



# **CONGENITAL HEART DISEASE**

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## **OBJECTIVES:**

- 1- Definition
- 2- Etiology
- 3- Pathophysiology
- 4- Eisenmenger's syndrome
- 5- Clinical presentation
- 6- Investigations
- 7- Management



- The result of defects in the formation of the heart or great vessels: the anatomical changes that occur during transition between the fetus and the newborn fail to proceed normally.
- Defects that are well tolerated, such as atrial septal defect, may cause no symptoms until adult life or may be detected **incidentally** on routine examination or chest X-ray.
- Patients with **surgically corrected defects** remain well for many years but subsequently re-present in later life with **related problems** such as arrhythmia or heart failure



## Etiology

- Infection : Maternal rubella infection : PDA , pulmonary valvular and/or artery stenosis, and atrial septal defect.
- Maternal alcohol misuse : septal defects
- Genetic or chromosomal abnormalities, such as Down's syndrome that cause septal defects



## Congenital cyanotic heart disease

- TGA ( Transposition of great arteries )
- TOF ( Tetralogy of fallot )
- TAPVD ( Total anomalous pulmonary venous drainage )
- Tricuspid atresia
- Truncus arteriosus
- Hypoplastic RV with or without pulmonary atresia

## Congenital acyanotic heart disease

- VSD
- ASD
- PDA
- CORACTATION OF AORTA

**i**

## 16.99 Incidence and relative frequency of congenital cardiac malformations

| Lesion                                   | % of all congenital heart defects |
|--|-----------------------------------|
| Ventricular septal defect                | 30                                |
| Atrial septal defect                     | 10                                |
| Persistent ductus arteriosus             | 10                                |
| Pulmonary stenosis                       | 7                                 |
| Coarctation of aorta                     | 7                                 |
| Aortic stenosis                          | 6                                 |
| Tetralogy of Fallot                      | 6                                 |
| Complete transposition of great arteries | 4                                 |
| Others                                   | 20                                |



# Clinical symptoms and signs vary with the severity and anatomical lesion



## i

### 16.100 Presentation of congenital heart disease throughout life

#### Birth and neonatal period

- Cyanosis
- Heart failure

#### Infancy and childhood

- Cyanosis
- Heart failure
- Arrhythmia
- Murmur
- Failure to thrive

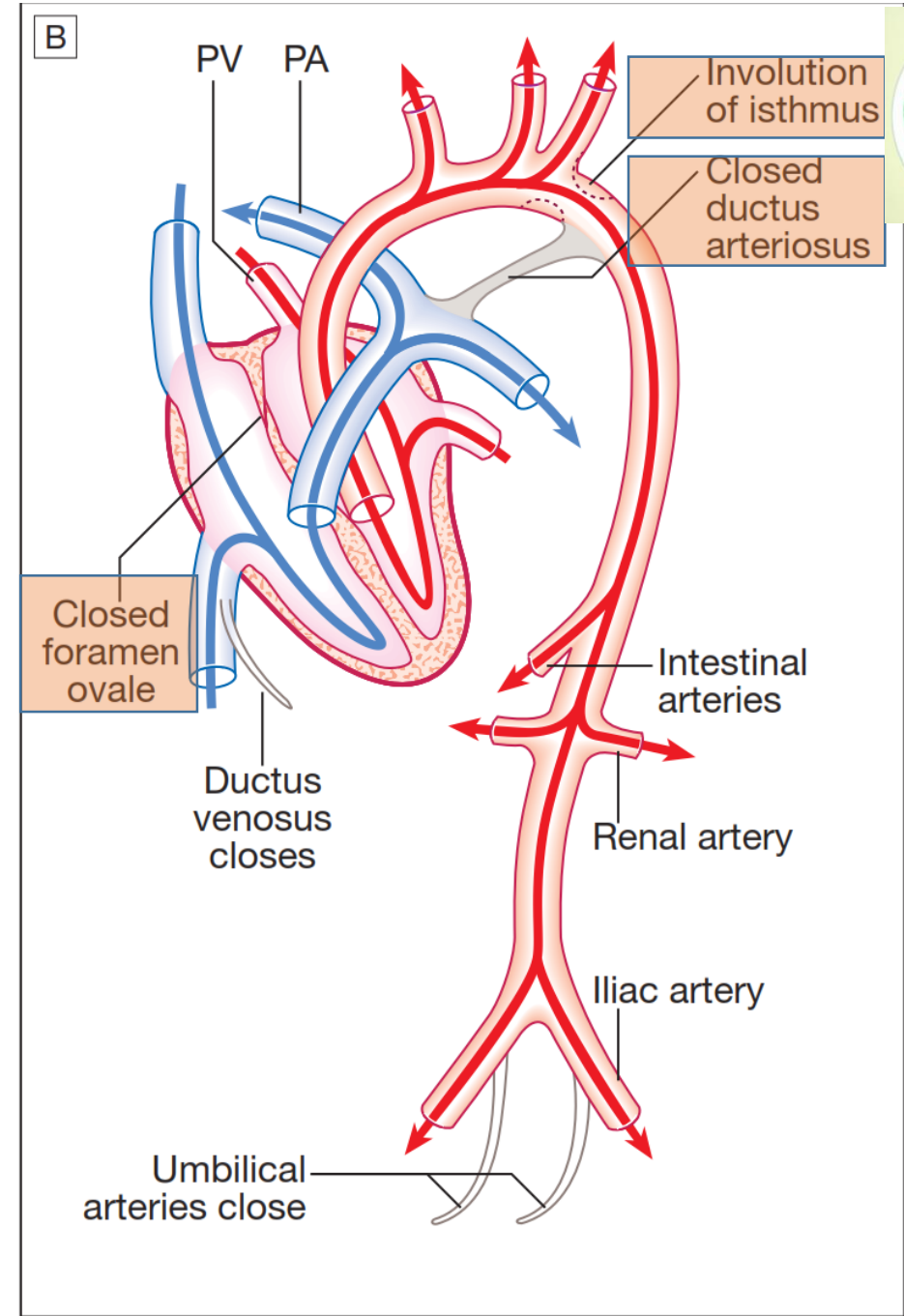
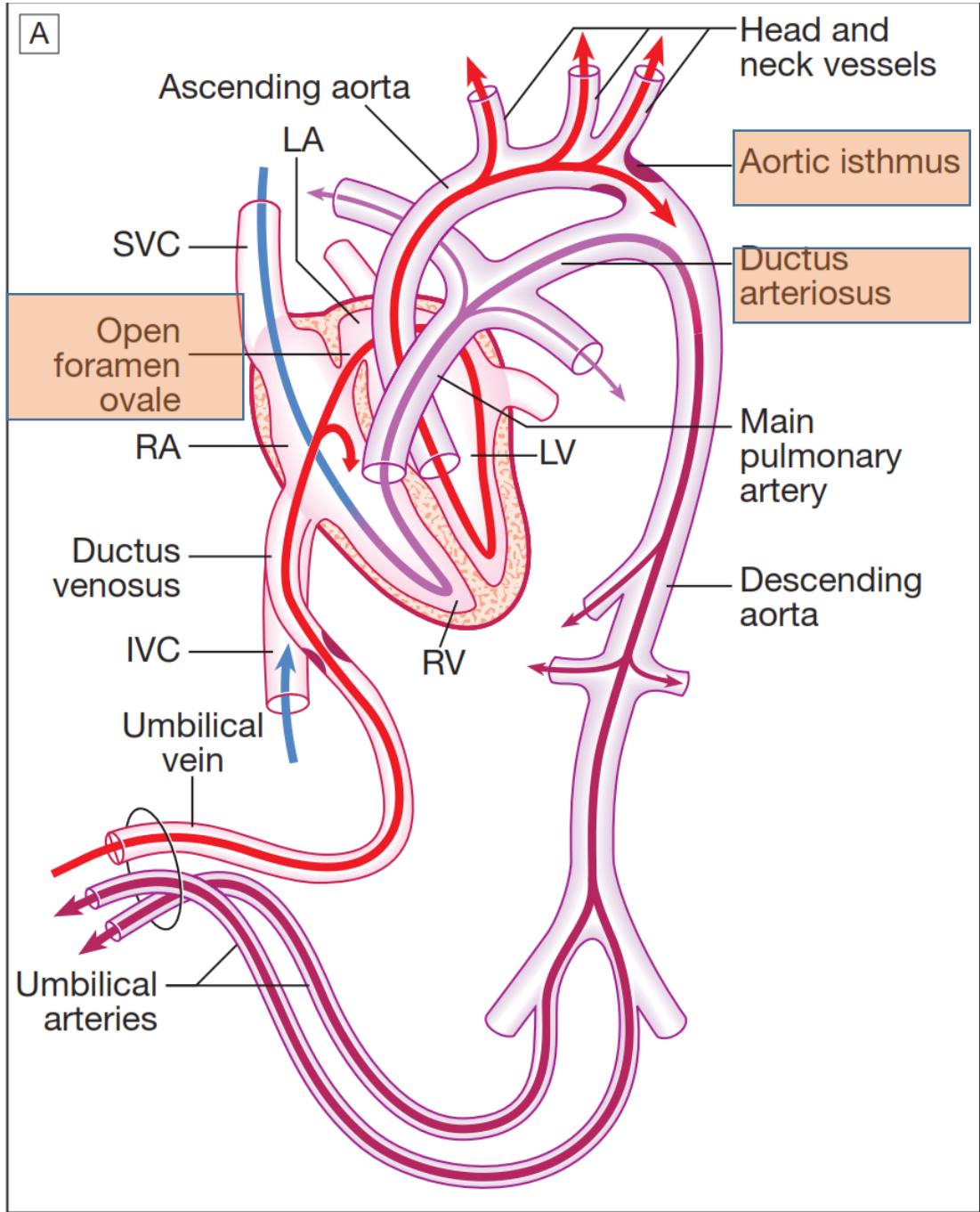
#### Adolescence and adulthood

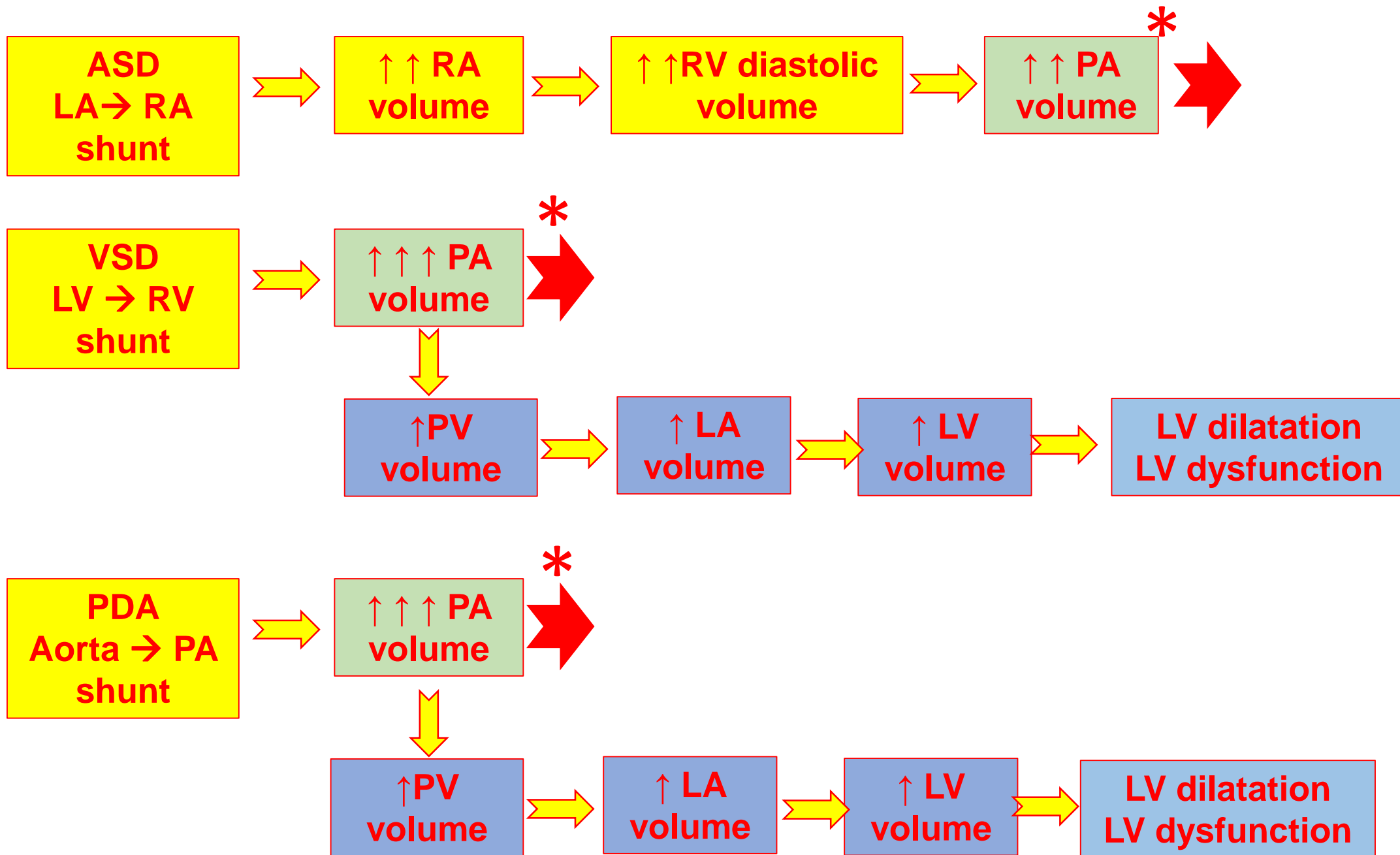
- Heart failure
- Murmur
- Arrhythmia
- Eisenmenger's syndrome
- Hypertension (coarctation)
- Complications of previous cardiac surgery:
  - Arrhythmia related to scarring
  - Heart failure secondary to scarring

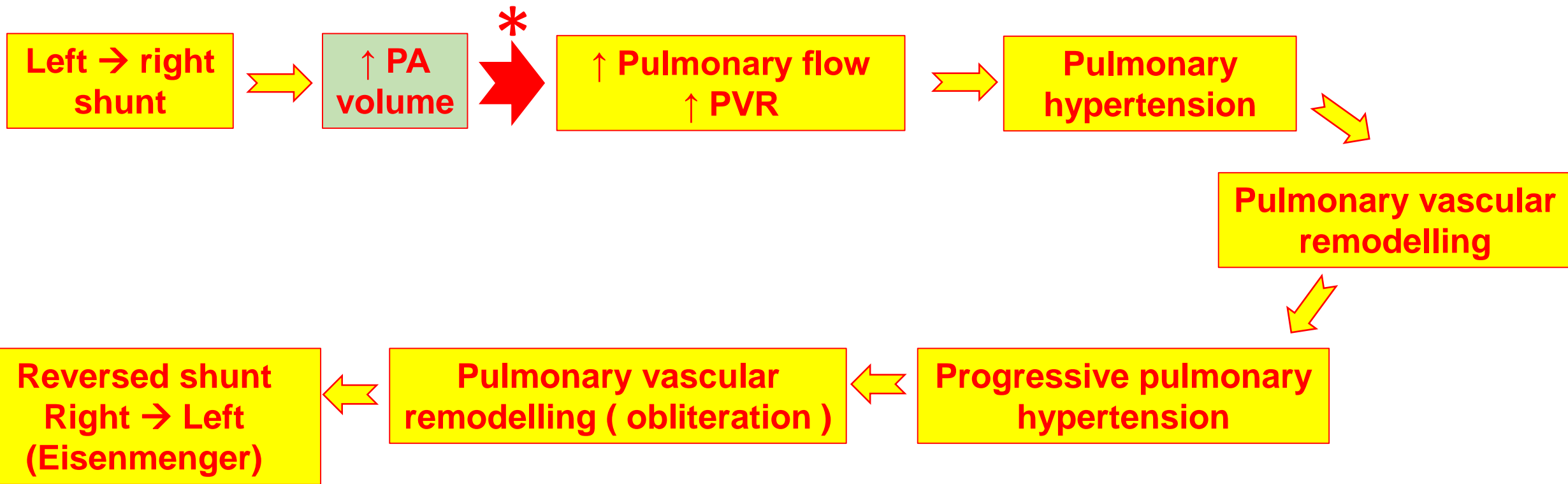


# Pathophysiology







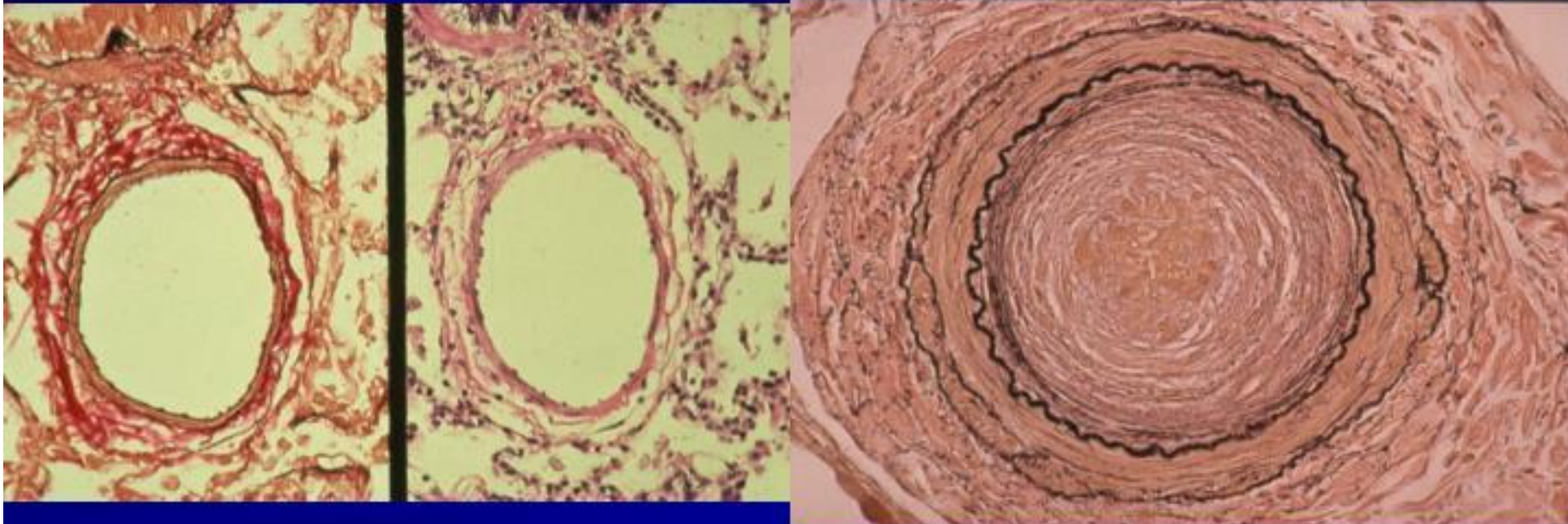


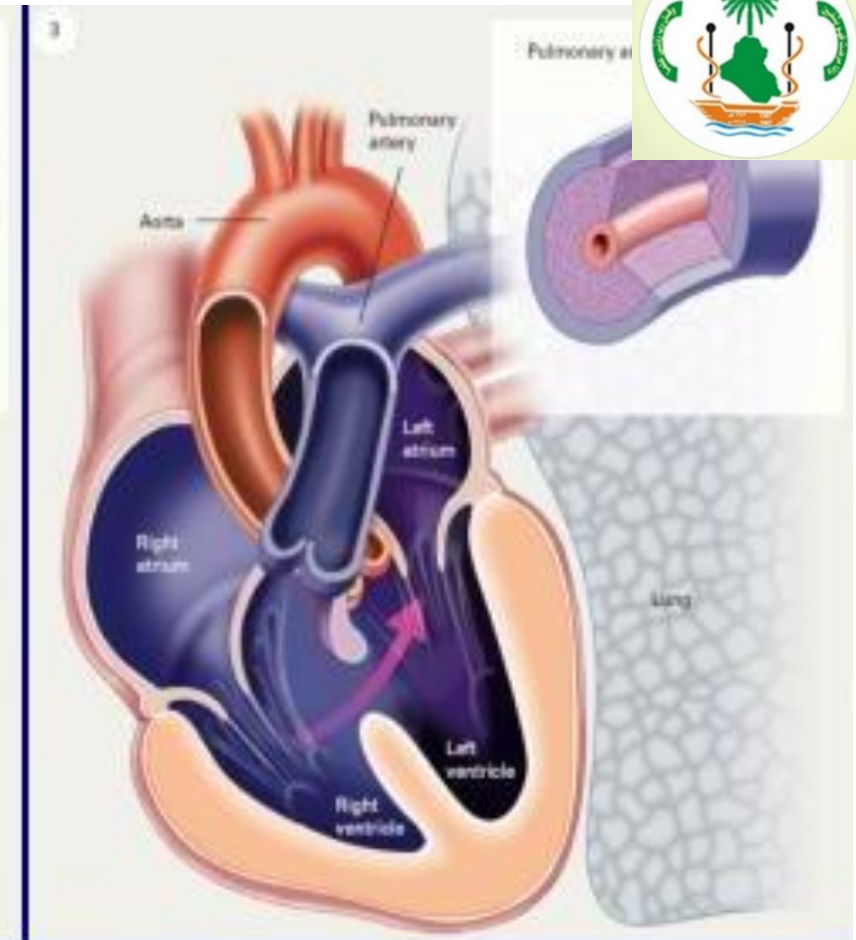
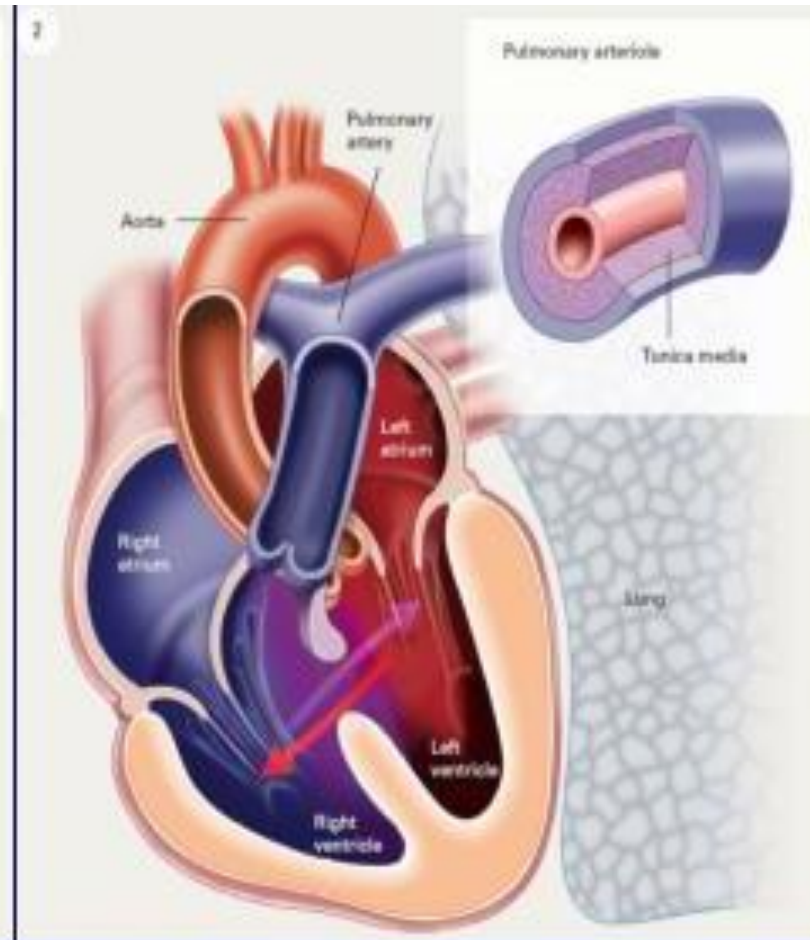
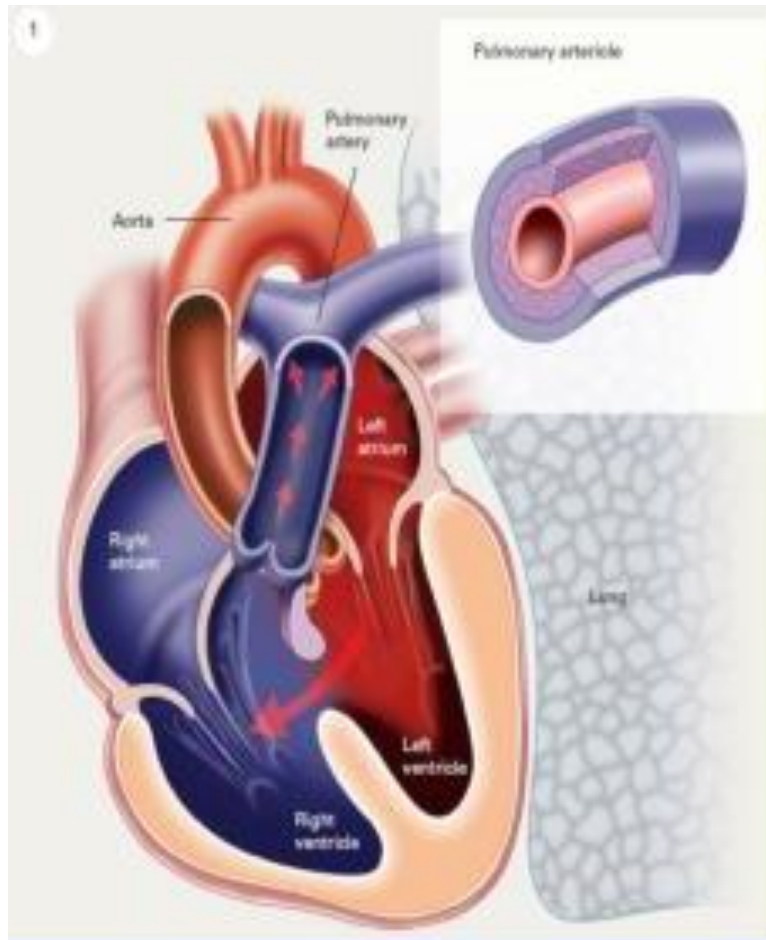
# Lung pathology



Normal pulmonary arteriols

Eisenmenger syndrome





**ASD, VSD or complex defect increases pulmonary blood flow via left-to-right shunt**

**Pulmonary resistance rises and results in bi-directional flow**

**Reversal of shunt: right-to-left → Eisenmenger syndrome**



## Eisenmenger's syndrome by anatomical location

**Age of onset : 25 – 49 years**

**Death : most in second or third decades, few cases survive to the fifth decade without transplantation.**

- **Complex anatomy** 44%
- **VSD** 37%
- **PDA** 12%
- **ASD** 7%



**Reversed shunt  
Right → Left  
(Eisenmenger)**

**Musculoskeletal**

**Clubbing → hypertrophic osteoarthropathy**

**Haematological**

**Secondary erythrocytosis**

**Hyperviscosity syndrome**

**Renal**

**Renal dysfunction**

**cardiac**

**Heart failure , Arrhythmias ( AF, AFL, VT)**

**Pulmonary**

**Pulmonary artery insitu thrombus , haemoptysis**

**Neurological**

**Paradoxical emboli ( septic or thrombus ) →  
CVA , brain abscess**



## What are the grades of clubbing?

- ❑ I. Softening of the nail beds and fluctuation
- ❑ II. Obliteration of the onychodermal angle
- ❑ III. Increased anteroposterior curvature
- ❑ IV. Increase in pulp tissue drumstick or parrot beak
- ❑ V. Hypertrophic osteoarthropathy









**Hyperviscosity  
syndrome**

**Musculoskeletal**

**Gout , myalgia**

**Haematological**

**Bleeding tendency**

**Renal**

**Uric acid stones**

**Hepatobiliary**

**Calcium bilirubinate gall stones**

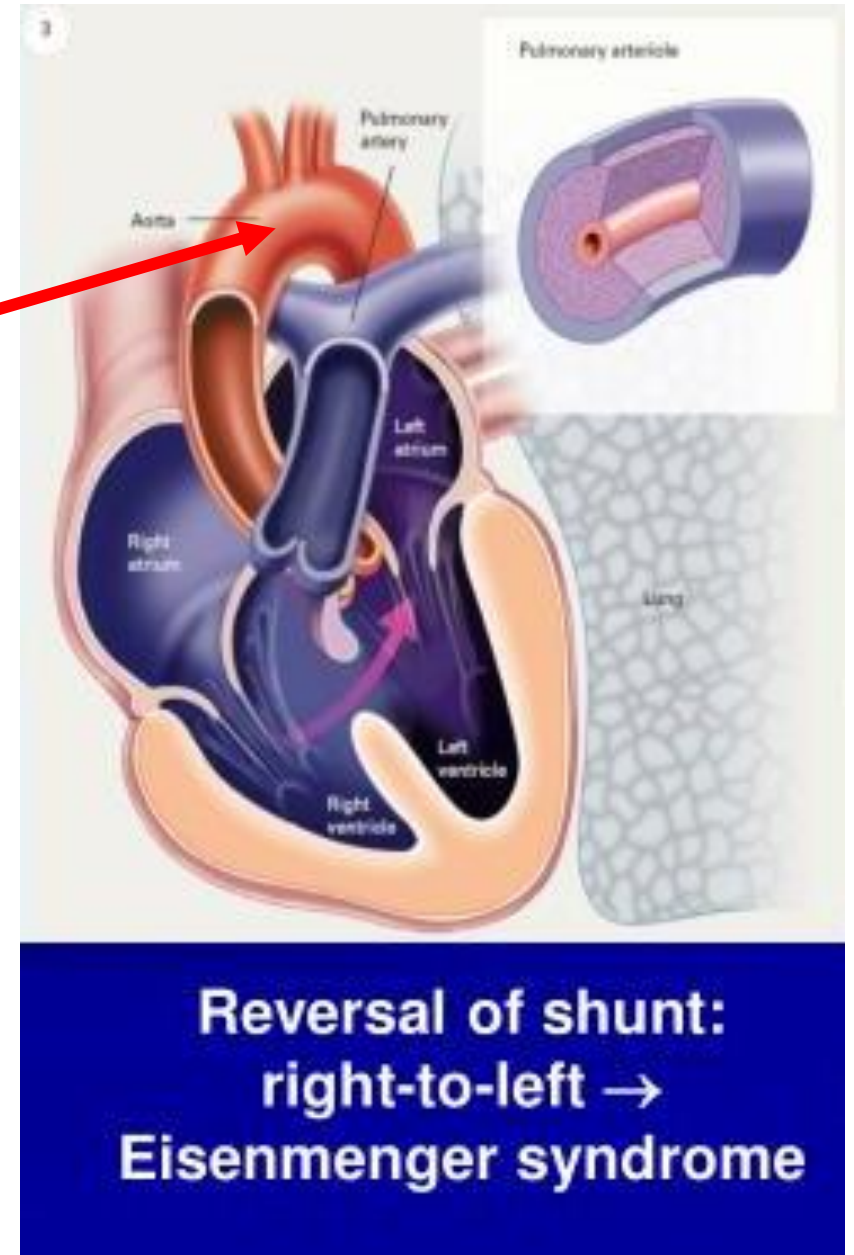
**Neurological**

**Slow mentation, irritability, headache,  
blurred vision, tinnitus , paraesthesia**

Vasodilatation, anaesthesia and pregnancy

abrupt decreases in left heart afterload →

Increase right-to-left shunting





## The causes of death in Eisenmenger pts:

- Sudden death (30%)
- Congestive heart failure (25%)
- Hemoptysis (15%)

## Enlarged chamber ( clinical , ECG , CXR , Echocardiography )



|             | RA | RV | PA  | LA     | LV     |
|-------------|----|----|-----|--------|--------|
| ASD         | ++ | ++ | ++  |        |        |
| VSD         |    |    | ++  | ++     | ++     |
| <b>PDA</b>  |    |    | ++  | ++     | ++     |
| Eisenmenger | ++ | ++ | +++ | + / ++ | + / ++ |

| MURMUR  | TIMING                          | SITE                             | RADIATION          | Thrill   | Other signs                               |
|---|---------------------------------|----------------------------------|--------------------|----------|---|
| <b>ASD</b>  | Ejection systolic               | pulmonary area                   |                    | -        | wide fixed spitted S2                     |
| <b>Often + severe secondary TR</b>                          | Pansystolic                     | Tricuspid area                   |                    |          |   |
| <b>VSD</b>  | Pansystolic                     | Left sternal border              | Across the sternum | +        |   |
| <b>PDA</b>  | Continuous machinery            | Pulmonary area                   |                    | +        | Large volume pulse<br>Wide pulse pressure |
| <b><u>Eisenmenger</u><br/>secondary TR<br/>secondary PR</b> | Pan-systolic<br>Early diastolic | Tricuspid area<br>Pulmonary area |                    | -/+<br>- |   |

**ASD VSD PDA :**

A small defect often produces a loud murmur

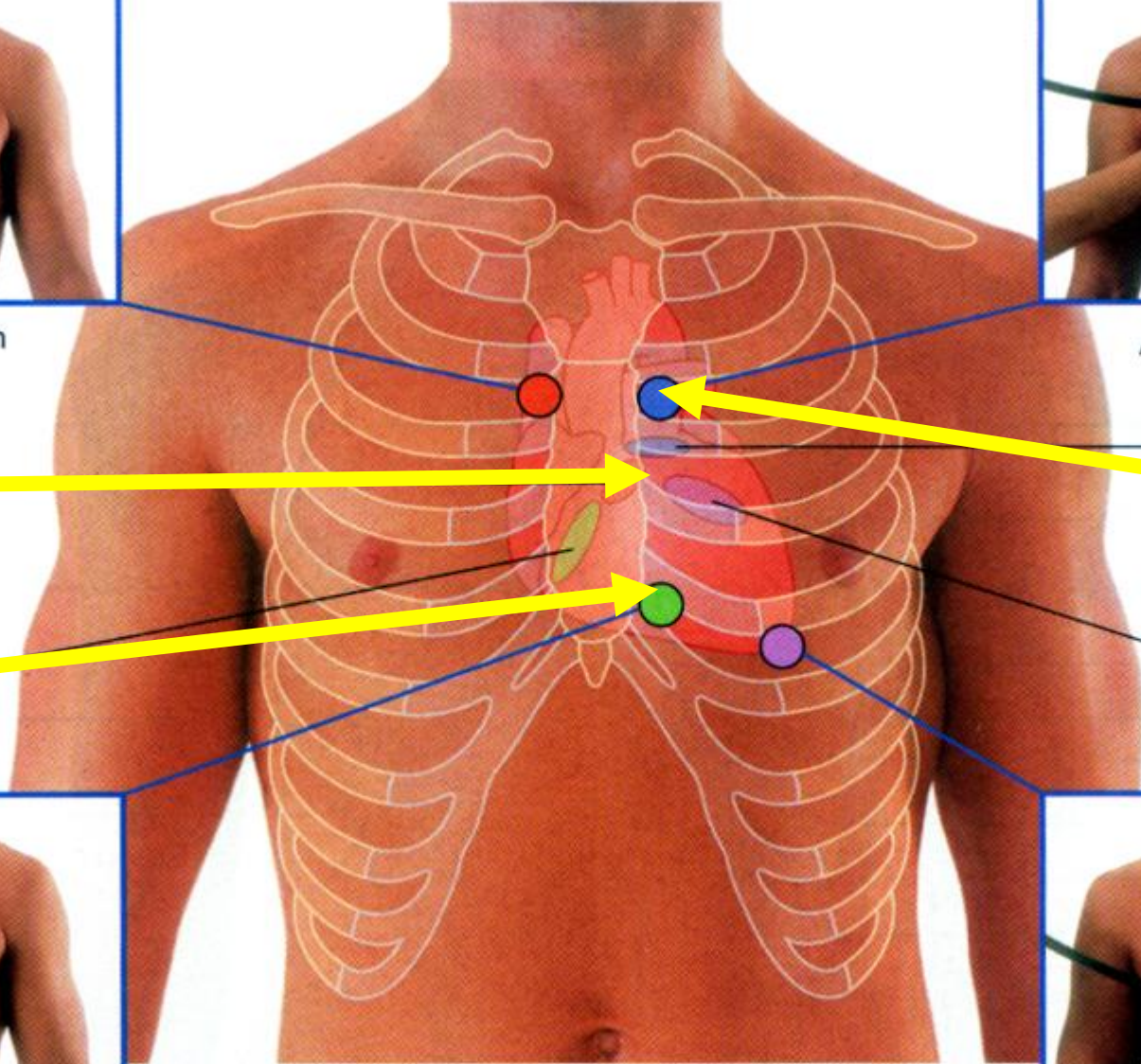
A large defect produces a softer murmur



Auscultation position for aortic valve



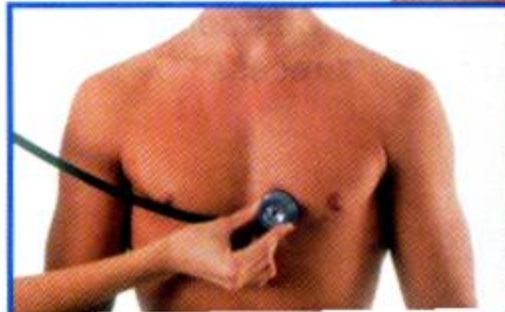
Auscultation position for pulmonary valve



**VSD**

**PDA  
ASD  
PS  
Innocent murmur**

**TR by Eisenmenger  
TR secondary to large ASD**



Mitral valve



# Investigations

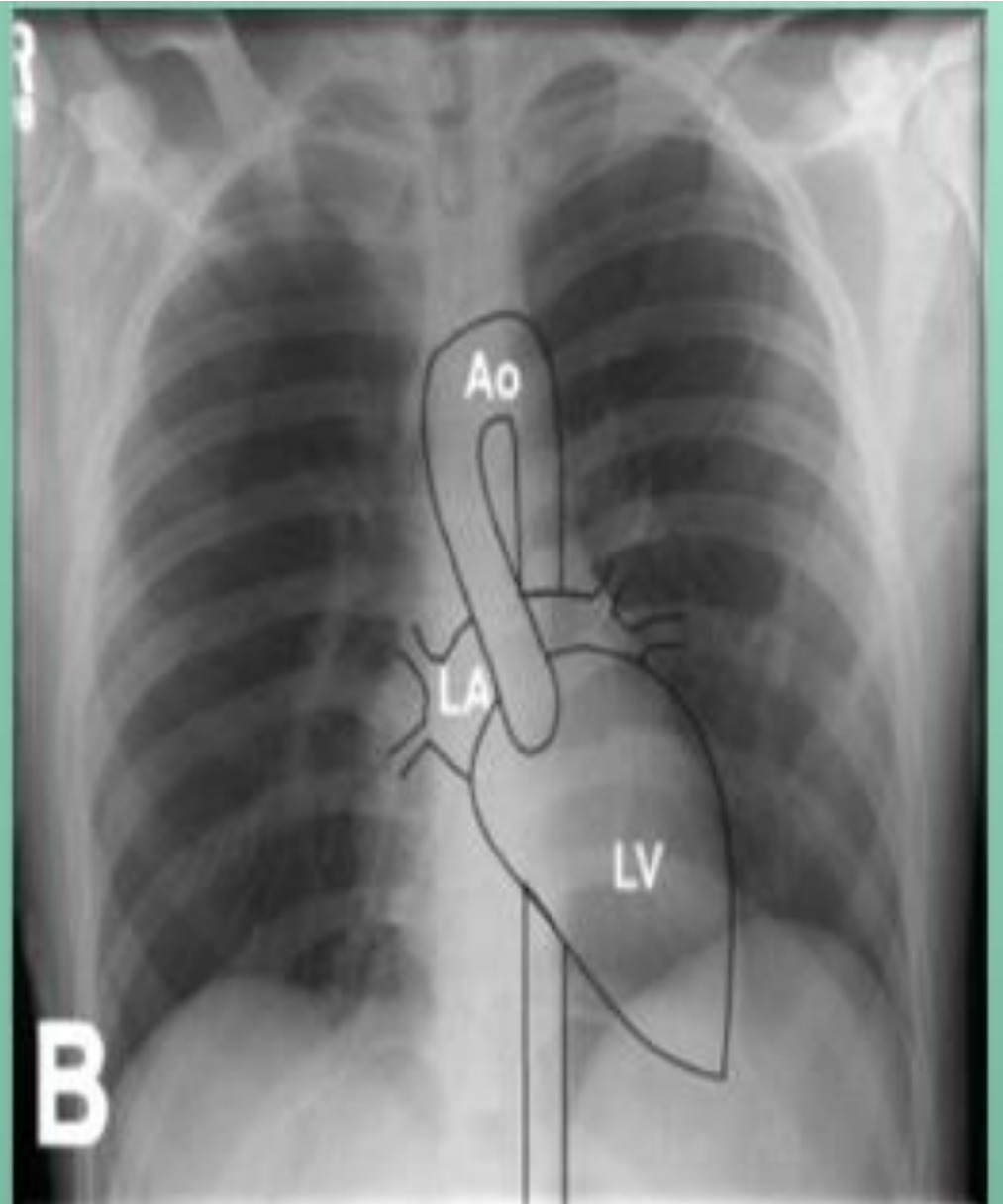
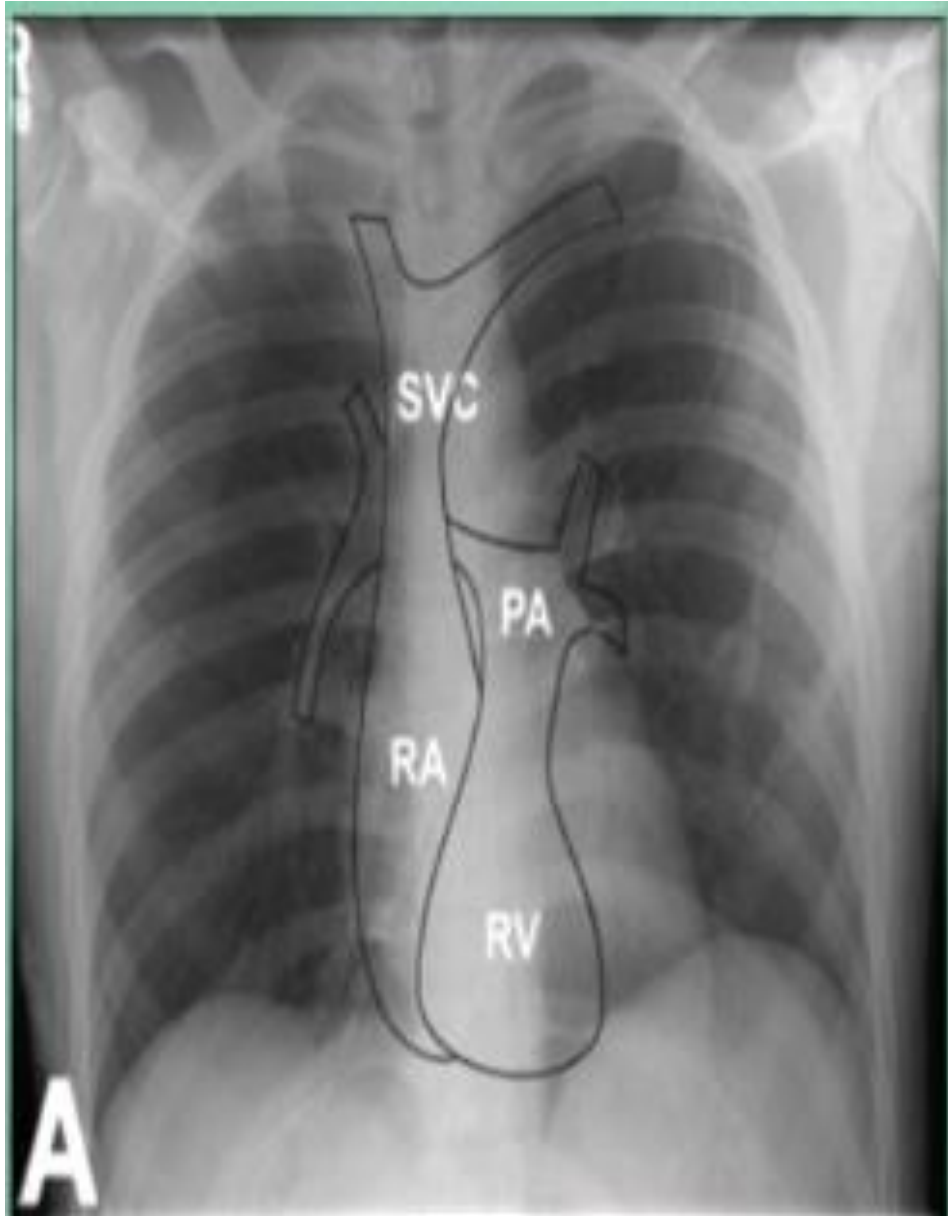
Doppler echocardiography

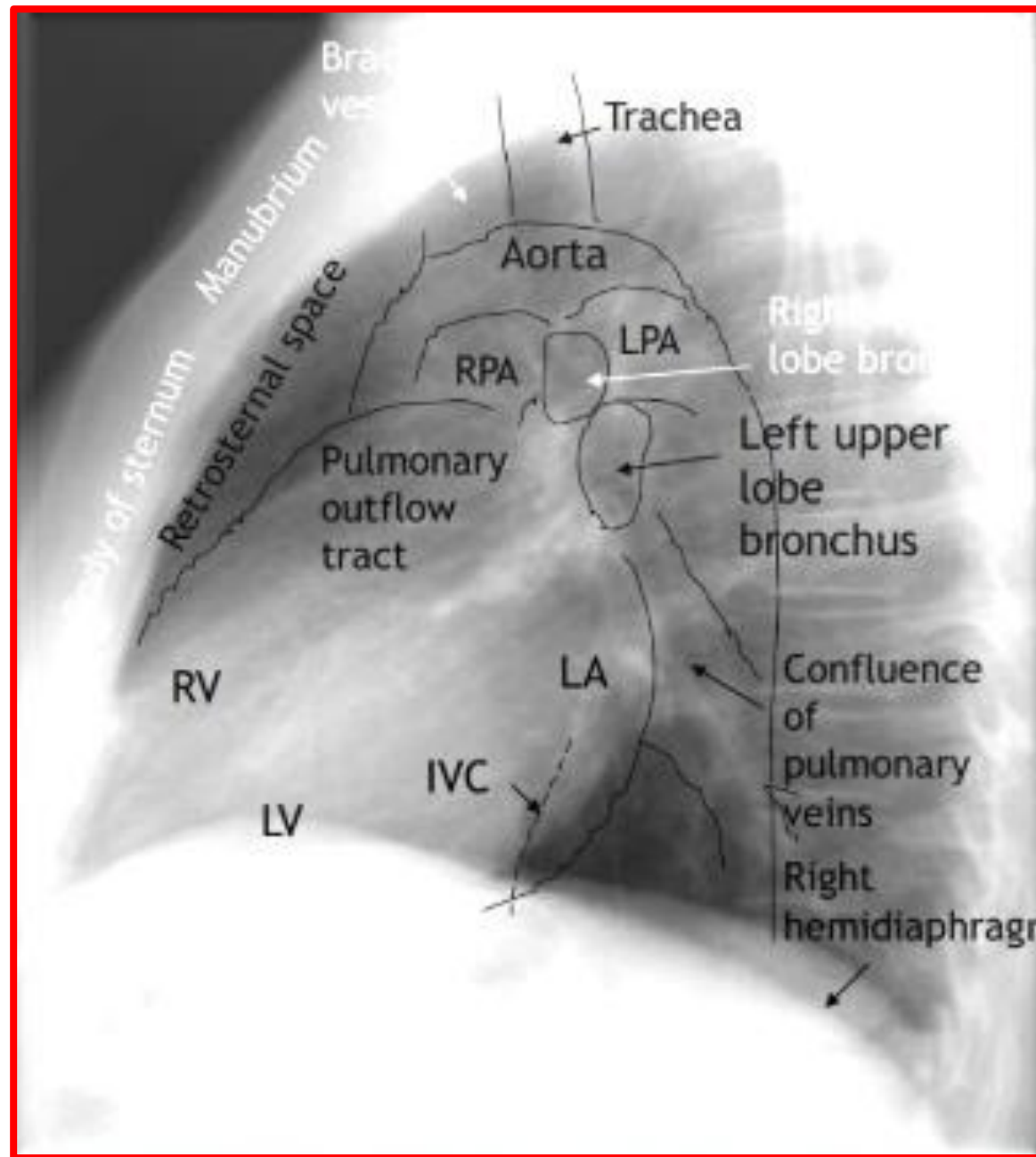
TOE

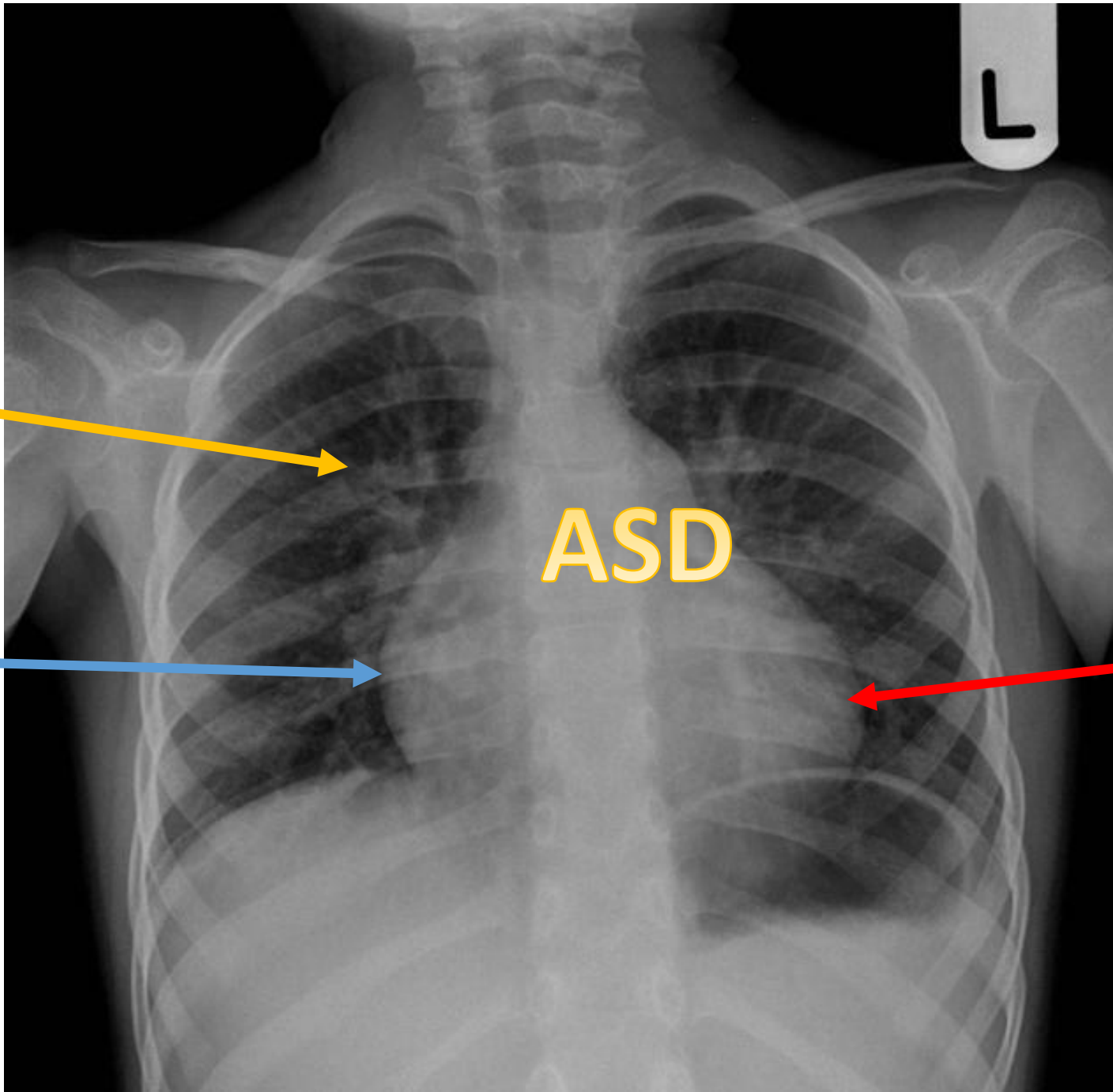
CXR

ECG

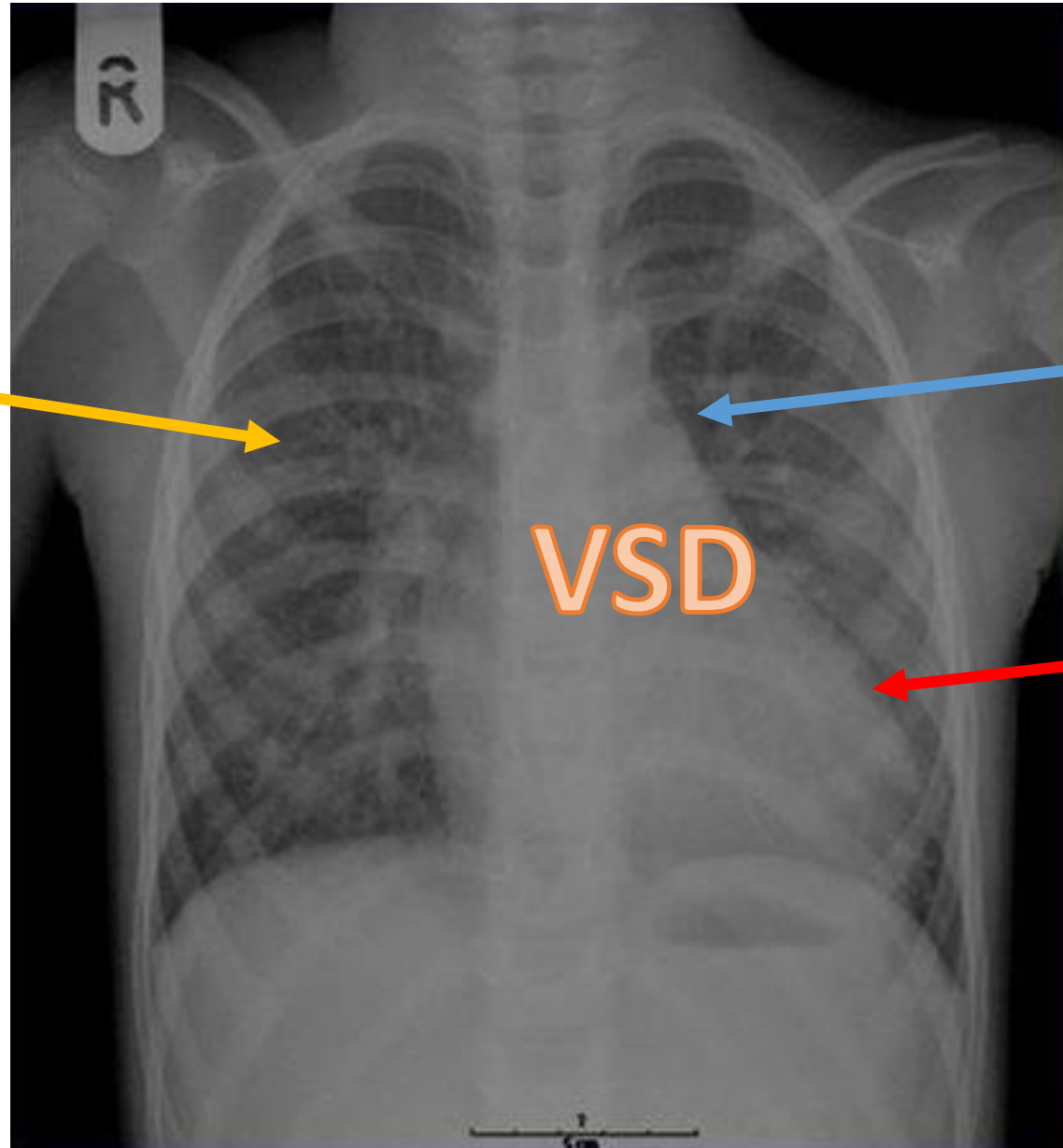




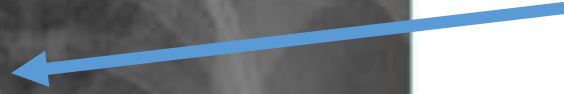


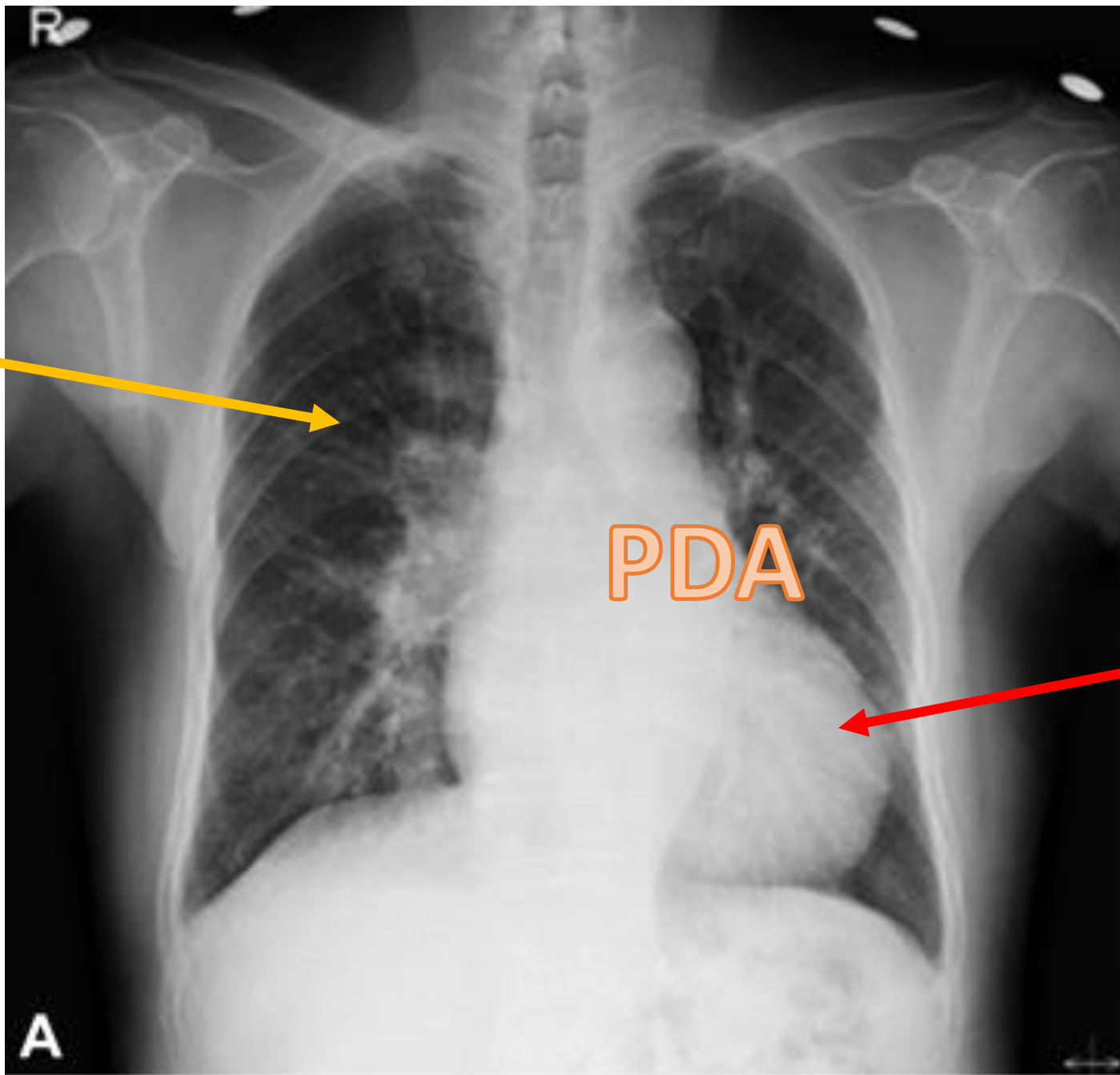


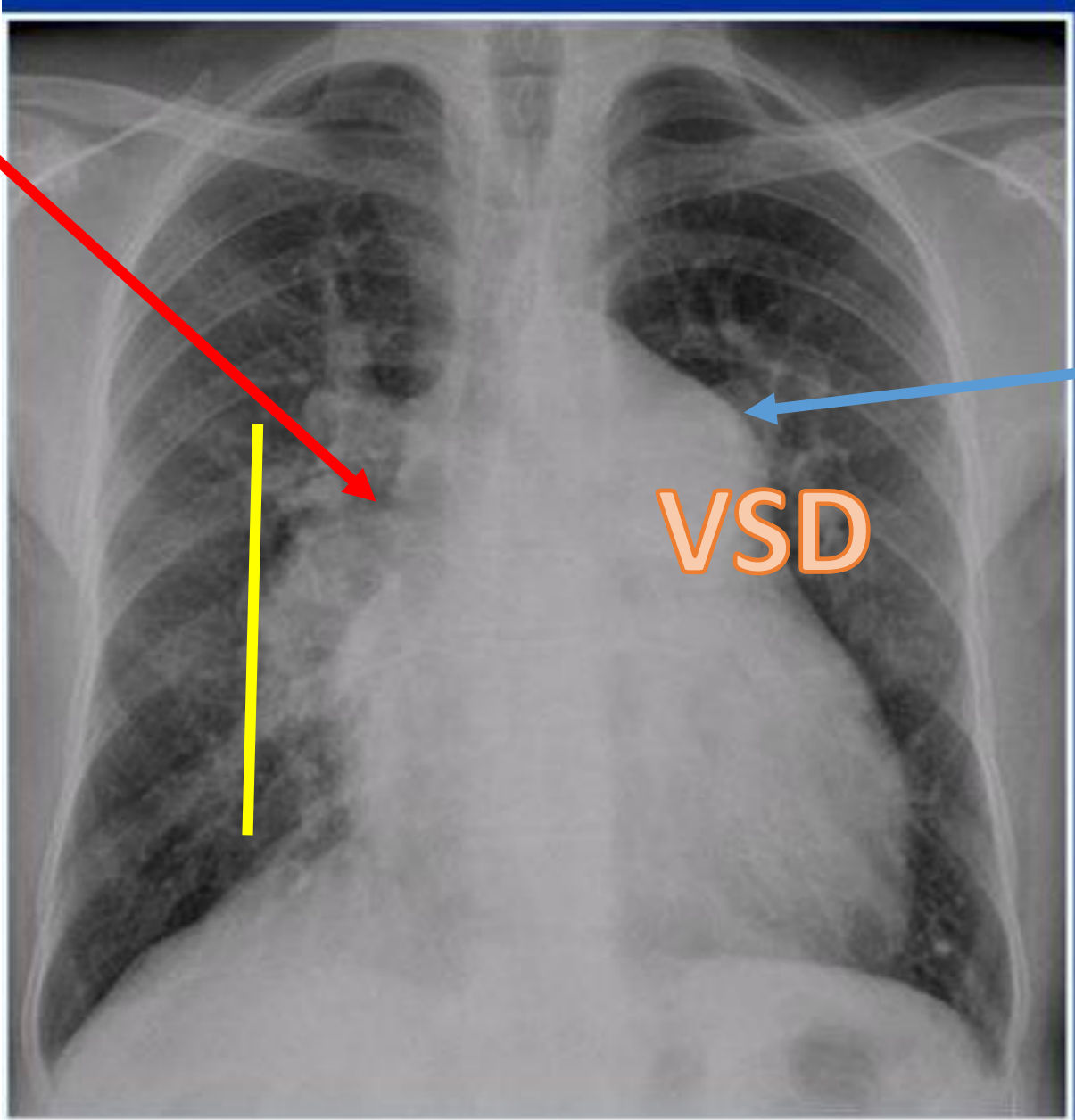
ASD



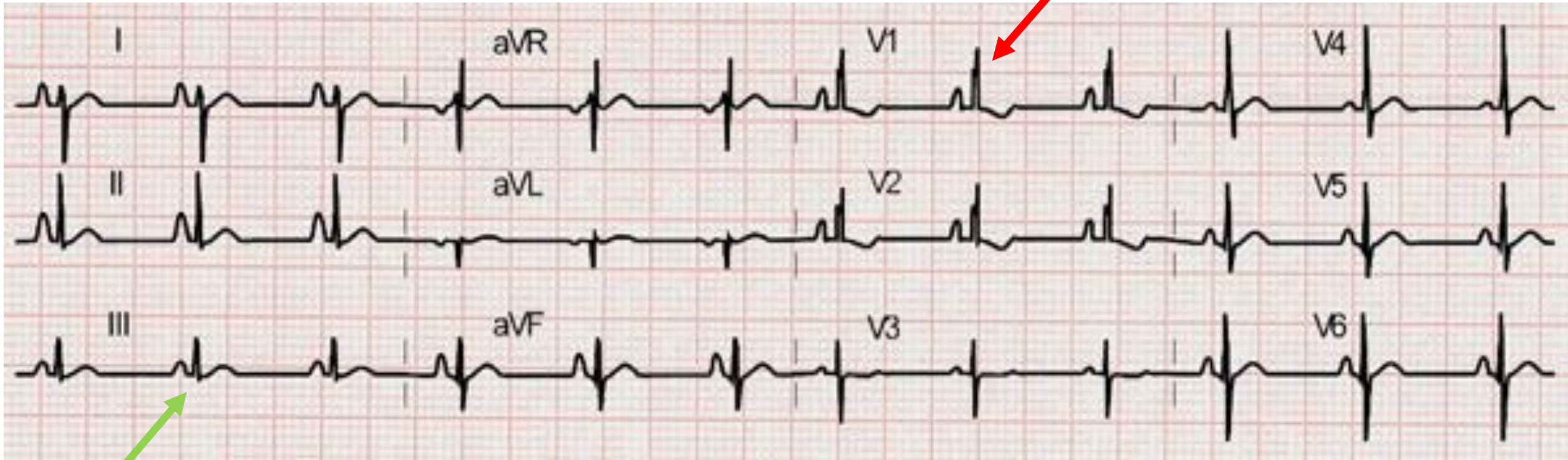
VSD



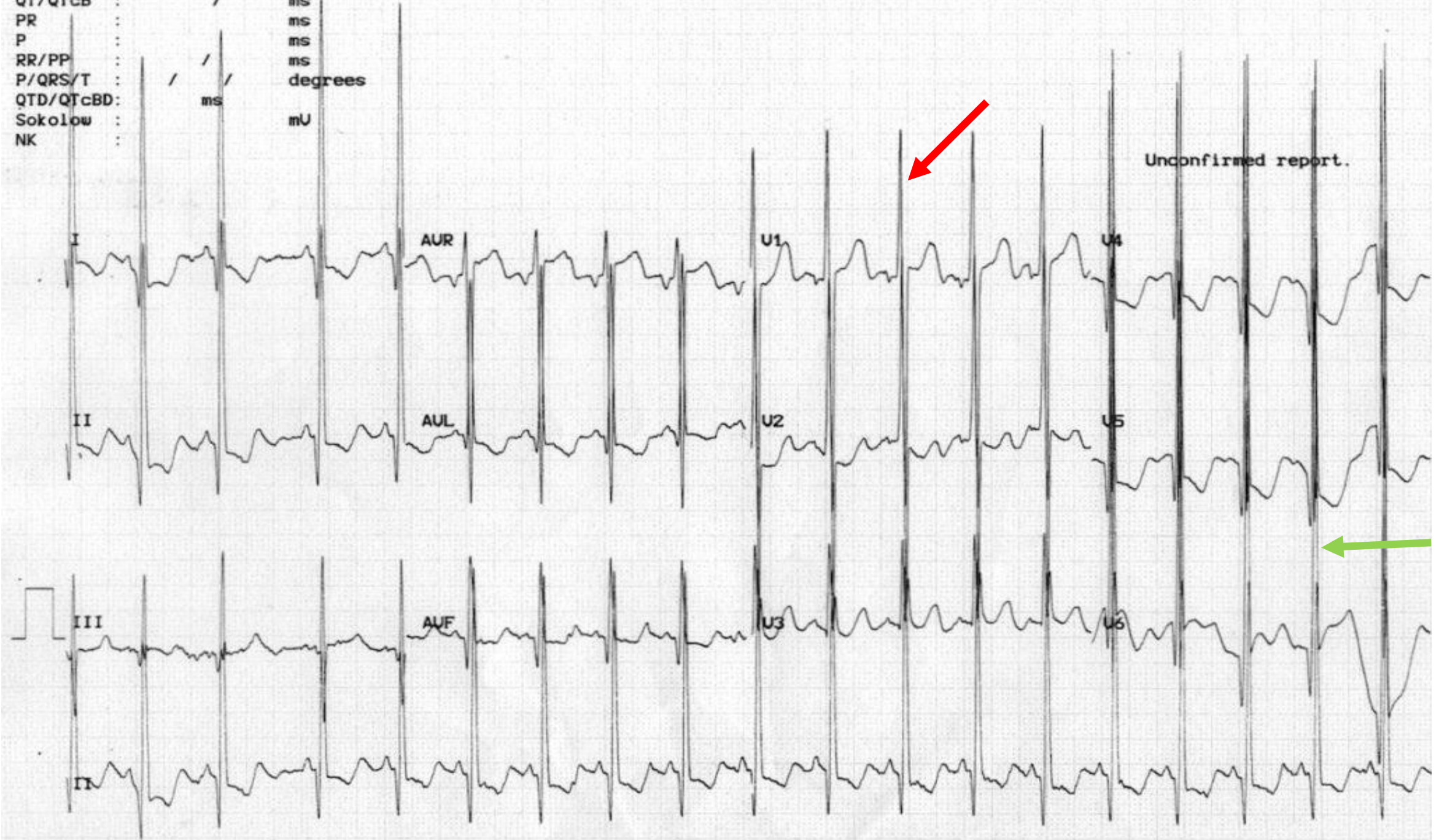




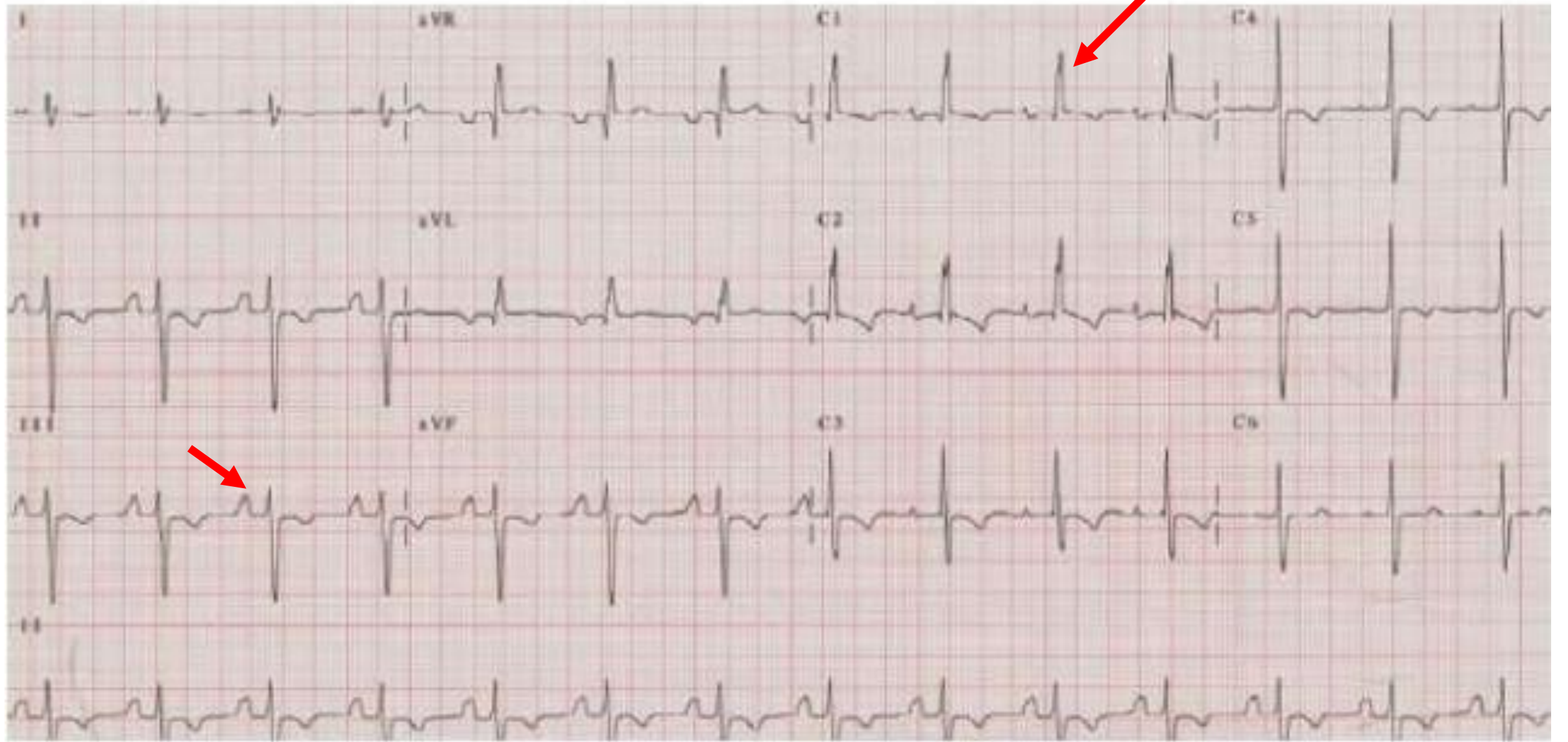
VSD



QT/QTcB : / ms  
PR : / ms  
P : / ms  
RR/PP : / ms  
P/QRS/T : / / degrees  
QTD/QTcBD : ms  
Sokolow : mU  
NK :







# Persistent ductus arteriosus

more common in females.

## Clinical features

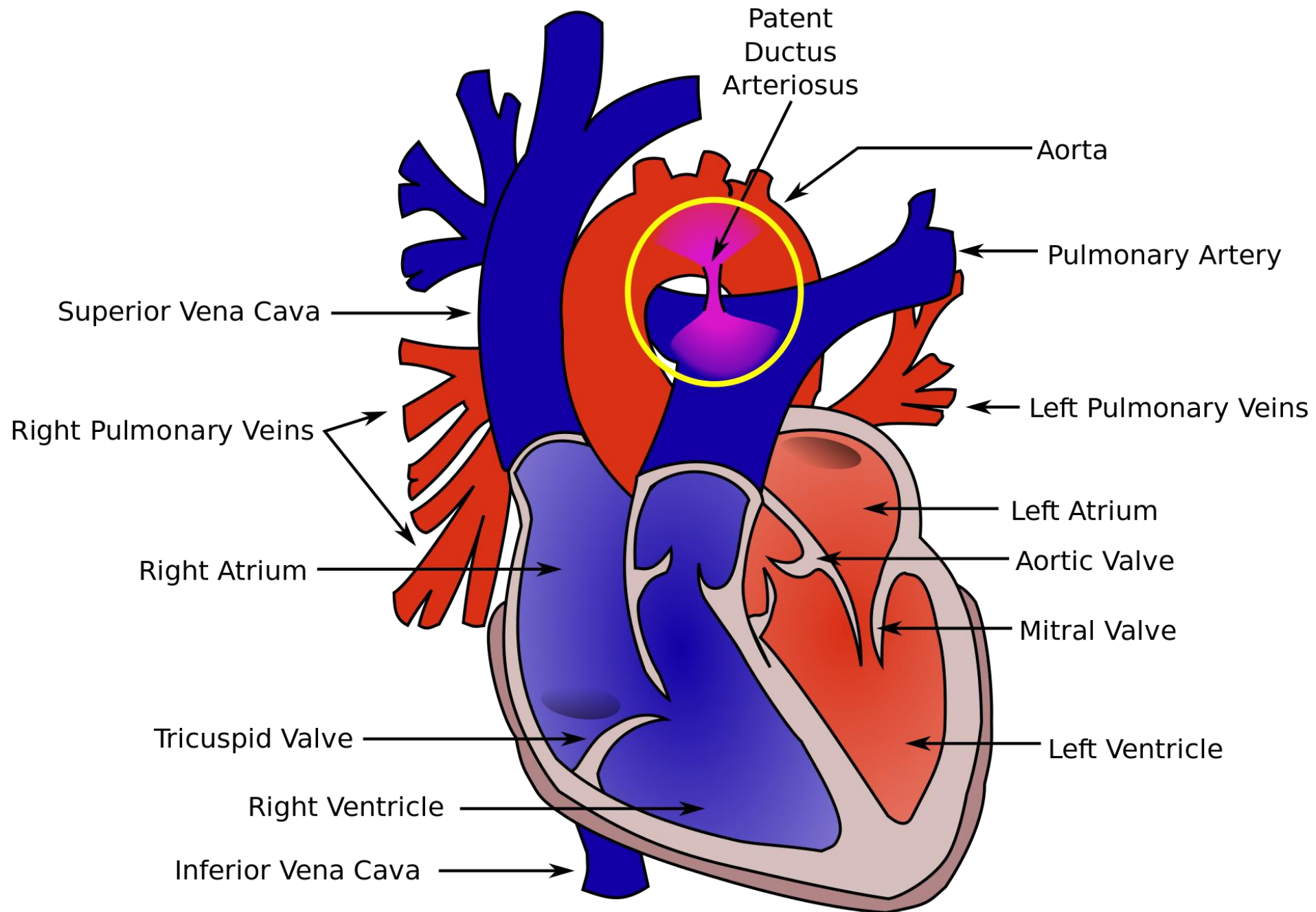
small shunts there → no symptoms for years

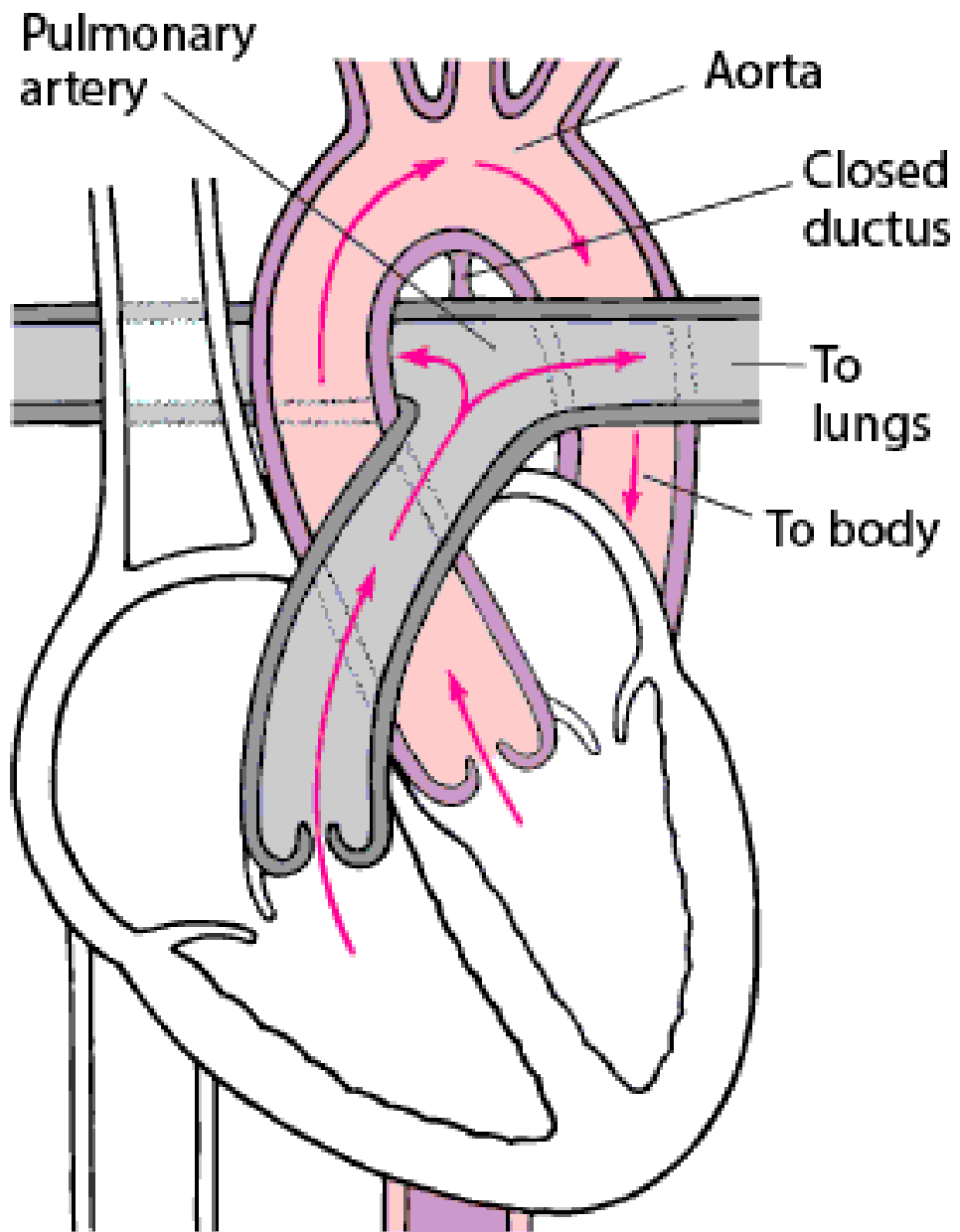
Large → growth and development may be retarded.

Dyspnoea being the first symptom.

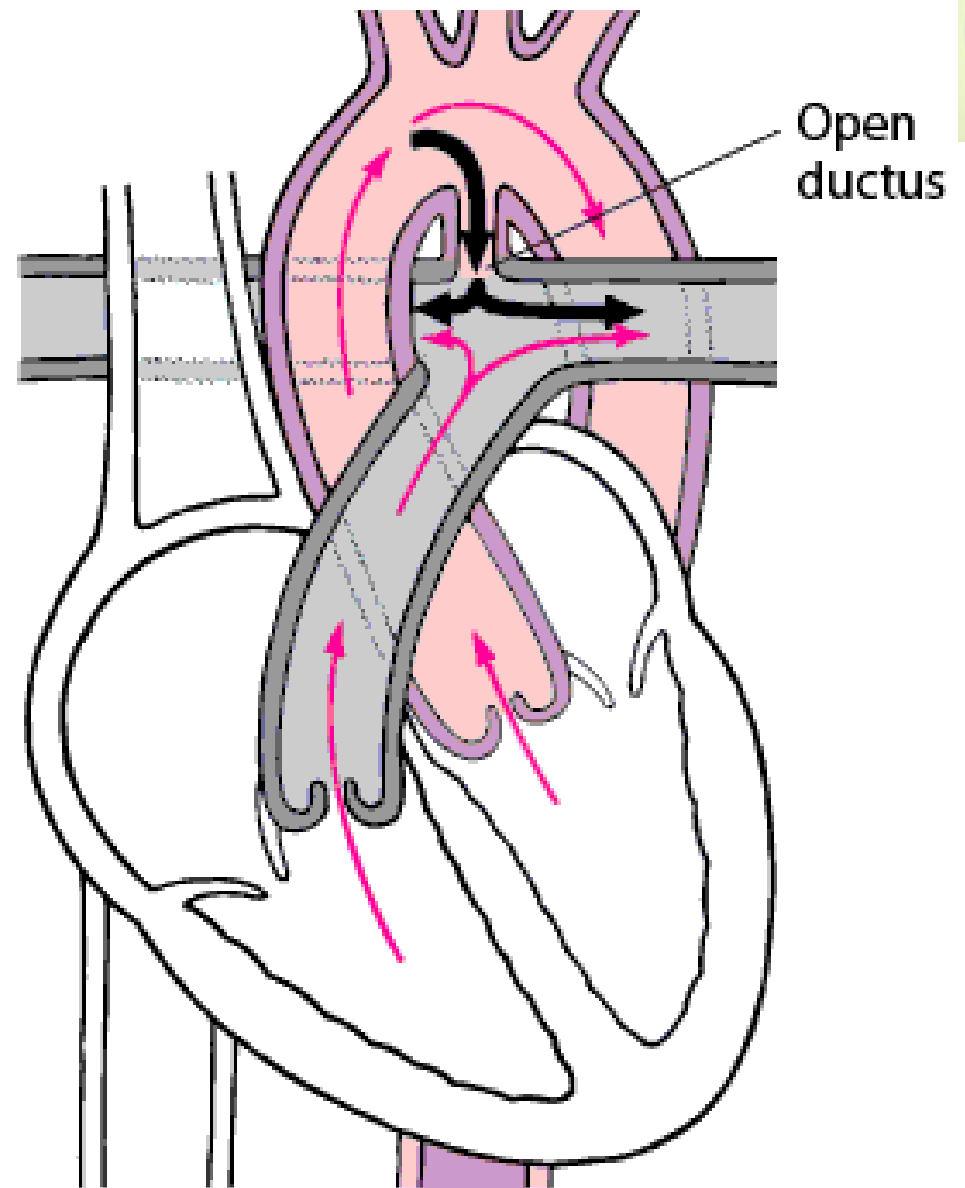
With progressive pulmonary hypertension the **murmur become quieter and shorter** then disappear when Eisenmenger syndrome develops



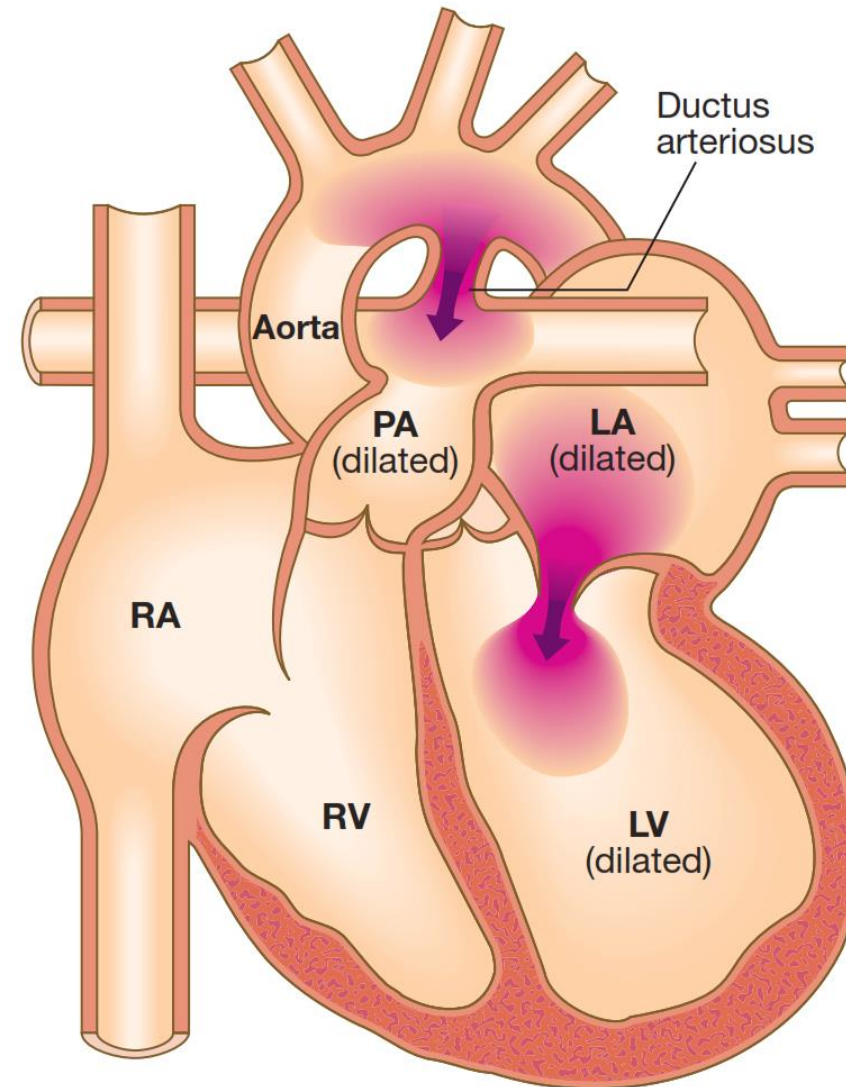




Normal Circulation



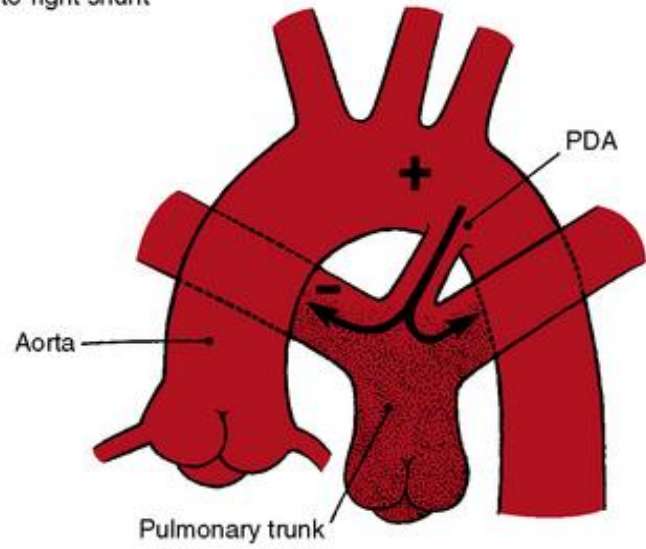
Patent Ductus Arteriosus



**Fig. 16.92 Persistent ductus arteriosus.** There is a connection between the aorta and the pulmonary artery with left-to-right shunting. (LA = left atrium; LV = left ventricle; PA = pulmonary artery; RA = right atrium; RV = right ventricle)



Left-to-right shunt



Right-to-left shunt

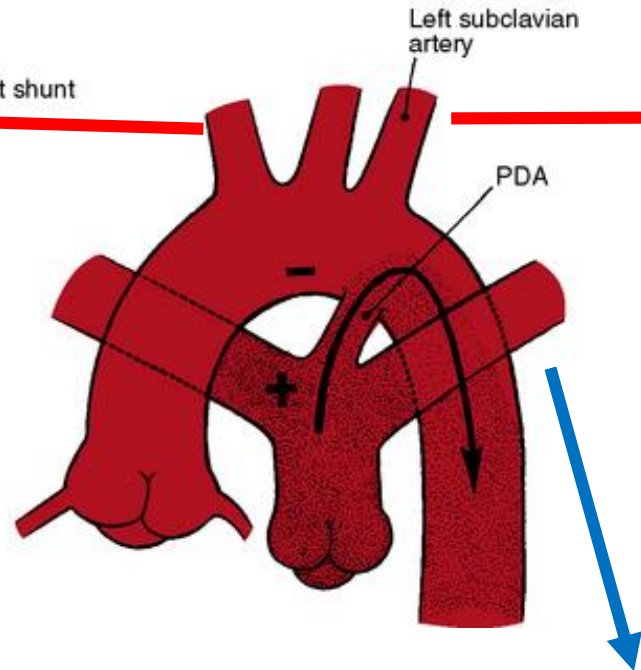




Figure 1. Differential cyanosis and clubbing in PDA Eisenmenger

Differential cyanosis and clubbing in PDA Eisenmenger

# Atrial septal defect

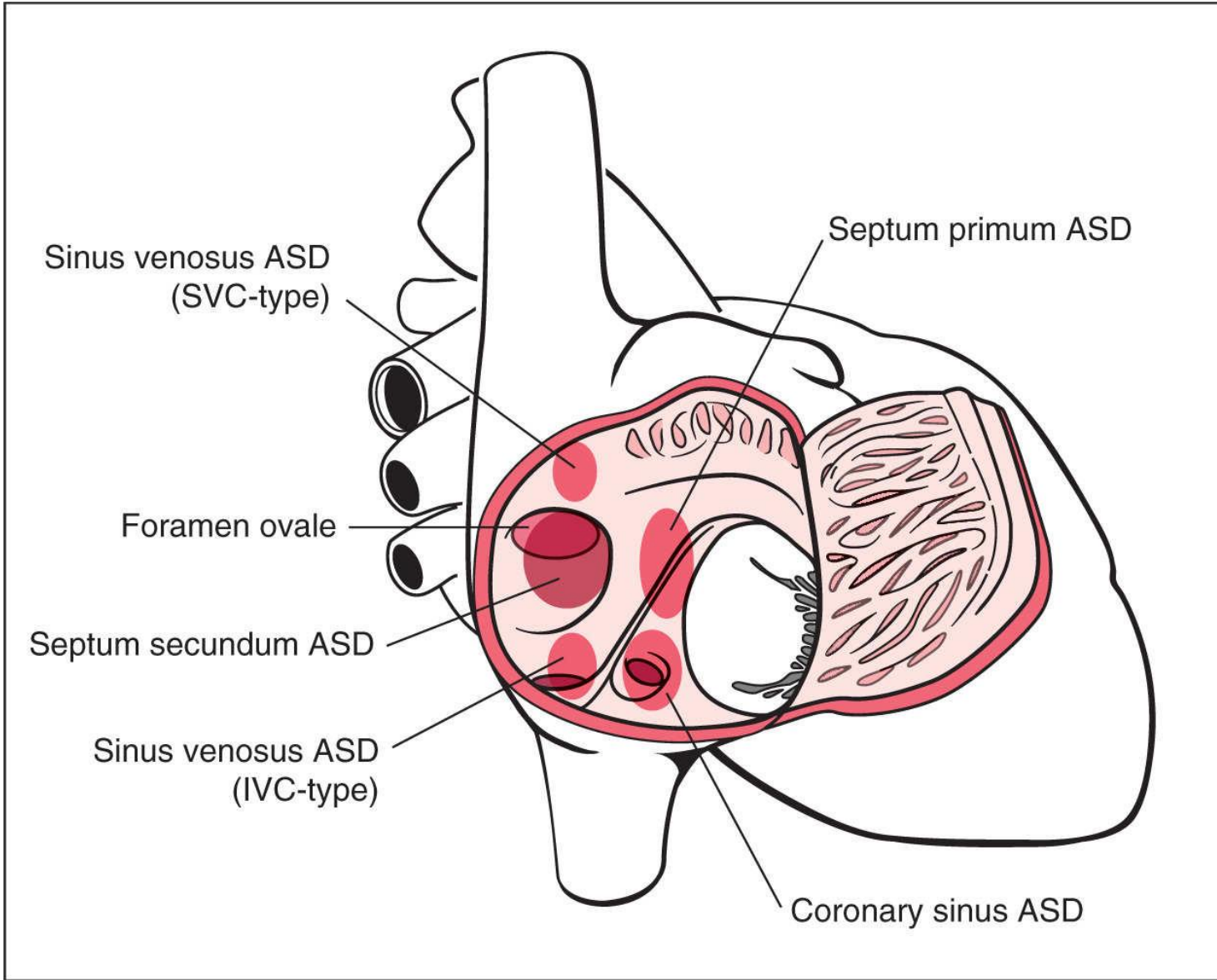
Twice as frequently in females.

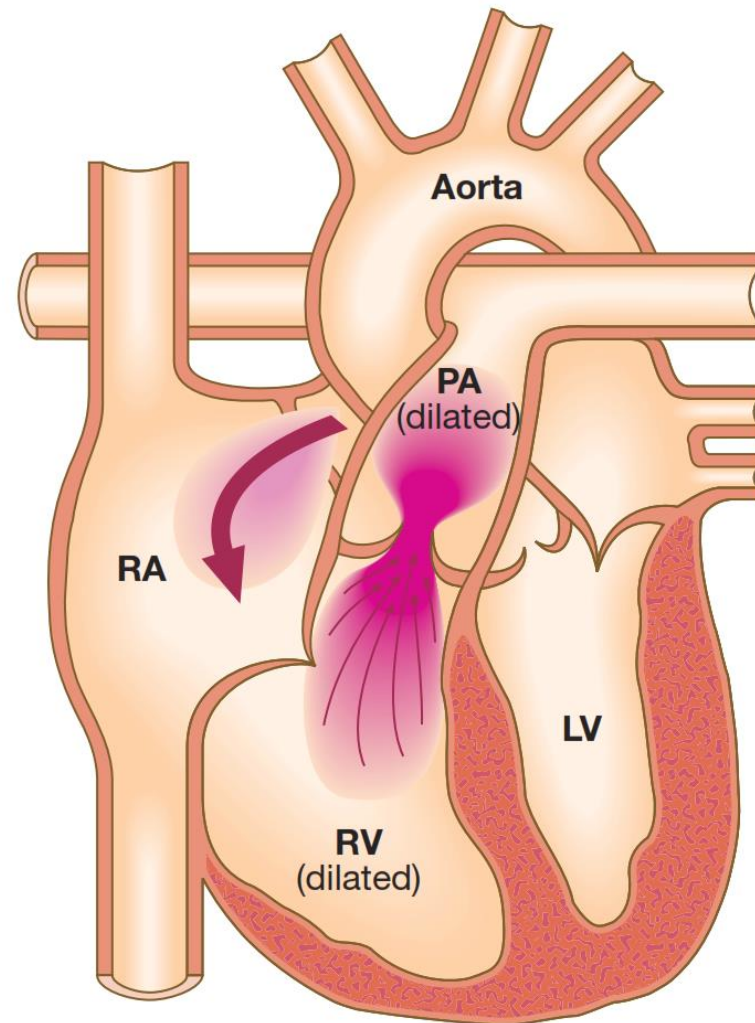


## Clinical features

Most children are asymptomatic for many years and the condition is often detected at routine clinical examination or following a chest X-ray.

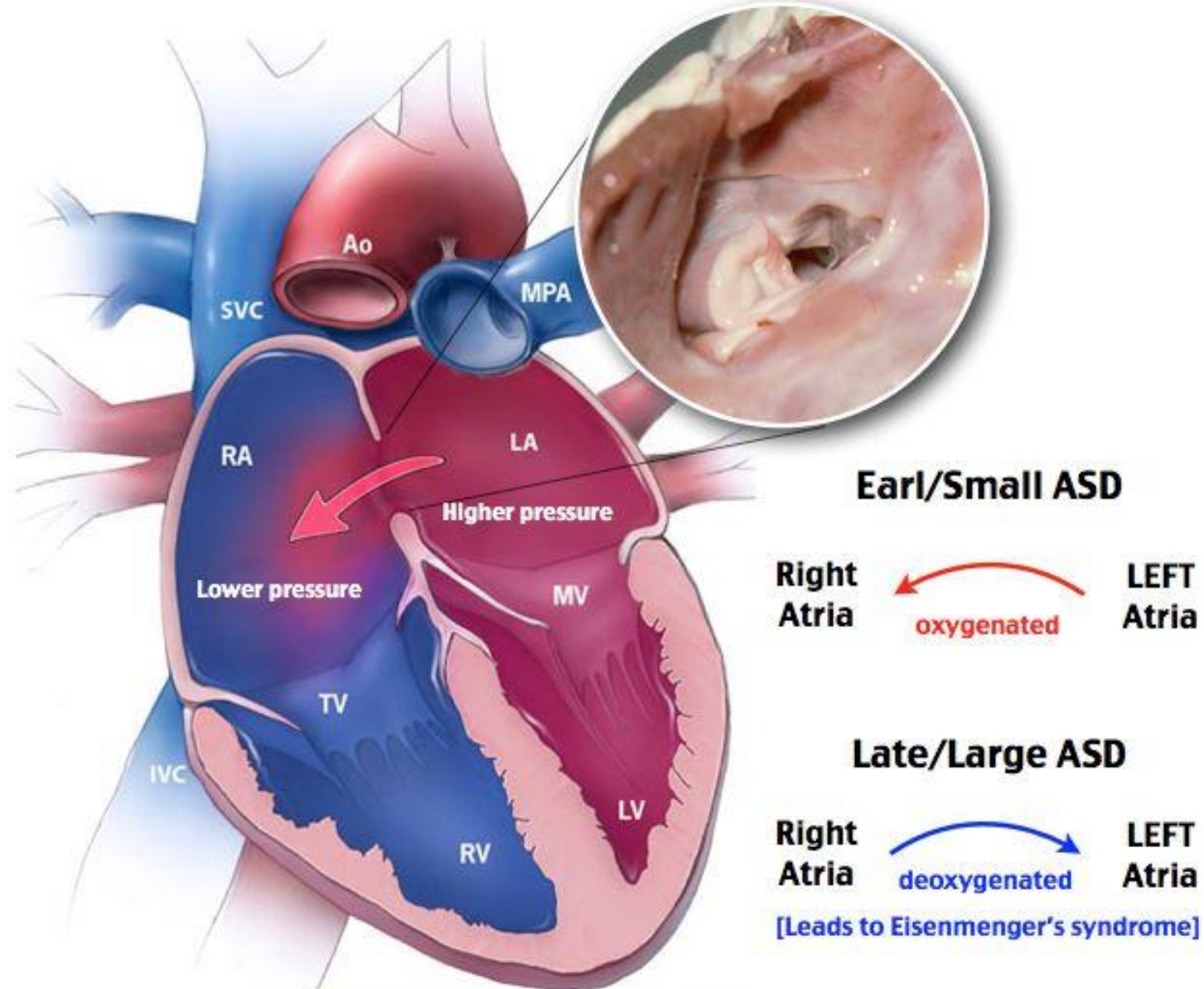






**Fig. 16.94 Atrial septal defect.** Blood flows across the atrial septum (arrow) from left to right. The murmur is produced by increased flow velocity across the pulmonary valve, as a result of left-to-right shunting and a large stroke volume. The density of shading is proportional to velocity of blood flow. (LV = left ventricle; PA = pulmonary artery; RA = right atrium; RV = right ventricle)

# Atrial Septal Defect (ASD)



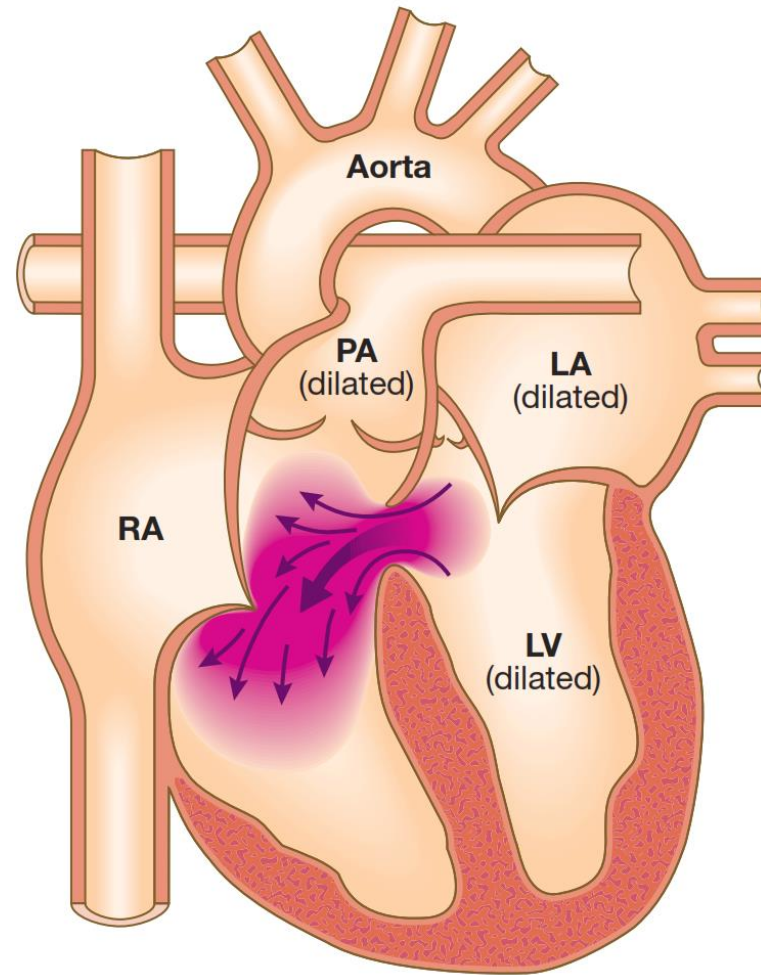
Fixed, Split S2 on cardiac auscultation



# Ventricular septal defect

the most common congenital cardiac defect

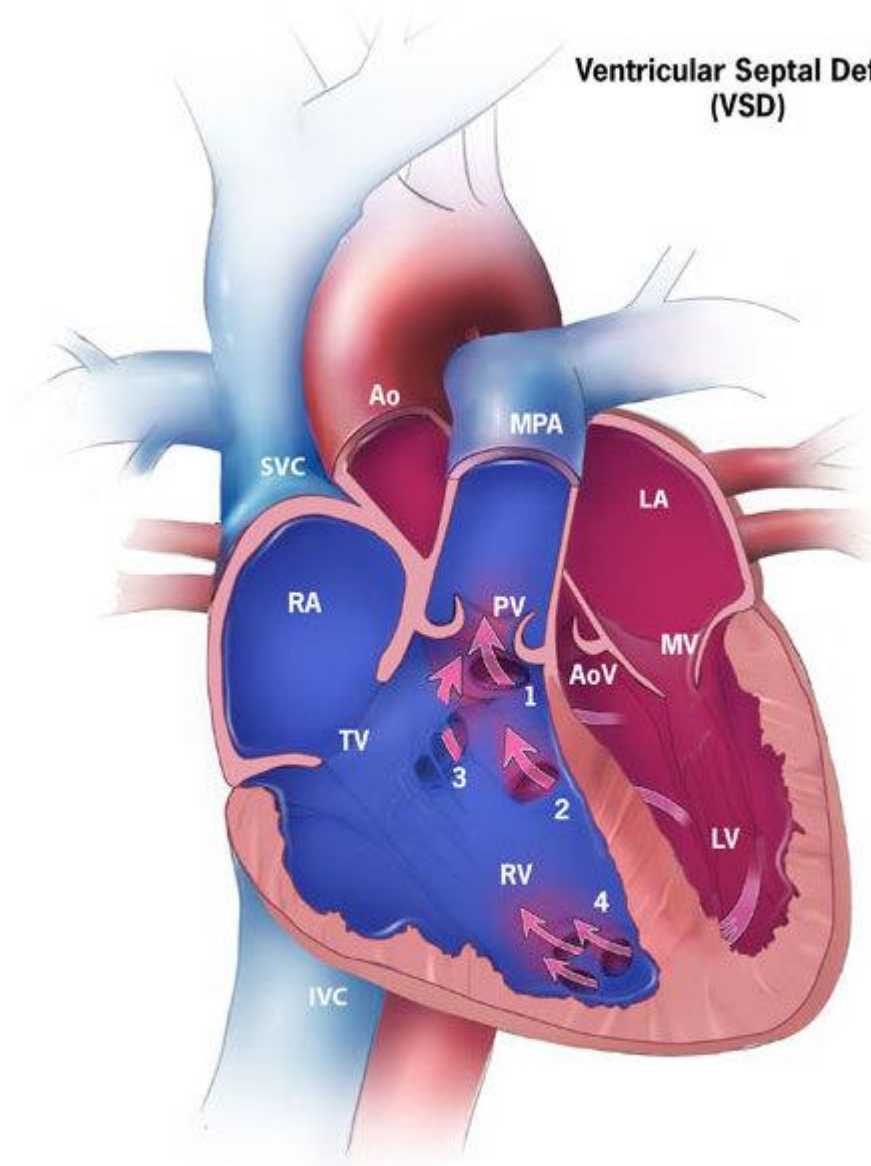
isolated or part of complex congenital heart disease.



**Fig. 16.97 Ventricular septal defect.** In this example, a large left-to-right shunt (arrows) has resulted in chamber enlargement. (LA = left atrium; LV = left ventricle; PA = pulmonary artery; RA = right atrium)



## Ventricular Septal Defect (VSD)



RA. Right Atrium  
RV. Right Ventricle  
LA. Left Atrium  
LV. Left Ventricle

SVC. Superior Vena Cava  
IVC. Inferior Vena Cava  
MPA. Main Pulmonary Artery  
Ao. Aorta

TV. Tricuspid Valve  
MV. Mitral Valve  
PV. Pulmonary Valve  
AoV. Aortic Valve

1. Conoventricular, malaligned  
2. perimembranous  
3. inlet  
4. muscular

## Management ( VSD, ASD, PDA )



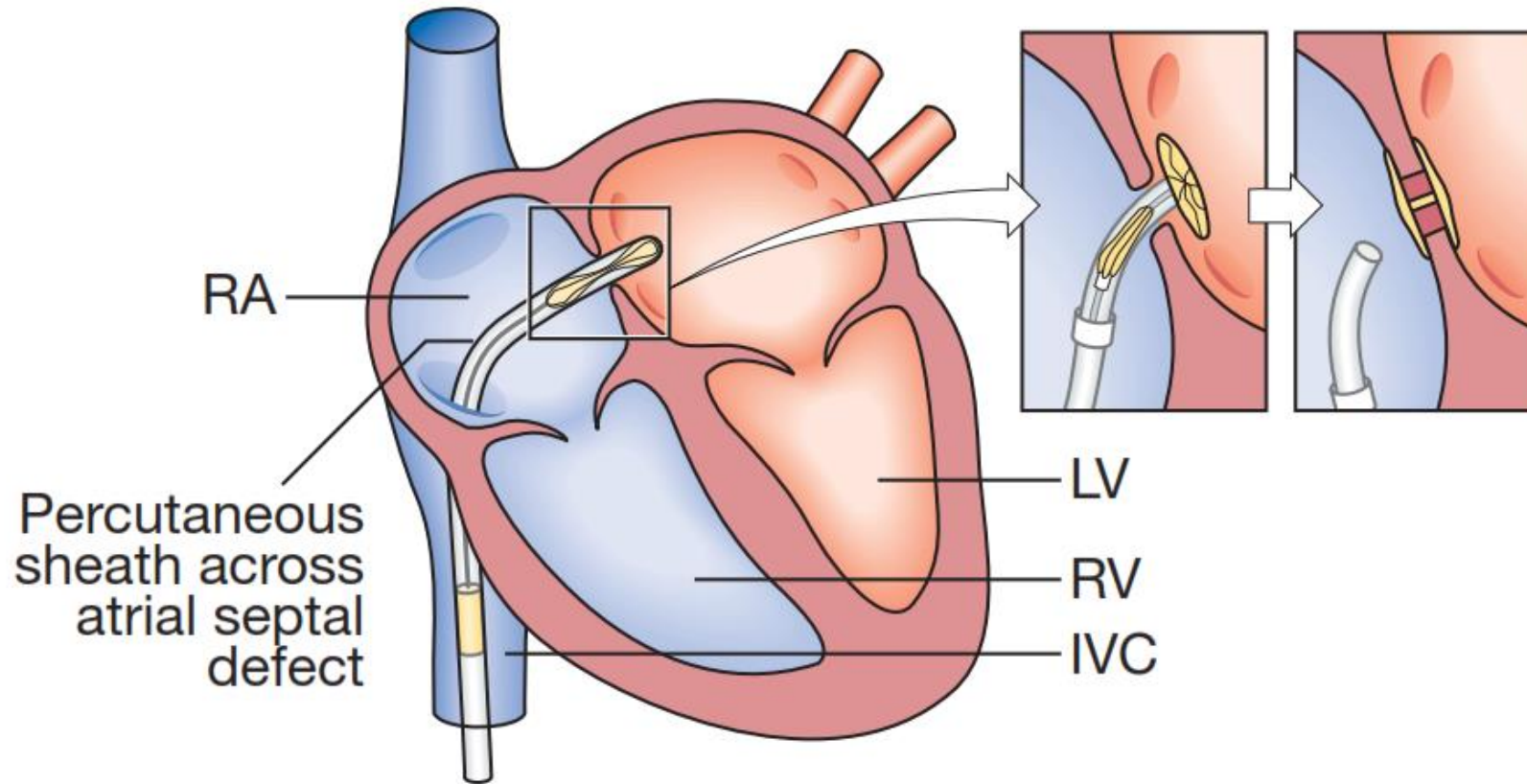
### Small defect

VSD PDA : follow-up

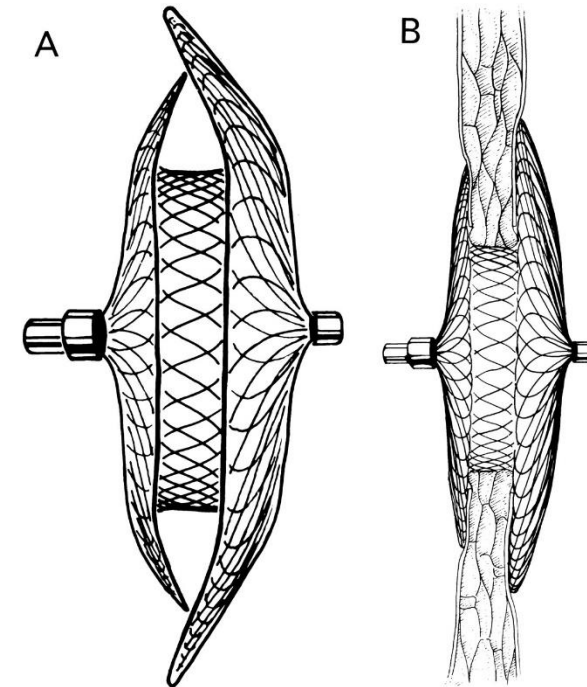
PDA : Closure to reduce the risk of endocarditis.

### Large defect:

- Percutaneous closure ( occlusive devices ) : ASD ( secundum only ) , most cases of PDA , some cases of VSD
- Surgical repair : Surgical closure is contraindicated in fully developed Eisenmenger's



**Fig. 16.96 Percutaneous closure of atrial septal defect.** The closure device is delivered across the interatrial septum and a disc deployed on either side to seal the defect. (IVC = inferior vena cava; LV = left ventricle; PA = pulmonary artery; RA = right atrium; RV = right ventricle)





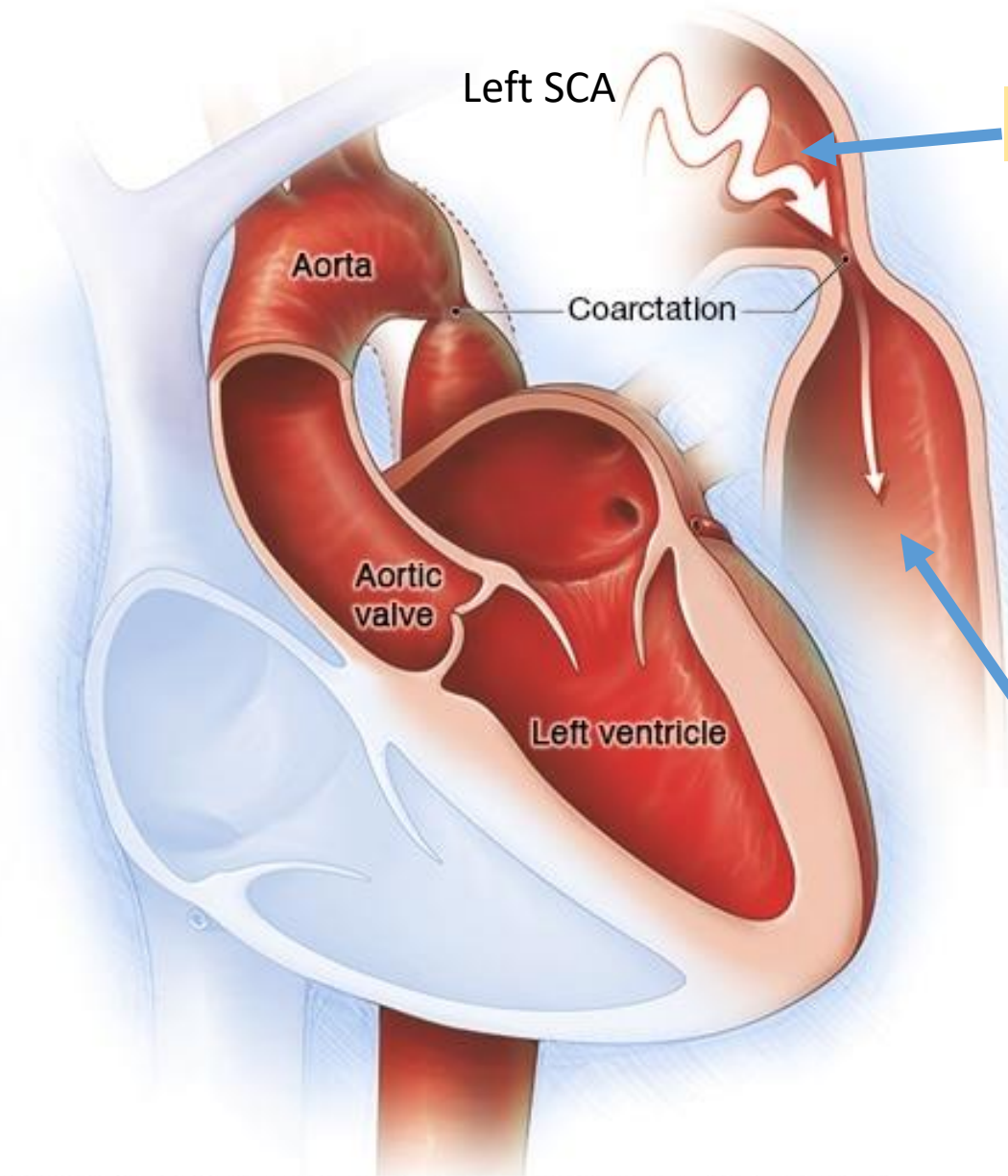
# Coarctation of the aorta



- **Genetic** : autosomal codominant : males 2:1 females  
Association : **bicuspid aortic valve ( upto 50% )** and cerebral '**berry**' aneurysms ( upto 10%)
- **Acquired** : rare : trauma , atherosclerosis or Takayasu's disease

## Other clinical features

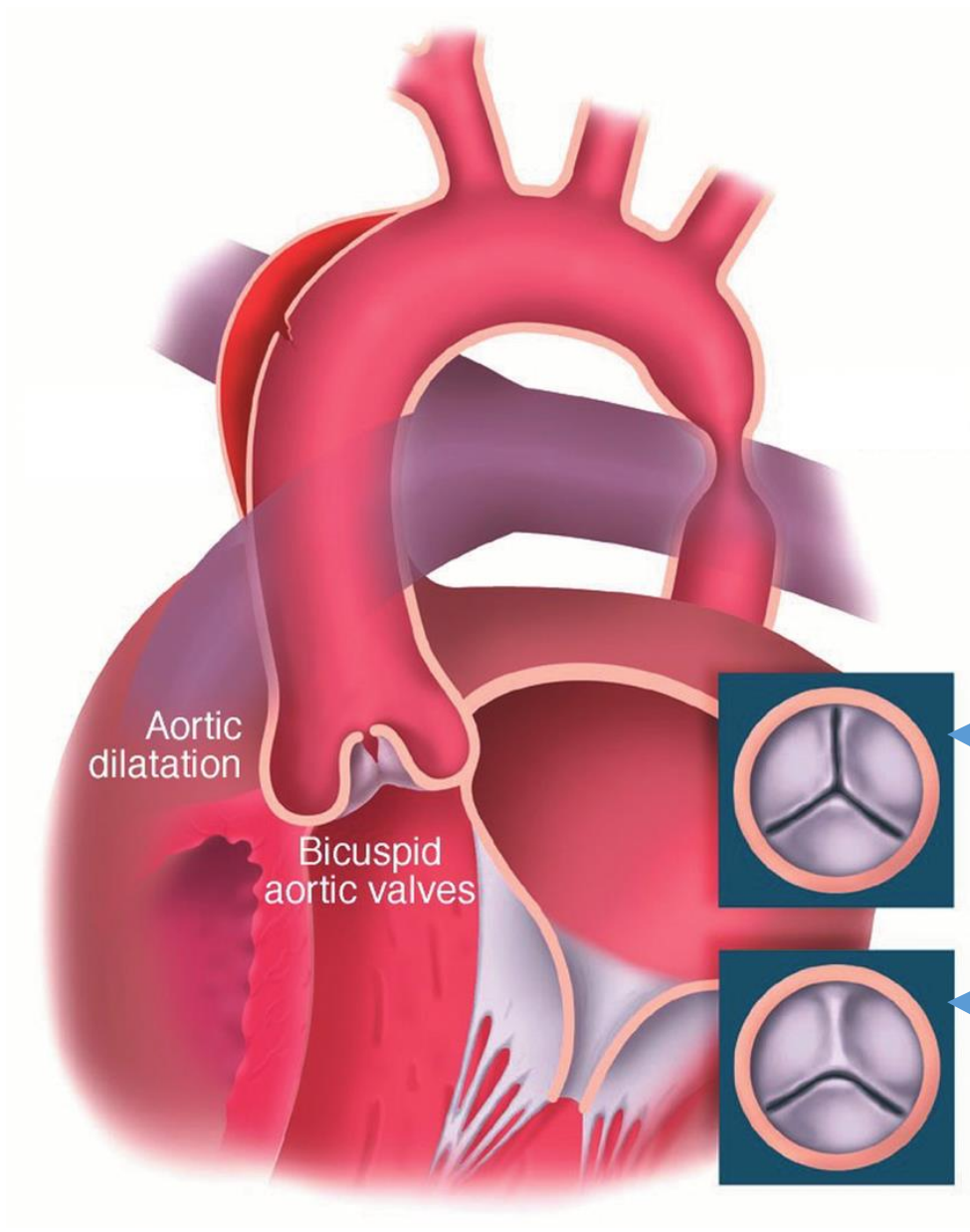
- A **systolic murmur** is usually heard posteriorly inter-scapular region
- Collaterals may result in **continuous murmur** best heard over the back
- If associated with bicuspid aortic valve : **ejection click, AS and/or AR murmurs**



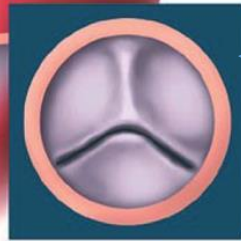
Hypertension → LVH → LV dysfunction

BP difference more than 20 mmHG  
RR delay  
RF delay

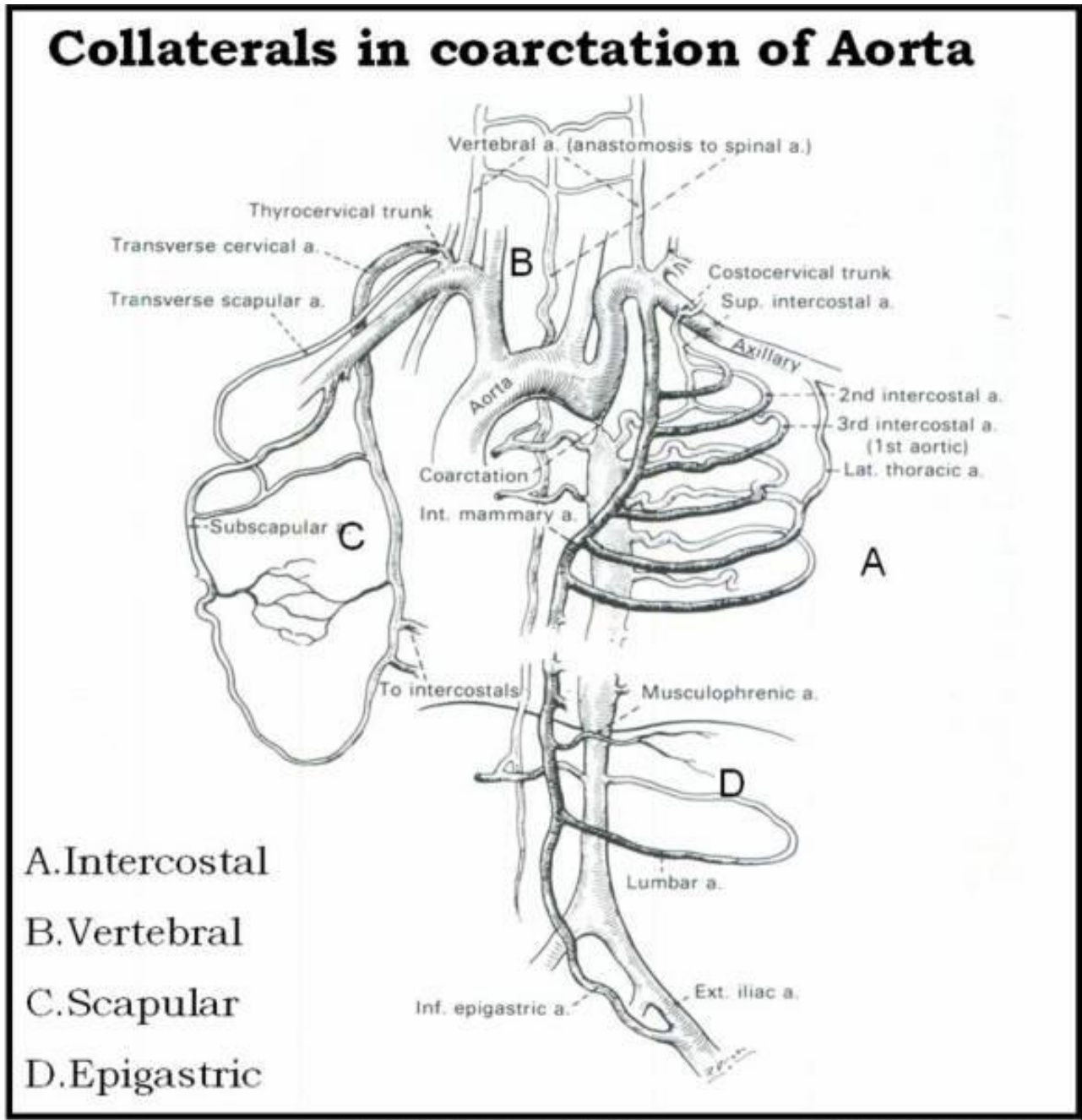
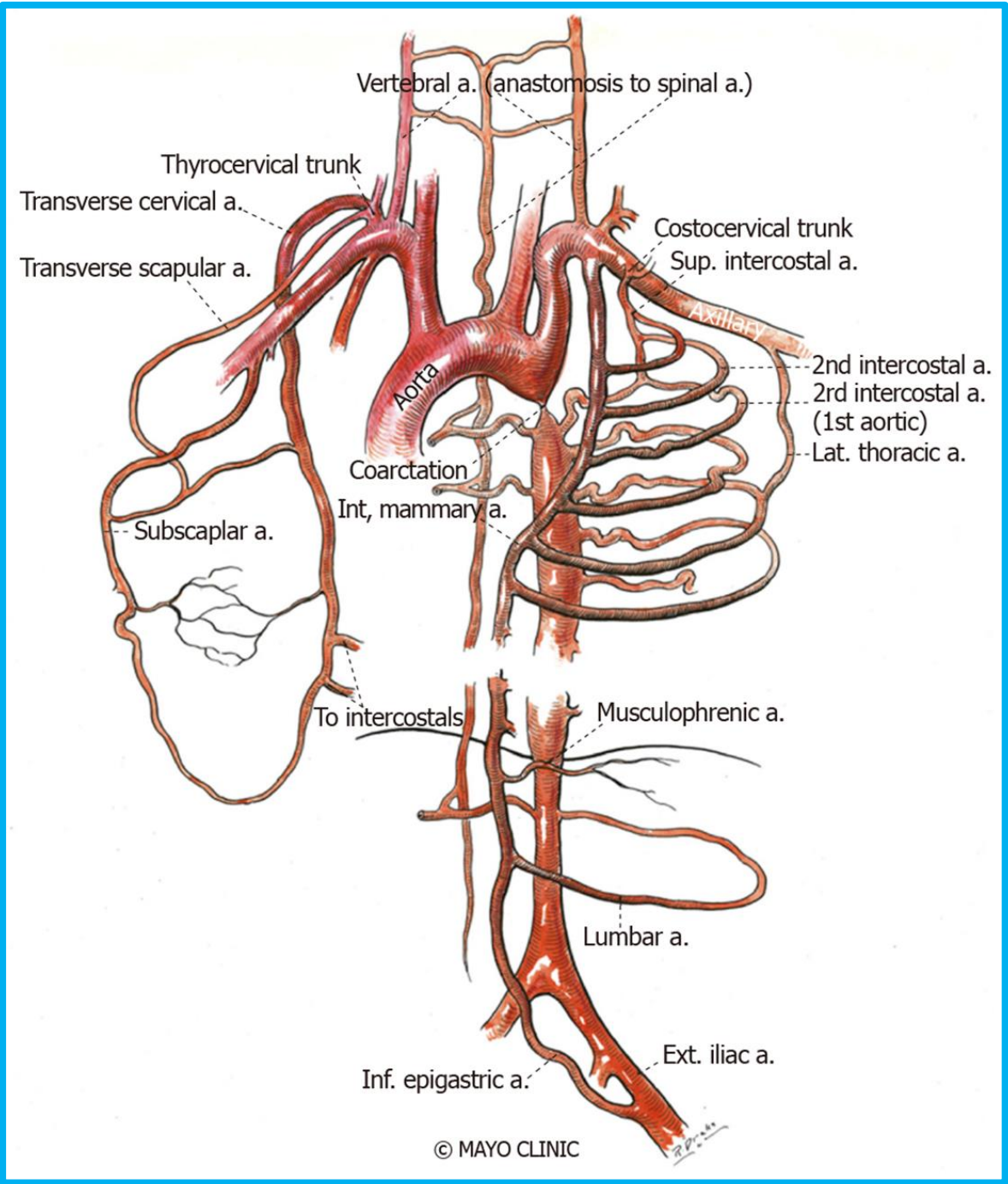
Hypo-perfusion : claudication  
Lower limbs hypotension



Tricuspid aortic valve



Bicuspid aortic valve



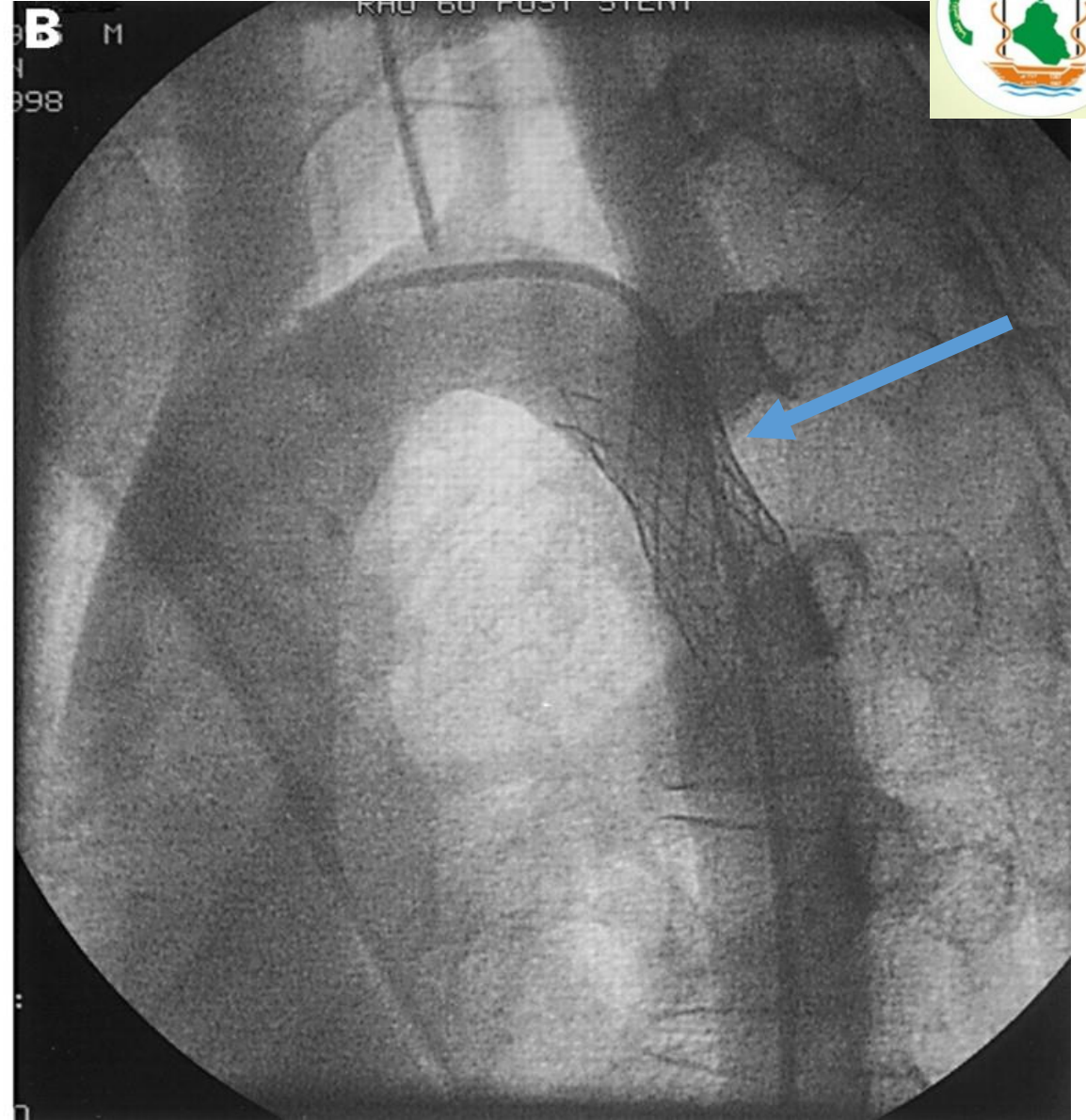
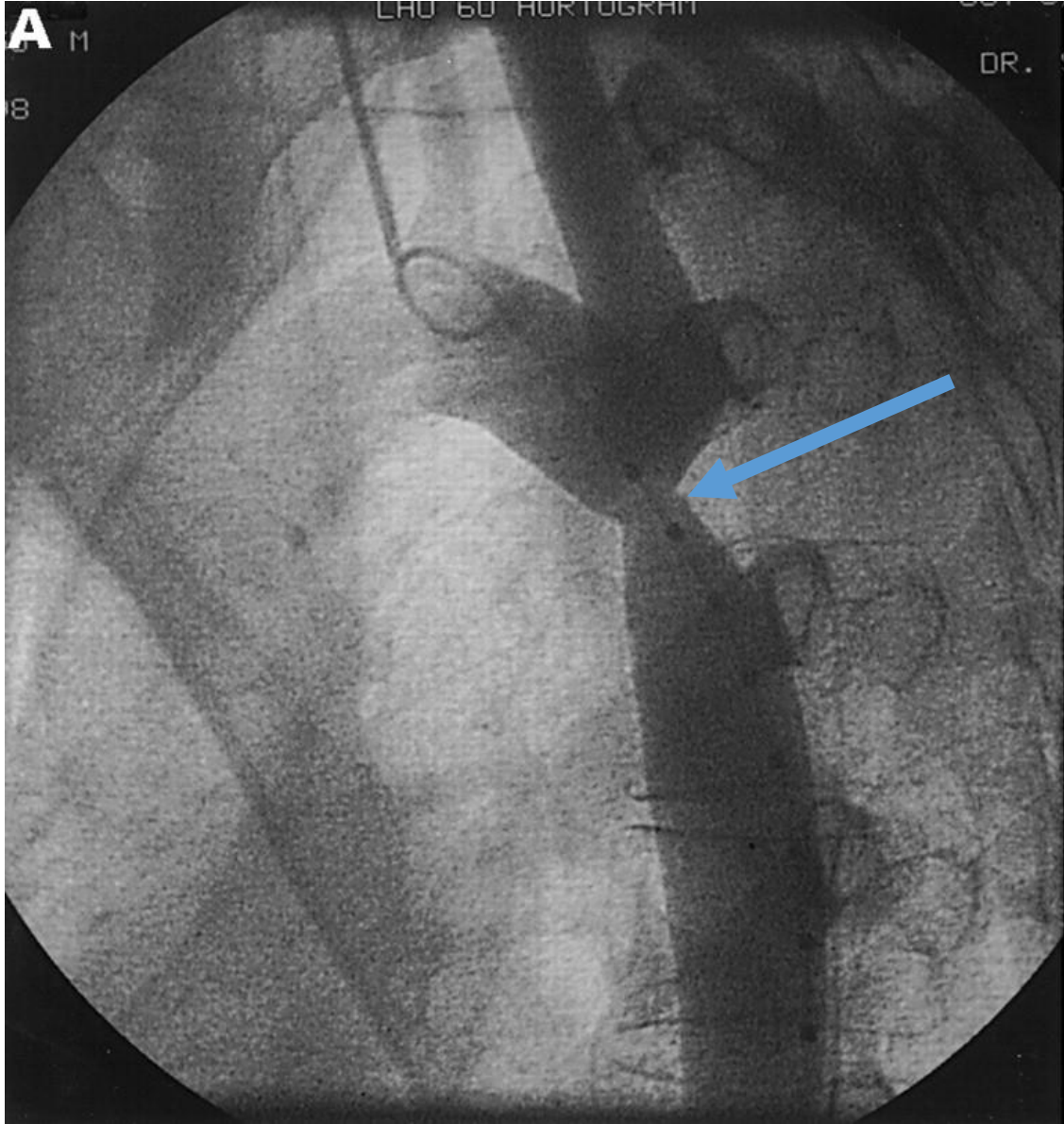


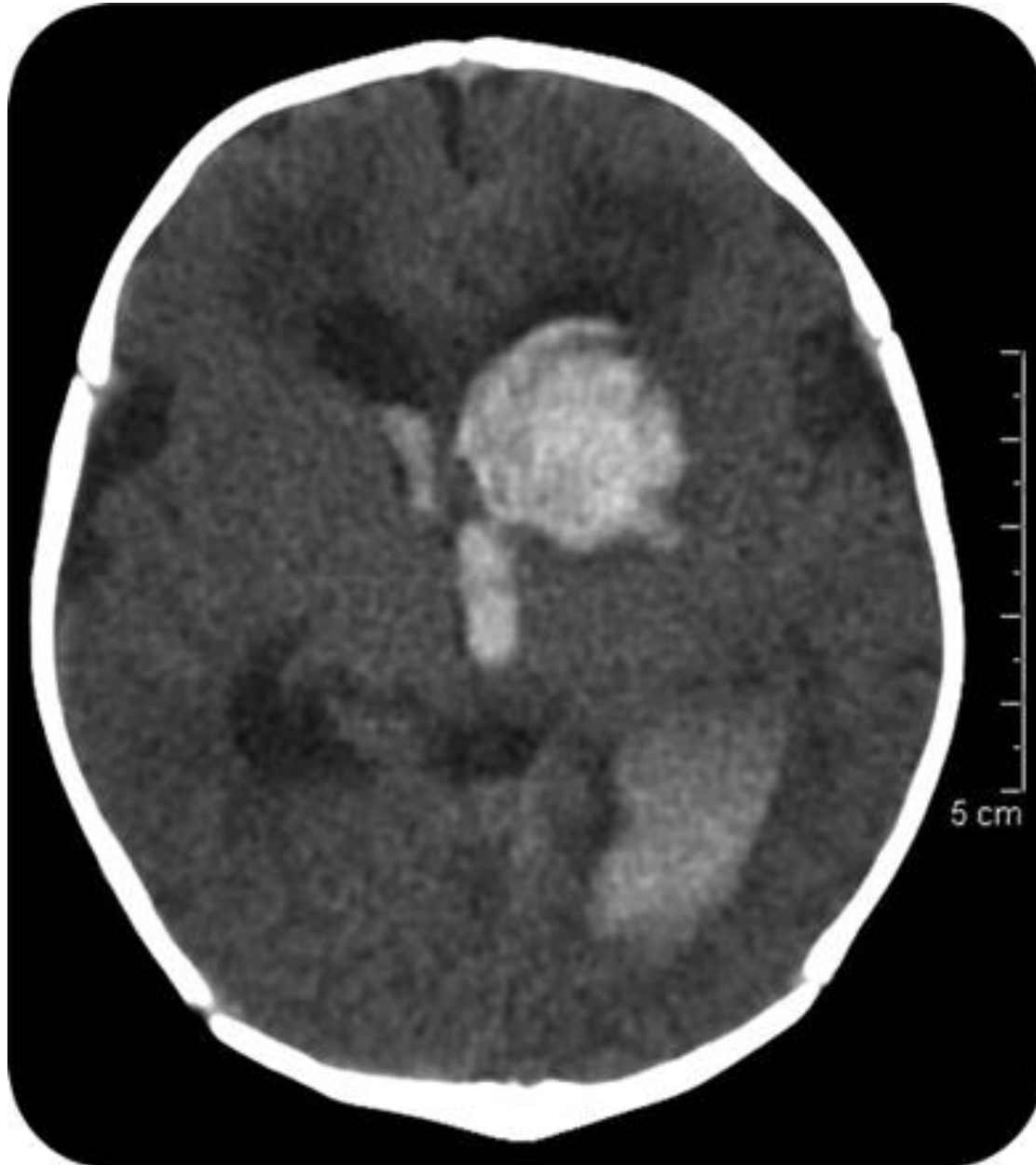
## Management

- **Surgical correction**
- **Trans- catheter:** Balloon dilatation -/+ stenting

**Early** → persistent hypertension can be avoided.

**Late ( in childhood or adult life )** → often remain hypertensive or develop recurrent hypertension later on.





ICH by rupture cerebral aneurysm in a patient with coarctation of aorta



**THANKS**