



SNS COLLEGE OF ALLIED HEALTH SCIENCES
SNS Kalvi Nagar, Coimbatore - 35
Affiliated to Dr MGR Medical University, Chennai



DEPARTMENT OF CARDIO PULMONARY PERFUSION CARE
TECHNOLOGY

COURSE NAME : PRINCIPLES OF PERFUSION TECHNOLOGY

II YEAR

PATHOLOGY OF HEART

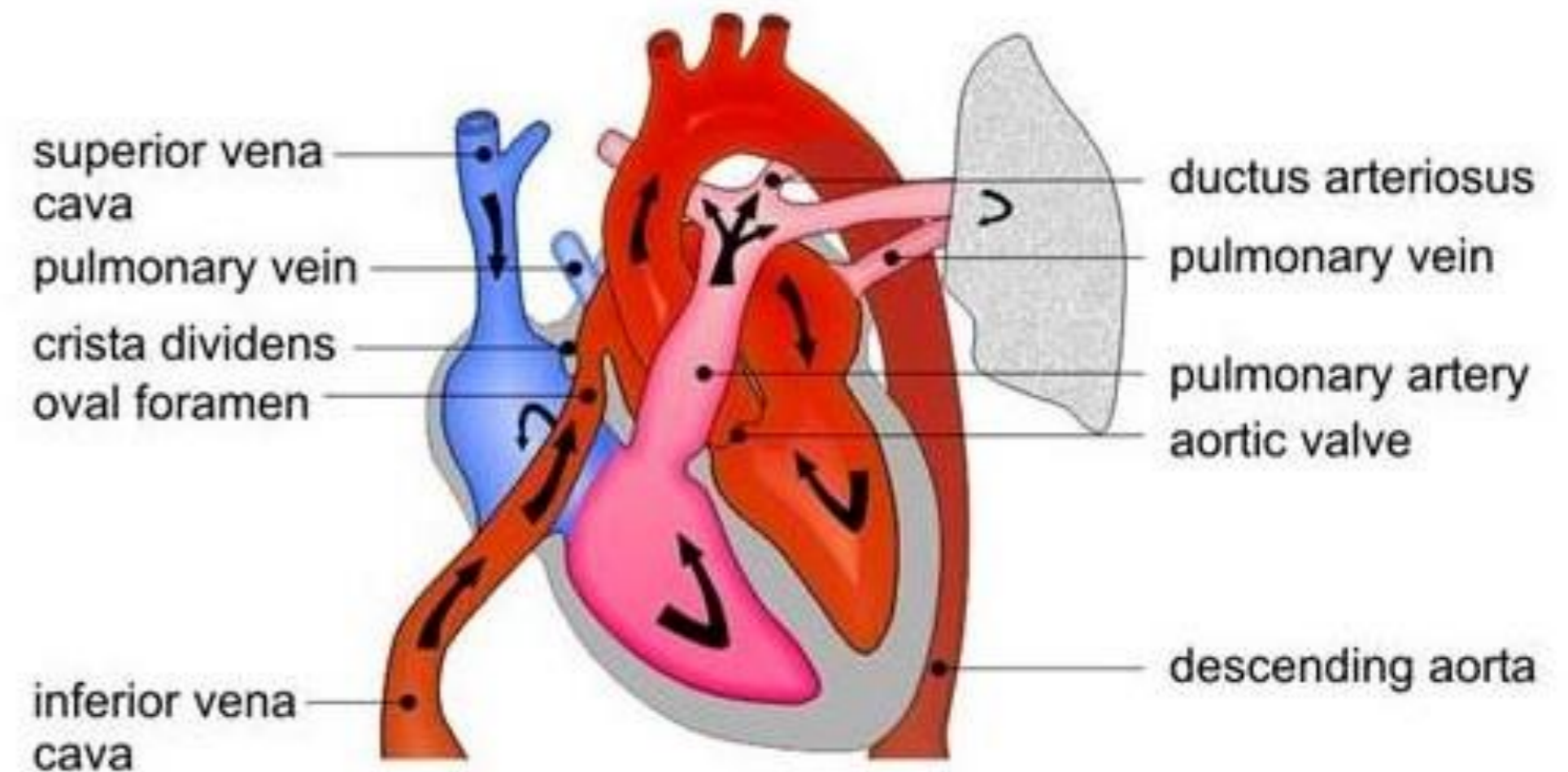
TOPIC : CONGENITAL HEART DISEASES



Congenital heart diseases



- Congenital heart disease is the **abnormality of the heart present from birth.**
- The incidence is higher in premature infants.
- It is attributed to multifactorial inheritance involving genetic and environmental influences.





Classification



Congenital anomalies of the heart may be either, ***shunts (left-to-right or right-to-left)***, or defects causing ***obstructions*** to flow, ***malposition*** of the heart.





Left - to - Right Shunts (A cyanotic or Late Cyanotic Group)



Left to Right Shunts (Acyanotic or Late Cyanotic Group)

It causes volume overload on the right heart



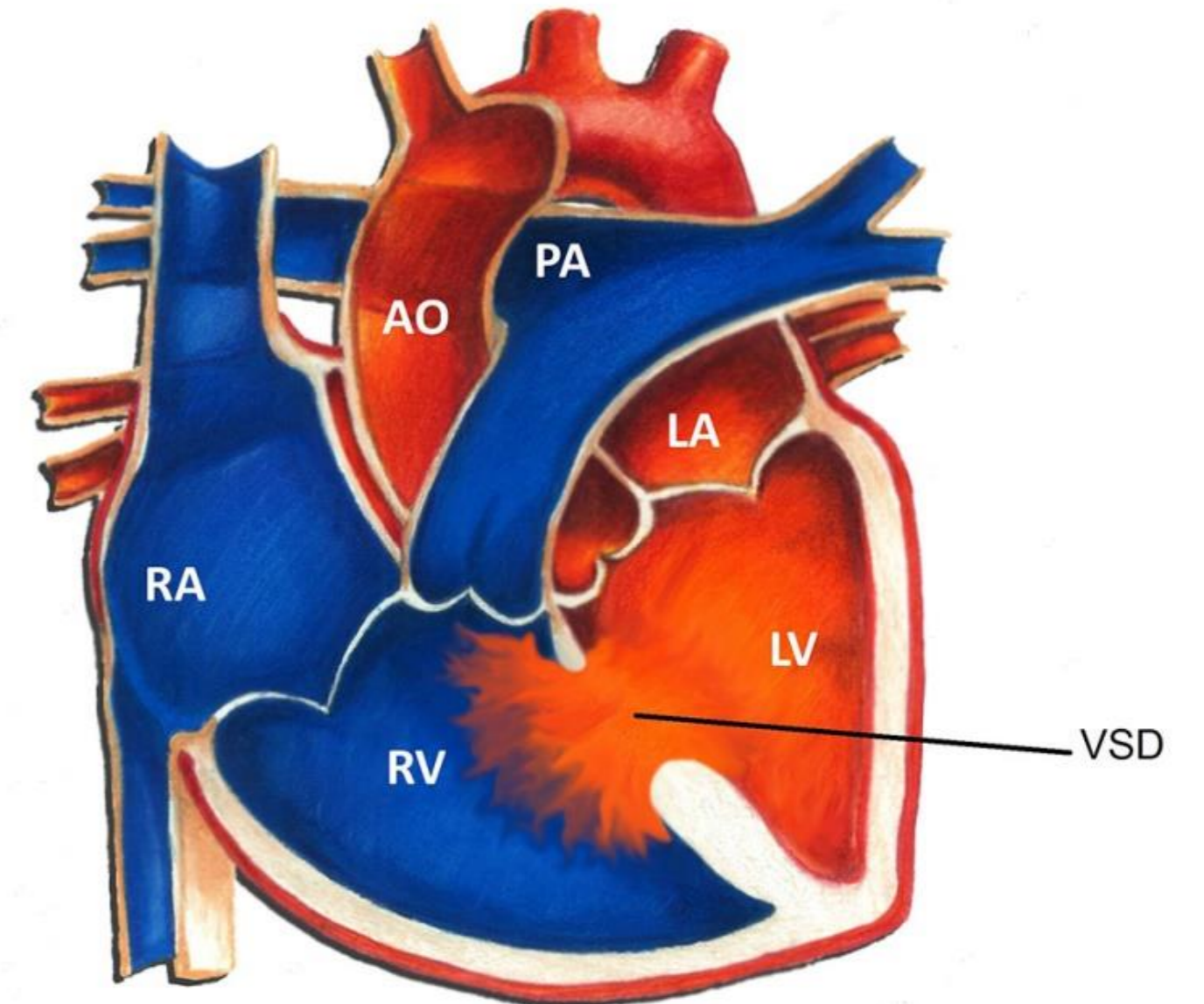
Pulmonary hypertension and right ventricular hypertrophy



Ventricular Septal Defect (VSD)

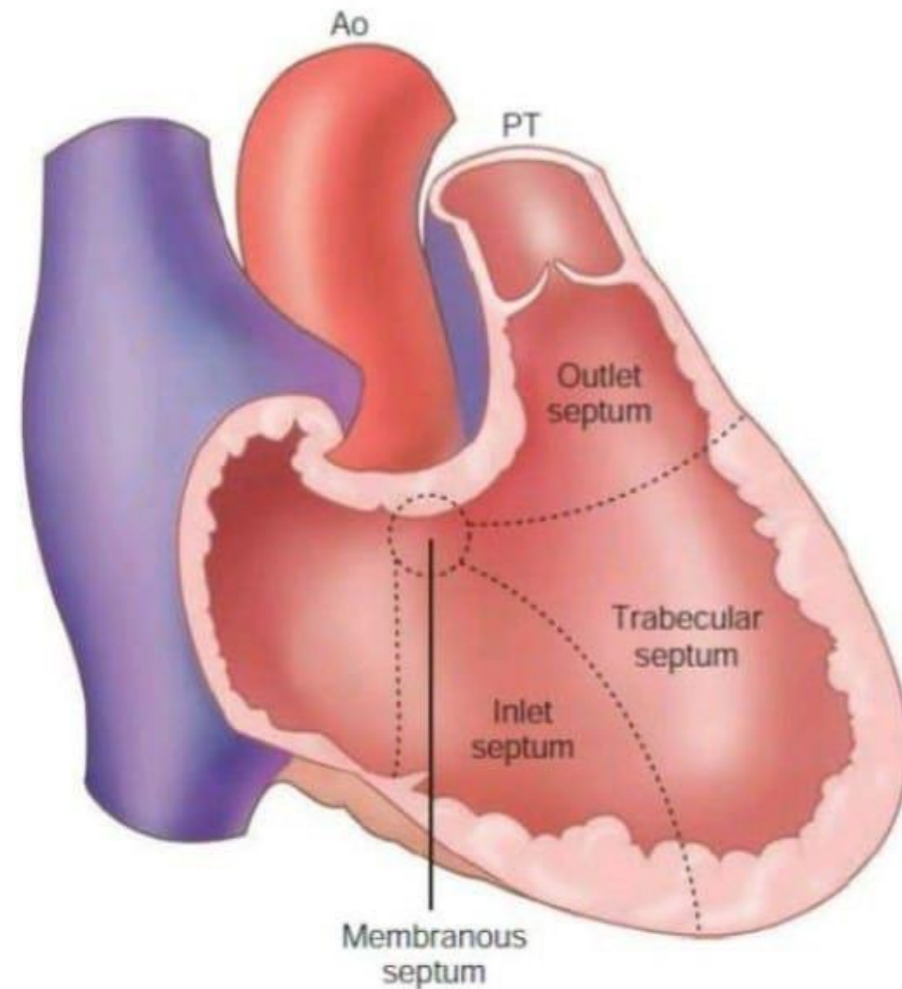


- VSD is the most common congenital anomaly of the heart and comprises about 30% of all congenital heart diseases.
- The smaller defects often close spontaneously, while larger defects remain patent and produce significant effects.



Types of VSD

Classification: Based on anatomical location



- a) A small membranous portion and
- b) A large muscular portion:
 - a) The inlet septum,
 - b) The outlet septum
 - c) The trabecular septum:
 - I. Anterior
 - II. Posterior
 - III. Mid
 - IV. Apical

Classification: Based on size

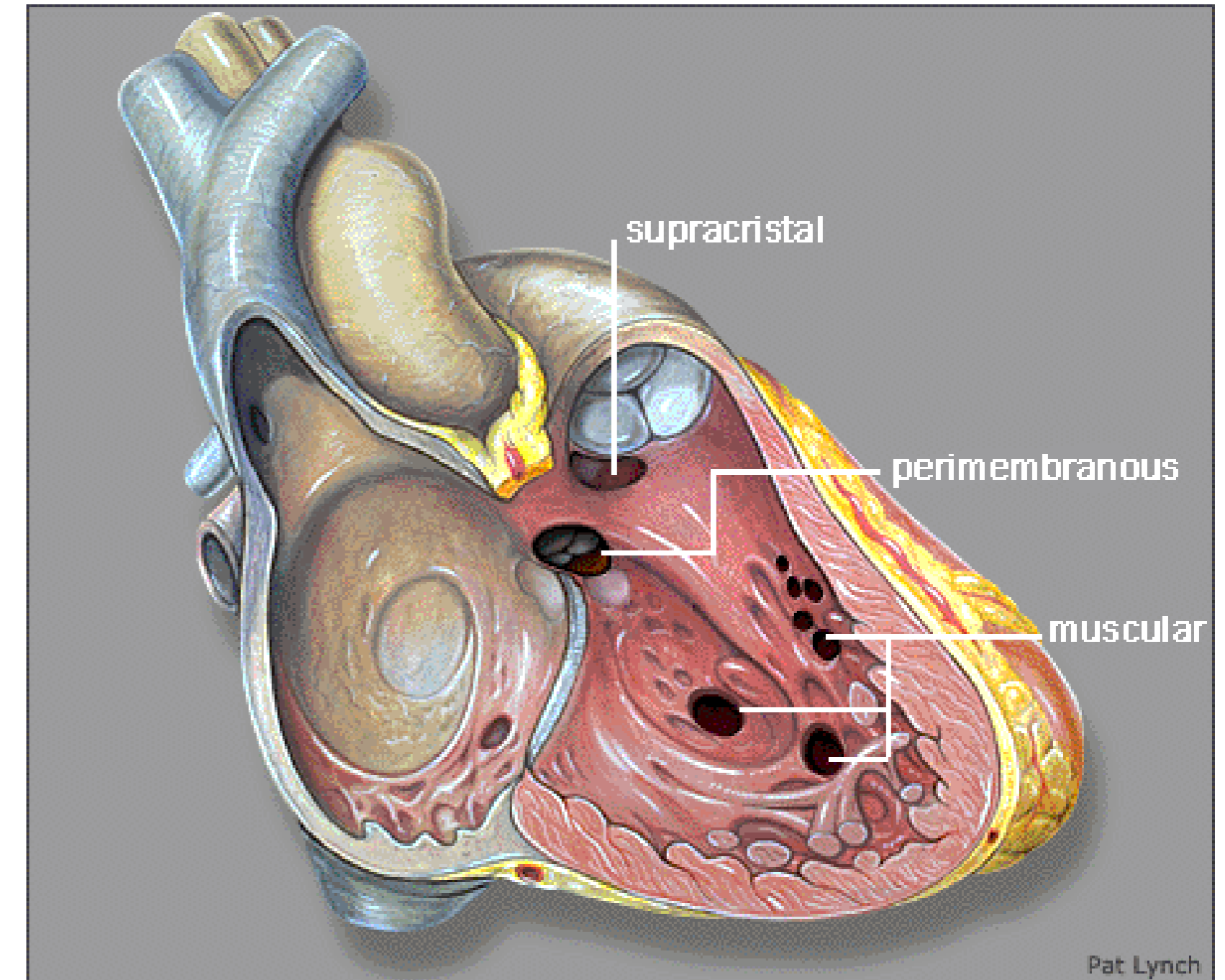
- **Small VSD:** defect size is less than one-third of the size of the aortic root,
- **Moderate VSD:** defect size is less than one-half of the size of the aortic root, and
- **Large VSD:** defect size is equal to or larger than the size of the aortic root.



Types of VSD



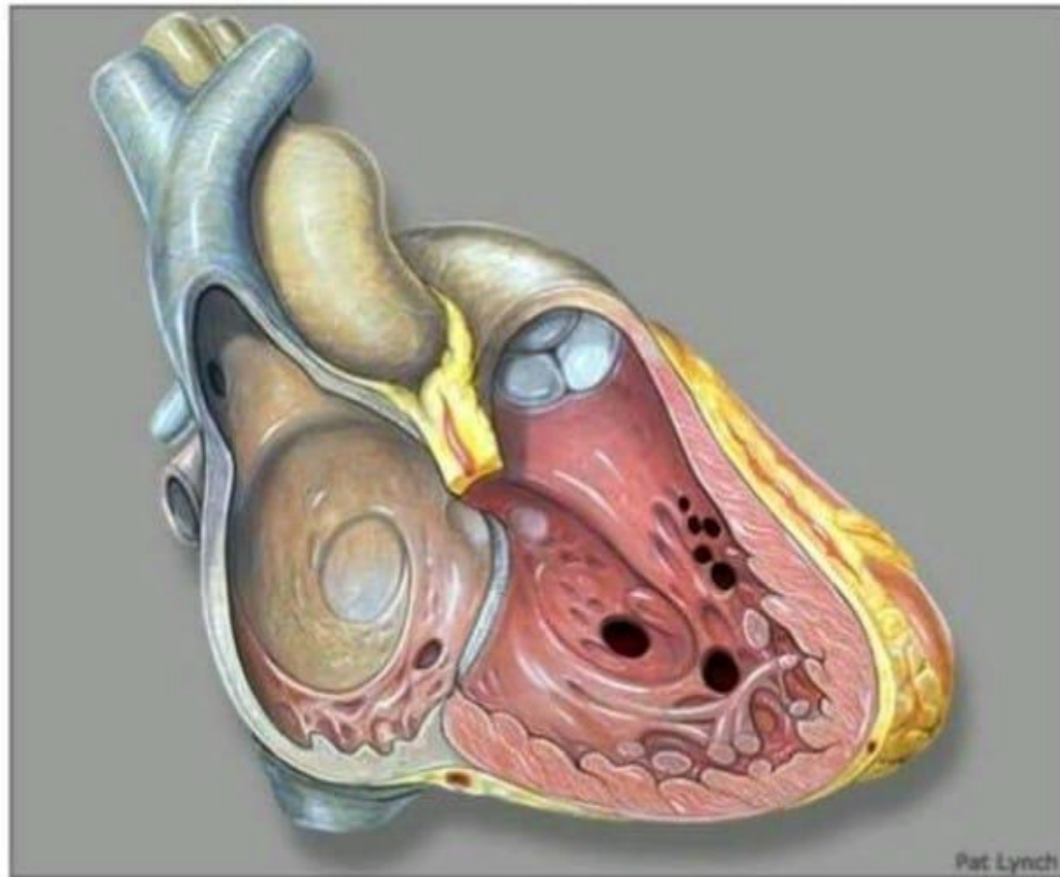
- In 90% of cases, the defect involves **membranous septum** and is very close to the bundle of His
- The remaining 10% cases have VSD **Infundibular VSD (10%)** immediately below the pulmonary valve (**sub-pulmonic**), below the aortic valve (**sub-aortic**), or exist in the form of multiple defects in the muscular septum.



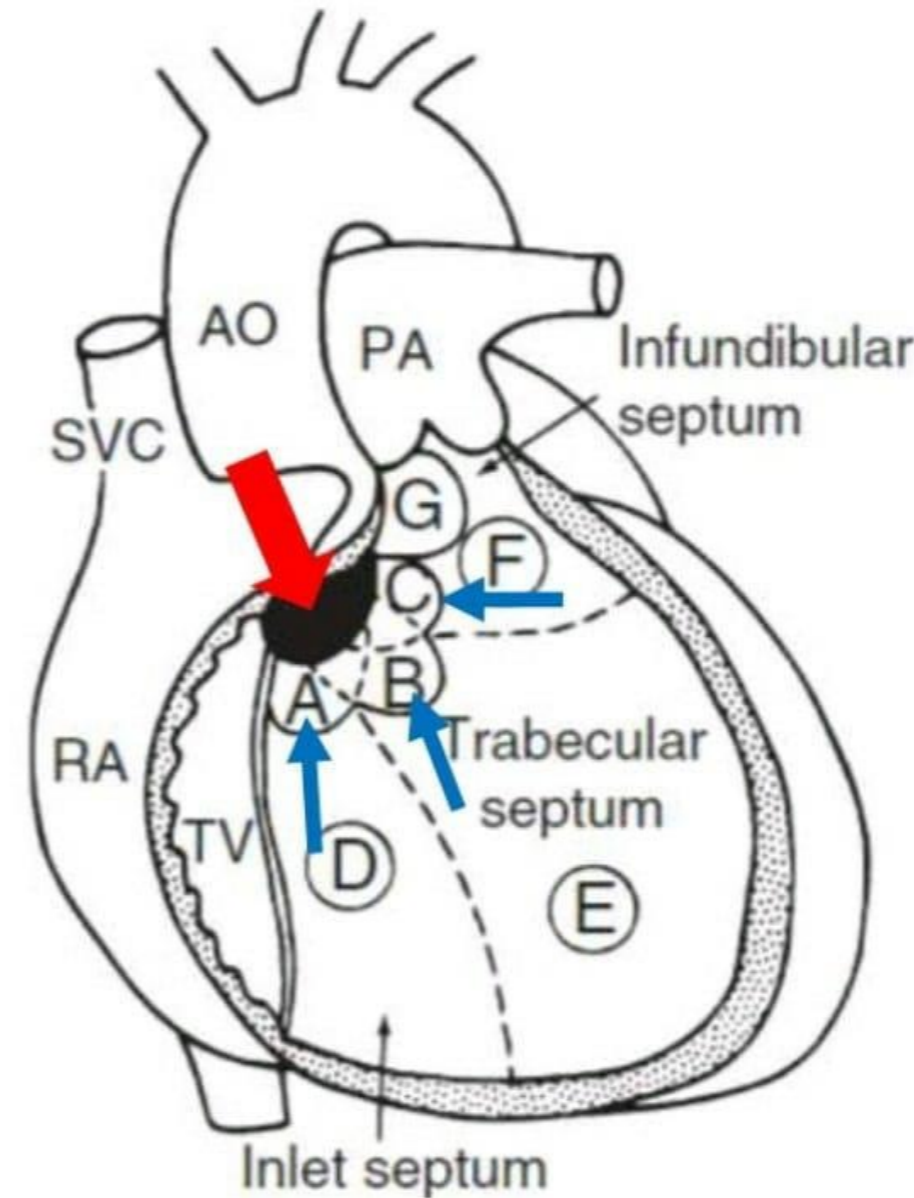
Pat Lynch

Types of VSD

Swiss-cheese VSD

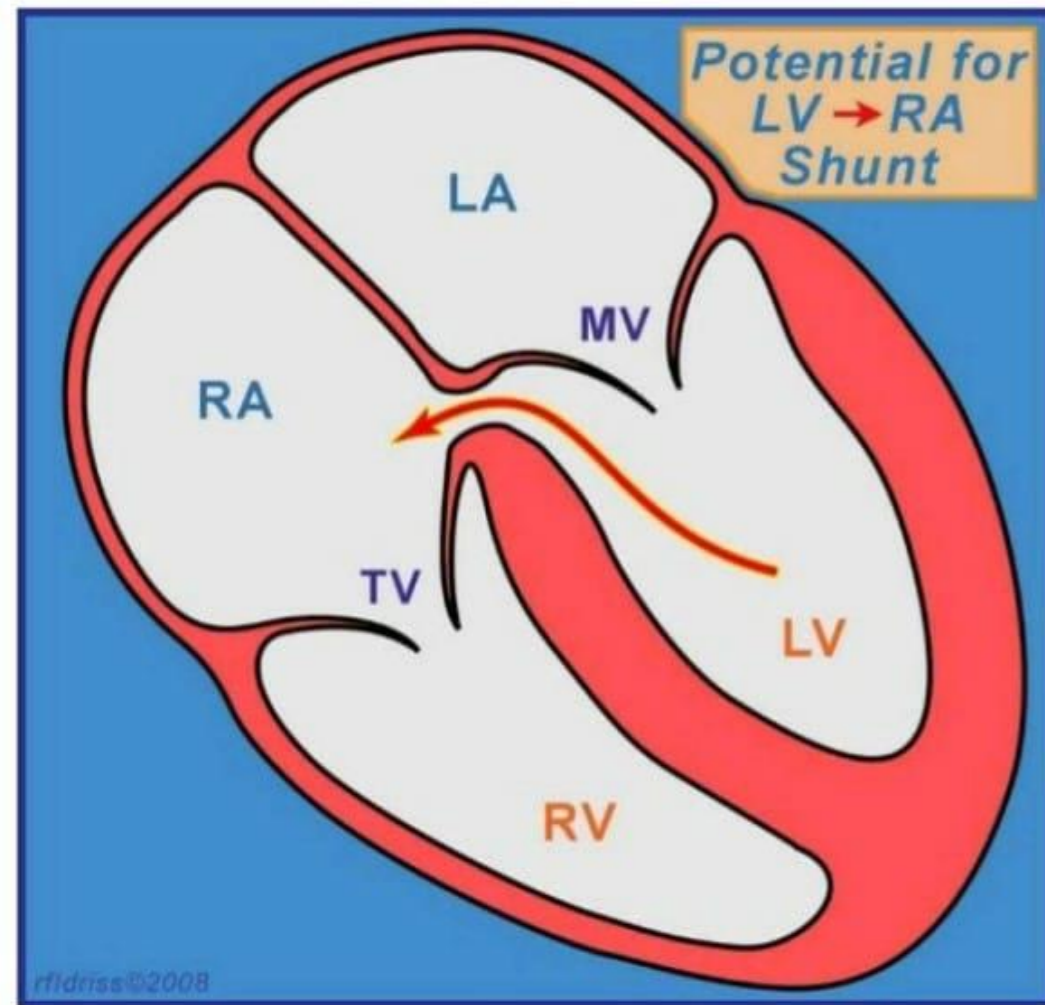


- The “Swiss cheese” type of multiple muscular defect (involving all components of the ventricular septum) is extremely difficult to close surgically.



- A. Perimembranous inlet (“AV canal-type”) VSD
- B. Perimembranous trabecular VSD
- C. Perimembranous infundibular VSD
- D. Inlet muscular VSD
- E. Trabecular muscular VSD
- F. Infundibular or outlet muscular VSD
- G. Subarterial infundibular (supracristal) VSD

Gerbode defect



- Located in the membranous portion of the atrioventricular septum.
- A Left Ventricular to Right Atrial defect.
- Uncommon, small.

The Gerbode defect is a ventriculo-atrial defect, the large systolic pressure gradient between the left ventricle and the right atrium is most likely the cause of the high velocity systolic flow from the left ventricle into the right atrium.



Pathophysiology

LV systolic pressure (120 mm Hg) is higher than the RV pressure (15 mm Hg)



Presence of VSD Leads to development of Left to Right Shunt



Overload to pulmonary circuit



And the volume overload occurs in LA and LV



Pulmonary Artery dilation occurs -----> Pulmonary pressure rises -----> PVR rises



RV and LV pressures become equal and shunt becomes reverse R to L - Eisenmenger Syndrome



Effects of VSD

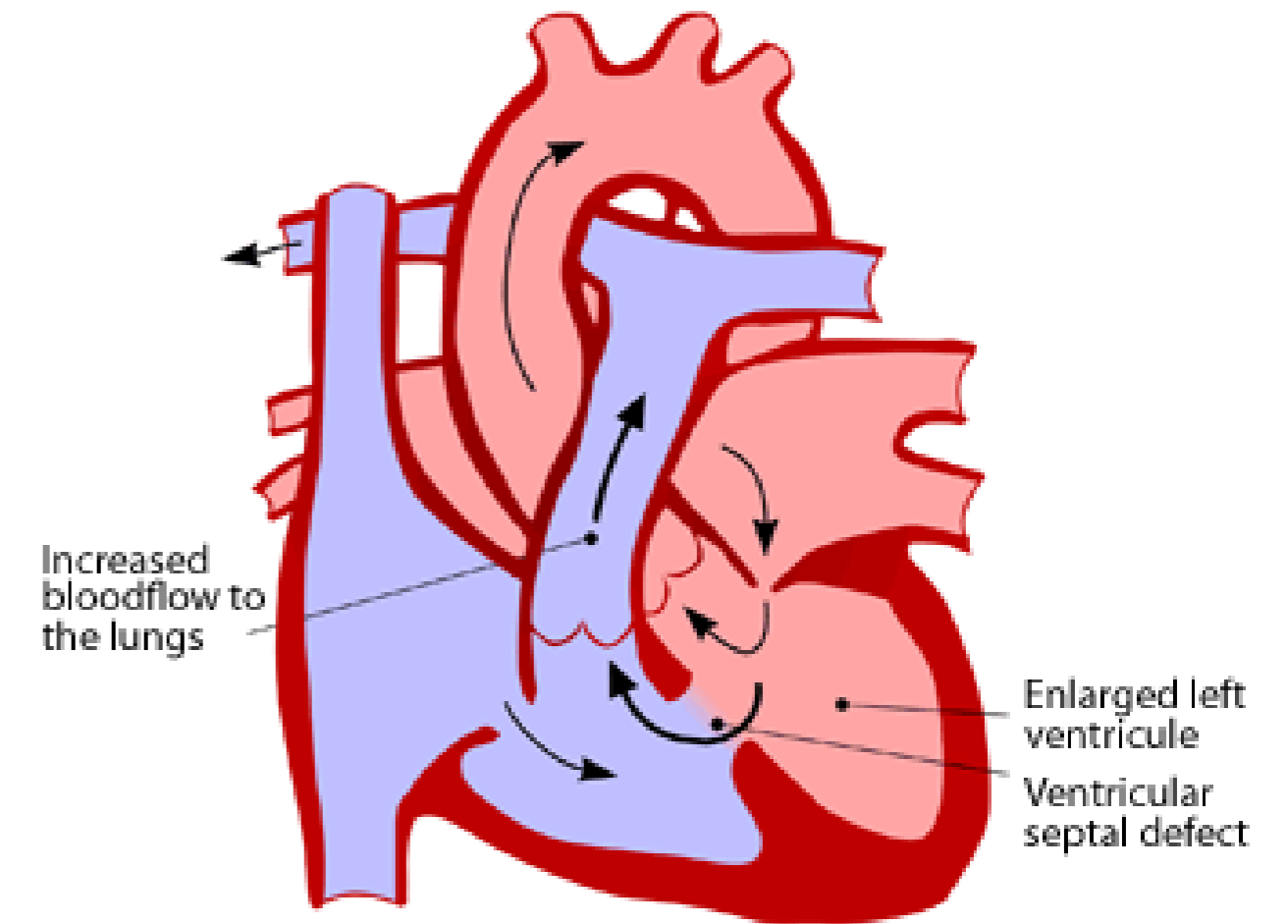


- VSD → increased pulmonary flow and increased volume in the left side of the heart

EFFECTS ON HEART

- Volume hypertrophy of RV
- Enlargement of TV & PV
- Endocardial hypertrophy of the RV
- Pressure hypertrophy of the RA

Large ventricular septal defect



ROLL OVER TO VIEW NORMAL CARDIAC ANATOMY

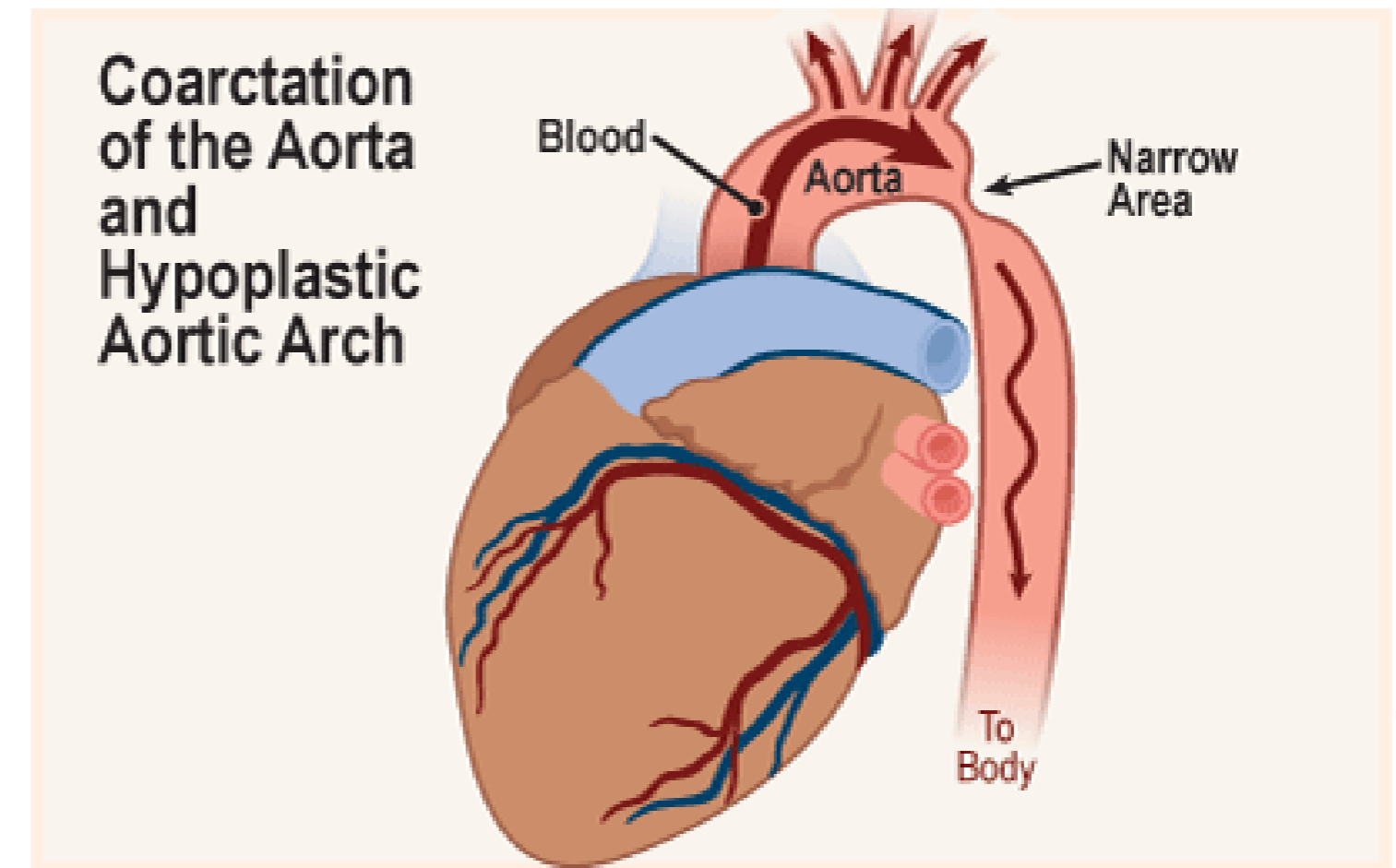


Associated Anomalies



Multiple Shunts ----- due to increased L to R Shunt

Coarctation of Aorta ----- due to increased resistance on the systemic circuit L to R shunt increases





Management



Conservative Treatment

No surgery is advised for a VSD with a diameter $<7\text{mm} / \text{m}^2$ BSA, without PH, AR or infective endocarditis

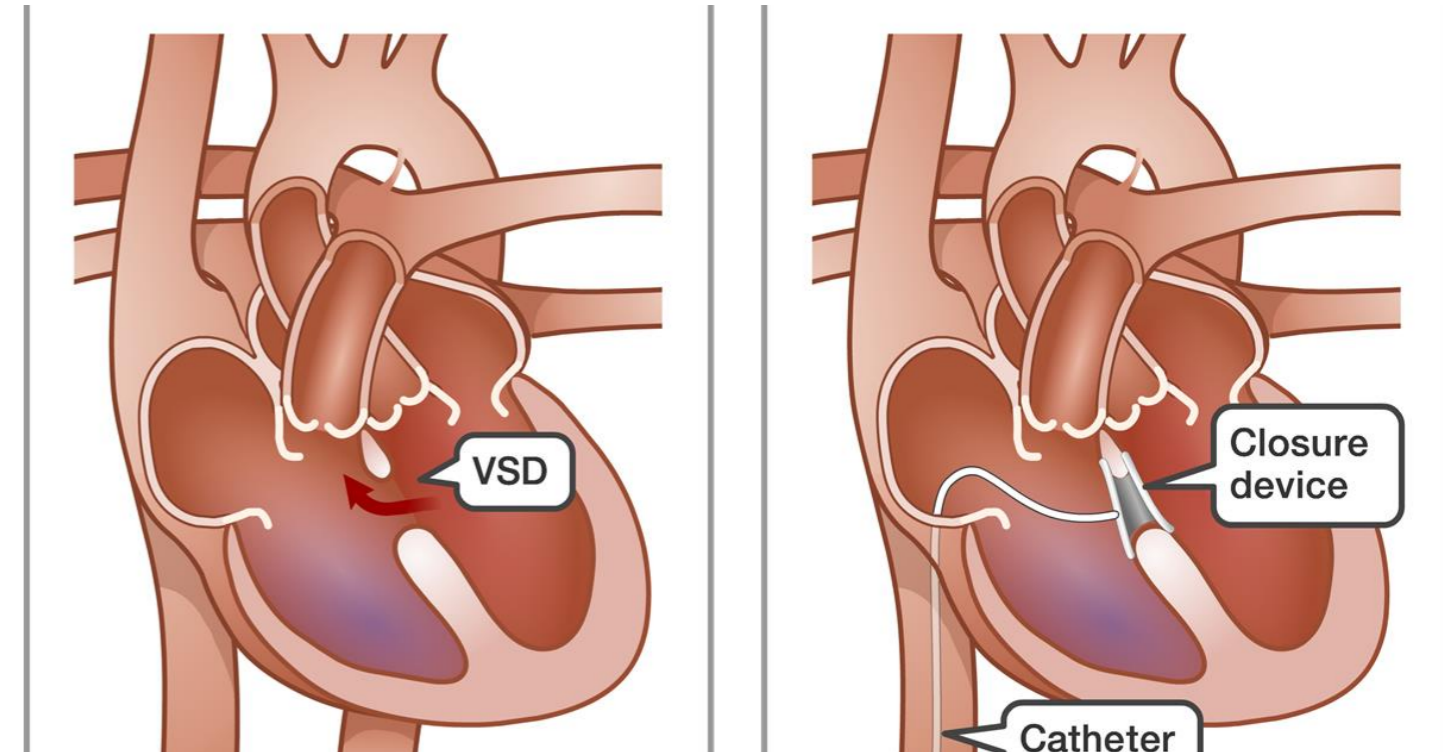
Medical Treatment

It is offered to babies who are awaiting for surgery.

Afterload decreasing agents like ACE Inhibitors, Diuretics

Decreased afterload improve LV output and reduce SVR

Interventional Treatment ---- Device closure of VSD



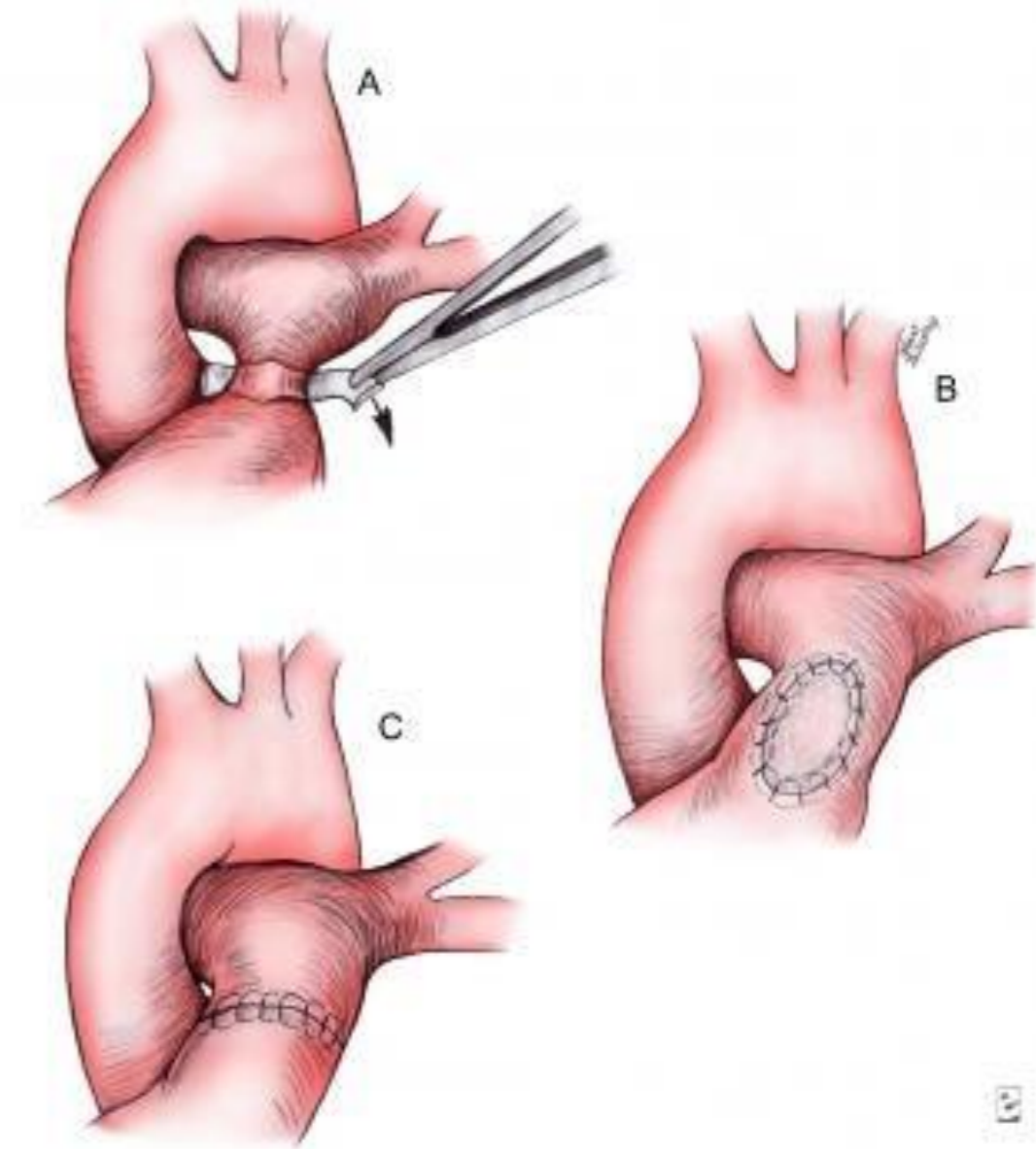


Management



Surgical Treatment - MPA Banding

- Pulmonary artery banding (PAB) creates a narrowing, or stenosing, of the main pulmonary artery (MPA) that decreases blood flow to the branch pulmonary arteries and reduces PBF and pulmonary artery pressure
- It is done for too small babies by weight and age
- It is also done for patients having multiple (Swiss Cheese Type) VSD.
- Closure of RVOT, creates an increased resistance to flow of blood from LV to PA, thus reduces the L to R shunt, and increase the forward flow through aorta.



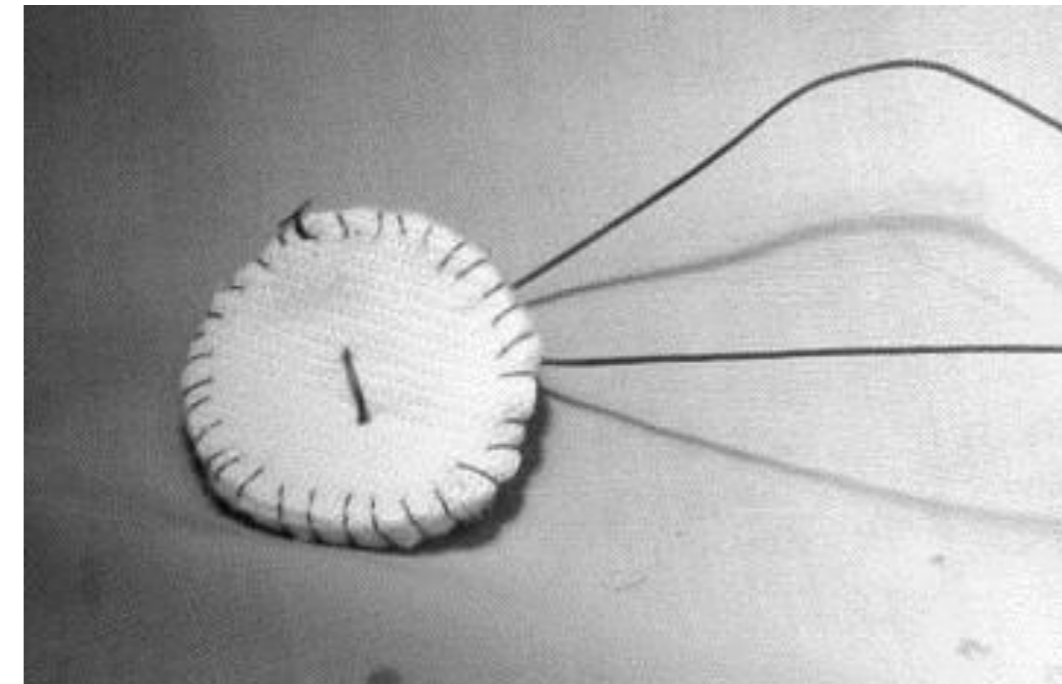
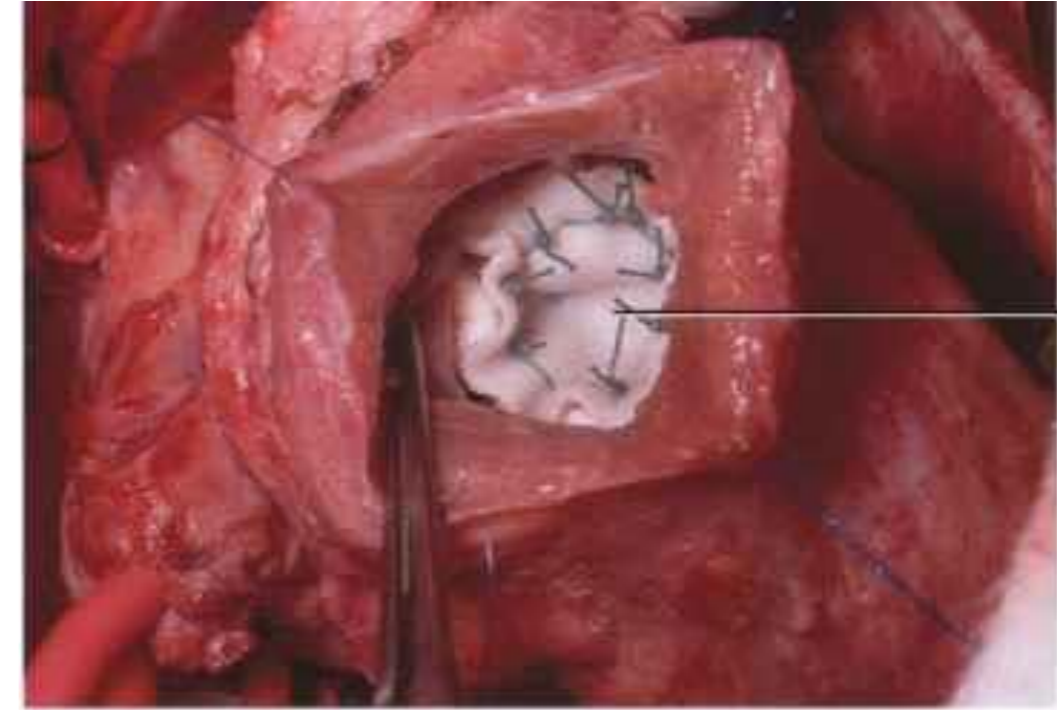


Management



VSD Closure

- Performed through median sternotomy
- VSD is approached through RA, RV, PA or aorta depending upon the situation of VSD
- VSD is closed directly if small or with a patch.
- If VSD is large, patch could be of autologous pericardium, Dacron or Poly Tetra Fluoro Ethylene



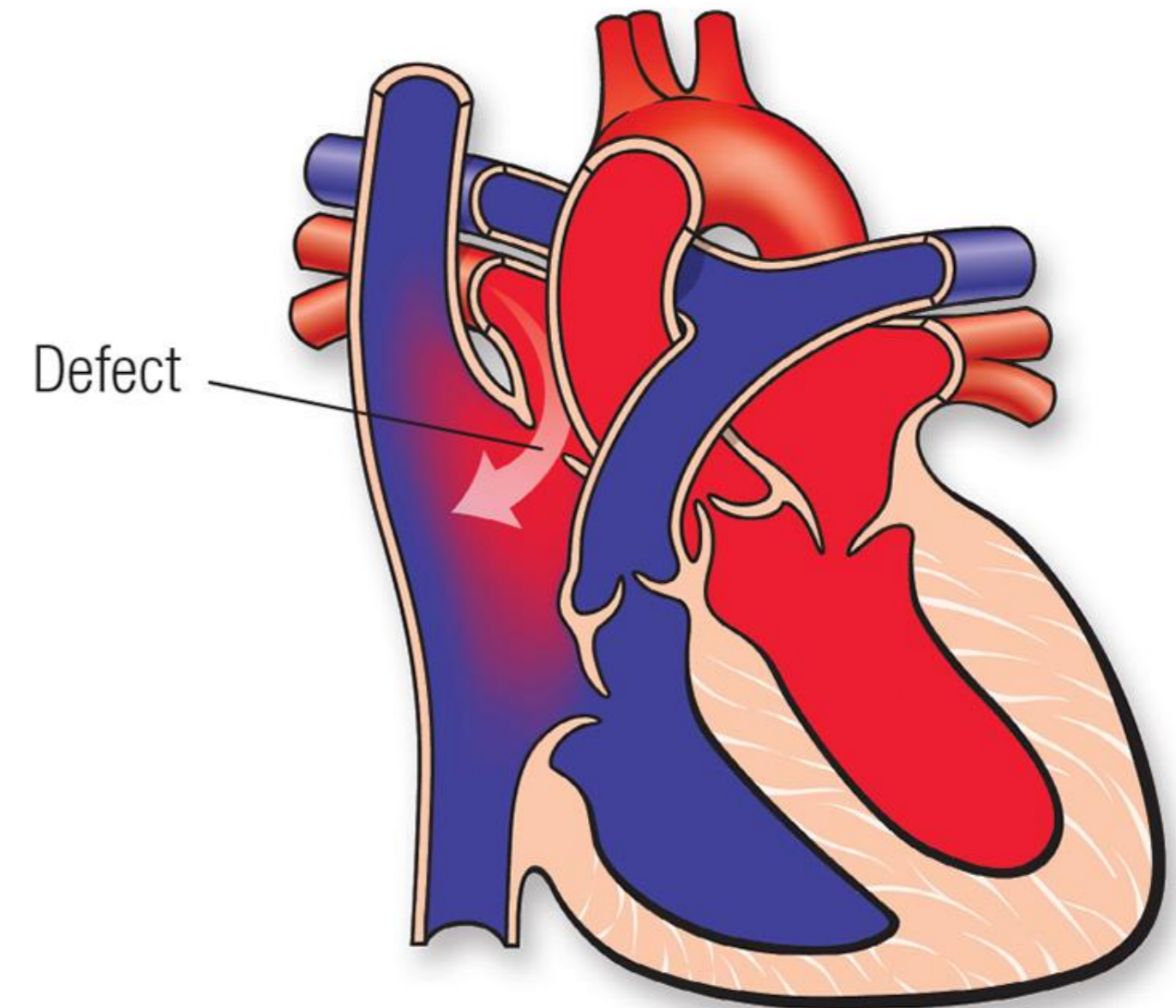


Atrial Septal Defect



- It comprises of 10% of congenital heart diseases
- The condition remains unnoticed in infancy and childhood till pulmonary hypertension is induced causing late cyanotic heart disease and right-sided heart failure.

Atrial Septal Defect





Types of ASD



Depending on location of defect, they are

- **Fossa ovalis type or ostium secundum type** is the most common form comprising about 90% cases of ASD (**located in fossa ovalis**)

- **Ostium primum type** comprises about 5% cases of ASD (**low in the interatrial septum**)



adjacent to atrioventricular valves

- **Sinus venosus type** accounts for about 5% cases of ASD (**high in the interatrial septum**)



near the entry of the superior vena cava.

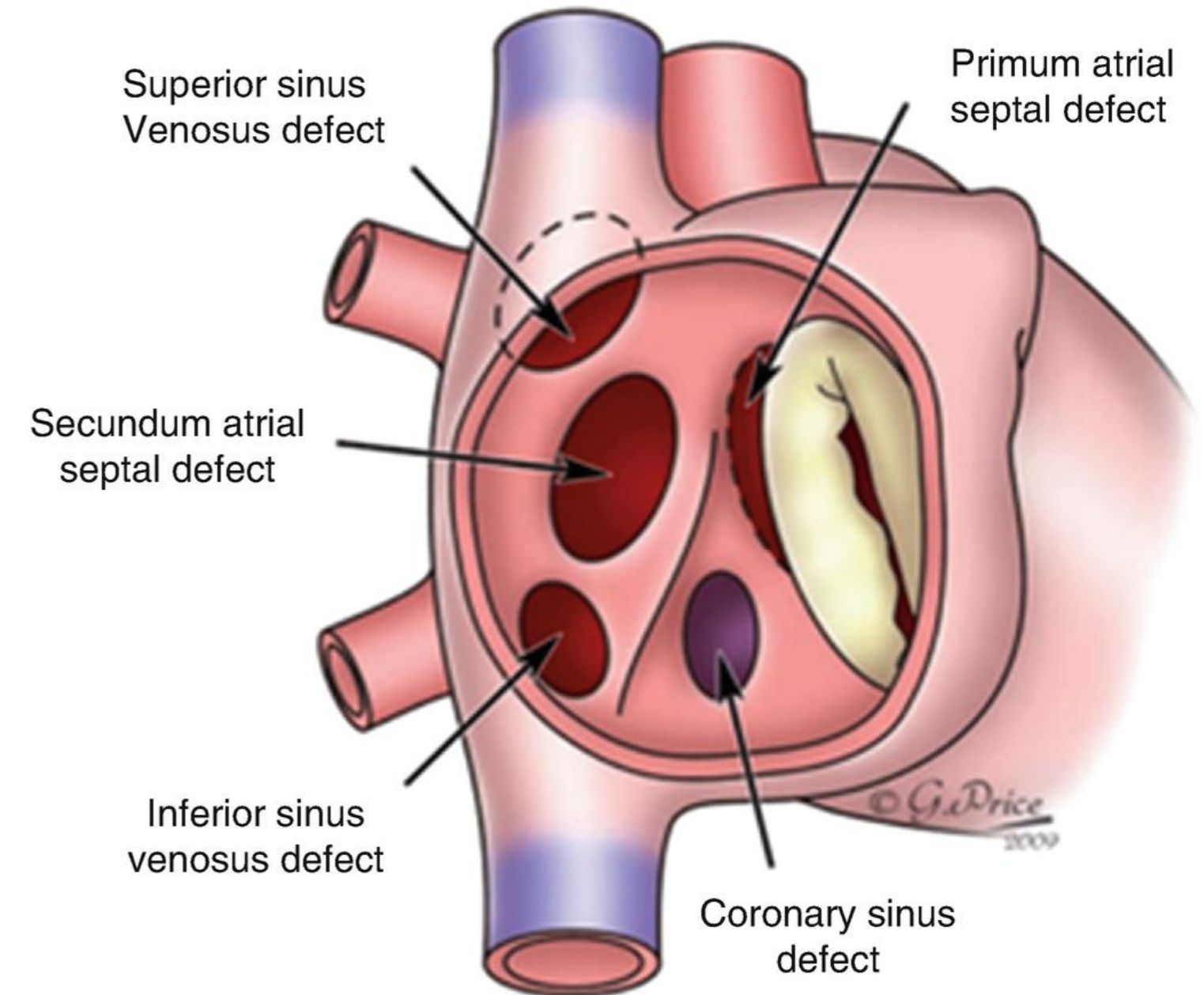


Types of ASD



The term single atrium is applied when Inter Atrial Septum (IAS) is absent

Fenestrated ASD: Some time there are multiple small (2-3 mm in diameter) ASD . Collectively they produce a significant hemodynamic load





Pathophysiology



Shunts through ASD are of two types:

Facultative Shunt ----- > It is seen in isolated ASDs. The shunt is not essential for sustaining life of the patient. The shunt is basically L to R shunts

Obligatory Shunt ----- > ASD is a part of a larger pathological complex. This shunt is essential for sustaining life of the patient.



Pathophysiology



- Defect ----- >IAS blood flows from Left Atrium to Right Atrium
- Blood flows from Left to Right,
- LA Pressure (4 – 12 mm Hg) is higher than RA Pressure (0 – 7 mm Hg)
- RV Compliance is much more than the LV Compliance, so blood will flow easily via LA-RA- RV
- The quantity of shunt is described as **Amount of blood (Q) passing through pulmonary circuit (Qp) to the amount of blood passing through systemic circuit (Qs).**
- The ratio in normal person is 1.0 : 1.0
- An ASD requiring surgery or intervention has Qp : Qs of more than 1.5 : 1.0



Pathophysiology

RA receives blood from SVC, IVC and CS

RV receives shunted blood from LV



RAVO and RVVO and volume load in PA occurs



RV dilation occurs, this increases pressure in PA



Pulmonary Hypertension occurs



Due to PH, RV has to generate more pressure to pump blood into pulmonary circuit.



Prolonged PH leads to RV Failure





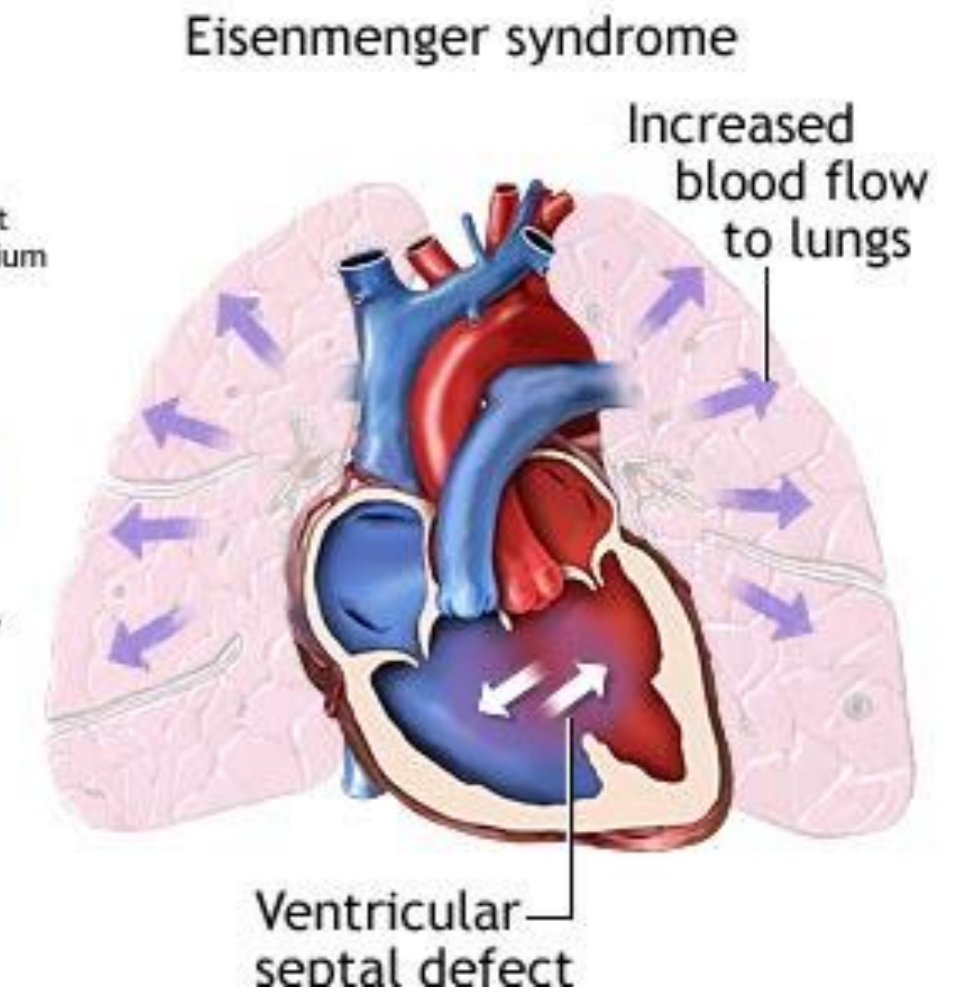
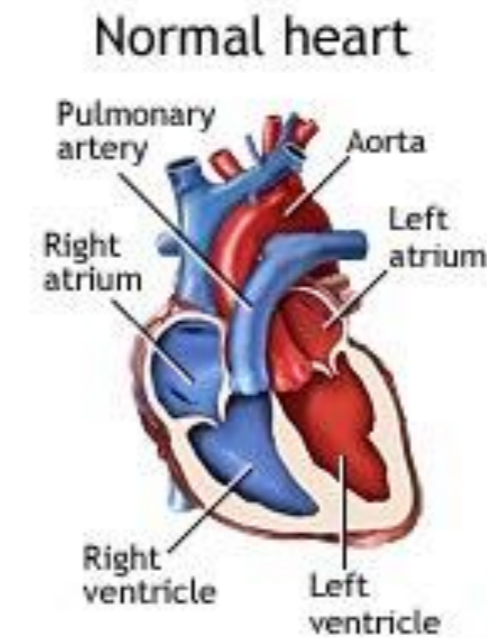
Eisenmenger Complex



• Eisenmenger syndrome is a irregular blood flow in the heart and lungs.

• This causes the blood vessels in the lungs to become stiff and narrow.

- 1 • Systemic to pulmonary circulation connection
- 2 • Left to right shunting of blood
- 3 • Increased pulmonary blood flow
- 4 • Irreversible pulmonary vascular injury
- 5 • Irreversible pulmonary vascular resistance
- 6 • Right to left shunting of blood
- Hypoxia and erythrocytosis





Associated Anomalies



Partial Anomalous Pulmonary Venous Connection (PAPVC)

- Associated with superior sinus venosus (SV ASD)
- One or two right pulmonary vein drain anomalously into RA or SVC

Mitral Valve Disease (called Leutembacher's syndrome) : Secundum ASD is associated with mitral stenosis or regurgitation

Multiple Shunts : ASD associated with additional VSD or PDA

Valvular Pulmonary Stenosis: In PS, the shunt through ASD is usually left to right, but when RV fails, shunt decreases and later becomes right to left

Left SVC : An additional L SVC producing bilateral SVC is found in many cardiac anomalies with bridging left innominate vein



Management



Medical:

- Drug therapy is mainly for symptomatic purposes
- Antibiotics: to control frequent lower respiratory infections (LRTI) which patients with ASD have due to excessive pulmonary flow

Interventional:

- Less than 2cm in diameter
- Has well defined margins around
- Has anomalies which can be treated by intervention (Absence of PAPVC & MR)

Surgical Management:

- Patch will be placed based on the size of the ASD - Autologous Pericardium, Dacron cloth or PTFE



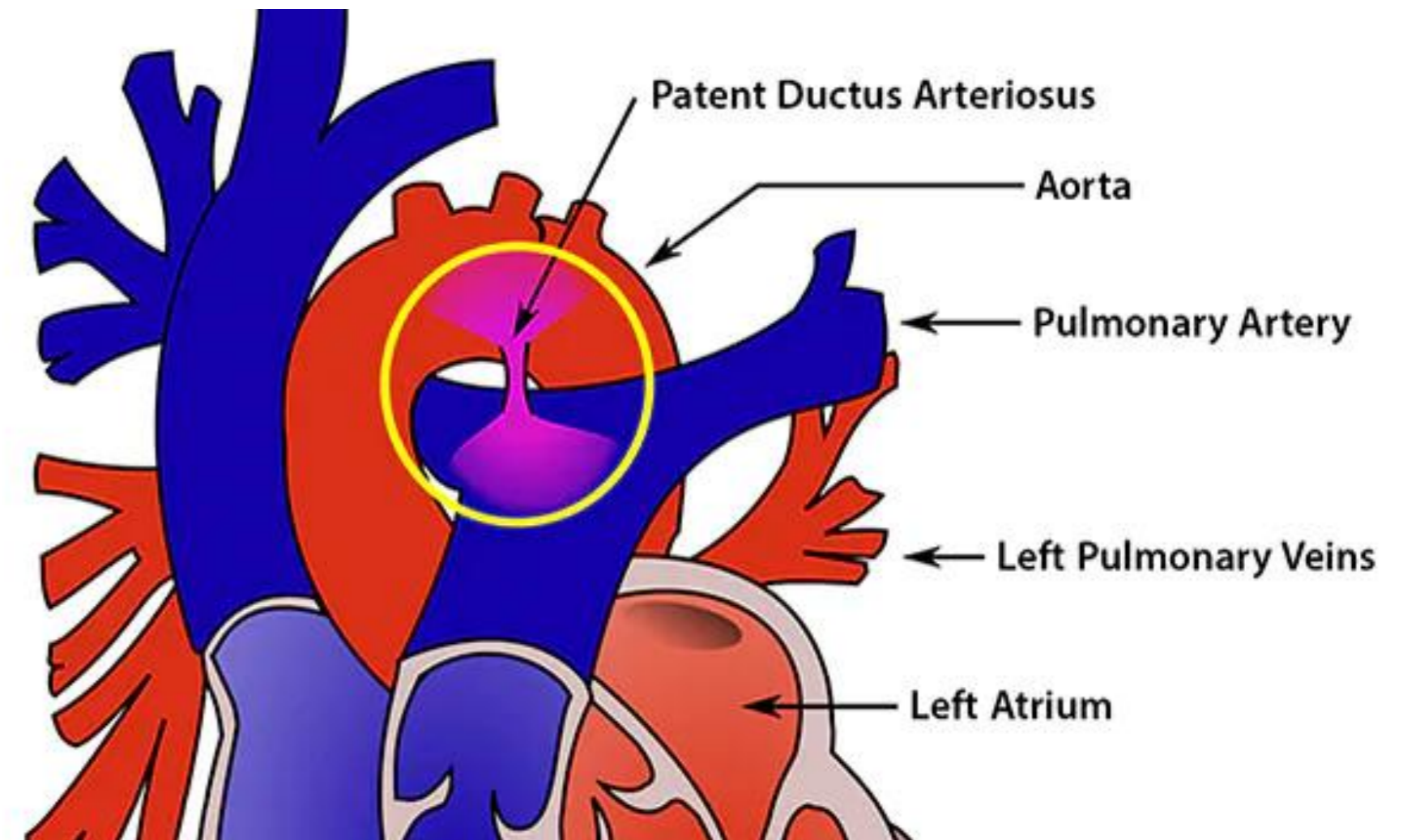
PDA



- The **ductus arteriosus** is a normal vascular connection between the aorta and the bifurcation of the pulmonary artery.
- Normally, ductus closes functionally within the first or second day of life
- More than 3 months of age ---- its abnormal

Etiology:

- The cause for patency of ductus arteriosus is not known, but possibly it is due to continued synthesis of PGE₂ after birth

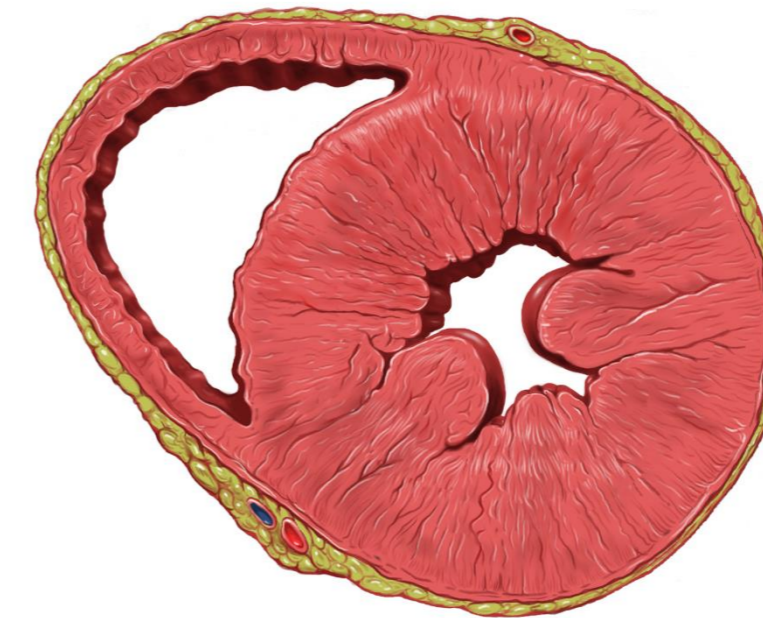




Morphological Changes in PDA



- Volume hypertrophy of the LA & LV
- Enlargement of the ascending aorta
- Enlargement of MV and PV

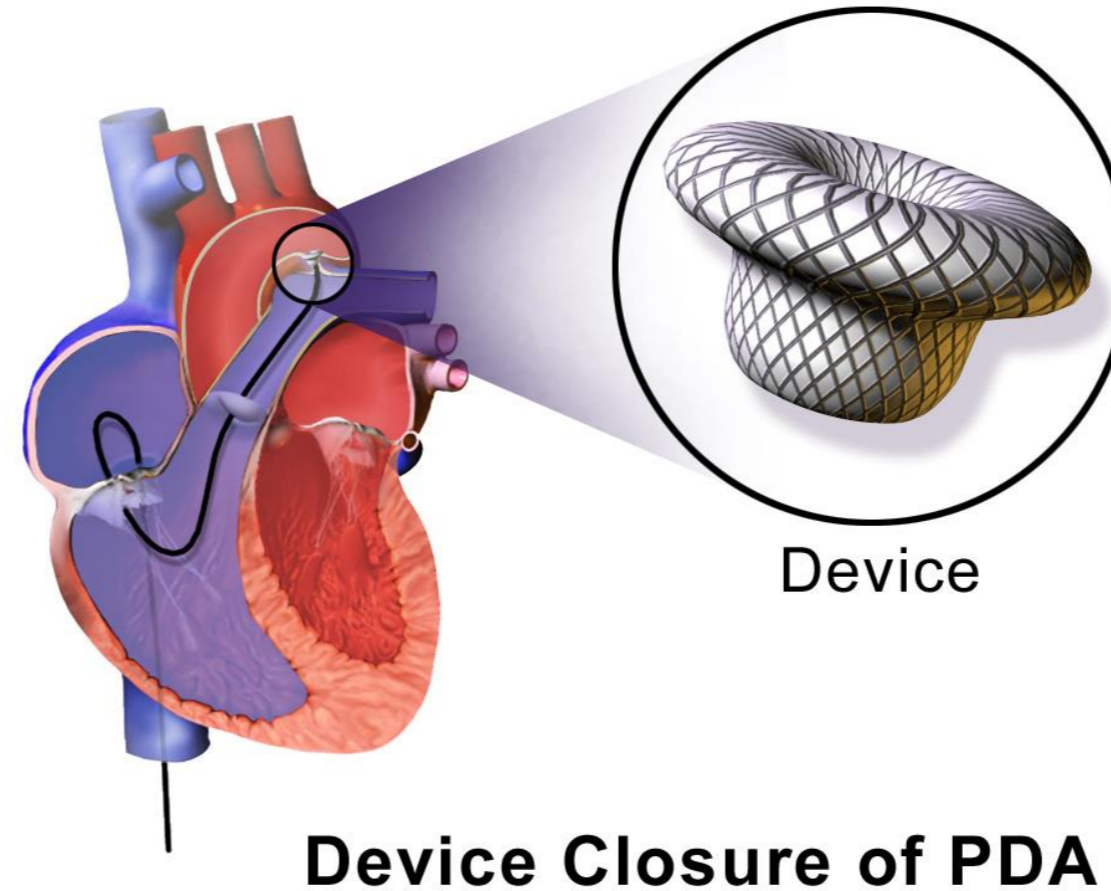




PDA



- Patient experience respiratory distress syndrome
- Pharmacologic closure of PDA with administration of indomethacin to suppress PGE₂ synthesis





Thank You



References:

Text book of pathology – Harsh Mohan

Principles of Cardiopulmonary Bypass – Medtronic MK. Brian