Congenital Heart Diseases

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Definition

Abnormalities of cardiac structure that are present from birth.

Such developmental abnormalities of the heart typically arise in the third to eighth week of gestation.

The first operation for congenital heart disease was patent ductus arteriosus (PDA) ligation by Gross in 1938.

With the development of neonatal CPB, improved myocardial protection and microsurgical techniques, an increasing number of corrective and palliative operations are possible.

Incidence Cardiac defects

the most common congenital abnormalities in the UK; the incidence of significant cardiac abnormalities is 8 per 1000 live births.

In neonates and children with congenital heart disease, 15% will have more than one cardiac abnormality and 15% will have another extracardiac abnormality.

how many live births happen in Iraq per day

According to demographic data based on **United Nations World Population Prospects**, Iraq has about **1,177,772 births per year**, which works out to roughly **3,227 live births per day**

How many CHD in IRAQ per year ?

How many from CHD need Surgery ?

Do you think surgery in Iraq cover all babys ?

Development of the heart and fetal circulation and circulatory changes at birth

By 12 weeks of fetal life the primitive vascular tube is fully developed

Fetal circulation differs from that of the adult in that the right and left ventricles pump blood in parallel rather than in series.

This arrangement allows the heart and head to receive more highly oxygenated blood.

This is possible because of the presence of three structural shunts:

- the ductus venosus,
- foramen ovale
- ductus arteriosus

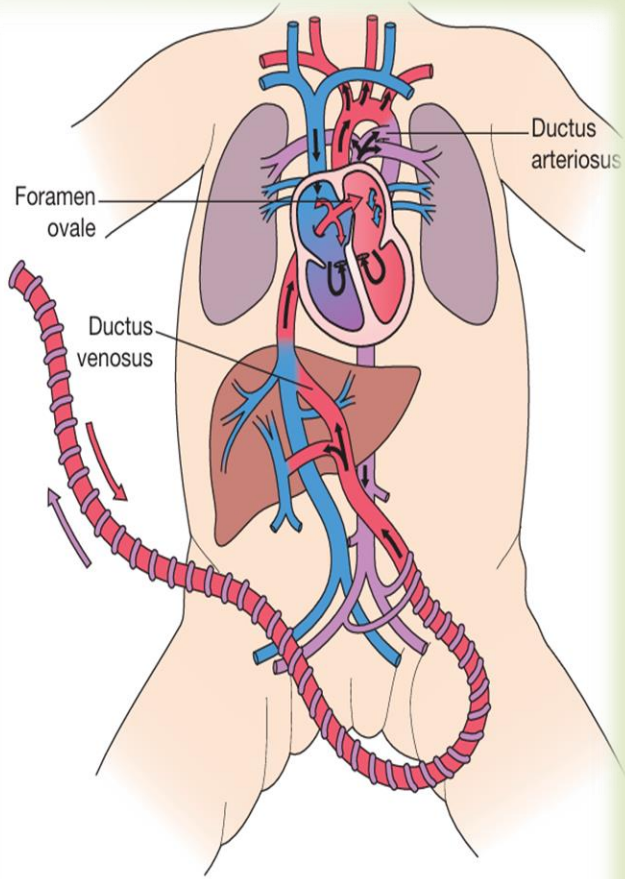


Figure 59.21 Fetal circulation.

After birth, cutting and tying of the umbilical cord stops venous blood flow from the placenta.

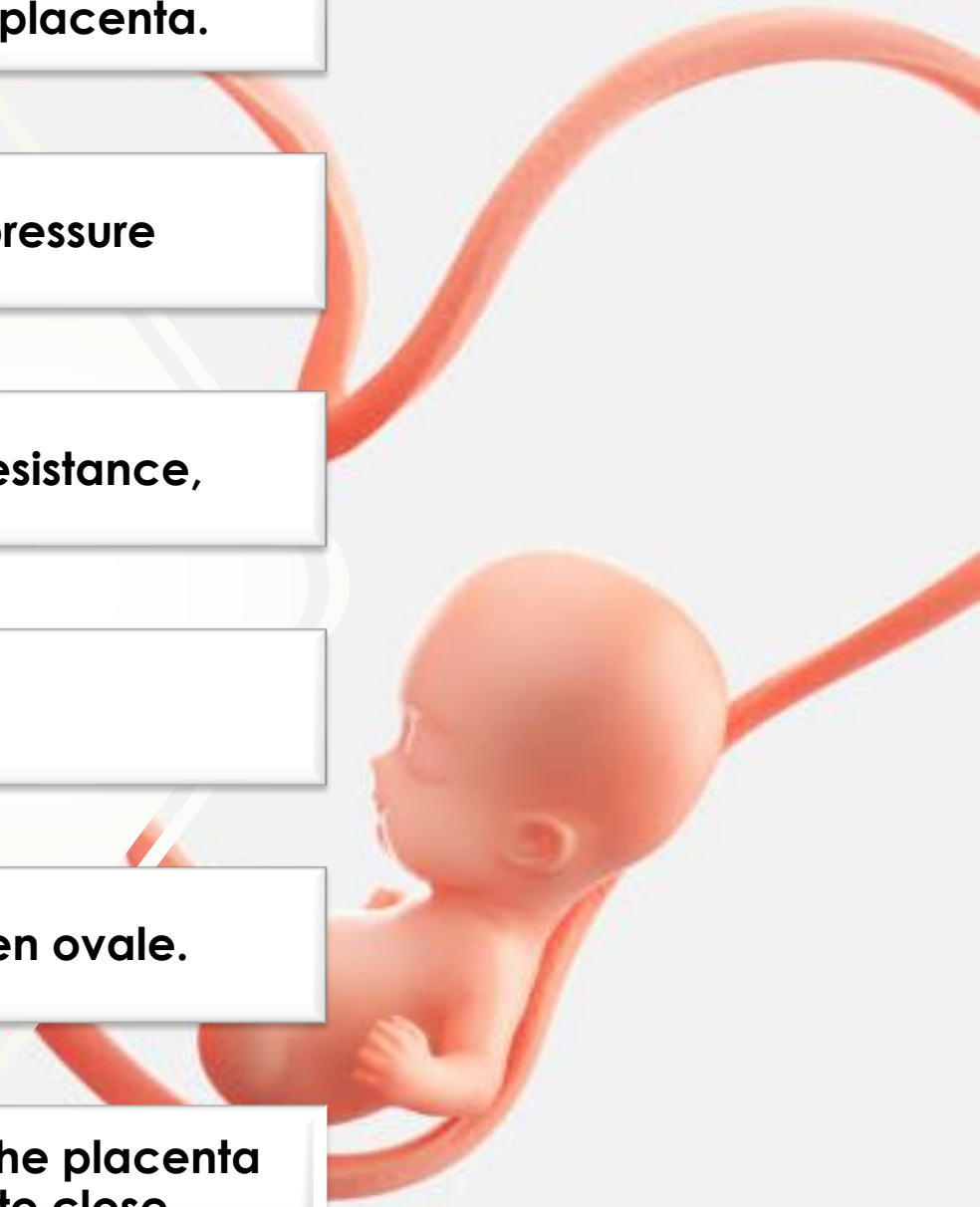
This lowers inferior vena cava pressure

with falling pulmonary vascular resistance,

right atrial pressure falls.

The result is closure of the foramen ovale.

The stopping of venous return from the placenta also causes the ductus venosus to close.



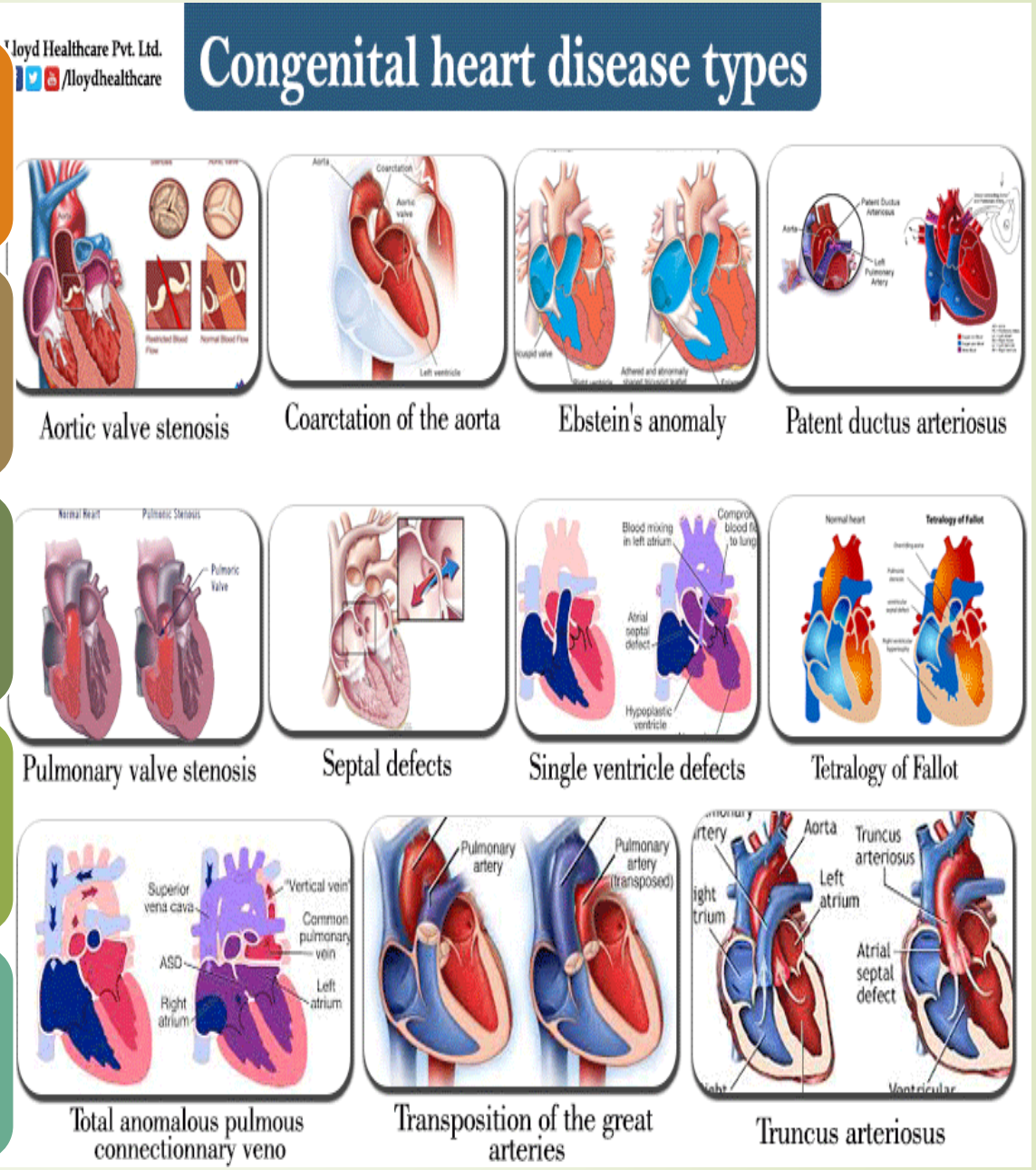
Abnormalities of cardiac structure may arise from the

persistence of normal fetal channels (PDA, patent foramen ovale)

failure of septation (atrial septal defect [ASD], VSD, tetralogy of Fallot)

stenosis (intracardiac, supravalvular, valvular, infravalvular or extracardiac coarctation of the aorta)

atresia or abnormal connections (transposition of the great vessels



Causes Recognized associations with congenital heart disease

Maternal (environmental) factors

- Infection: rubella
- Disease: systemic lupus erythematosus, diabetes mellitus, maternal phenylketonuria

Genetic factors

- **Drugs/medications**: alcohol abuse, warfarin, phenytoin, lithium, thalidomide
- **Single gene defects**: Marfan, Noonan and Holt–Oram syndromes; numerous single-gene disorders
- **Chromosomal defects**: trisomy 21 (Down syndrome), trisomy 18 (Edwards syndrome), trisomy 13 (Patau syndrome), Turner syndrome, Klinefelter syndrome
- **Deletions**: DiGeorge and Williams syndromes

Classification Congenital heart disease

Cyanotic congenital heart diseases make up 25% of cases (8 or 9/1000 live births) and are usually more complex, although they do include simple defects.

Classification of CHD

Cyanotic Heart Disease

- Decreased pulmonary flow:
  - Tetralogy of Fallot
  - Tricuspid atresia
  - Other univentricular heart with pulmonary stenosis.
- Increased pulmonary flow:
  - Transposition of great arteries
  - Total anomalous pulmonary venous return.

Acyanotic Heart Disease

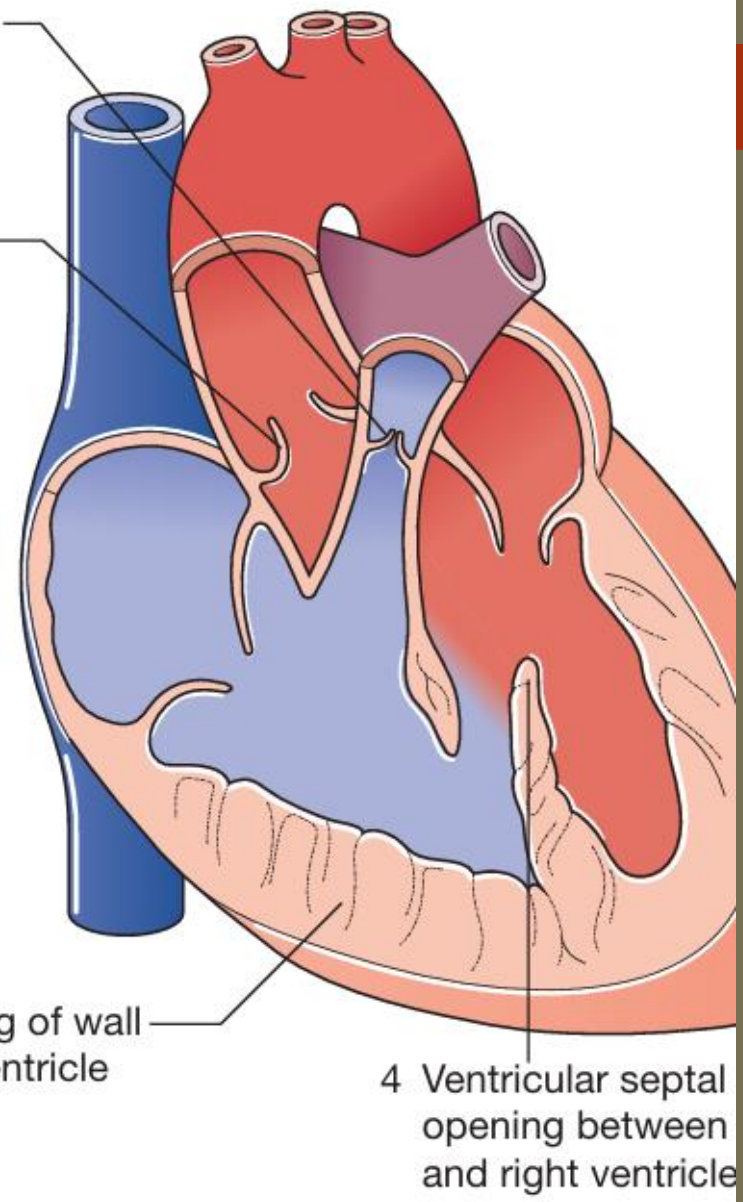
- Left – Right shunt lesions:
  - Ventricular septal defect
  - Atrial Septal Defect
  - Atrio-ventricular Septal Defect
  - Patent Ductus Arteriosus
- Obstructive lesions:
  - Aortic stenosis
  - Pulmonary valve stenosis
  - Coarctation of Aorta

	Cyanotic	A cyanotic
Definition	CHD with systemic desaturation due to mixing of deoxygenated blood	CHD with normal systemic oxygenation initially
Cyanosis	Present (central)	Absent initially
Type of shunt	Right → Left or parallel circulation	Left → Right
Arterial O_2 saturation	↓ (<85–90%)	Normal
Pulmonary blood flow	↓ (TOF) or ↑ (TAPVR)	↑
Heart failure	Less common early	Common
Chest infections	Uncommon	Common
Clubbing	Common (chronic)	Rare
Polycythemia	Common	Absent
Squatting	Typical (TOF)	Absent
Main complication	Brain abscess, stroke, endocarditis	Pulmonary HTN, Eisenmenger
Reversibility	Often fixed anatomy	May reverse (Eisenmenger)
Prostaglandin E1	Often lifesaving	Rarely needed

Cyanotic congenital heart diseases

Cyanotic congenital heart diseases involve defects that cause **poorly oxygenated blood to mix with oxygenated blood**, resulting in low oxygen levels in the blood and tissues. These conditions often present with cyanosis (a bluish tint to the skin and mucous membranes).

- **Tetralogy of Fallot (TOF)**:
  - **Ventricular Septal Defect (VSD)**: A hole between the ventricles.
  - **Pulmonary Stenosis**: Narrowing of the pulmonary valve, obstructing blood flow to the lungs.
  - **Overriding Aorta**: The aorta is positioned over the VSD, allowing oxygen-poor blood to enter the systemic circulation.
  - **Right Ventricular Hypertrophy**: Thickening of the right ventricle due to increased workload.
- **Transposition of the Great Arteries (TGA):**
  - The aorta and pulmonary artery are switched, causing oxygen-poor blood to be pumped to the body and oxygen-rich blood to be pumped back to the lungs.
- **Tricuspid Atresia:**
  - Absence of the tricuspid valve, obstructing blood flow from the right atrium to the right ventricle.
  - Blood flows through an ASD and then through the left side of the heart, mixing oxygen-poor and oxygen-rich blood.
- **Total Anomalous Pulmonary Venous Return (TAPVR):**
  - The pulmonary veins do not connect normally to the left atrium, causing oxygen-rich blood to mix with oxygen-poor blood in the right side of the heart.
- **Hypoplastic Left Heart Syndrome (HLHS):**
  - Underdevelopment of the left side of the heart, resulting in a weak left ventricle that cannot effectively pump blood to the body.
  - The right side of the heart must pump blood to both the lungs and the body, often through a PDA or ASD.

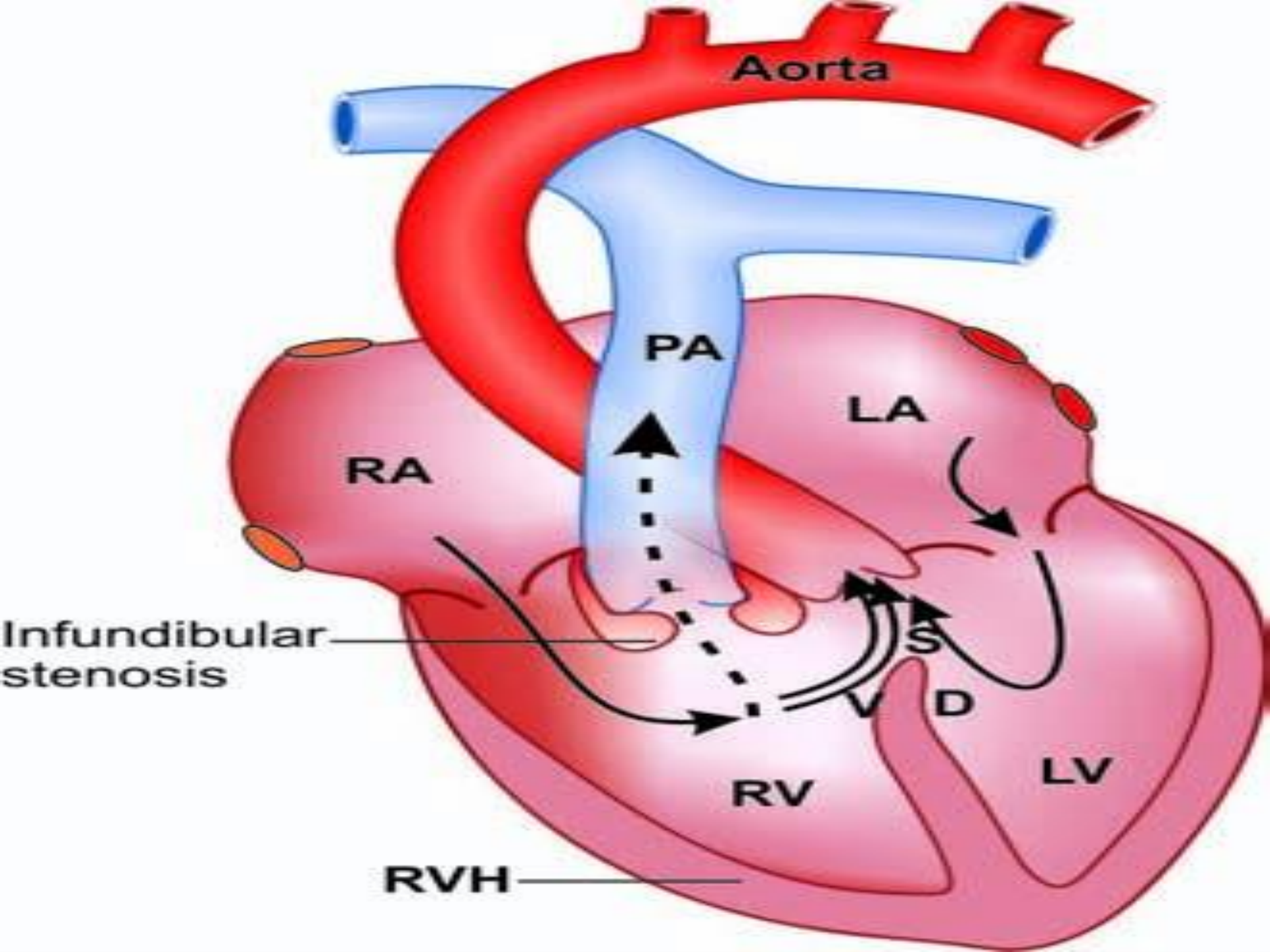


s tetralogy. Four abnormalities that result i
blood being pumped to the body.

Tetralogy of Fallot

This is the most common cyanotic congenital heart disease in children surviving to 1 year and accounts for about 4–6% of all congenital heart diseases. The four intracardiac lesions originally described

- **VSD**
- **overriding aorta**
- **pulmonary (infundibular or sub pulmonary) stenosis**
- **right ventricular hypertrophy**


Aorta
PA
LA
RA
Infundibular stenosis
S
V
D
RV
LV
RVH

- It is a cyanotic congenital heart disease.

- Components:
  - Ventricular septal defect
  - Infundibular and pulmonary stenosis causing right ventricular outflow obstruction
  - Right ventricular hypertrophy
  - Dextroposition of aorta

- Because of RV outflow obstruction, venous blood is diverted through the existing VSD in to the LV and so to the aorta.

- So systemic circulation is provided with unoxygenated venous blood causing cyanosis.

Clinical Features:

- Cyanosis with dyspnoea. Ejection systolic murmur which diminishes during cyanotic attack.
- Echocardiography, cardiac catheterization are the essential investigations.

Timing of Surgery

Patient	Recommended Approach
Stable infant	**Primary total repair** at **3–6 months**
Severe cyanosis / hypercyanotic spells	**Early repair** or **palliative shunt**
Low birth weight / small PAs	**Staged approach**

Definitive (Total) Surgical Repair – Gold Standard

VSD closure

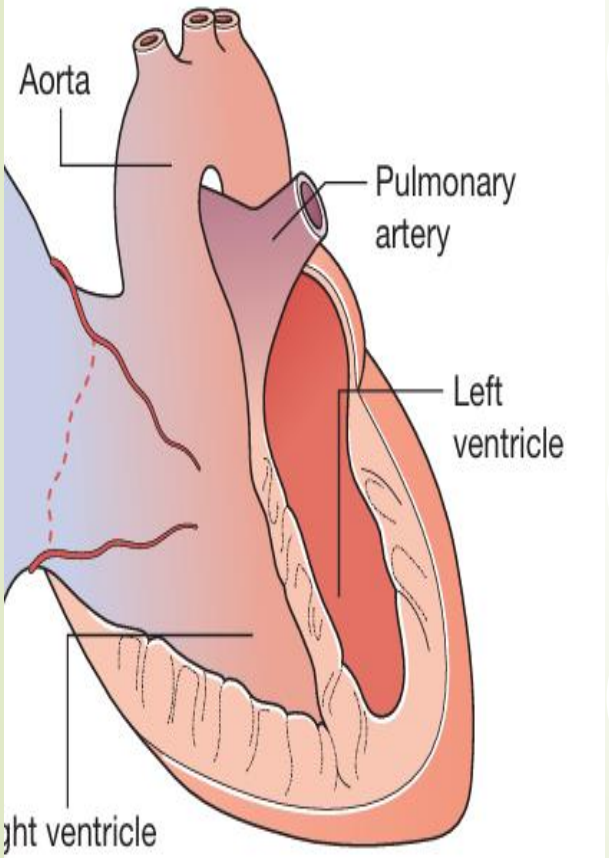
- Patch closure (Dacron / PTFE)
- Routed LV → aorta

Relief of RVOT obstruction

- Infundibular muscle resection
- Pulmonary valvotomy or valvectomy
- RVOT patch or **Transannular patch (TAP)** if needed

Pulmonary artery reconstruction (if hypoplastic)

Transposition of the great vessels



sition of the great vessels.

This is the second most common cyanotic congenital heart disease TGV results from abnormal development, with the aorta arising from the right ventricle and the pulmonary artery from the left ventricle

The situation is incompatible with life and mixing of the Patients often present with severe central cyanosis occur ring within 48 hours of birth.

The resulting transposition causes pulmonary and systemic circulations to run in parallel rather than in series; oxygenated pulmonary venous blood returns to the lungs and desaturated systemic venous blood is pumped around the body.

The chest radiograph shows pulmonary plethora, with the heart having an ‘egg on its side’ appearance

Many infants will die without treatment within 1 month of birth. Initial stabilization can be achieved by performing percutaneous balloon septostomy to increase the systemic arterial oxygen saturation. Arterial switch repair is currently the standard operation and is typically carried out within the frst few weeks of life. Long-term outcomes of the operation are excellent and many patients achieve good exercise tolerance; however

TREATMENT

Immediate palliative method is Rashkind balloon septostomy.

- A balloon is passed in to the RA and through the septum in to the LA. Balloon is inflated and rapidly pulled out through the atrial septum causing widening of the defect in the septum. It allows flow of oxygenated blood from pulmonary veins through the septum and so to the RV and aorta.

Total anomalous pulmonary venous drainage TAPVD

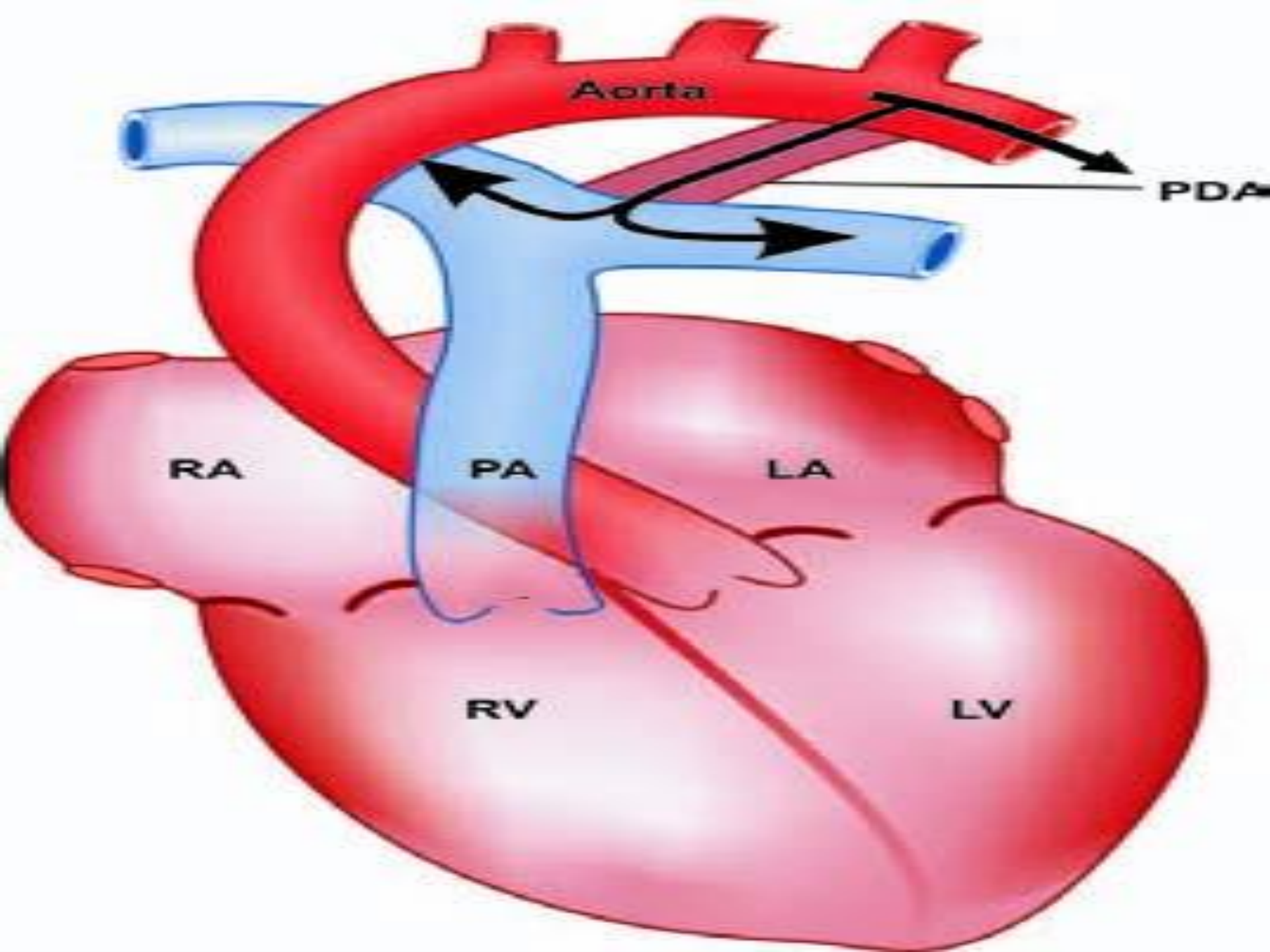
- **accounts for l–2% of congenital heart disease.**
- **In TAPVD, the pulmonary venous drainage has disconnected from the left atrium and drains into the systemic**
- **TAPVD presents after the first week of life with cyanosis that is mild to moderate depending on pulmonary flow. Infants with high pulmonary flow develop cardiac failure, recurrent chest infections, failure to thrive and feeding difficulties.**
- **Echocardiography and cardiac (pulmonary) angiography are necessary to confirm the diagnosis and delineate the anomalous drainage.**
- **The surgical principle is to re-establish the pulmonary venous drainage into the left atrium. The exact operative technique depends on the anatomy and type of TAPVD.**
- **The long-term results for survivors of the operation are generally good. Late death following repair is uncommon**

Patent ductus arteriosus(PDA 10%)

- The ductus arteriosus, a normal fetal communication, facilitates the shunting of oxygenated blood from the pulmonary artery to the aorta, away from the lungs.
- Normally, functional closure of the ductus occurs within **a few hours of birth;** it is abnormal if it persists beyond the neonatal period.
- The ductus closes in response to:

1. an increase in peripheral oxygen saturation
2. a drop in the resistance of the pulmonary circulation as the lungs expand; this causes the ductal tissue to contract through a prostaglandin inhibition mechanism.

- A cyclo-oxygenase inhibitor (e.g. **indomethacin)** may be used therapeutically to close the ductus in the first few weeks of life. In premature babies the ductus is more likely to remain patent for longer or permanently. In the isolated case of PDA,
- the **continuous machinery murmur in the left second intercostal space**.


Aorta
PDA
RA
PA
LA
RV
LV

Larger ducts cause **CARDIAC FAILURE** and can uncommonly lead to shunt reversal The diagnosis is best confirmed by **ECHOCARDIOGRAPHY** with colour flow Doppler imaging.

- After 6 months of age, PDA closure is rare. Most should be closed by preschool age, regardless of symptoms, if the risks of infective endocarditis, left ventricular failure or, rarely, Eisenmenger syndrome are to be avoided.
- In the adult, surgical treatment is indicated if there is a persistent left-to-right shunt, even in the presence of reversible pulmonary hypertension.
- The PDA may be treated by **PERCUTANEOUS INTERVENTIONAL CARDIOLOGY TECHNIQUES USING AN UMBRELLA OR COIL DUCT OCCLUSION DEVICE.**
- If the PDA is very large or the patient very small, surgical closure via a left thoracotomy is preferred. This can be accomplished by either ligation or division of the PDA.
- The operative mortality rate is low and outcome generally very good.

Indications for Surgical Closure of Patent Ductus Arteriosus (PDA)

1. Hemodynamically Significant PDA: Left ventricular volume overload, Left atrial and left ventricular dilatation on echocardiography, Cardiomegaly on chest X-ray, Even if symptoms are mild, significant shunt physiology warrants closure to prevent long-term complications.

2. Heart Failure: Congestive heart failure not responding adequately to medical therapy, Tachypnea, feeding difficulty, sweating, or poor weight gain in infants

3. Pulmonary Hypertension (Reversible): PDA should be closed when pulmonary hypertension is **present but still reversible:** Early closure prevents progression to Eisenmenger syndrome,

4. Failure to Thrive: Poor growth and inadequate weight gain due to chronic volume overload, Especially important in infants and young children

5. Recurrent Respiratory Tract Infections : Frequent chest infections due to pulmonary overcirculation: (Common indication in pediatric practice,

6. Endarteritis or Risk of Endocarditis: (Previous or active infective endarteritis is an **absolute indication,** Even small but audible PDAs are closed to prevent this complication

7. Large PDA(Large PDAs should be closed even if the patient is asymptomatic, Risk of heart failure and pulmonary hypertension is high)

8. Symptomatic PDA in Preterm Neonates: Medical treatment (indomethacin or ibuprofen) fails, NSAIDs are contraindicated (renal failure, NEC, thrombocytopenia), The infant remains ventilator-dependent

9. When Catheter Closure Is Not Feasible : (PDA anatomy is unsuitable for device closure, Device closure has failed or caused complications)

COARCTATION OF AORTA(5-7%)

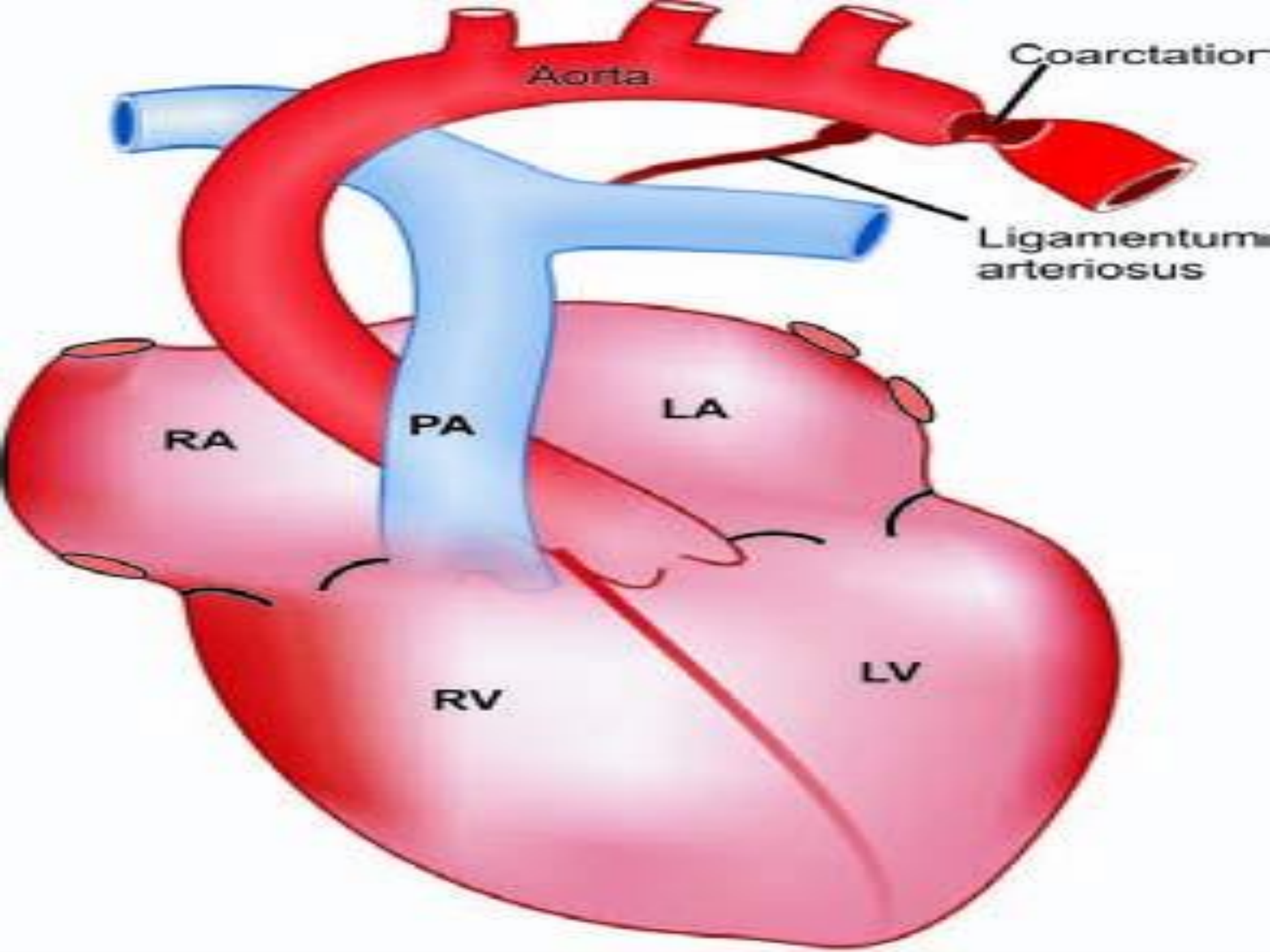
Cardio Thoracic & Vascular Surgery

Narrowing of the aorta just distal to the origin of left subclavian artery. It is often associated with PDA, VSD.

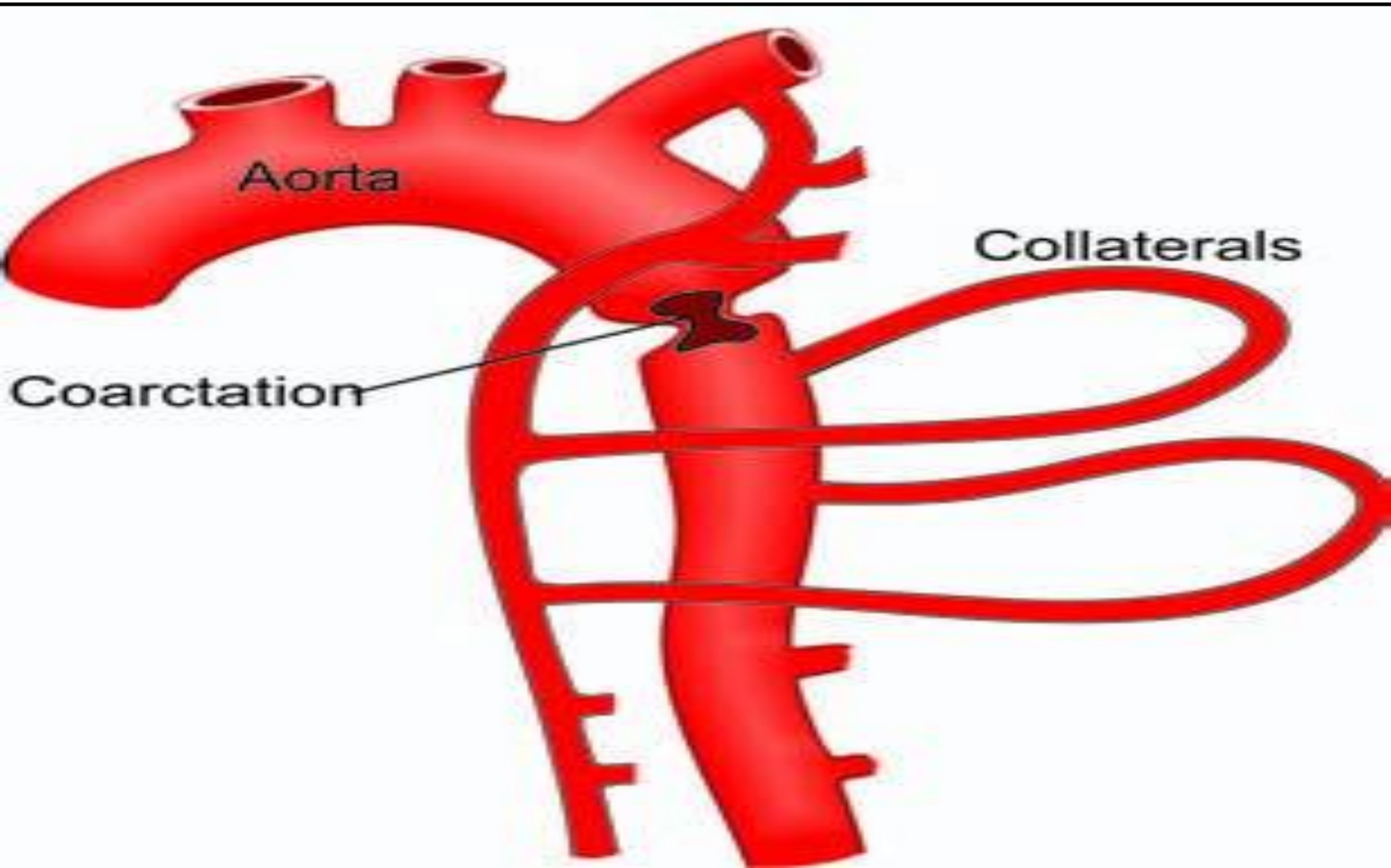
There is increased perfusion of upper limb, cranium, and face. But less blood supply to lower limb, kidneys.

It causes left ventricular hypertrophy, differential bd. pr. (pressure upper limb is higher but pressure in lower limb is lower). Often there is differential cyanosis.

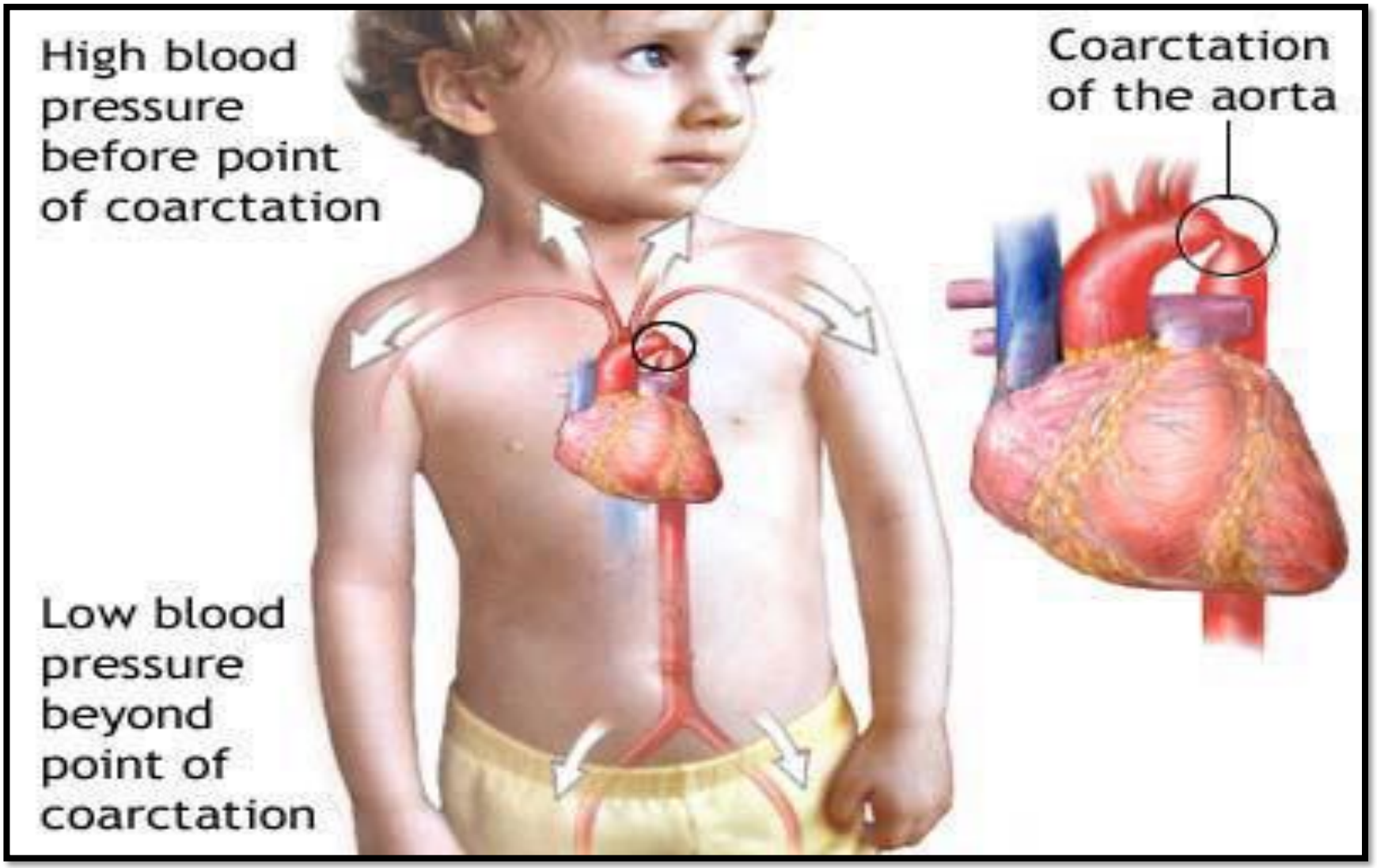
Visible dilated intercostal vessels through collateral perfusion is more obvious when patient leans forwards.

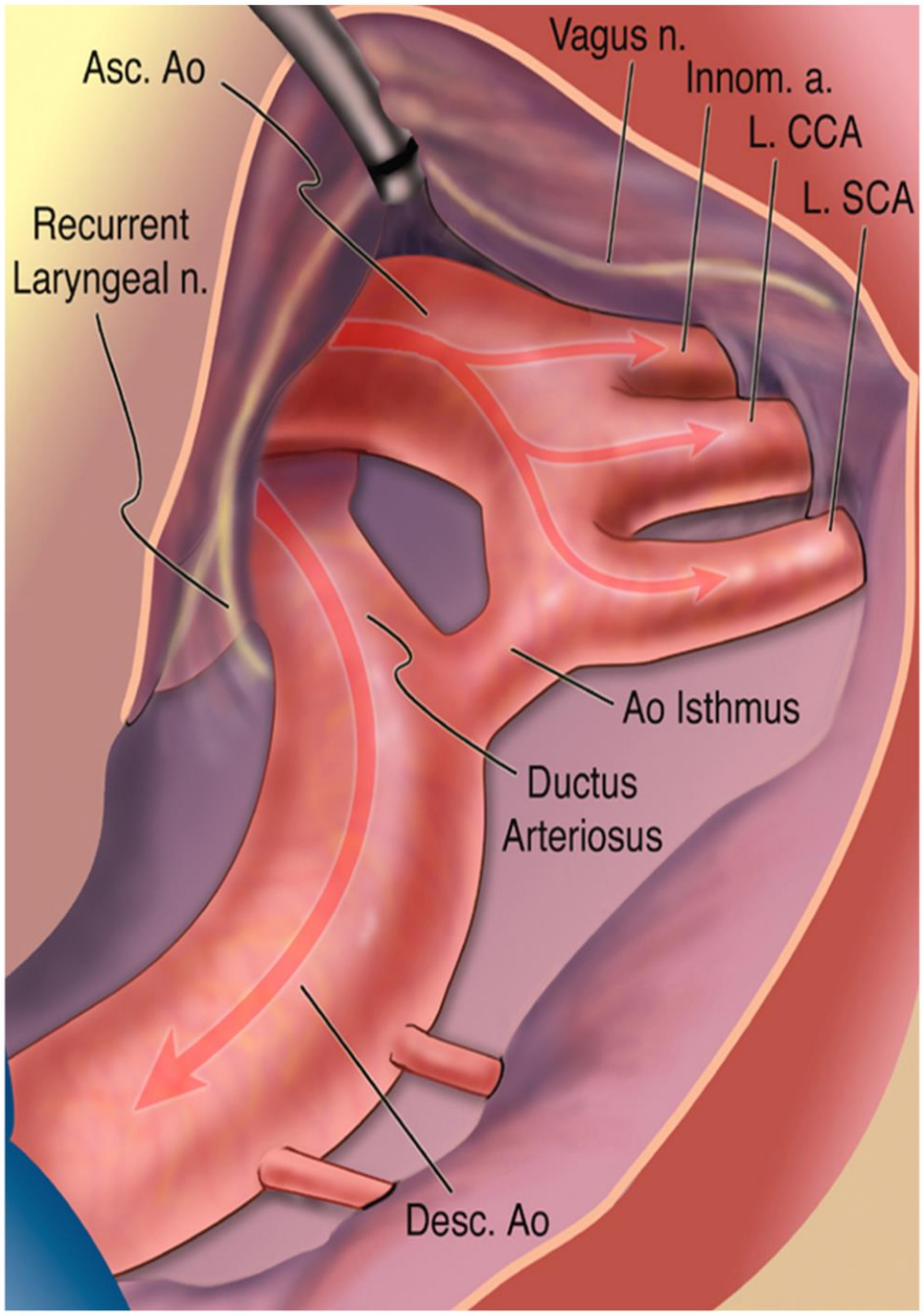

Aorta
Coarctation
Ligamentum
arteriosus
RA
PA
LA
RV
LV

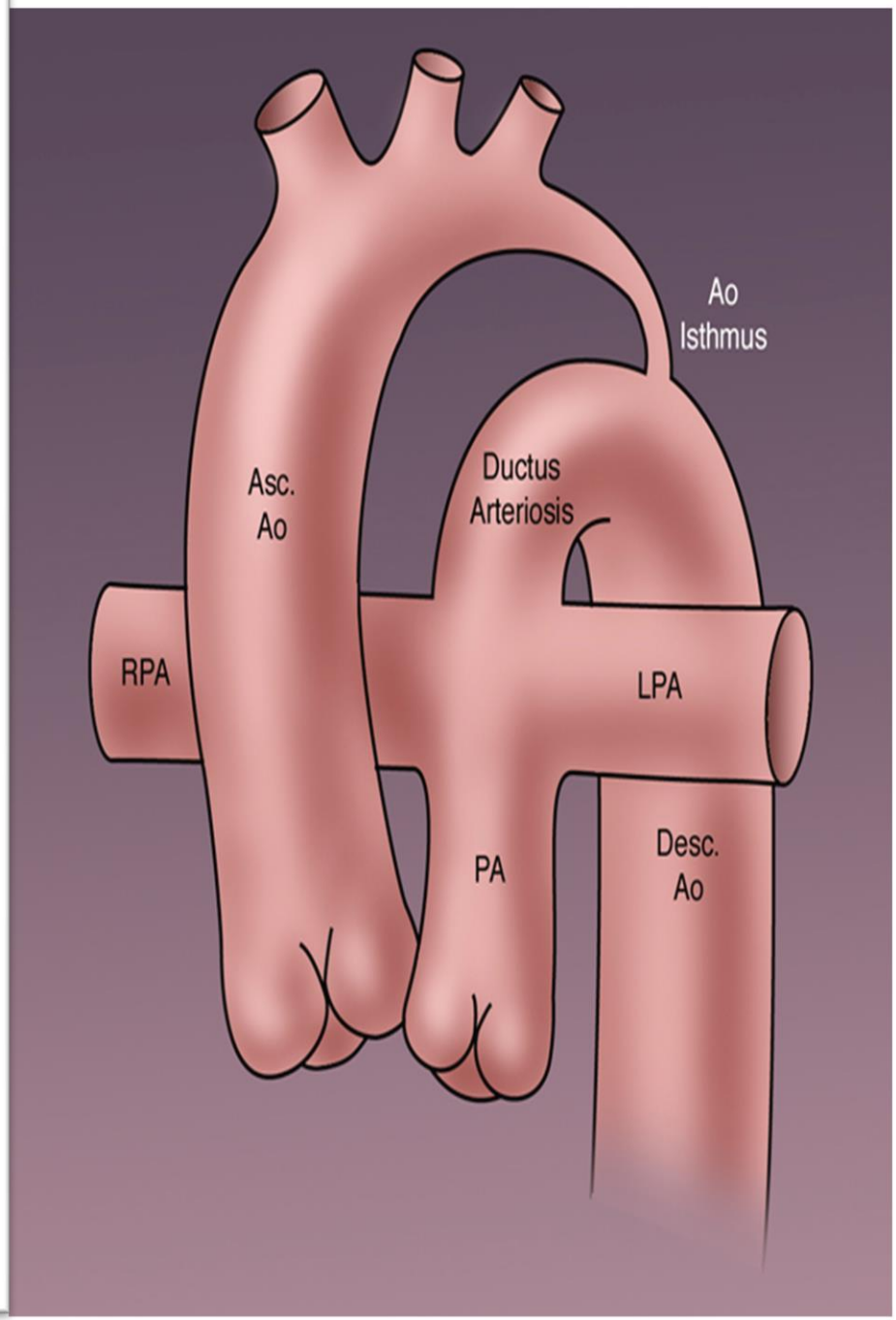
COLLATERALS DEVELOPMENT IN INTERCOSTAL VESSELS IN COARCTATION OF AORTA.

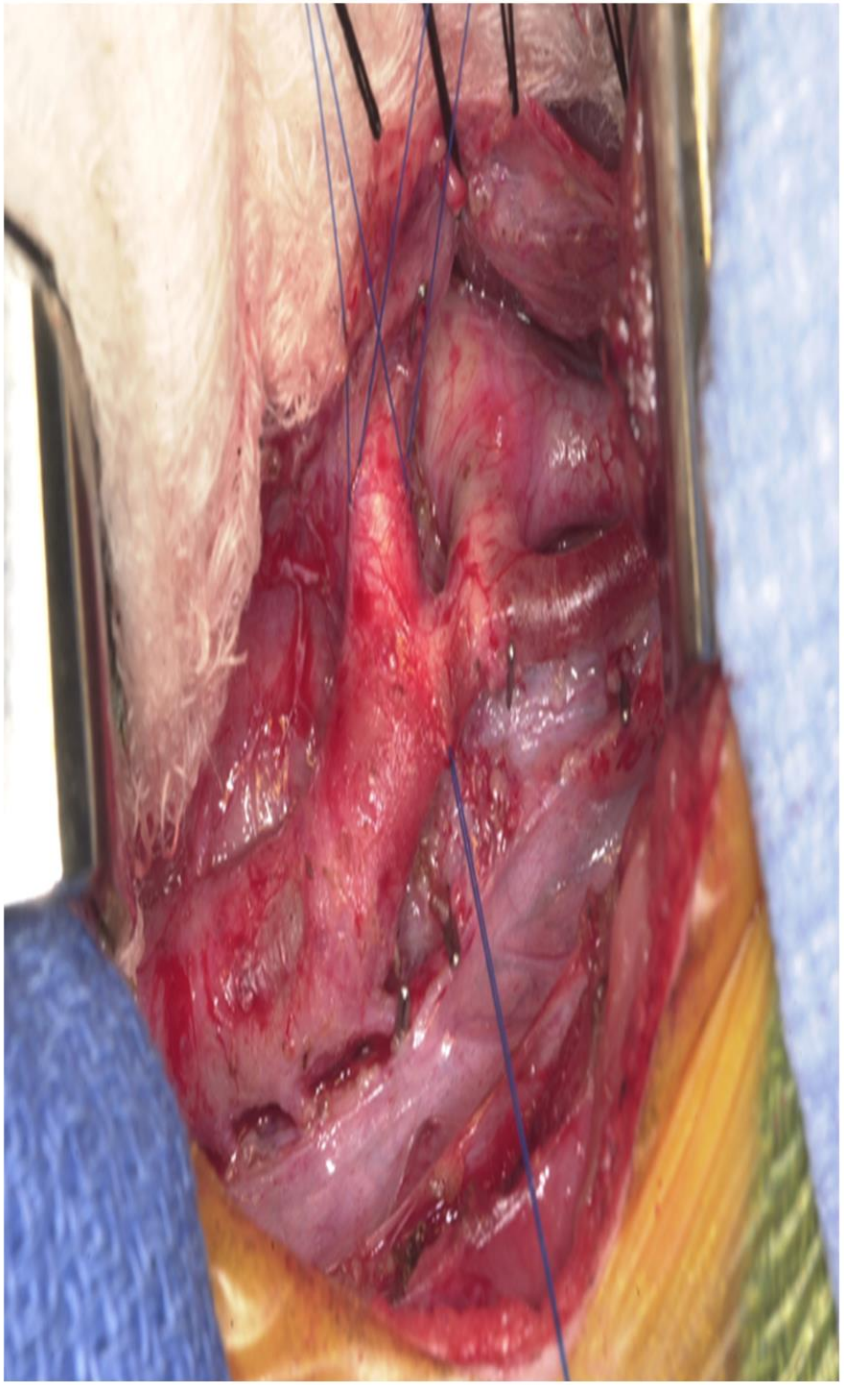


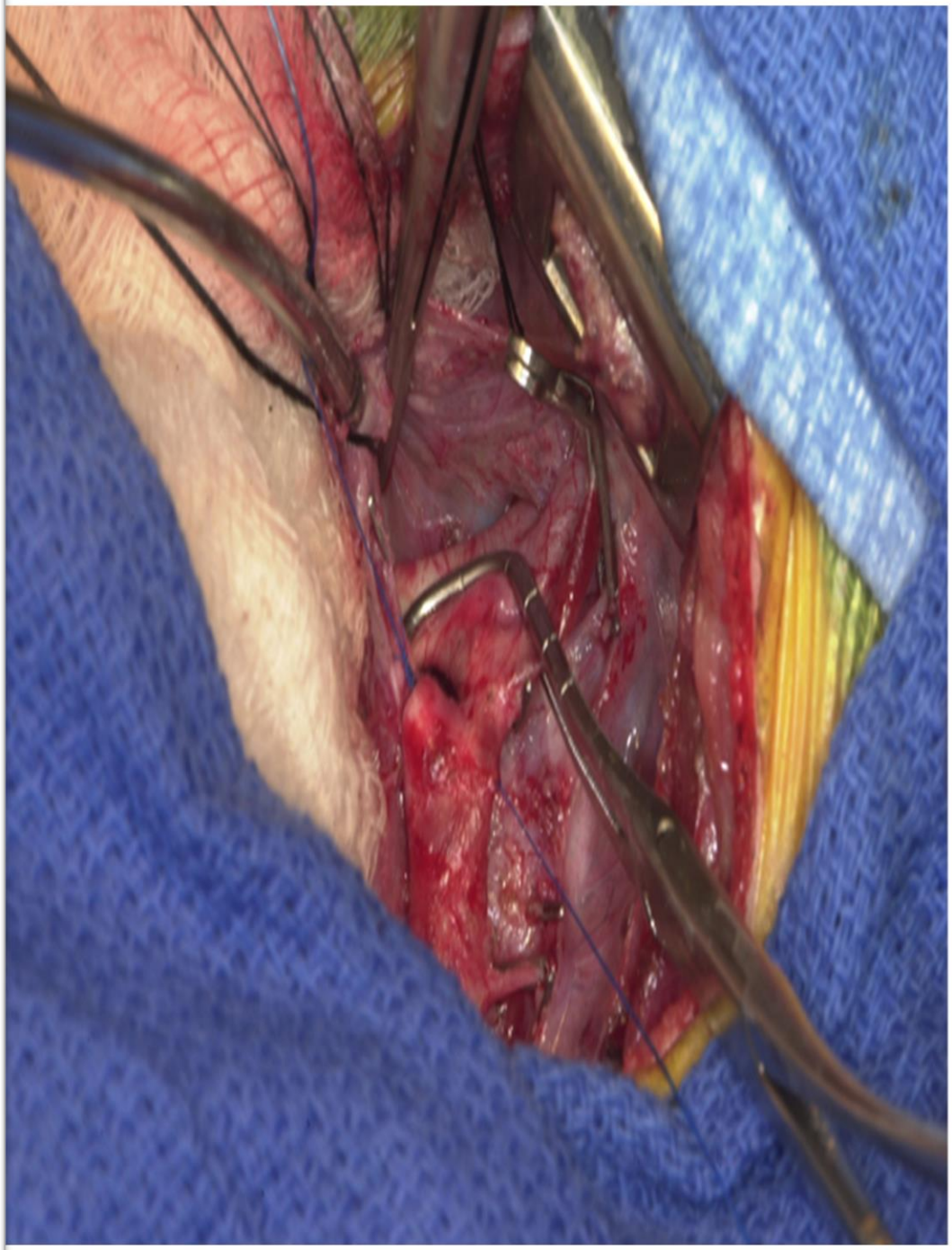
Coarctation of aorta(5-7%)

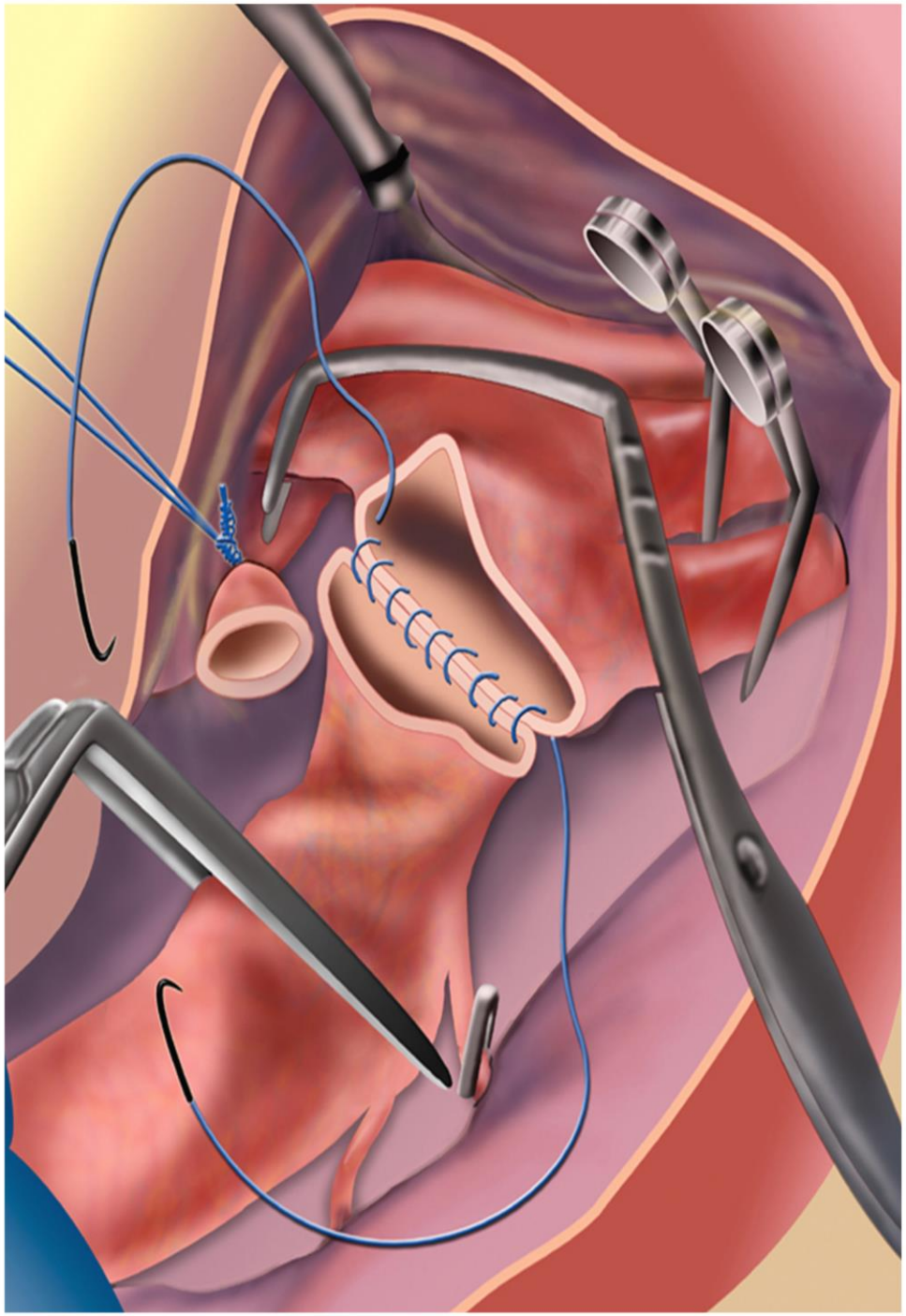


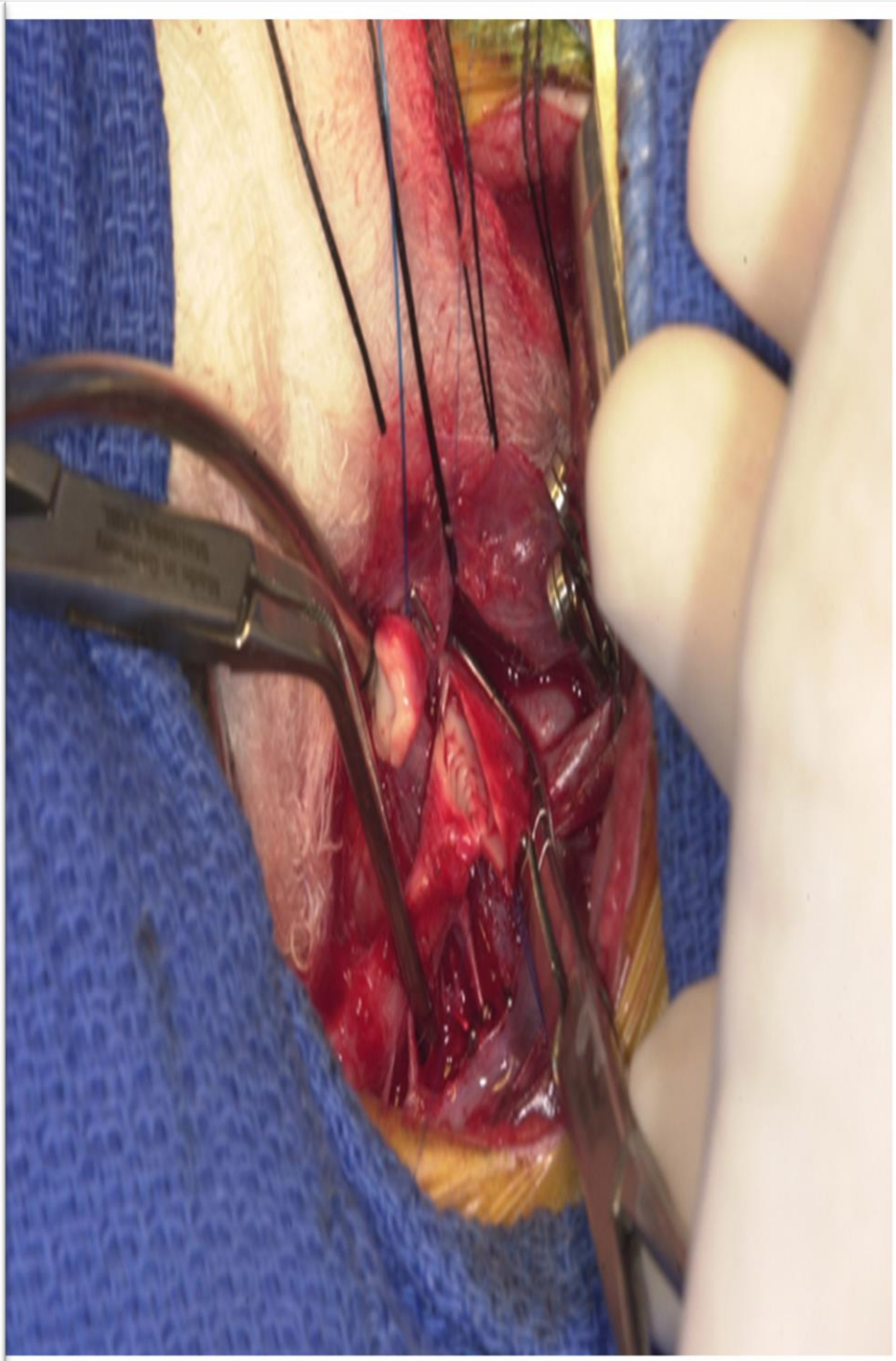

Asc. Ao
Vagus n.
Innom. a.
L. CCA
L. SCA
Recurrent
Laryngeal n.
Ao Isthmus
Ductus
Arteriosus
Desc. Ao


Ao
Isthmus
Asc.
Ao
Ductus
Arteriosis
RPA
LPA
PA
Desc.
Ao









X-ray will show dilated proximal aorta, notching of ribs due to erosion of the intercostals vessels.

Aortogram is diagnostic.

Types: It can be preductal or postductal depending on relation to the ligamentum/ductus arteriosus.

Treatment: is widening of the narrowed segment of aorta with Dacron patch graft, or removal of stenosed segment and arterial graft.

Complications: Congestive cardiac failure, bacterial endocarditis, aortic rupture.

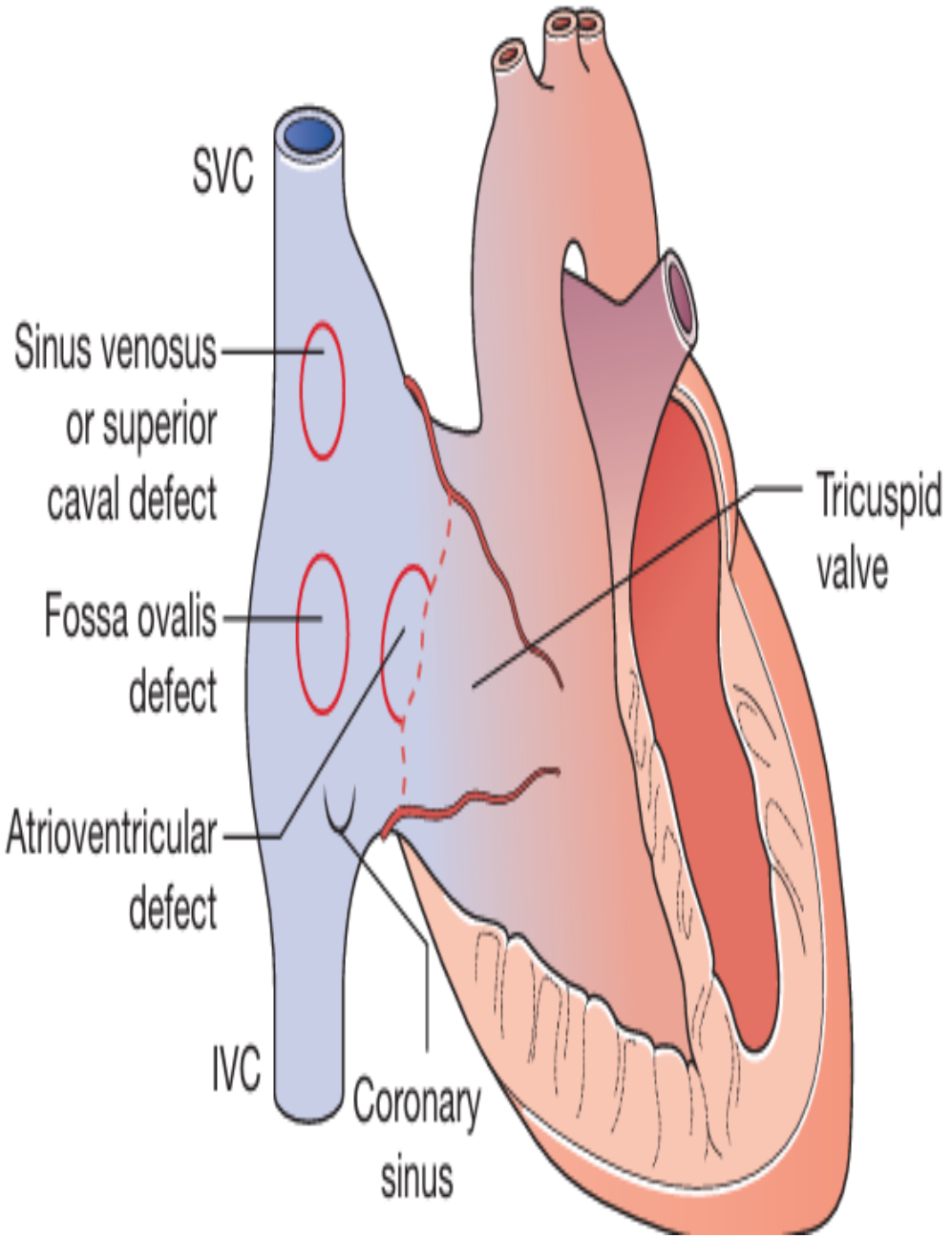
Atrial Septal defect (ASD 6-8%)

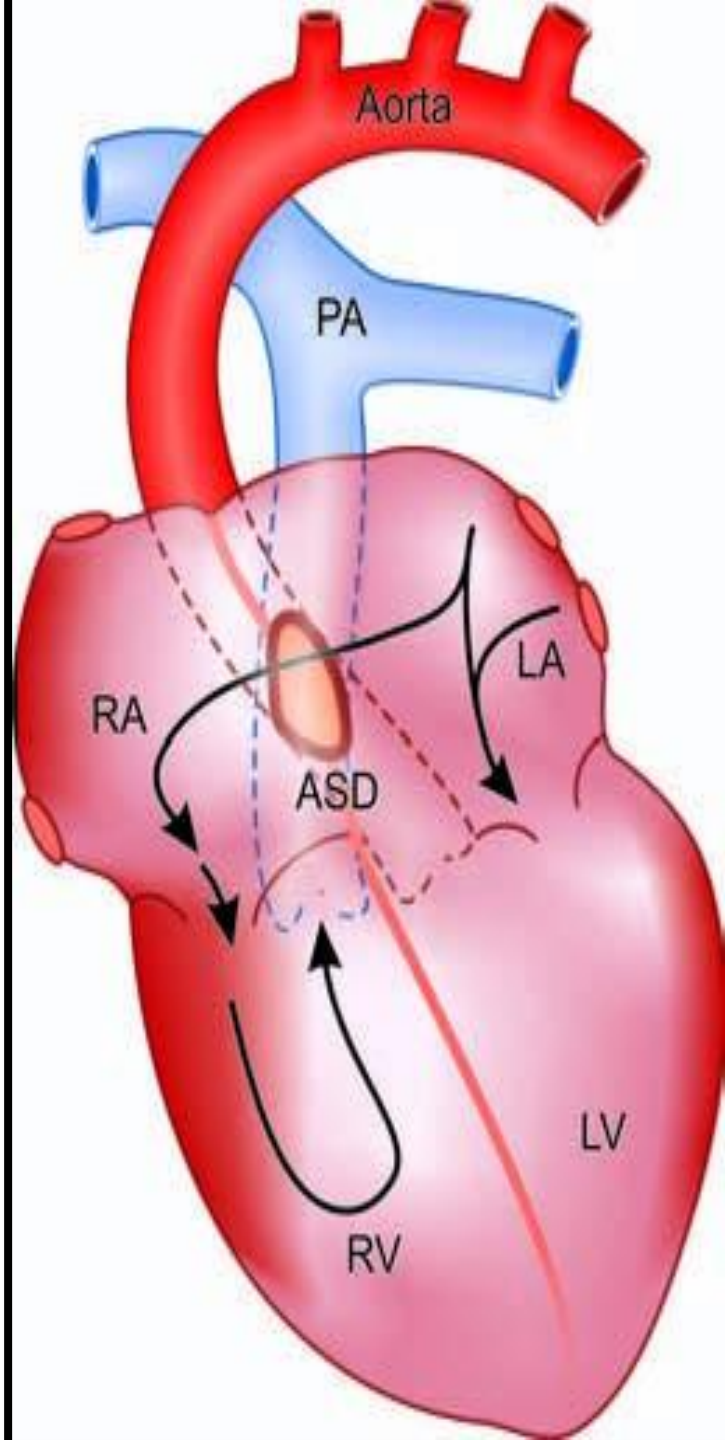
An ASD is the hole in the atrial septum thus causing communication between left and right atrium.

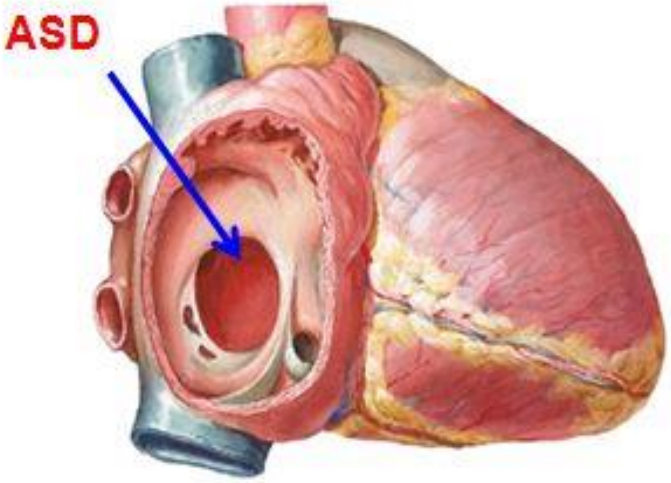
There is shunting of blood from LA to RA. It causes LV hypertrophy, pul. H.T, eventually occasionally reversal of shunt as right to left → cyanosis

Dyspnoea, fixed split of second heart sound, systolic murmur due to increased blood flow through the pulmonary valve are the clinical features

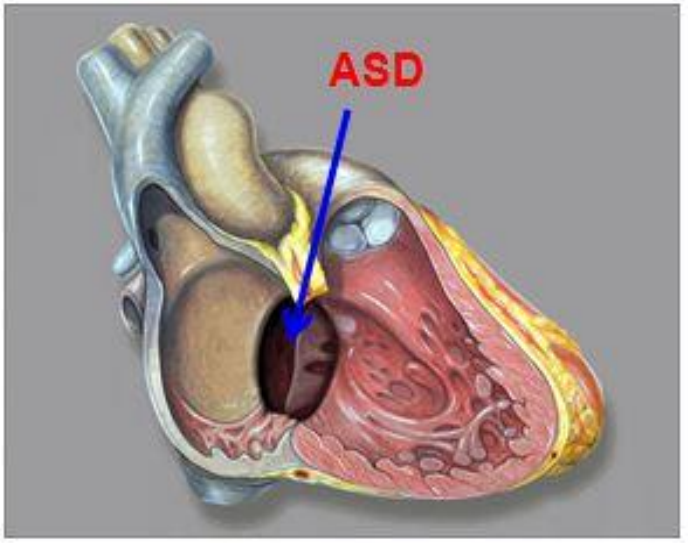
It is diagnosed by echocardiography, cardiac catheterisation and angiocardiogram.


SVC
Sinus venosus or superior caval defect
Fossa ovalis defect
Atrioventricular defect
IVC
Coronary sinus
Tricuspid valve

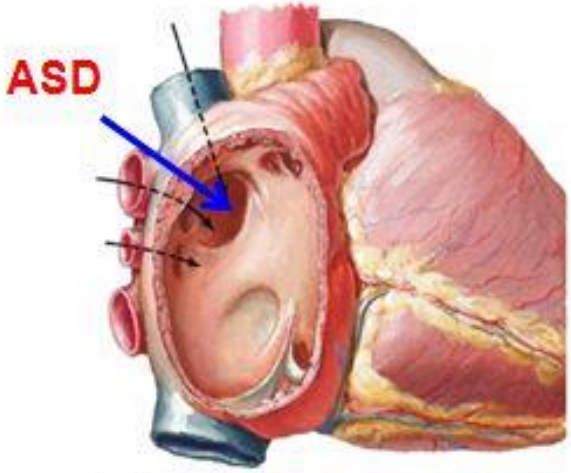

Aorta
PA
RA
LA
ASD
LV
RV



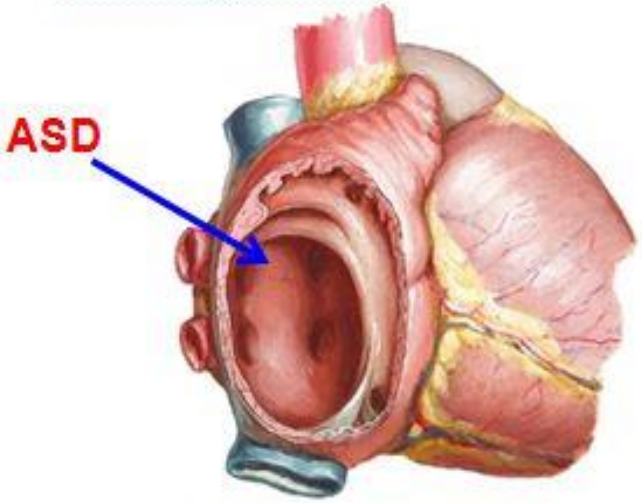
Ostium secundum defect



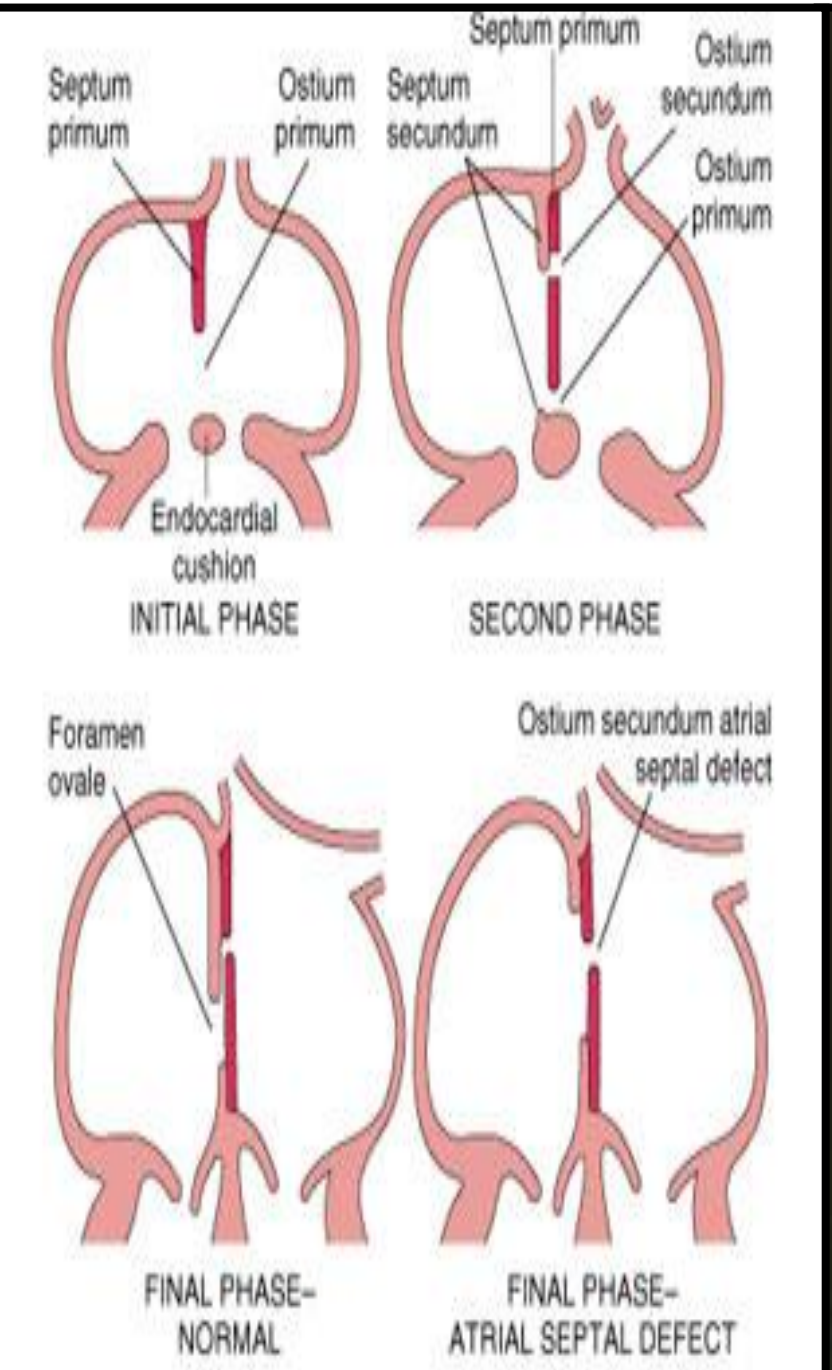
Ostium primum defect



Sinus venosus defect



Common atrium


Septum primum
Ostium primum
Endocardial cushion
INITIAL PHASE
Septum primum
Septum secundum
Ostium secundum
Ostium primum
SECOND PHASE
Foramen ovale
FINAL PHASE– NORMAL
Ostium secundum atrial septal defect
FINAL PHASE– ATRIAL SEPTAL DEFECT
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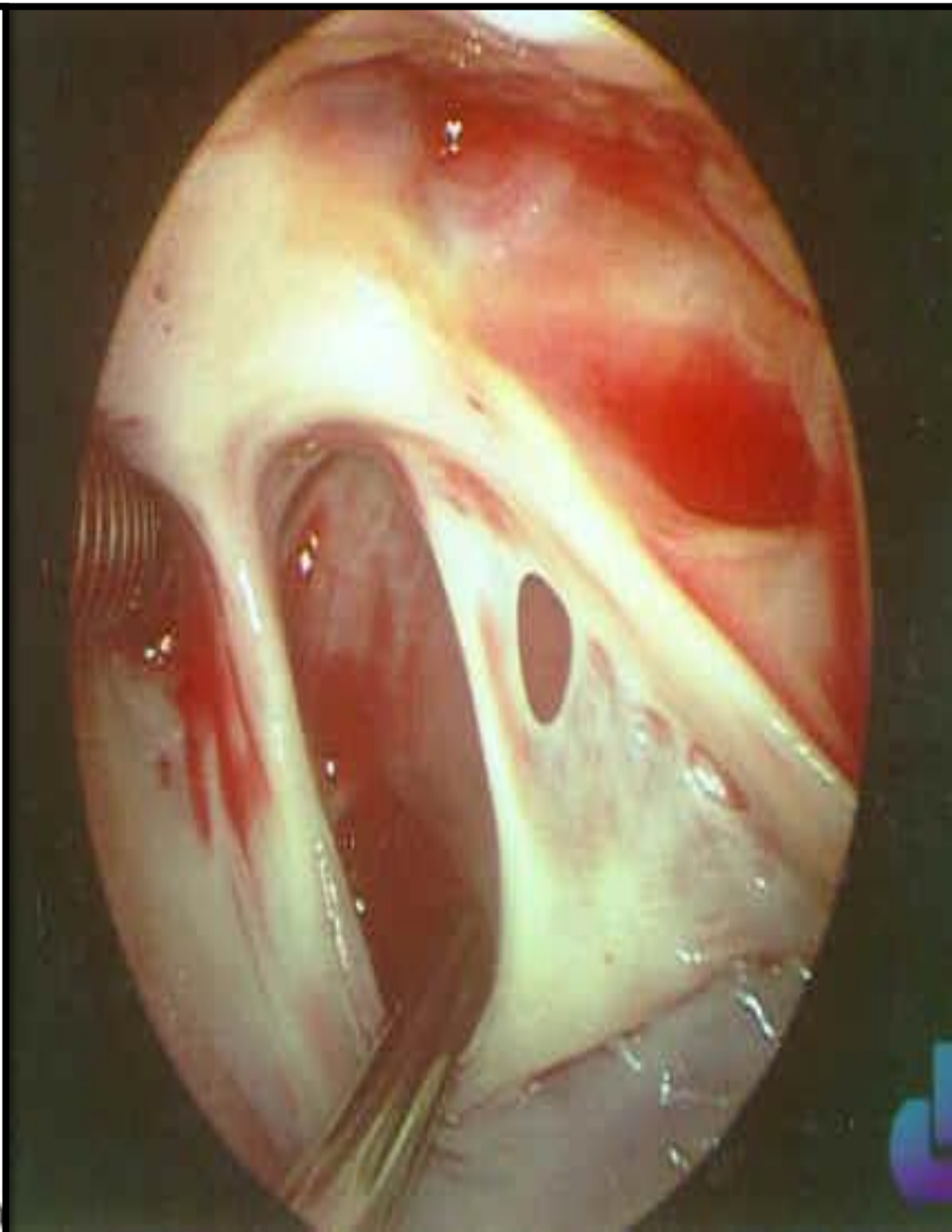
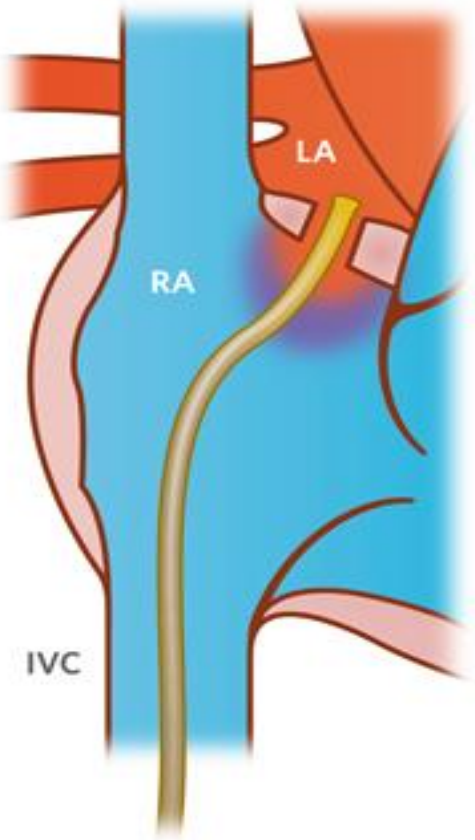
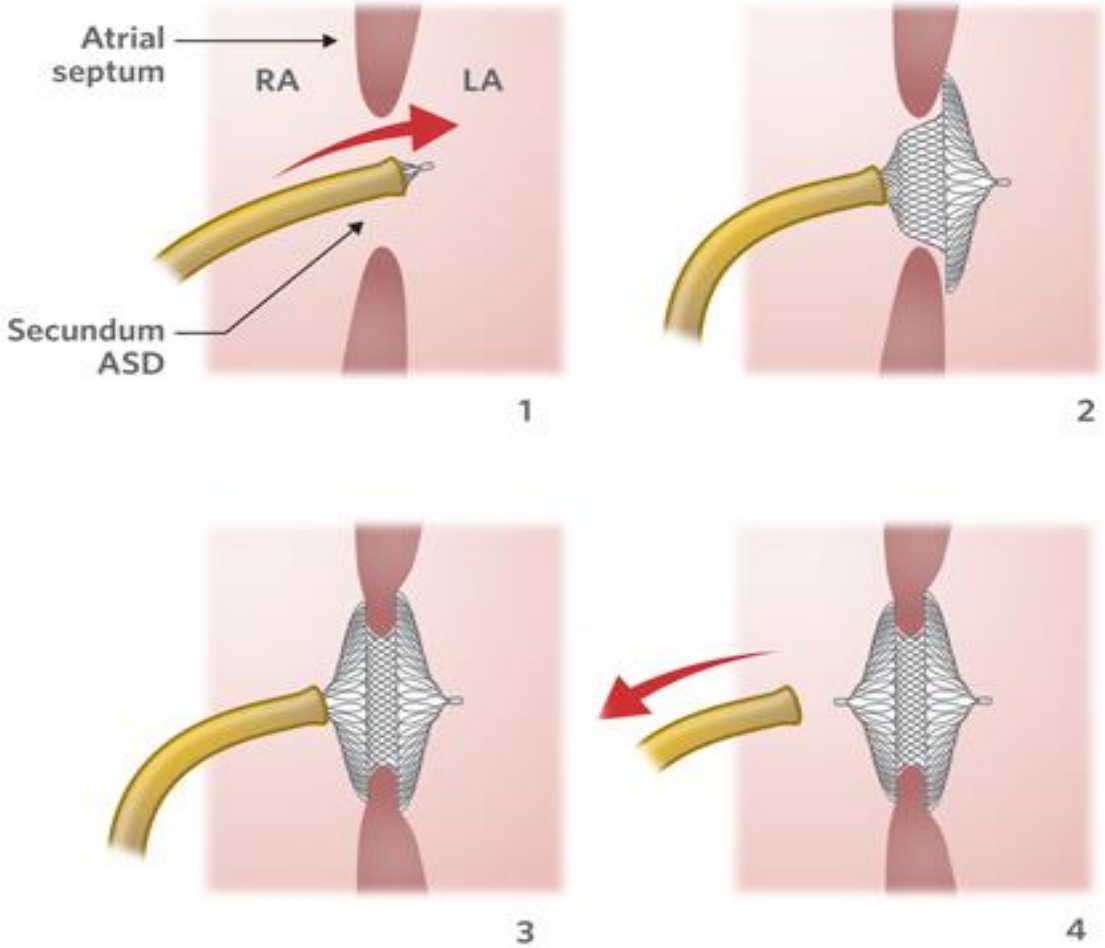


Atrial septal defect closure using an expanding device



Device passed up inferior vena cava, into the right atrium and into septal defect

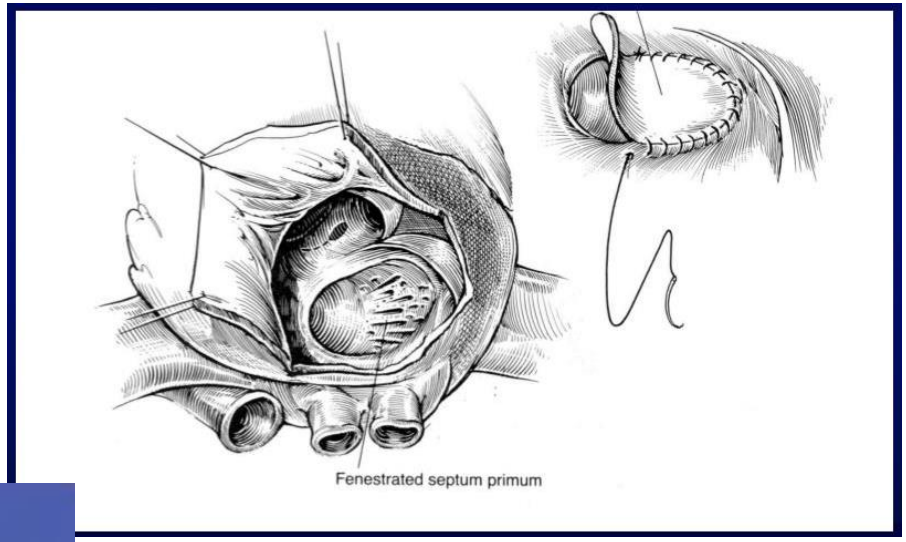


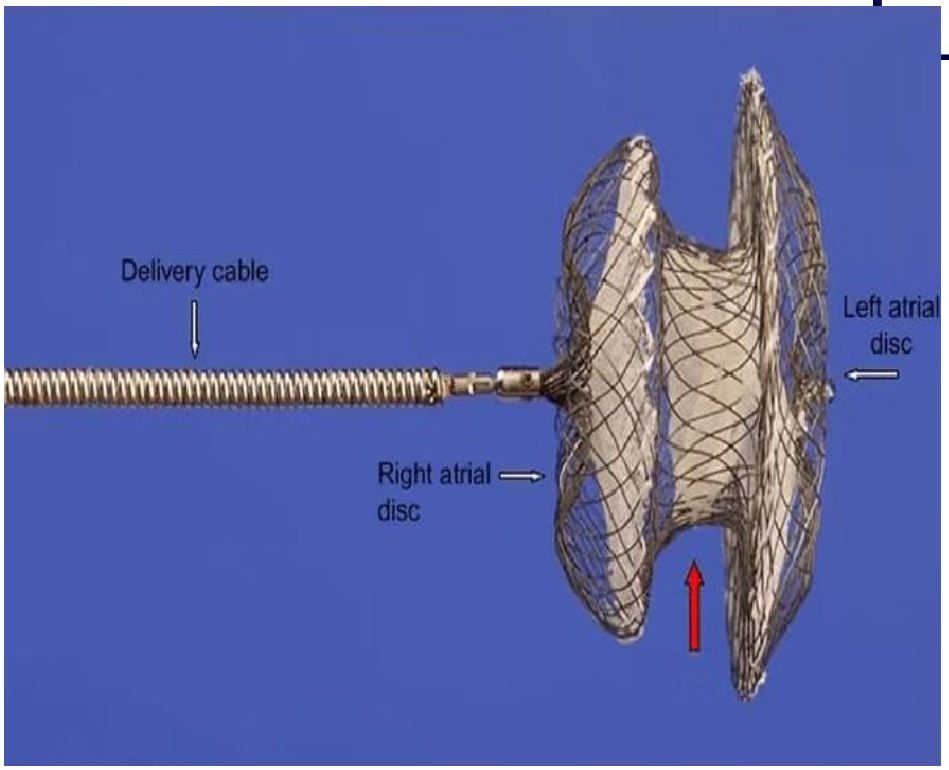
Types:

- **Secundum type** is the commonest one. It is elliptical, in the middle of the atrial septum and is due to defect in the site of foramen ovale
- **Primum type** is lower part. It is rare
- **Sinus venosus type** is near SVC opening

Treatment:

- **Closure of the defect by direct suturing or using patch either pericardium or Dacron.**
- **Transcatheter occlude**


Fenestrated septum primum


Delivery cable
Left atrial disc
Right atrial disc

Atrial septal defects (ASDs)

Common defects

- Ostium secundum: fossa ovalis defect (approximately 70% of ASDs)
- Ostium primum: atrioventricular septal defect (approximately 20% of ASDs)
- Sinus venosus defect: often associated with anomalous pulmonary venous drainage (approximately 10% of ASDs)
- Patent foramen ovale: common in isolation, usually no left-to-right shunt (not strictly an ASD)

Rarer defects

- Inferior vena cava defects: a low sinus venosus defect and may allow shunting of blood into the left atrium
- Coronary sinus septal defect: also known as unroofed coronary sinus, with the left superior vena cava draining to the left atrium as part of a more complex lesion

ventricular Septal defect (vsd 20-30%)

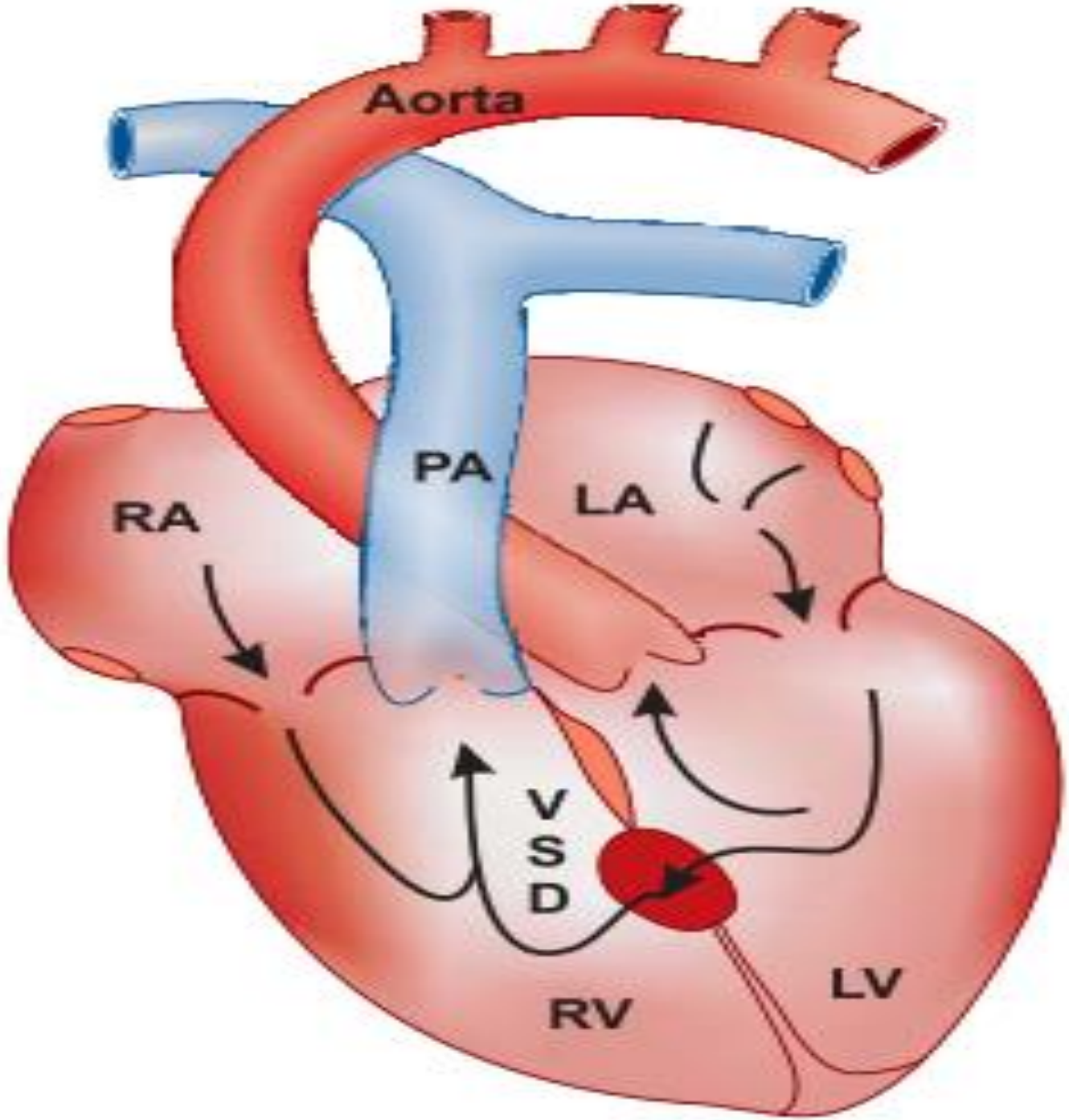
VSD is defect in the ventricular septum causing left to right shunt. There is pulmonary hypertension, ventricular hypertrophy.

Types:

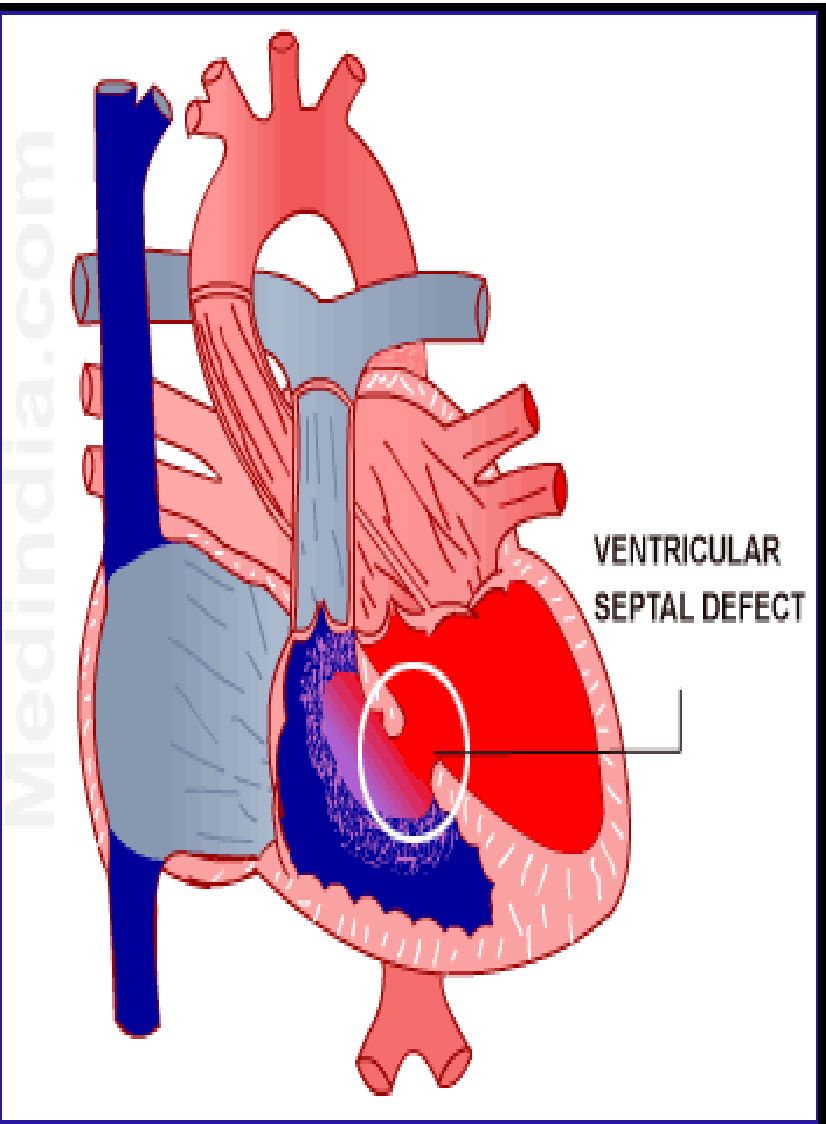
- Four types of VSD are present depending on location of the defect. Defect can be small or large.

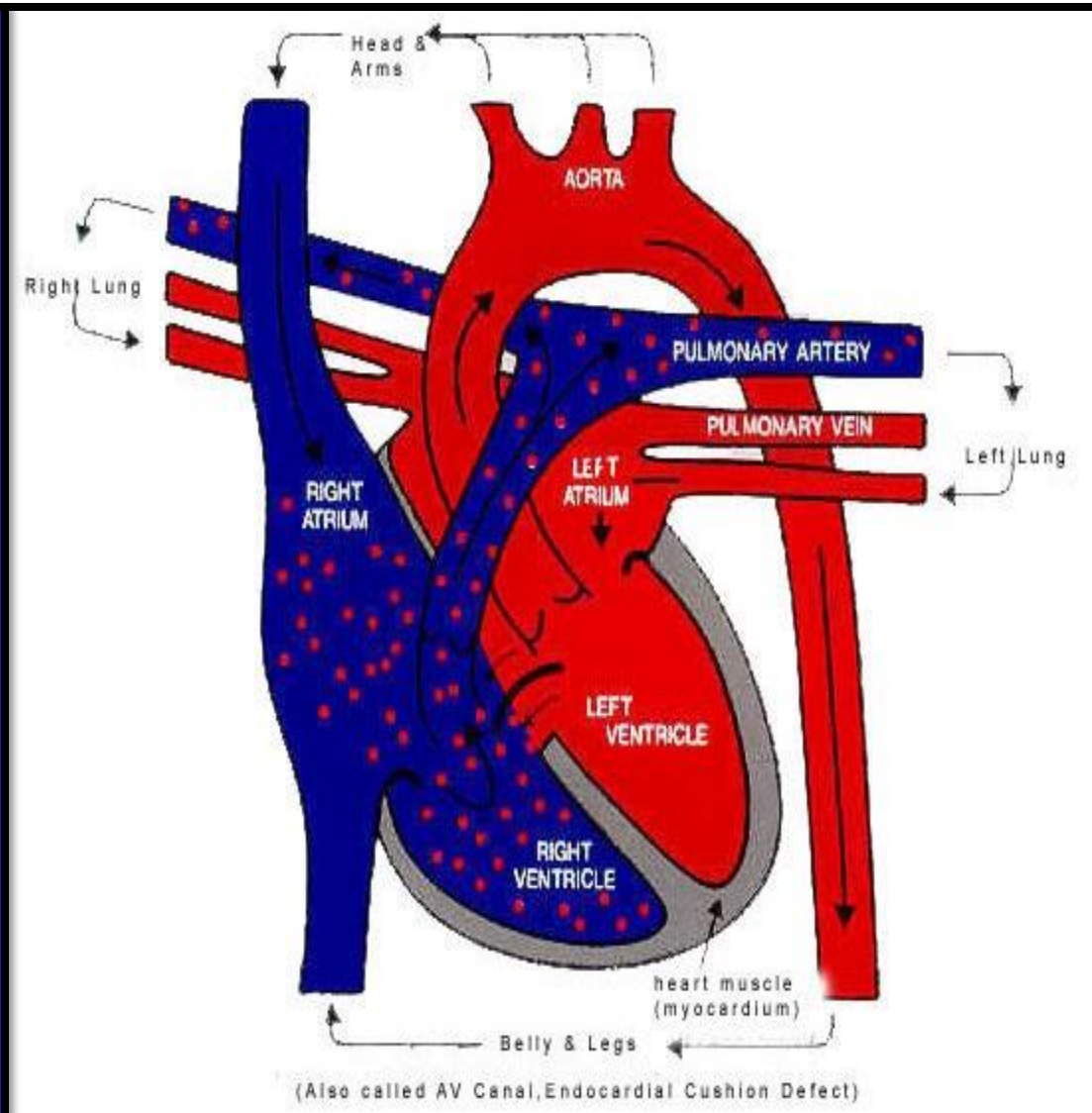
Most common type is perimembranous

- It may be associated with tetralogy of Fallot.
- Severe cases of shunt become reversal causing Eisenmenger's syndrome.


Aorta
PA
RA
LA
VSD
RV
LV

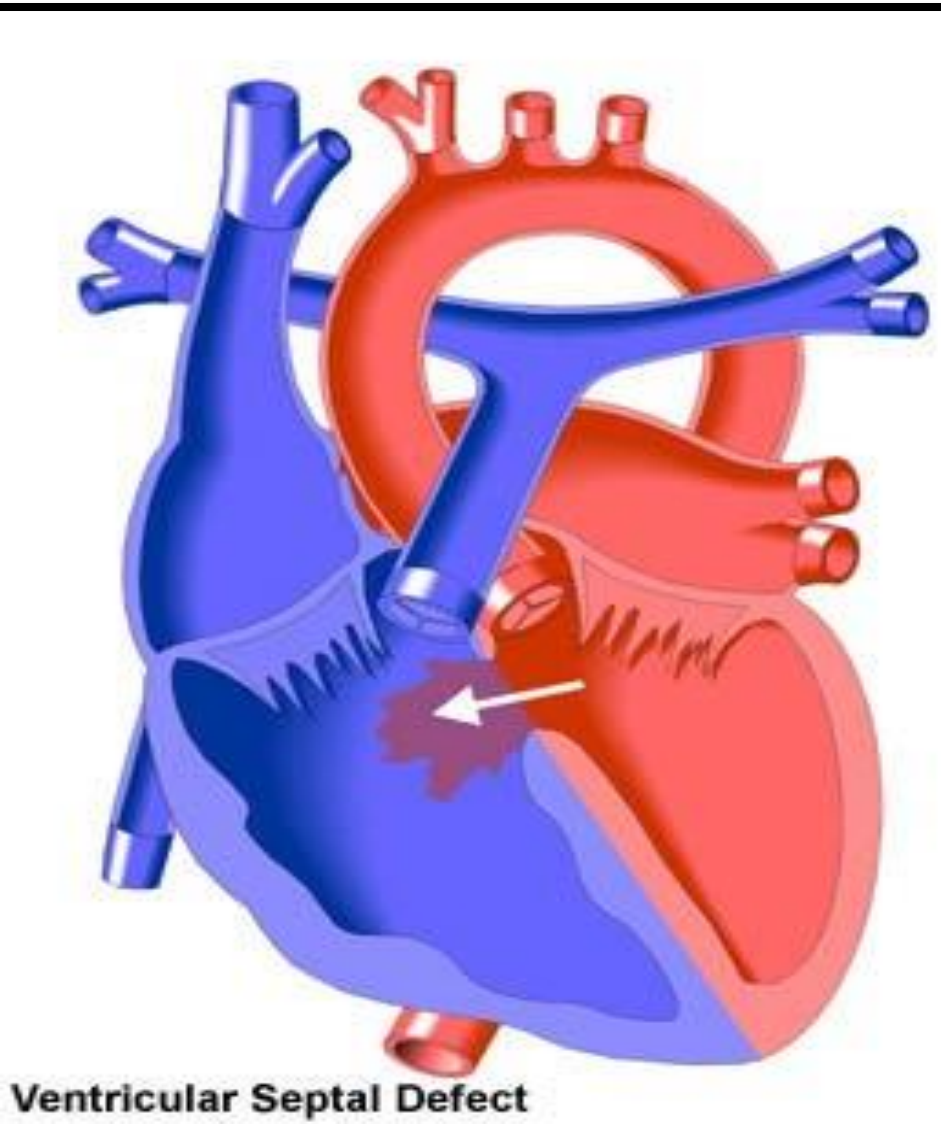
VENTRICULAR SEPTAL DEFECT

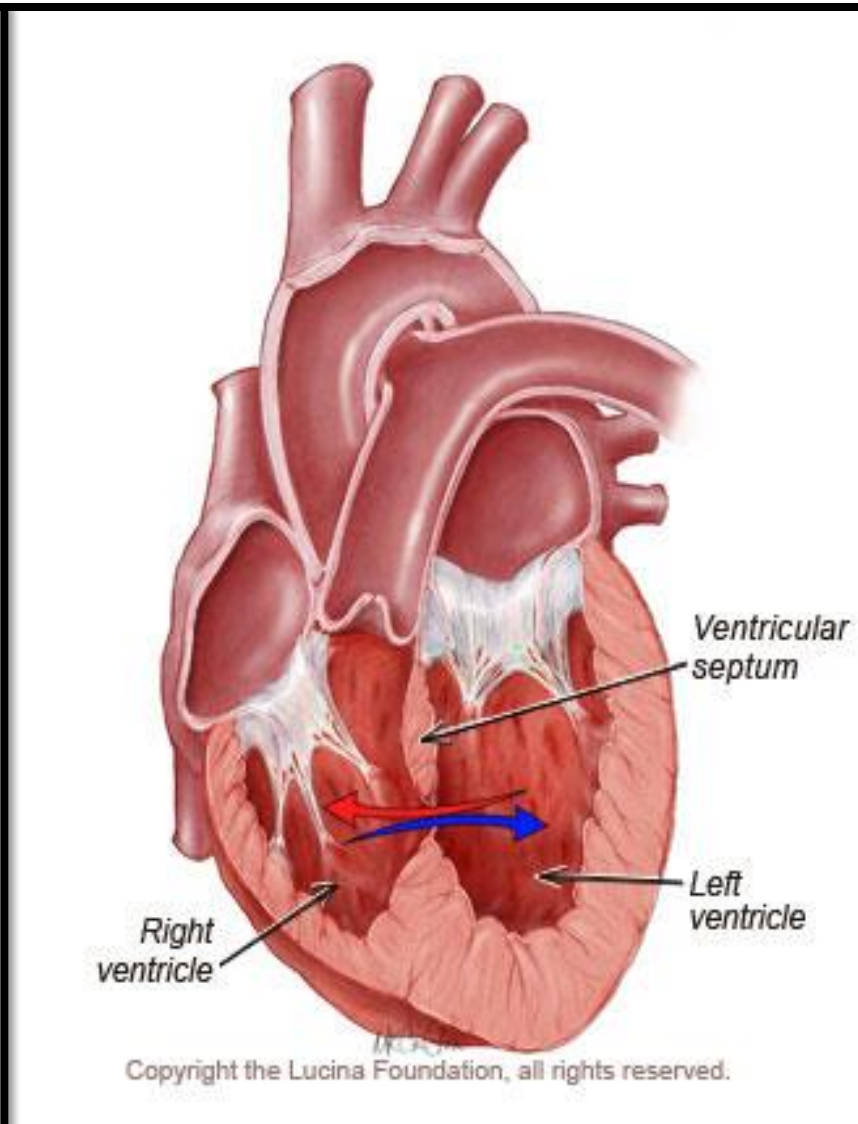




(Also called AV Canal, Endocardial Cushion Defect)

VENTRICULAR SEPTAL DEFECT





VENTRICULAR SEPTAL DEFECT (VSD)

CLINICAL FEATURES : PANSYSTOLIC MURMUR, PALPABLE THRILL, SPLIT SECOND SOUND WITH PULMONARY ACCENTUATION, RECURRENT RESPIRATORY INFECTIONS.

- 40-50% OF DEFECTS ESPECIALLY WHEN THEY ARE SMALL, WILL CLOSE SPONTANEOUSLY. SPONTANEOUS CLOSURE WILL NOT OCCUR AFTER THE AGE OF 6.

- CLOSURE IS INVERSELY RELATED TO THE AGE

- THUS, INFANTS AT 1 MONTH OF AGE HAVE AN 80% INCIDENCE OF SPONTANEOUS CLOSURE, WHEREAS A CHILD AT 12 MONTHS OF AGE HAS ONLY A 25% CHANCE OF CLOSURE

- ***INVESTIGATIONS:*** ECHOCARDIOGRAPHY, CARD. CATHETERIZATION.
- ***TREATMENT:*** CLOSURE OF THE DEFECT USING PERICARDIAL PATCH OR DACRON.
- ***COMPLICATIONS OF VSD:*** REC. RESPIRATORY INFECTION, PULMONARY HYPERTENSION, CARDIAC FAILURE.

Summary box 59.13

Types of ventricular septal defects (VSD)

Perimembranous (conoventricular) defect

- The most common defect (70–80%), usually located within the membranous septum and may extend to the tricuspid valve annulus or the base of the aortic valve

Muscular (trabecular) defect

- Occurs in 10% of cases and is located within the membranous septum and can be multiple

Atrioventricular (inlet) defect

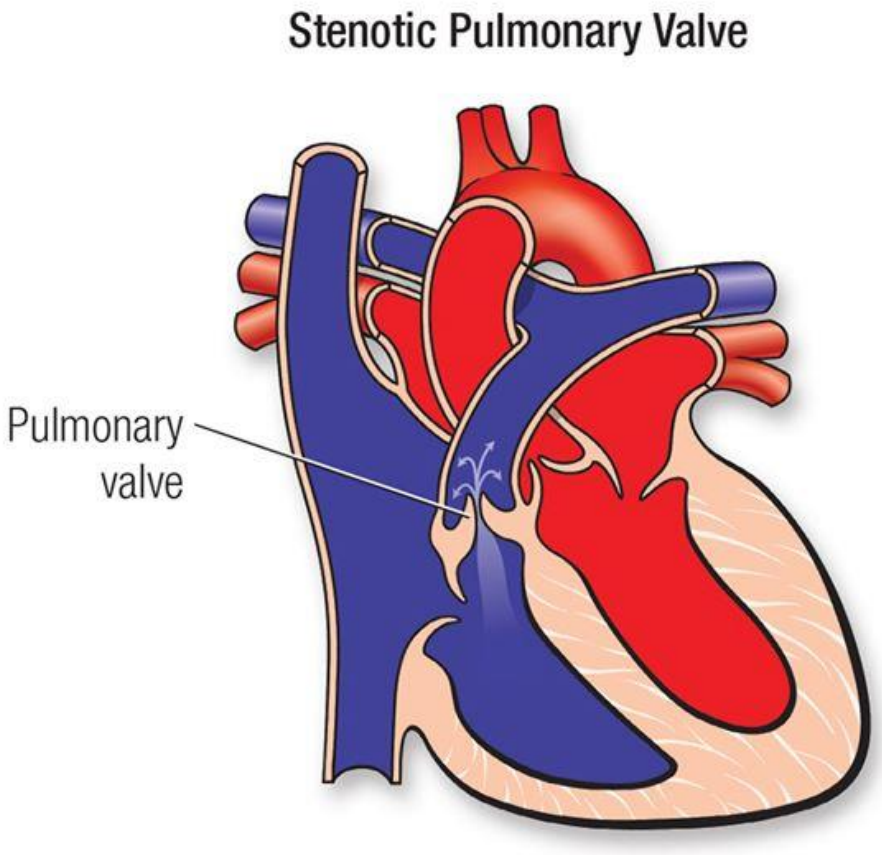
- Also called an atrioventricular canal-type defect; occurs in 5% of cases and is located in the atrioventricular canal beneath the tricuspid valve

Subarterial (outlet) defect

- Occurs in 5–10% of cases and lies within the conal septum immediately beneath the pulmonary valve annulus

Pulmonary stenosis (7-12%)

- It is narrowing of the pulm. valves d fusion of cusps. Narrowing may be a 2-4 mm.
- Patient develops right ventricular hypertrophy and later right ventric failure.
- Pressure in the RV is higher, in PA is
- Treatment is pulmonary valvotomy.




Thank
You