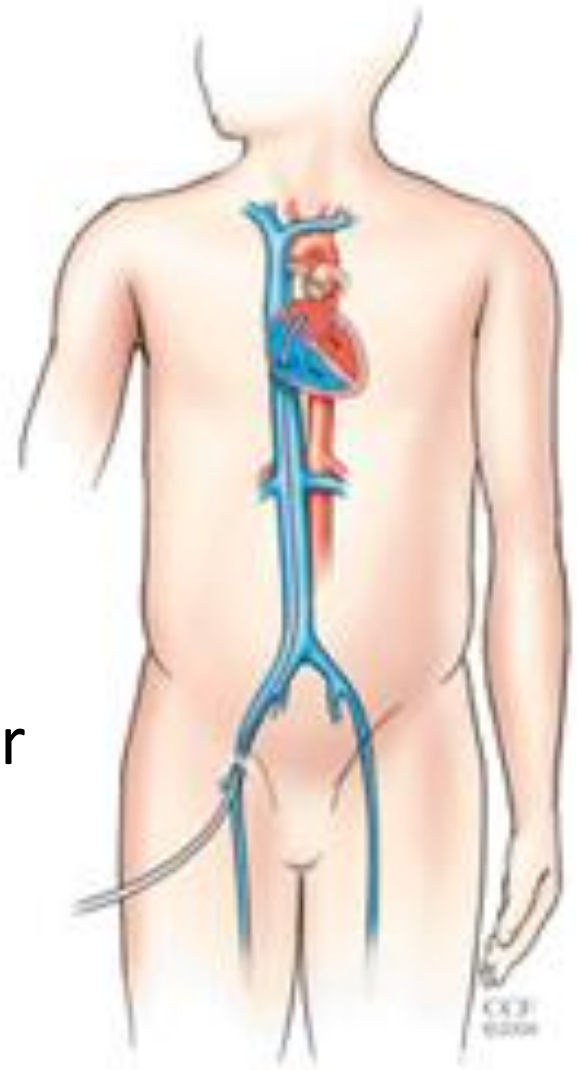
Management

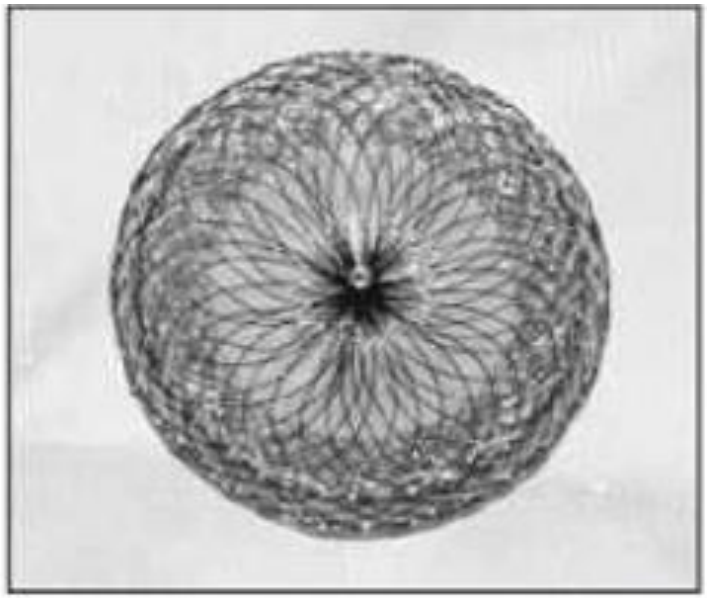
Medical

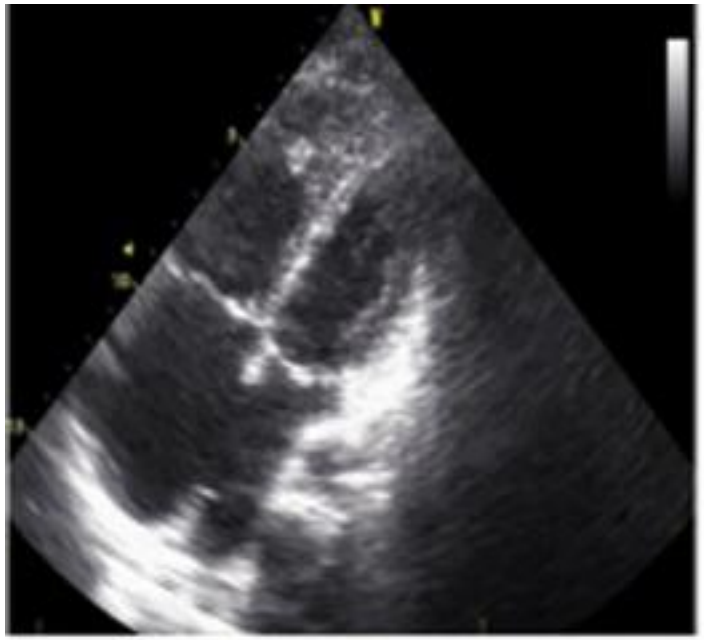
1. Exercise restriction is not required, unless symptomatic.

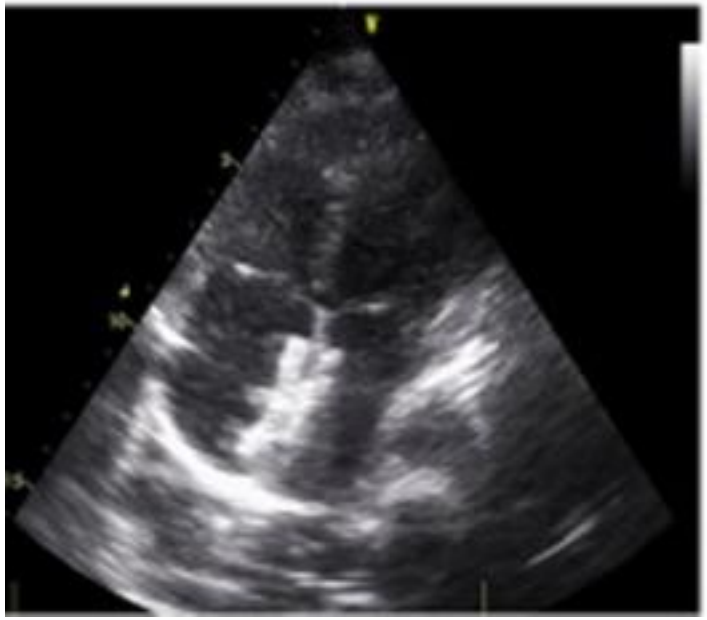
2. Antifailure if CHF developed (Rarely)

3. Transcatheter closure of the defect using a catheter-delivered closure device has become a preferred method for secundum ASD.









- Advantages of nonsurgical closure would include a <24-hr hospital stay, rapid recovery, and no residual thoracotomy scar.
- Closure rates are excellent.

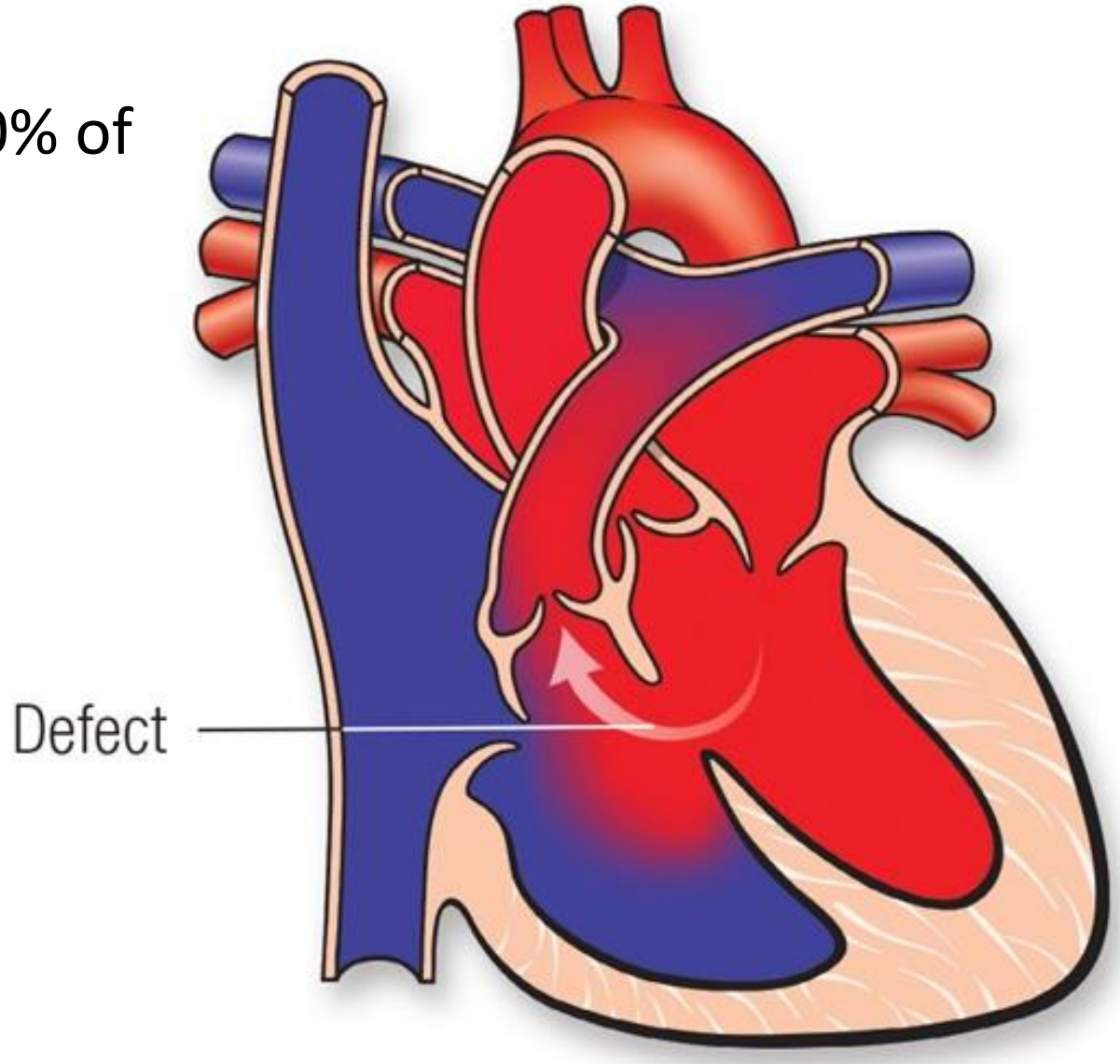
Surgical

- For patients with primum ASD and sinus venosus defect, and some patients with secundum ASD for which the device closure is considered inappropriate.

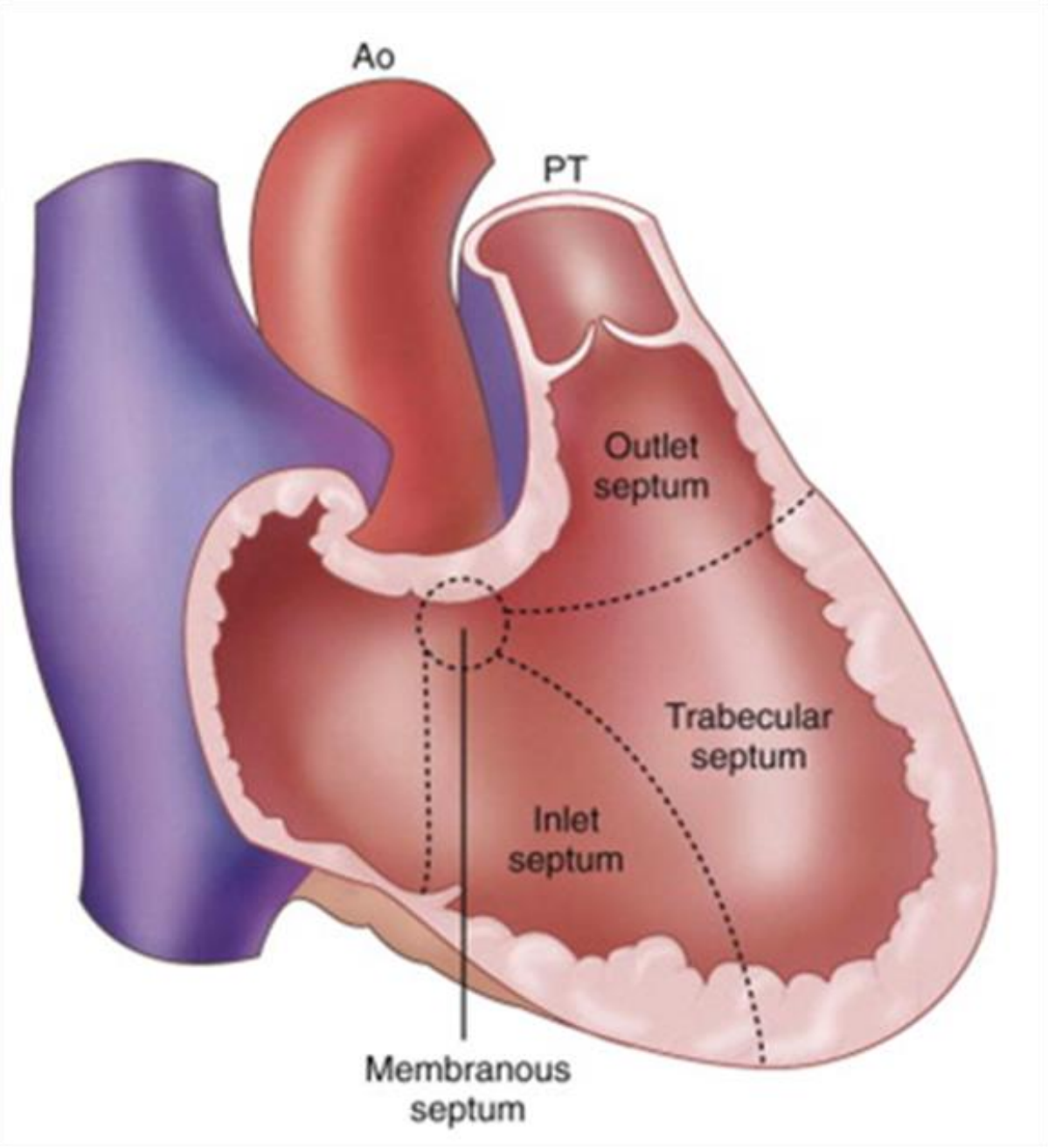
- Surgical closure is indicated when there is a significant L-R shunt with Qp/Qs of 1.5:1 or greater.

VENTRICULAR SEPTAL DEFECT

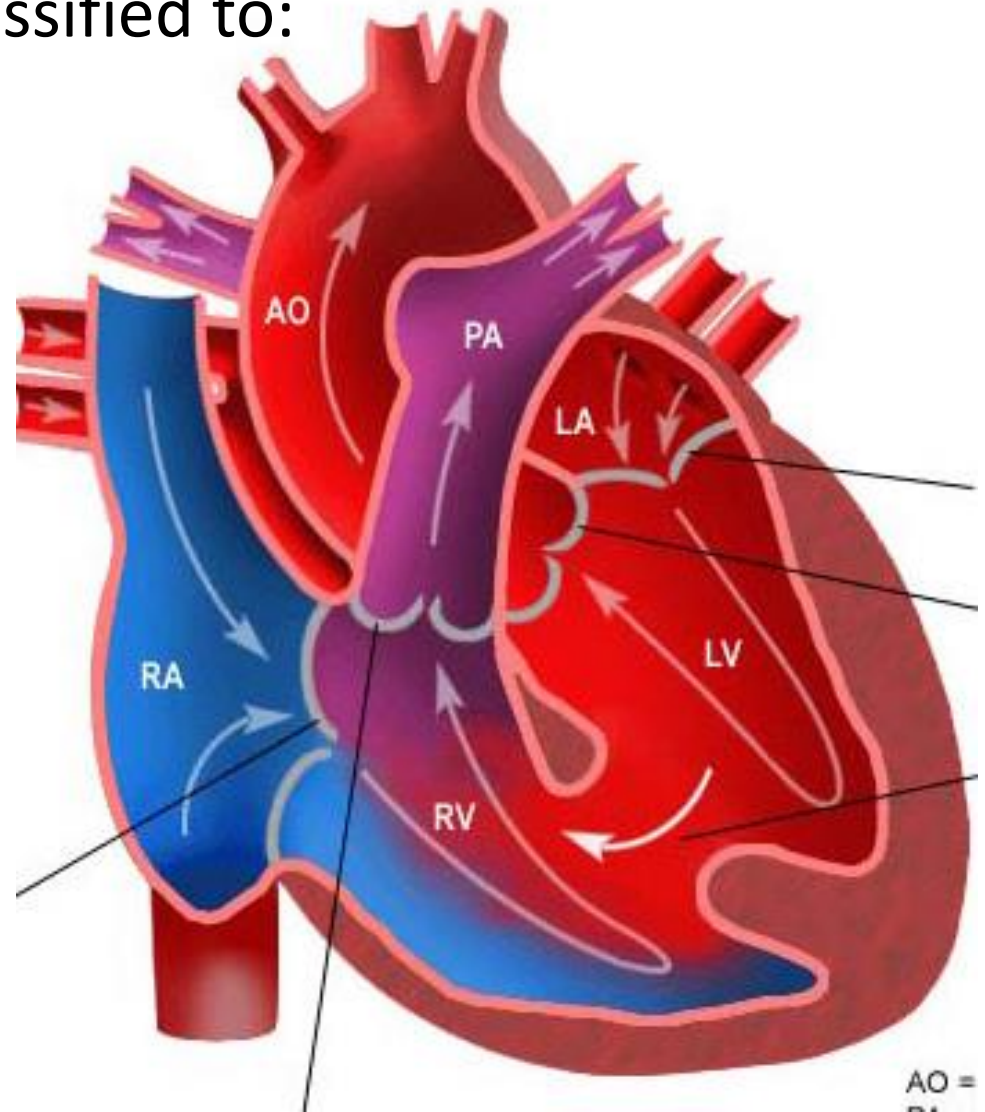
- VSDs are common, accounting for 30% of all cases of CHD.



- **There are 4 types:**
  1. Perimembaranous VSD.
  2. Muscular VSD.
  3. Inlet VSD.
  4. Outlet VSD.



- According to the size of the defect, VSD classified to:
  - **Small size**: less than 1/3 of the aorta size.
  - **Moderate size**: 1/3-1/2 of aorta size.
  - **Large size**: more than 1/2 of the aorta size.



Clinical Manifestations

- Patients with small VSDs are asymptomatic, with normal growth and development.

- Patients with Large VSDs, delayed growth and development, repeated pulmonary infections, CHF, and decreased exercise tolerance are relatively common.

- Patients with PVOD, cyanosis and a decreased level of activity may result.

- **Small VSD →**
  - loud pansystolic murmur at lower left sternal edge.
  - Quiet pulmonary second sound (P2).

- **Large VSD →**
  - Tachypnoea, tachycardia and enlarged liver.
  - Active precordium.
  - Soft pansystolic murmur (implying large defect).
  - Apical mid-diastolic murmur.
  - Loud pulmonary second sound (P2).

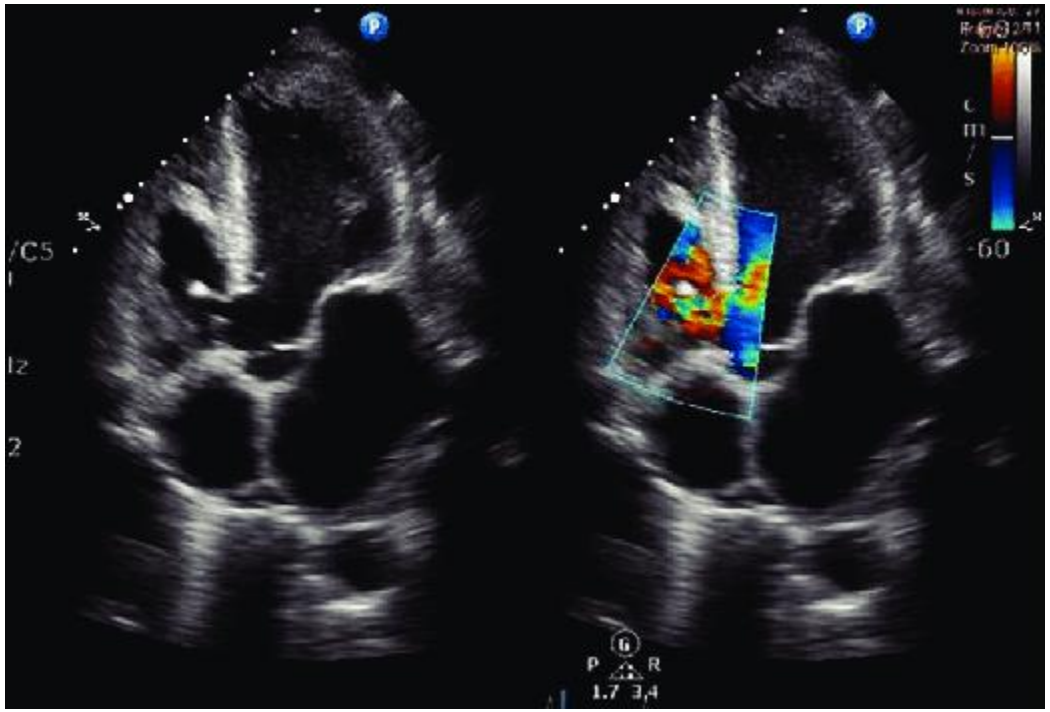
- **ECG** → **Small VSD** → normal
  → **Moderate VSD** → LVH and LAE (±)
  → **Large VSD** → biventricular hypertrophy (BVH) and LAE (±)
  → **PVOD** → pure RVH

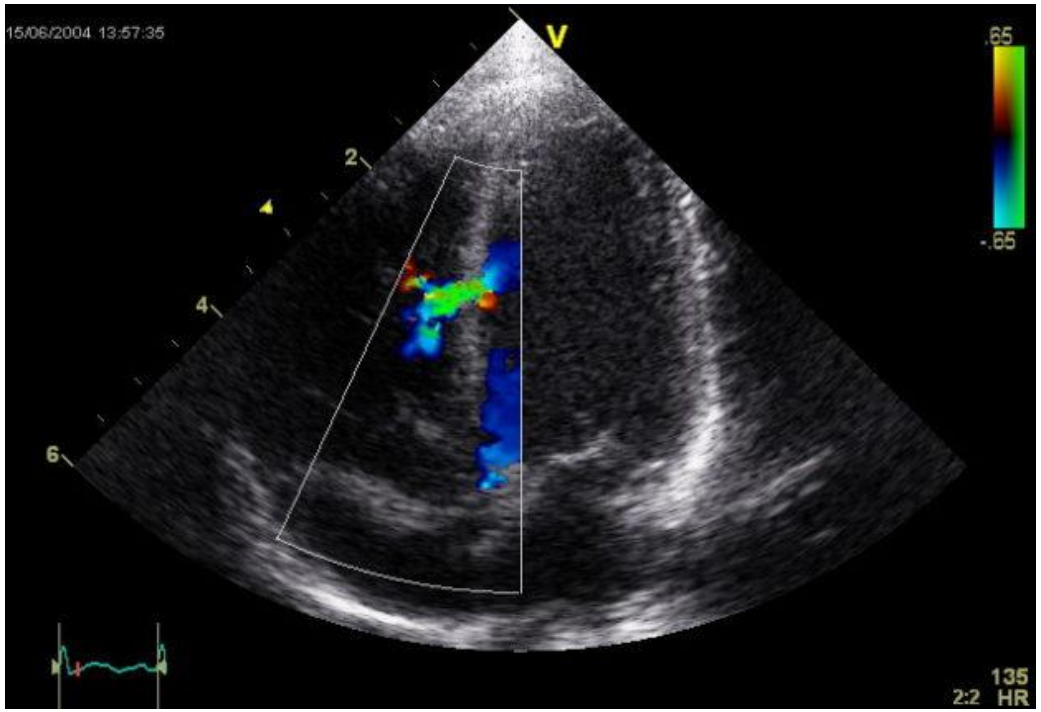
- **CXR** → Cardiomegaly of varying degrees

  → Pulmonary vascular markings (PVMs) are increased.

  → In PVOD, the heart is no longer enlarged and the MPA and the hilar pulmonary arteries are notably enlarged, but the peripheral lung fields are oligemic.

- **ECHO** → accurate diagnosis of the position and size of the VSD.





Natural history

- Spontaneous closure occurs in 30% to 40% of all VSDs, most often in muscular VSDs (up to 80%), more frequently in small defects than in large defects, and more often in the first year of life than thereafter.

- CHF develops in infants with a large VSD but usually not until 6-8 weeks of age, when the PVR drops below a critical level.

- PVOD may begin to develop as early as 6 to 12 months of age in Patients with a large VSD.

Management

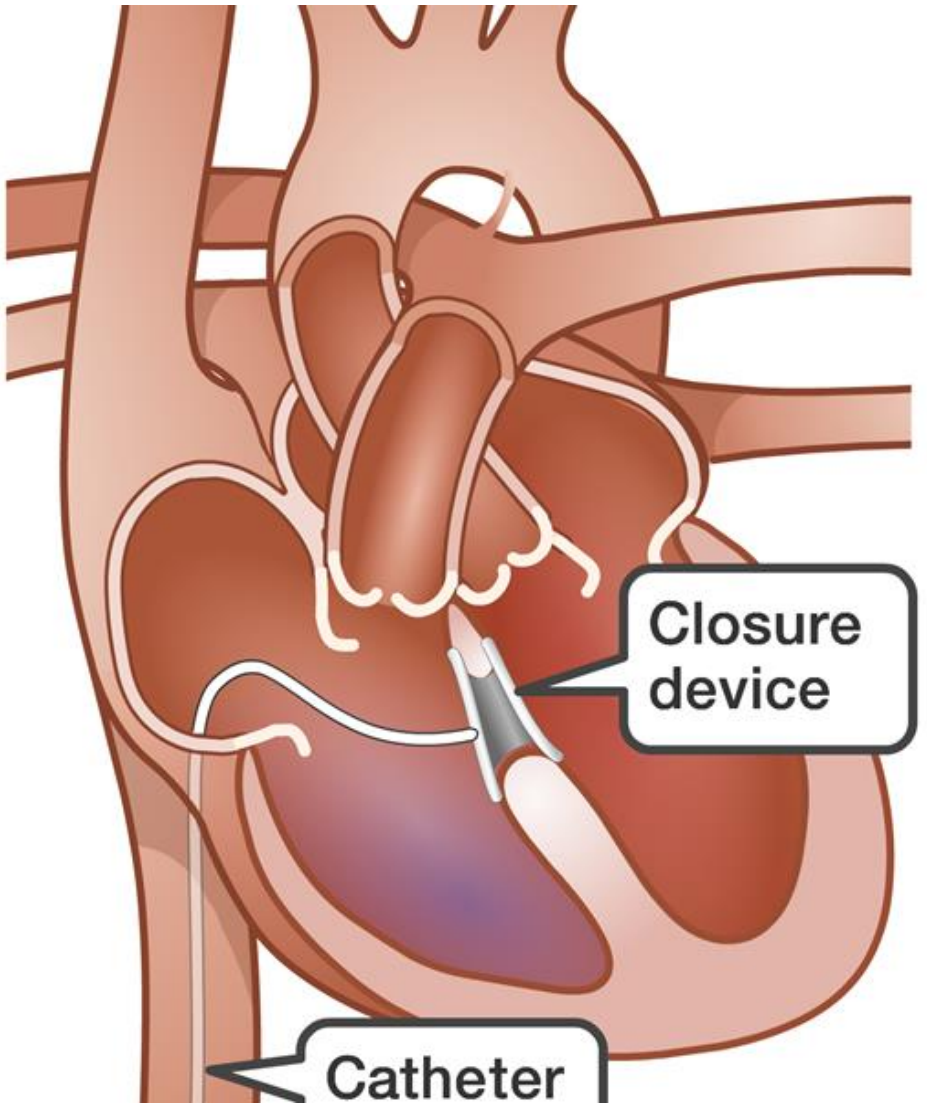
Medical

1. Treatment of CHF with diuretics, after-load reducers, and sometimes digoxin.

2. No exercise restriction is required in the absence of pulmonary hypertension.

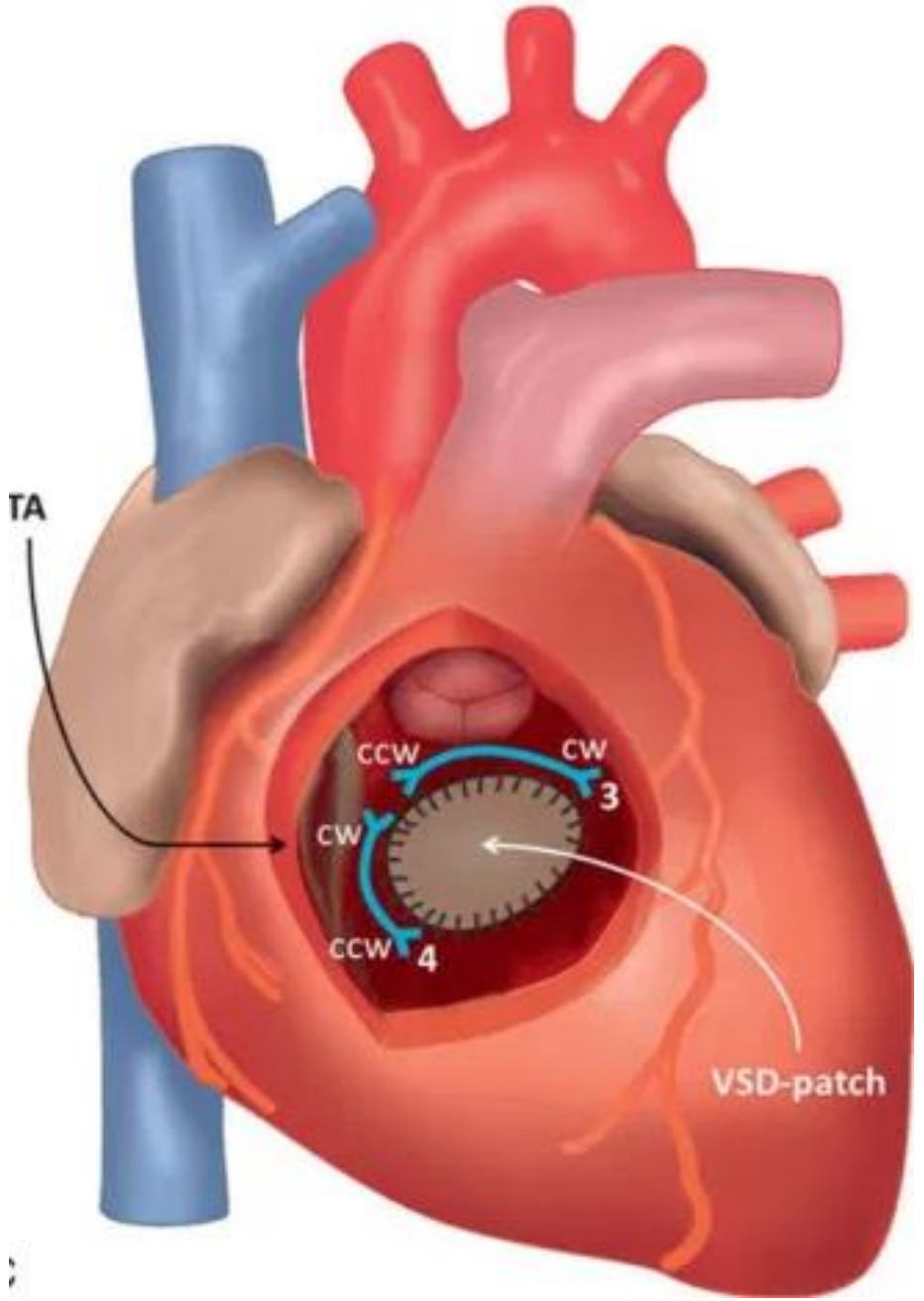
3. Proper dental hygiene.

4. Transcatheter device closure of VSD is possible when the defect is anatomically amenable or when it is difficult to access surgically.



Surgical

- Direct closure of the defect is performed using pericardial or prosthetic patch.



Timing

(1) Infants with CHF and FTT unresponsive to medical therapy should be closed on at any age, including early infancy.

(2) Infants with a large VSD and evidence of increasing PVR should be closed on as soon as possible.

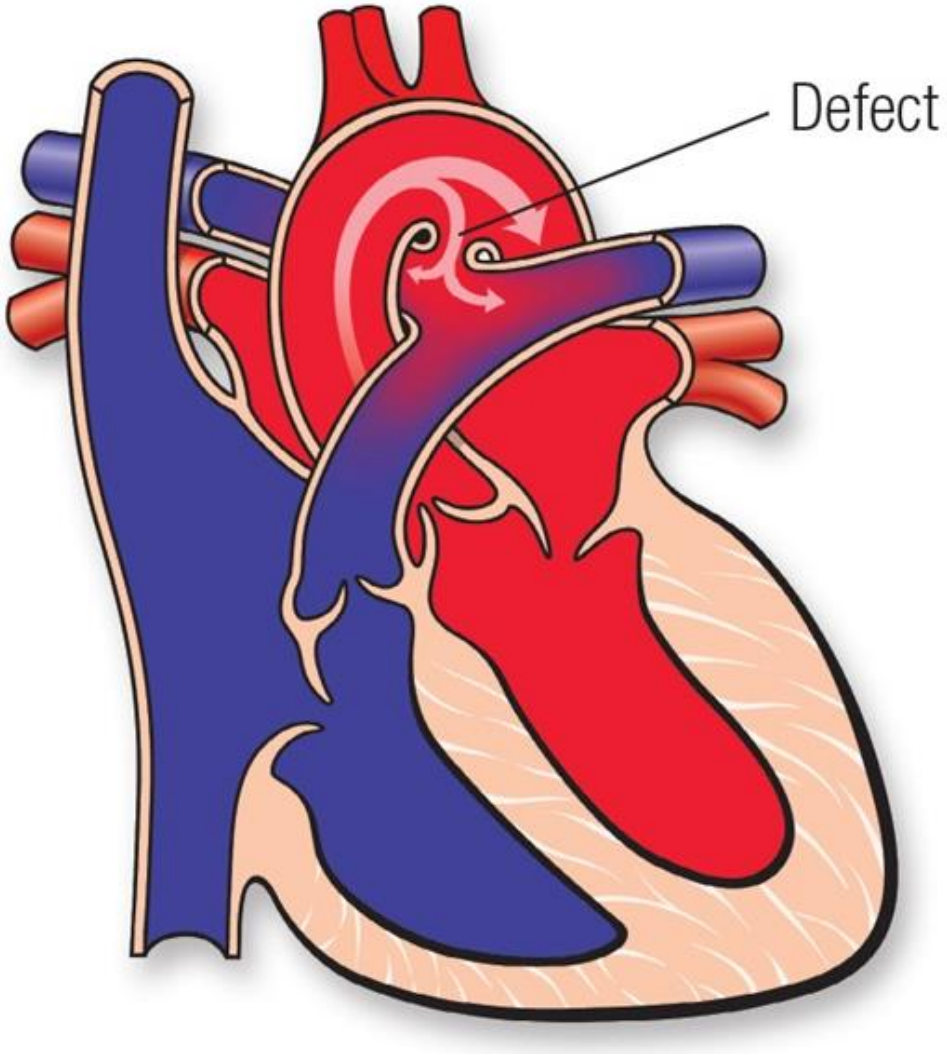
(3) Infants who respond to medical therapy may be closed on by the age of 12 to 18 months.

(4) Asymptomatic children may be closed on between 2 and 4 years of age.

Patent ductus arteriosus

- The ductus arteriosus connects the pulmonary artery to the descending aorta. In term infants, it normally closes shortly after birth.

- In PDA it has failed to close by 1 month after the expected date of delivery due to a defect in the constrictor mechanism of the duct.



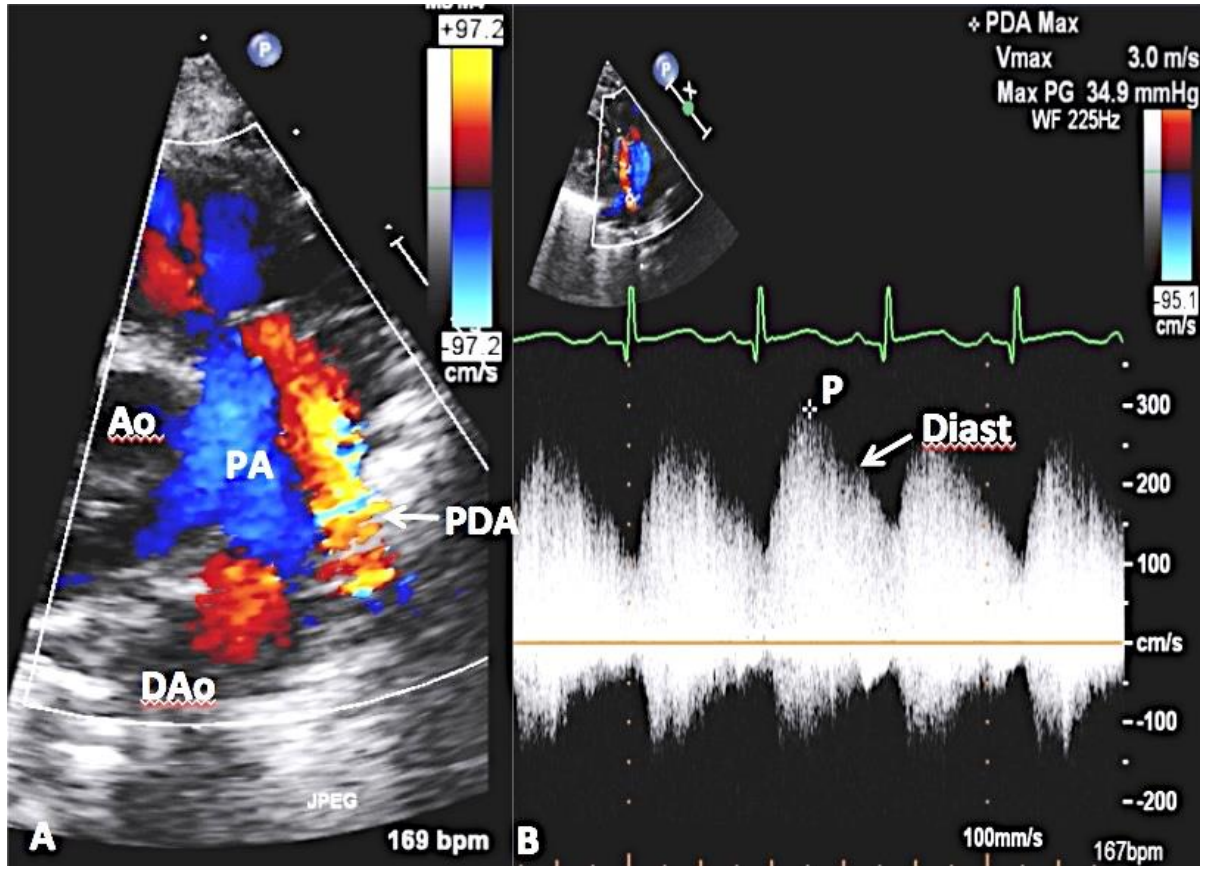
Clinical features

- Depending on the size of PDA, presentation range from asymptomatic to CHF, FTT, PHT.

- Most children present with a **continuous murmur**.

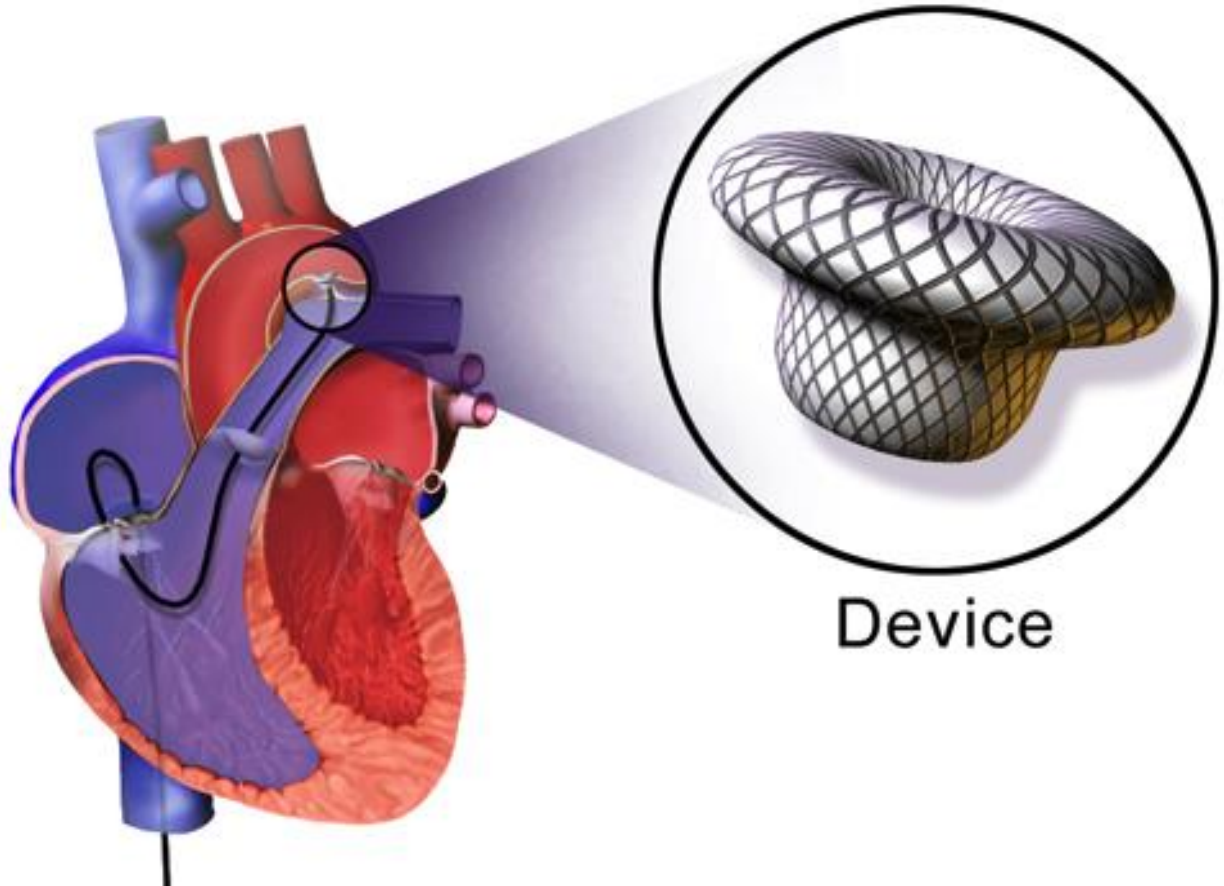
- The pulse pressure is increased, causing a collapsing or bounding pulse.

- The chest radiograph and ECG are usually normal, but if the PDA is large and symptomatic the features on chest radiograph and ECG are as in patient with a large VSD.
- Echocardiography to confirm diagnosis.



Management

- Closure is recommended to abolish the lifelong risk of bacterial endocarditis and of PHT.
- Medical treatment to control heart failure
- Transcatheter closure using coil or occluding device is the method of choice.
- Occasionally, surgical ligation is required.



FEEL FREE TO ASK ANY QUESTION