

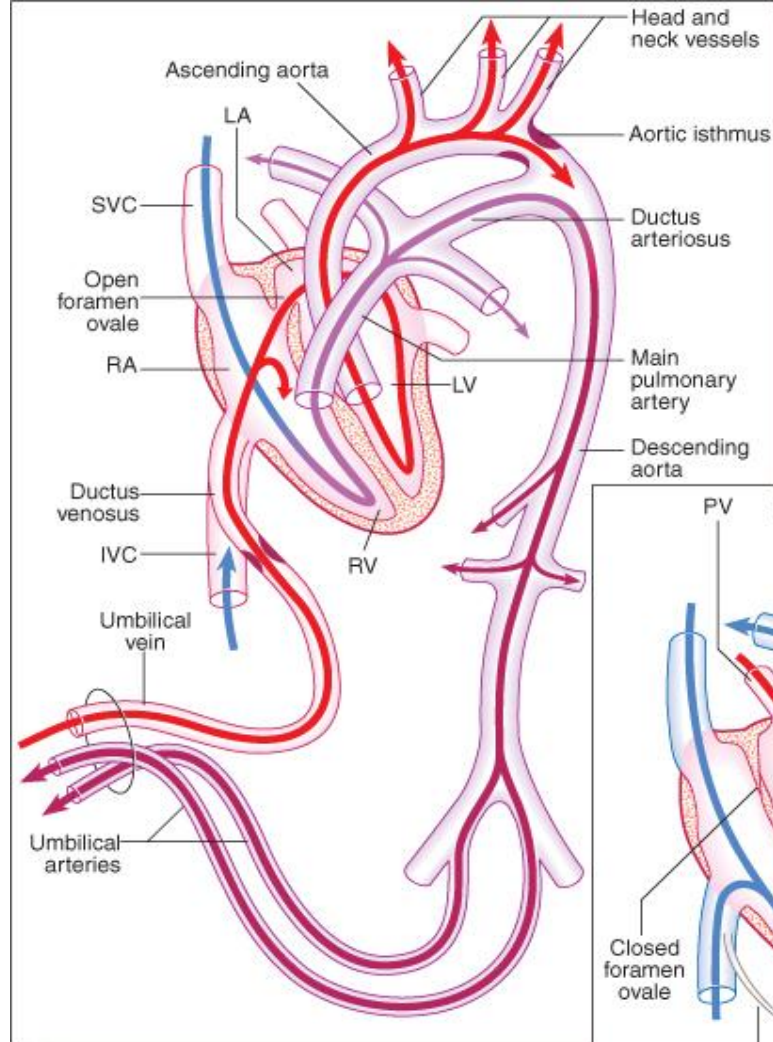
Congenital Heart Disease



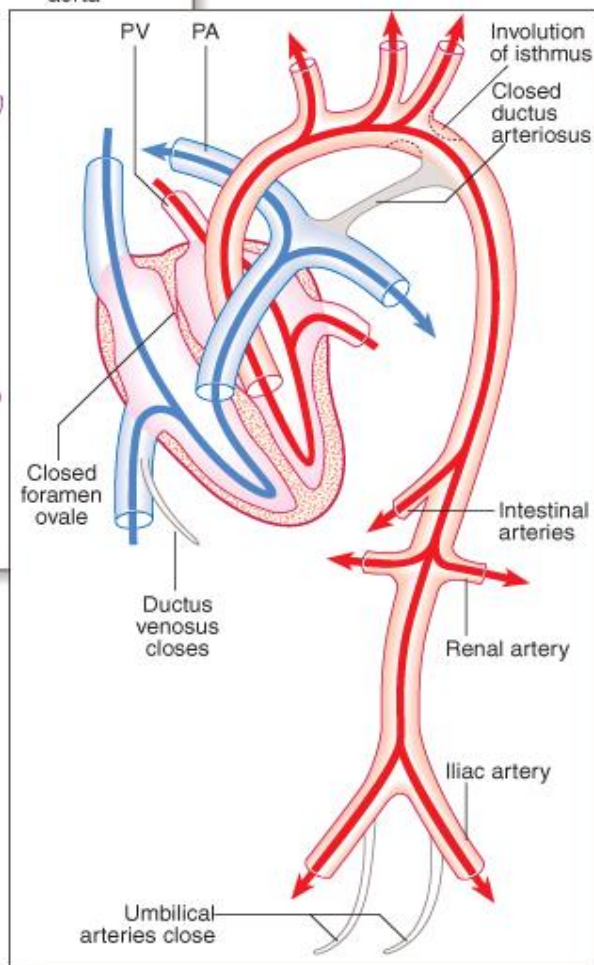
Congenital Heart Disease

General Consideration

1. Severe forms appear in early childhood while mild form seen during adolescence & adulthood
2. Incidence is 0.8% of live births
3. Causes are exposure of mother to infections , drugs & toxins



A



B

INCIDENCE AND RELATIVE FREQUENCY OF CONGENITAL CARDIAC MALFORMATIONS

Lesion	% of all CHD defects
Ventricular septal defect VSD	30
Atrial septal defect ASD	10
Patent ductus arteriosus PDA	10
Pulmonary stenosis PS	7
Coarctation of aorta Co A	7
Aortic stenosis AS	6
Tetralogy of Fallot TOF	6
Complete transposition of great arteries TGV	4
Others	20

Congenital Heart Disease

Associtions

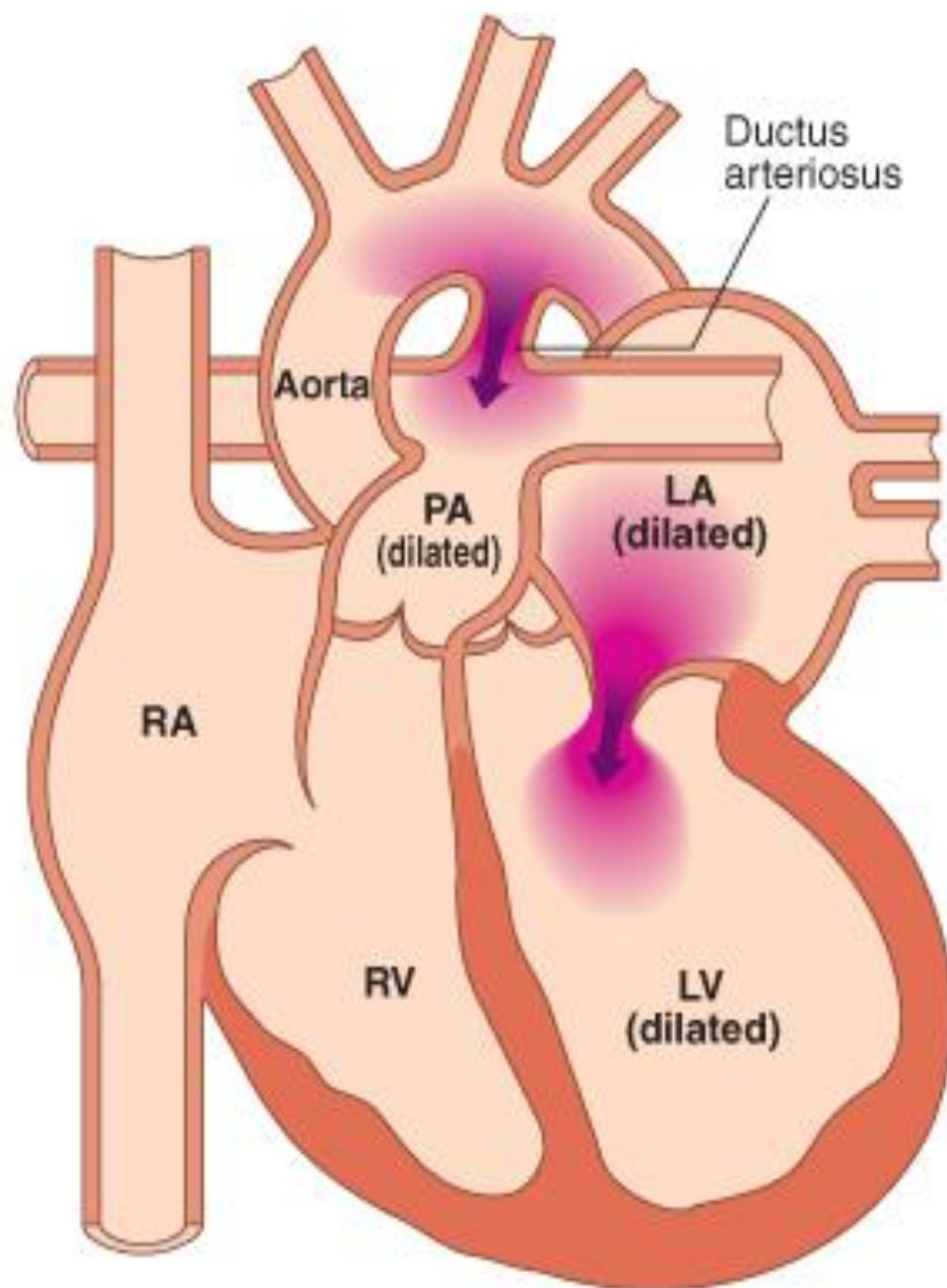
- **Rubella ... PDA , ASD , VSD**
- **Alcoholic mother.... ASD, VSD**
- **SLE Congenital Heart Block**
- **Down's syndromeASD, VSD**

Cyanotic Congenital HD

- **Transposition of great vessels**
- **Tetralogy of Fallot (TOF)**
- **VSD with reverse shunt**
- **Tricuspid atresia**
- **Ebstein's anomaly**

Patent Ductus Arteriosus (PDA)

- Ductus arteriosus which should be closed after birth remains open giving rise to left to right shunt between aorta at the level of L. subclavian artery & pulmonary artery.
- Symptoms depend on size of shunt and pulmonary vascular resistance.



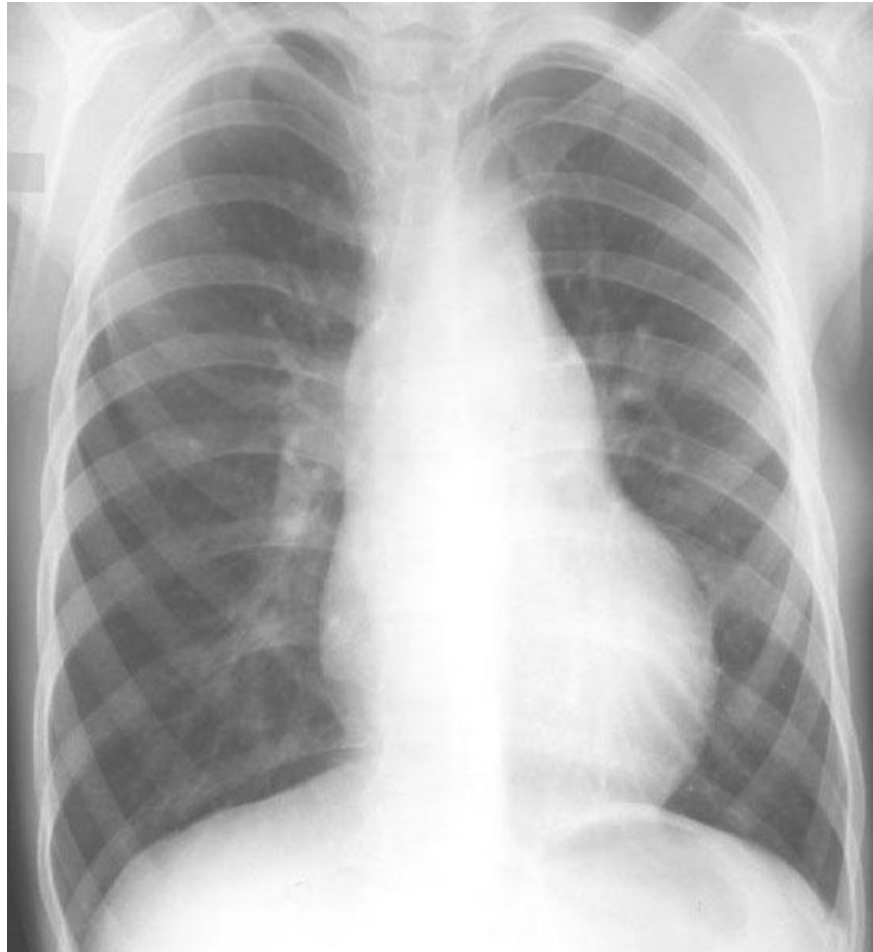
PDA – Clinical Features

- **Symptoms :-** Asymptomatic if small..
If large : growth retardation , dyspnea.
- **Signs :-** Wide pulse pressure, loud S2, continuous (machinery) murmur over pulmonary area. Thrill is common.
When R. to L. shunt , differential cyanosis appears.

PDA - Investigations

- * CXR :- The heart is of normal size & contour or slightly enlarged with LV & LA enlargement. PA & aorta may be prominent
- * ECG :- Non-specific findings
- Echocardiography/Doppler is helpful, but the lesion is best visualized by MRI, CT, or contrast angiography.

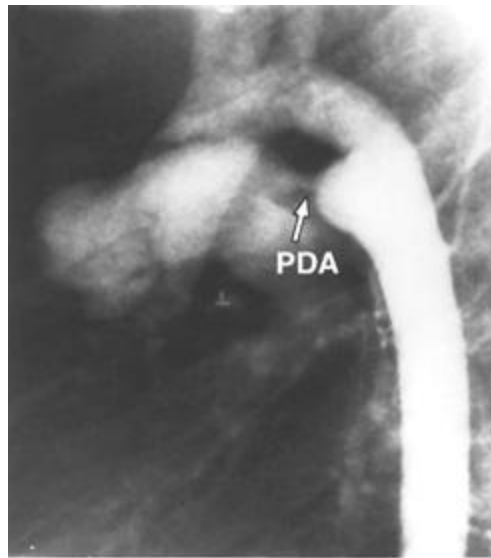
PDA - CXR



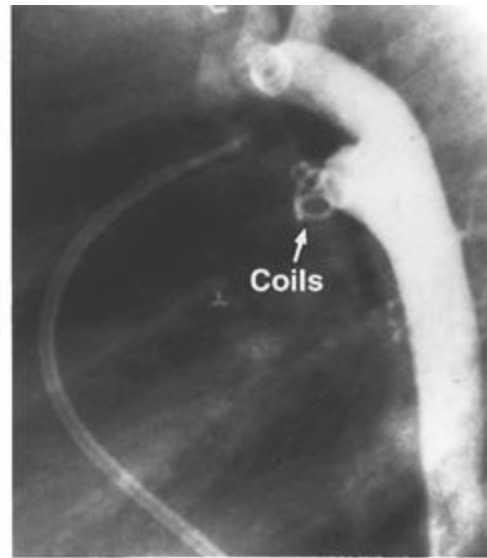
PDA - Treatment

- **Duct closure** is recommended if the **murmur** is **audible** & there is no pulmonary HT & no R to L. shunt.
- **Closure by surgical operation or by transcatheter approach using device**
- **Use of Indometacin** in first week of life may be helpful.

PDA – Treatment by coil



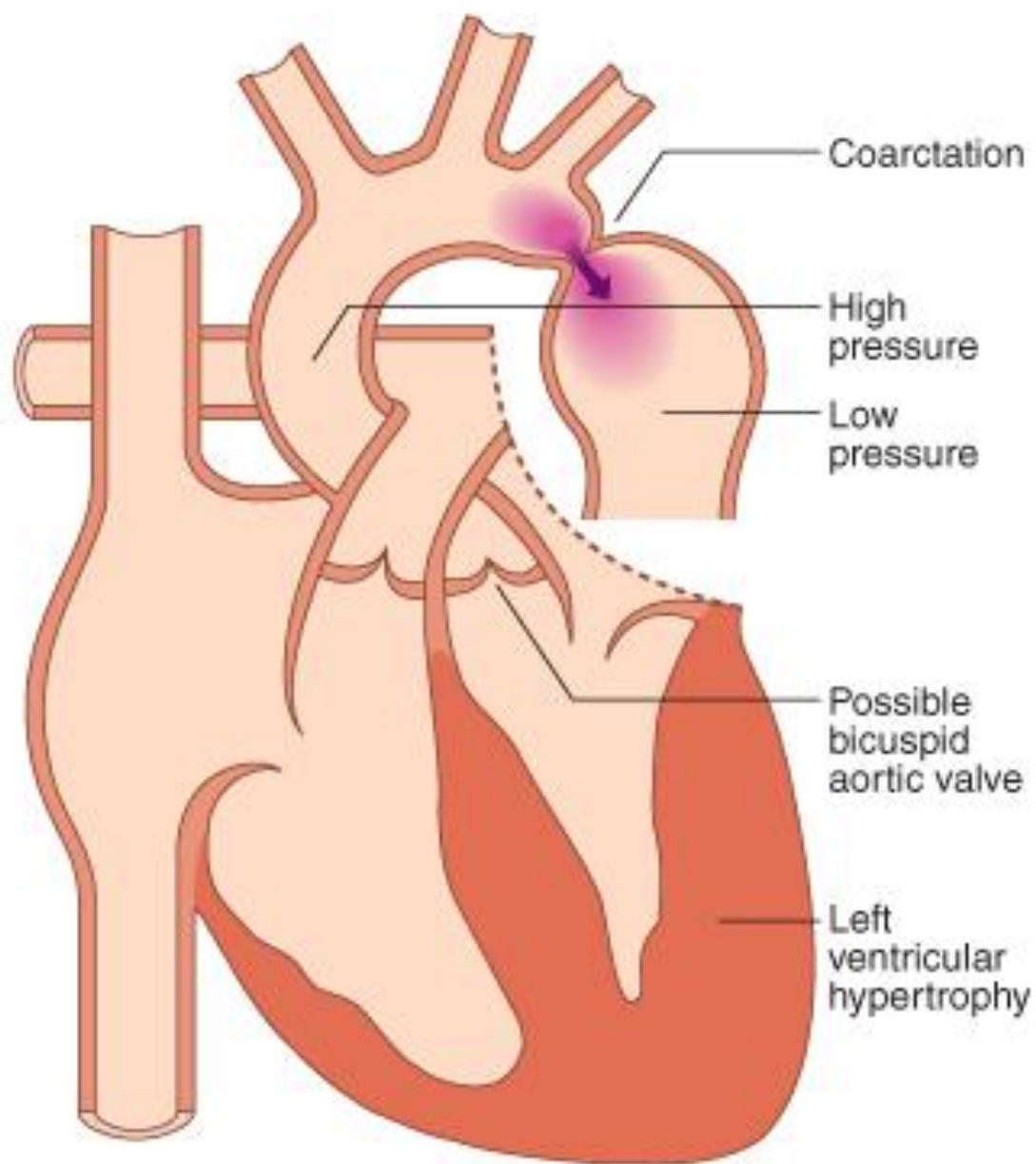
A



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Coarctation of the Aorta

- * It is a constriction of lumen of aorta most common site is distal to the origin of the L. subclavian artery near the insertion of ligamentum arteriosum.
- * Occurs more in males, seen with Turner's syndrome. Associated with bicuspid AV (50%)



Coarctation – Clinical Features

- Symptoms :- May be asymptomatic at early childhood but symptoms of HT & intermittent claudication appear later.
- Signs :- High BP in upper extremities & low in the lowers. Systolic murmur at L. intercostal spaces, axilla & back. Tortuous... collaterals may be seen. Signs of LVH. Radial-femoral pulse delay.

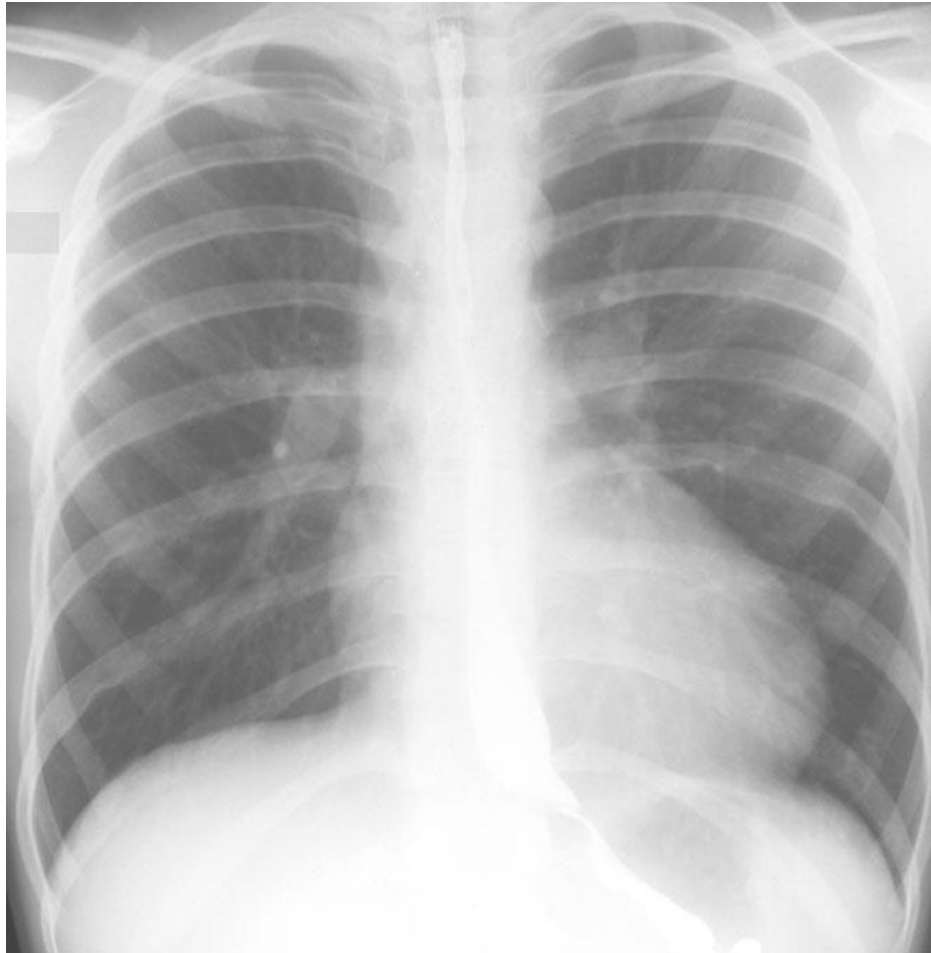
Coarctation - Investigations

- **ECG : - LVH**
- **2 D Echo & Doppler**
- **TEE**
- **MRI**
- **3 dimension CT Scan**
- **Cardiac cath. In adults to assess coronaries**

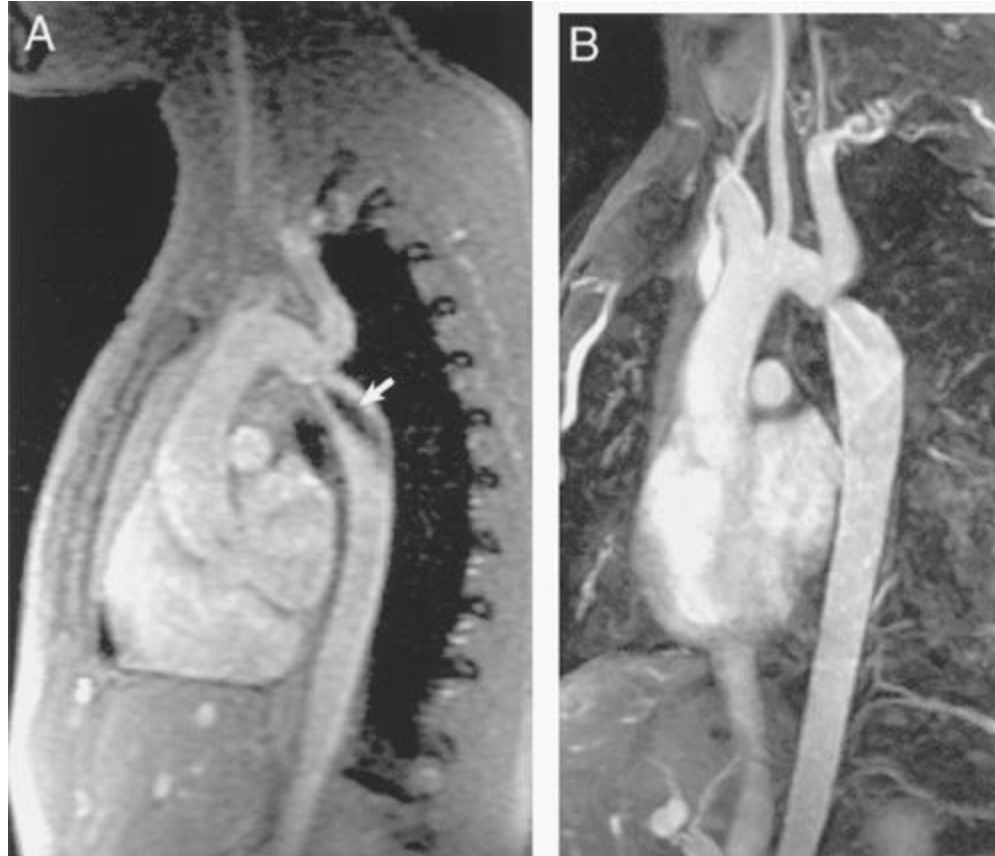
Coarctation - Investigations

- CXR :
- Indentation of the aorta at the site of coarctation and pre- and poststenotic dilatation (the “3” sign)
- Notching of the 3rd to 9th ribs, is due to inferior rib erosion by dilated collateral vessels.

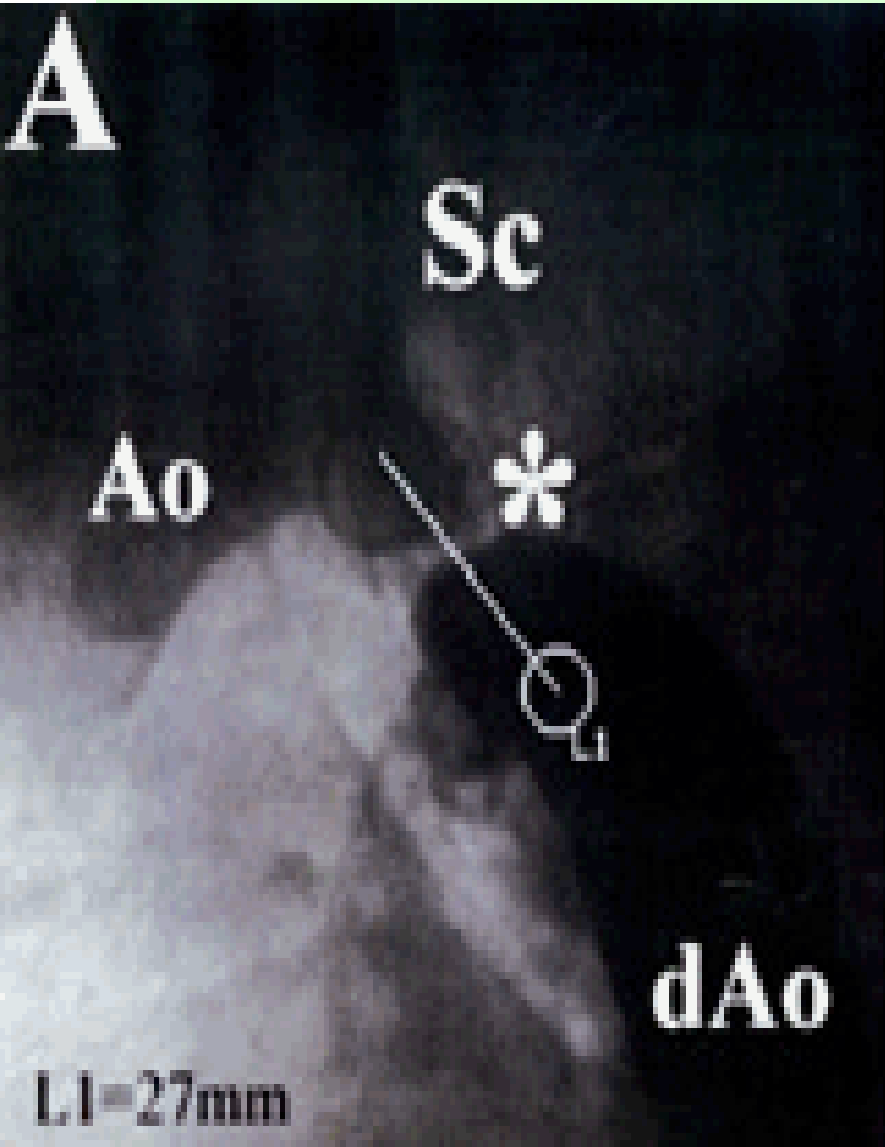
Coaractation - Rib Notching



Coarctation - MRI



Coarctation



Coaractation - Management

- **Complications:** include HF, rupture of aorta, dissecting aneurysm, rupture cerebral aneurysm & endocarditis
- * **Treatment:** By surgical repair or stent implantation
- * **Follow up:** treatment of HT

Atrial Septal Defect (ASD)

- Common congenital HD
- More common in females
- Often asymptomatic & discovered by physical exam.
- Types :-
 - ostium secundum
 - ostium primum (with cleft mitral valve)

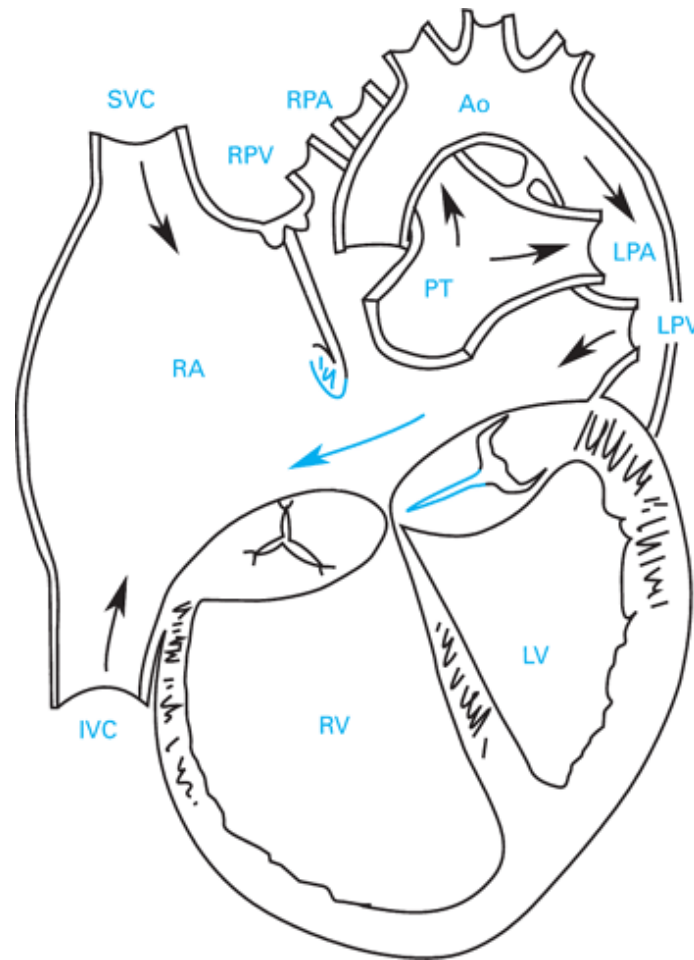
ASD – Ostium Secundum

- **The most common type of ASD.
Almost 85% of cases of ASD**
- **Involves fossa ovalis**
- **Size of defect is variable**

ASD – Ostium Primum

- **Low down in the septum near the mitral & tricuspid valves, associated with deformed valve(s) or regurgitation.**
- **High incidence of VSD**
- **Common with Down's syndrome...**

ASD - ostium primum



ASD - Pathophysiology

- **Left to right shunt** (degree depends upon size of defect & pulmonary vasc. rest.)
- **Diastolic RV load**
- **Increase PVS & PA pressure**
- **Bidirectional & finally right to left shunt**
(Eisenmenger's syndrome)

ASD – Clinical Features

- **Symptoms:** depend on the severity & duration.
- At first asymptomatic but with increase of shunt there is recurrent respiratory symptoms.
- Arrhythmia is common especially atrial
- Symptoms of heart failure appear later
- Cyanosis appears at last (R to L shunt)

ASD- Signs

- RV impulse
- Loud S1
- Wide fixed splitting of S2
- Pulmonary mid-systolic murmur
- Functional mid-diastolic tricuspid murmur.
- Cyanosis if R. to L. Shunt

ASD - Investigations

- ECG :-

Incomplete RBBB , AF

In addition LAD in ostium primum

- * CXR :-

RAH, RVH, prominent PA & increase pulmonary vascular marking

- * ECHO (TEE)

ASD – CXR (prominent PA & RVH)

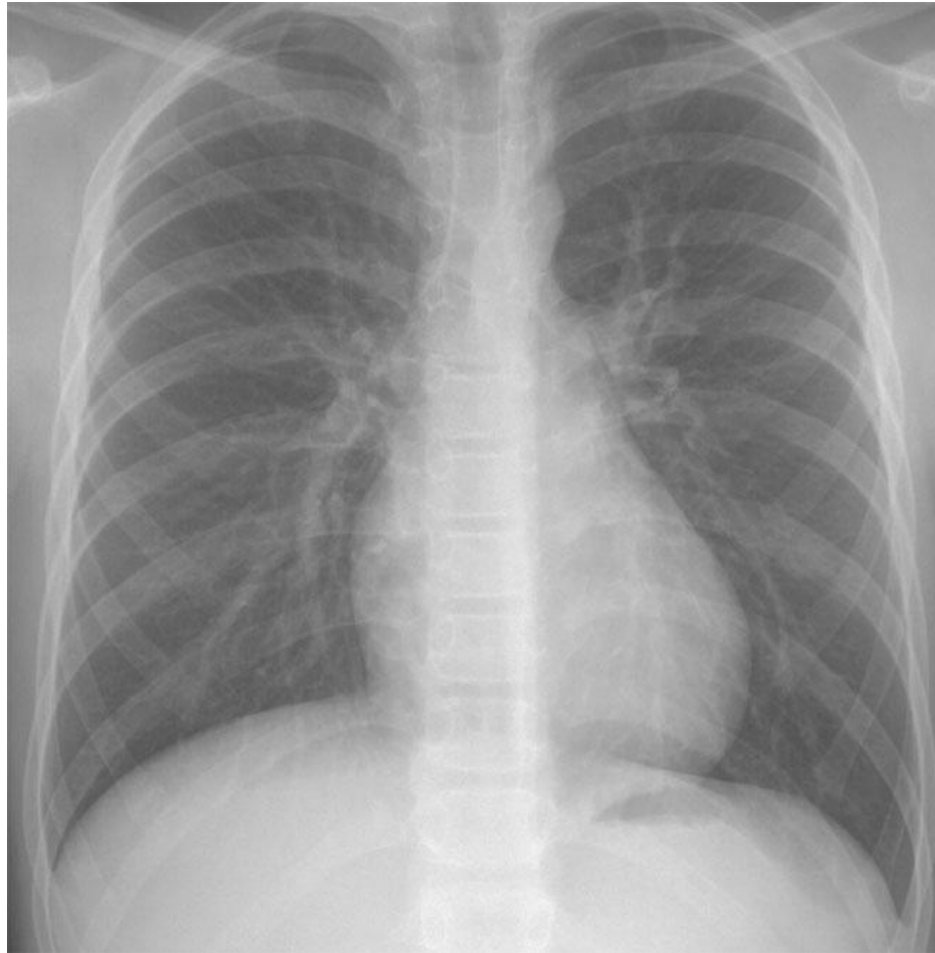


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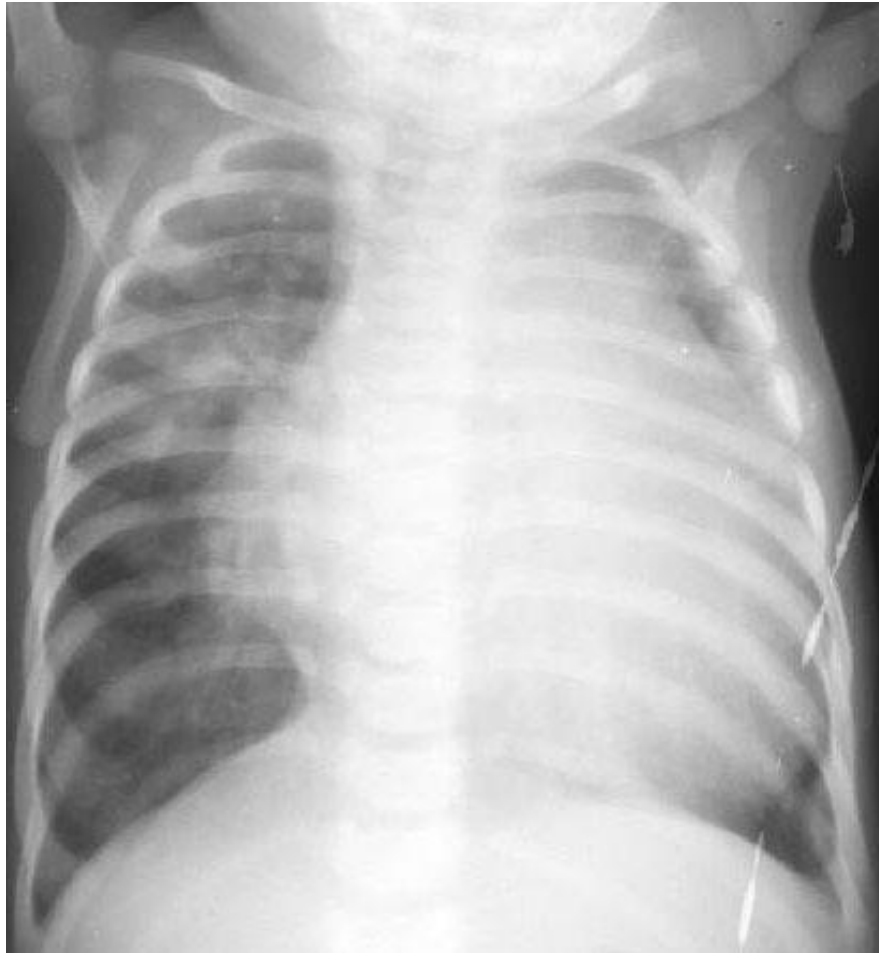


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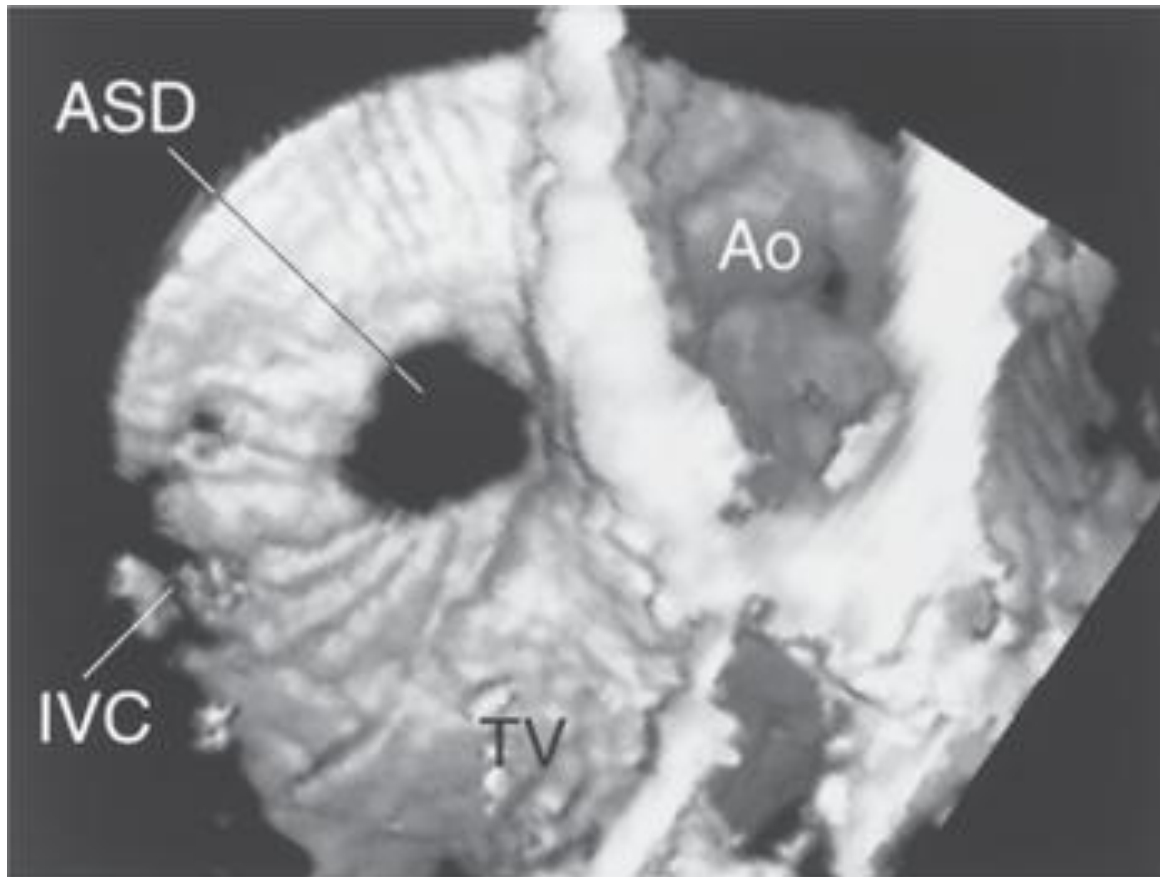
ASD - CXR (increase plethora)



Ostium primum & AV canal



ASD – Three dimension Echo



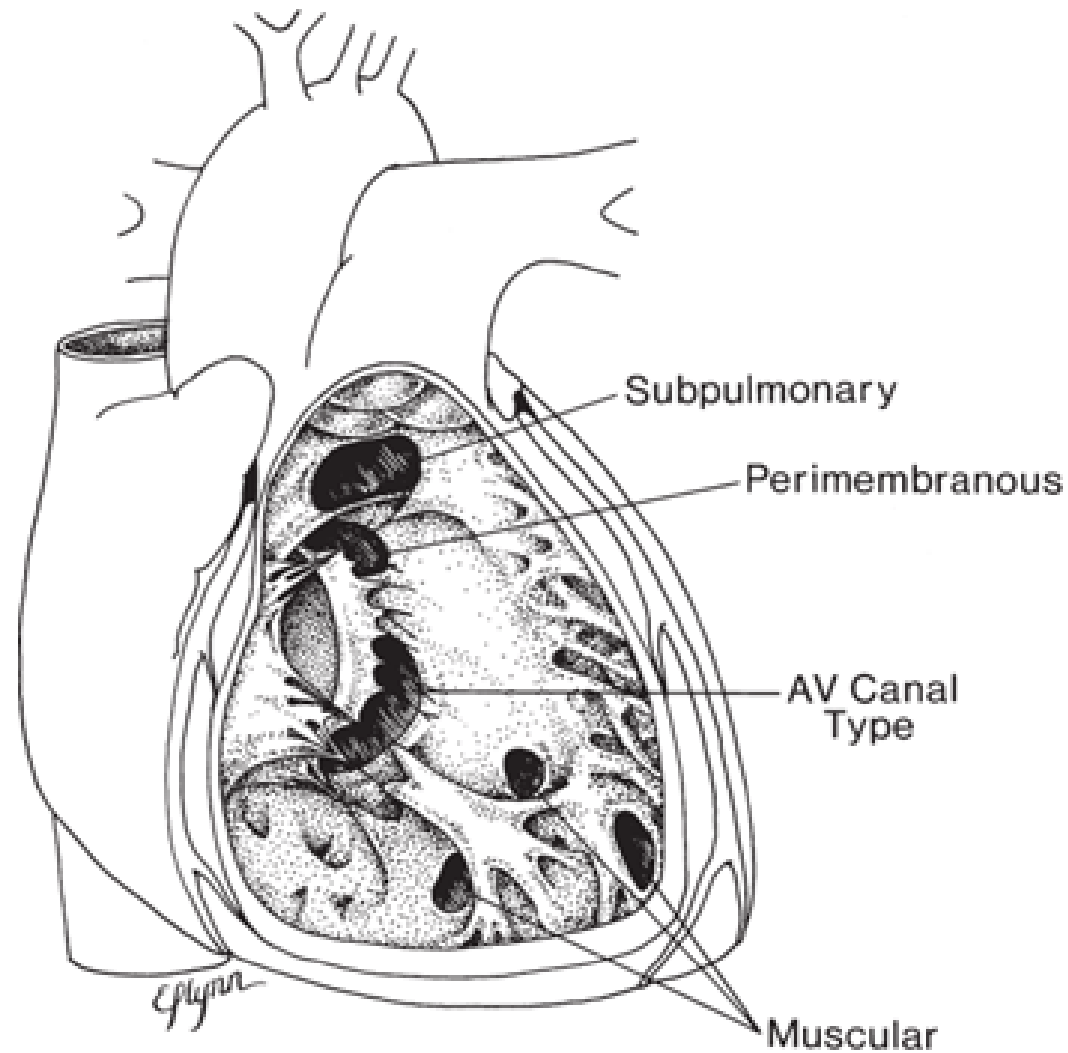
ASD - Treatment

- **No OP for small (endocarditis is rare)**
- **Device or repair for symptomatic or asymptomatic ASD if shunt is more than 1.5:1**
- **Valve repair + closure of primum**
- **Operation is contraindicated if R to L shunt**

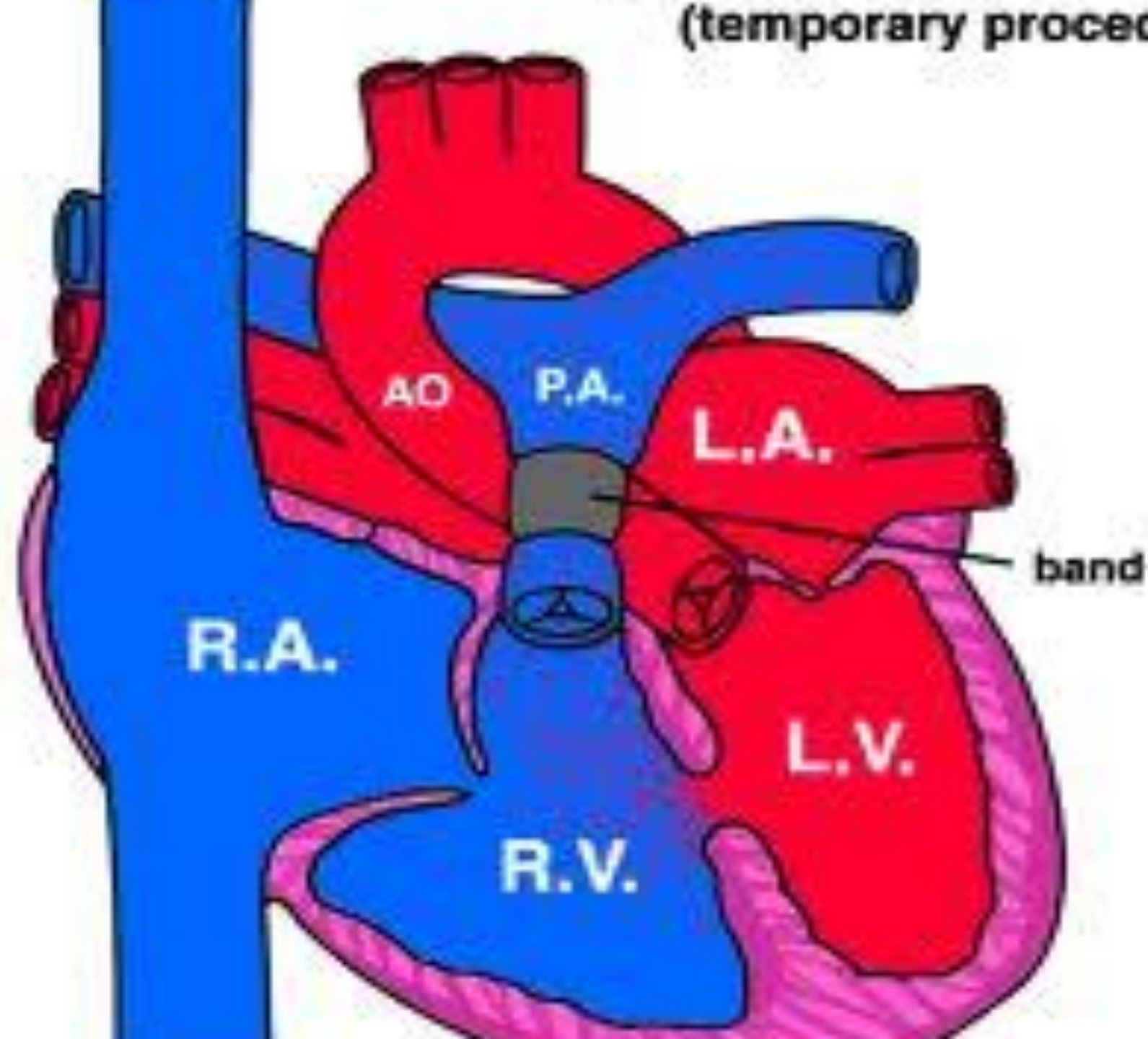
Ventricular Septal Defect (VSD)

- The **most common** congenital HD
- Defect may be single or multiple. Most common site is the **membranous** part.
- May be associated with other lesions (AR is seen in 5% of cases).
- Small VSD may close spontaneously in early childhood.

VSD – Multiple Sites



(temporary procedure)



VSD - Pathophysiology

- Hemodynamically depends upon size & pulmonary vascular resistance.
- Left to right shunt leads to increase blood of pulmonary circulation leading to congestion
- Right to left shunt at last (Eisenmenger's)

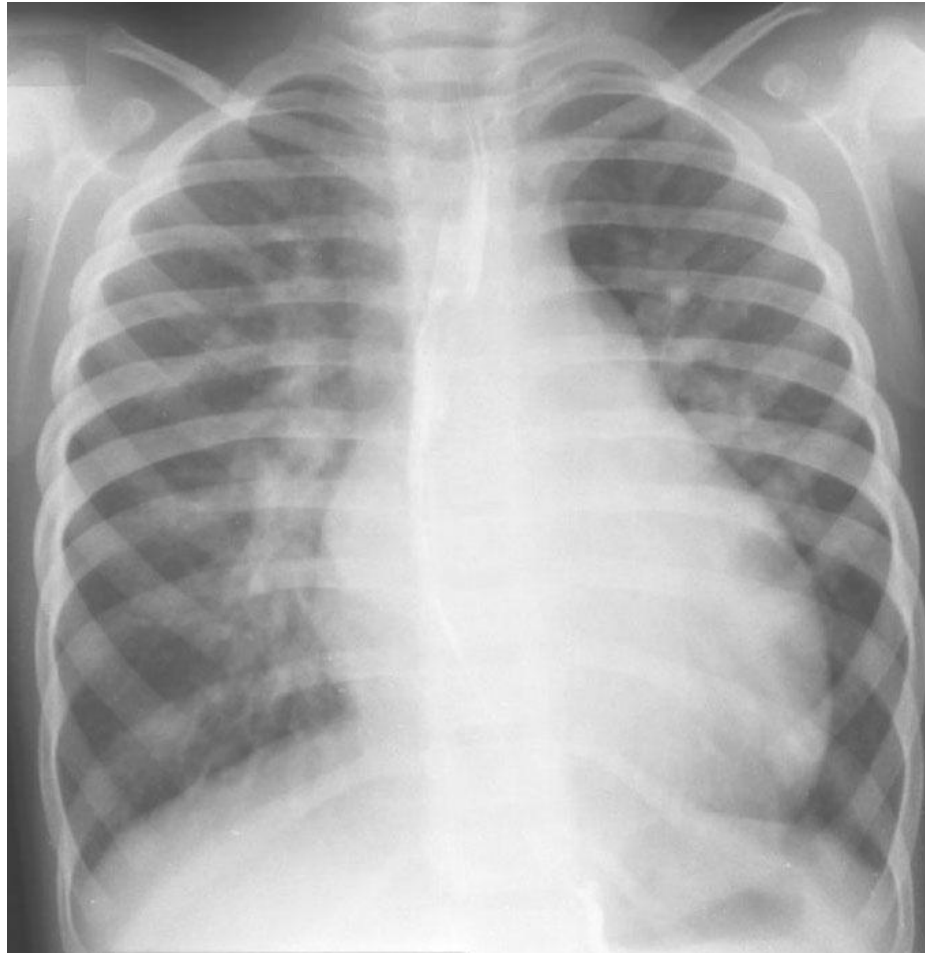
VSD – Clinical Features

- **Symptoms** :- Dyspnoea, hemoptysis, chest pain & syncope.
- **Signs** :- systolic murmur at 3rd or 4th left space...Small VSD gives louder murmur & large VSD less intensity.

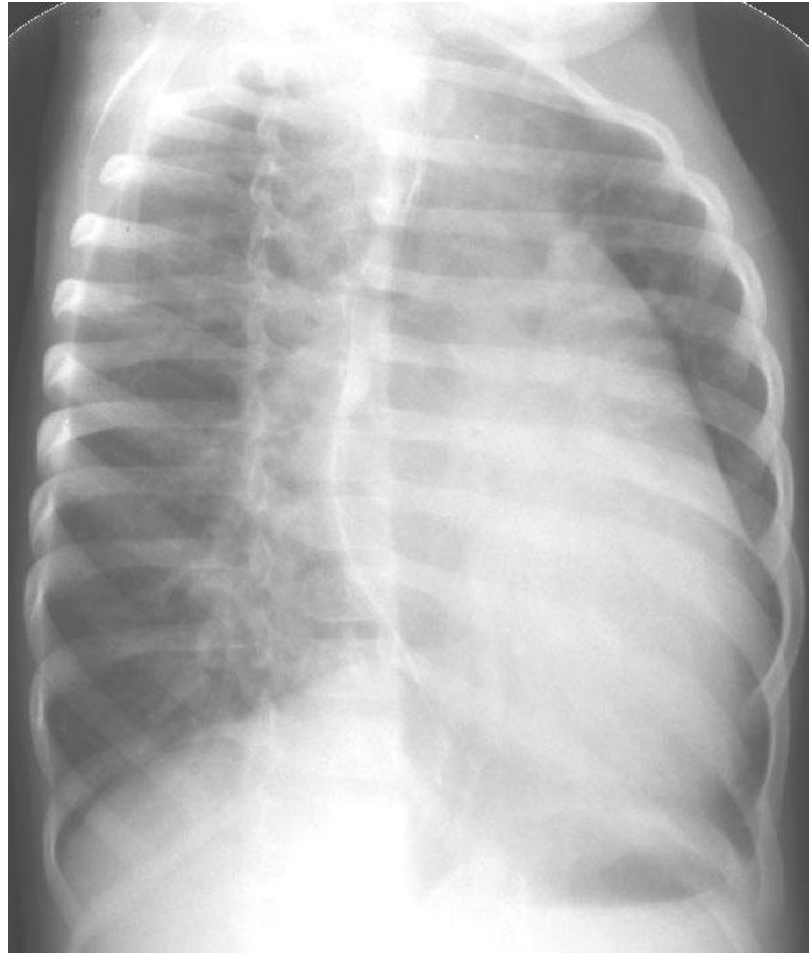
VSD – Investigations

- ECG :- None specific. Normal or R. or L. ventricular hypertrophy or both.
- * CXR :- increase pul. vascular marking with prominent PA , LVH & RVH
- Echo – doppler can establish diagnosis

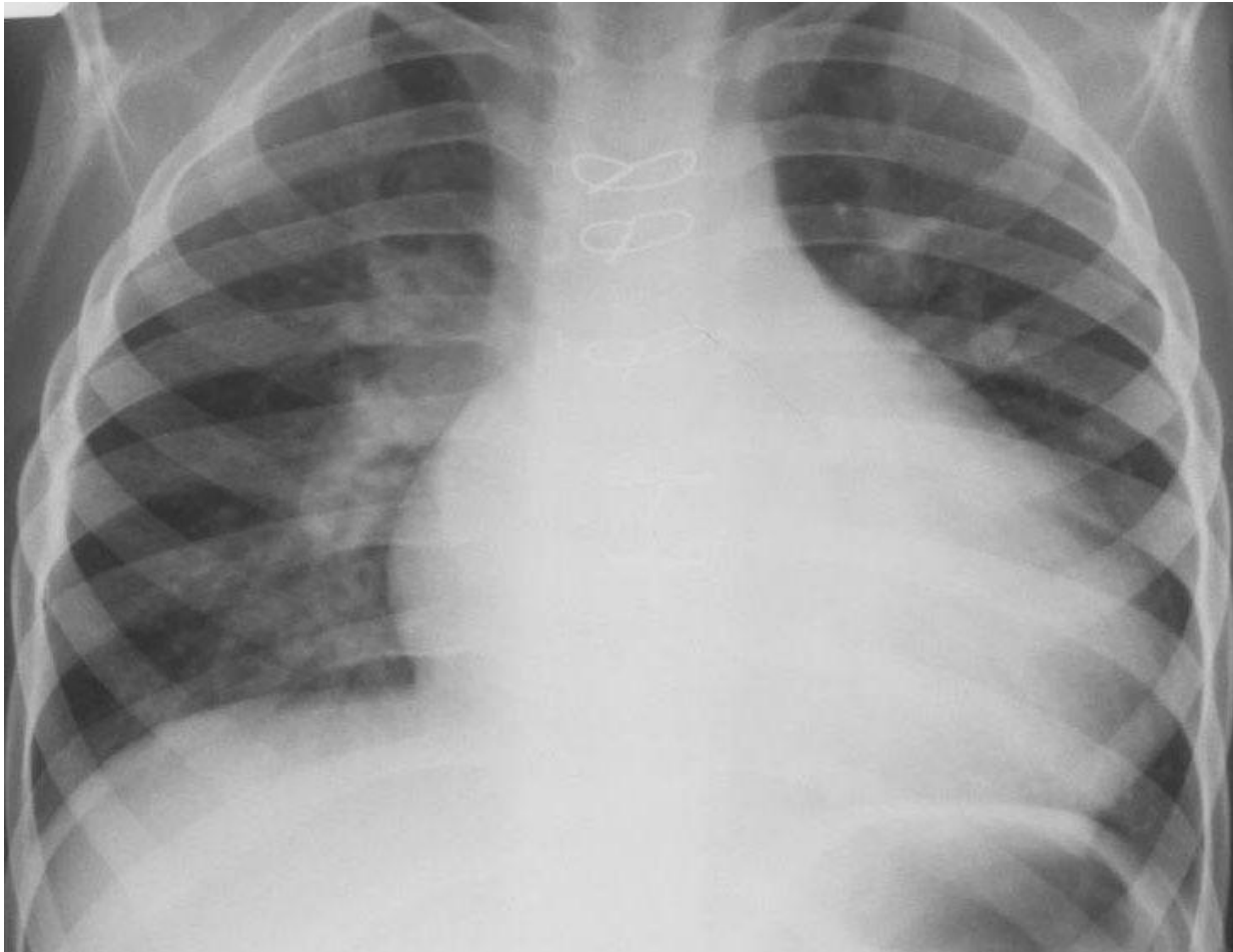
VSD - CXR



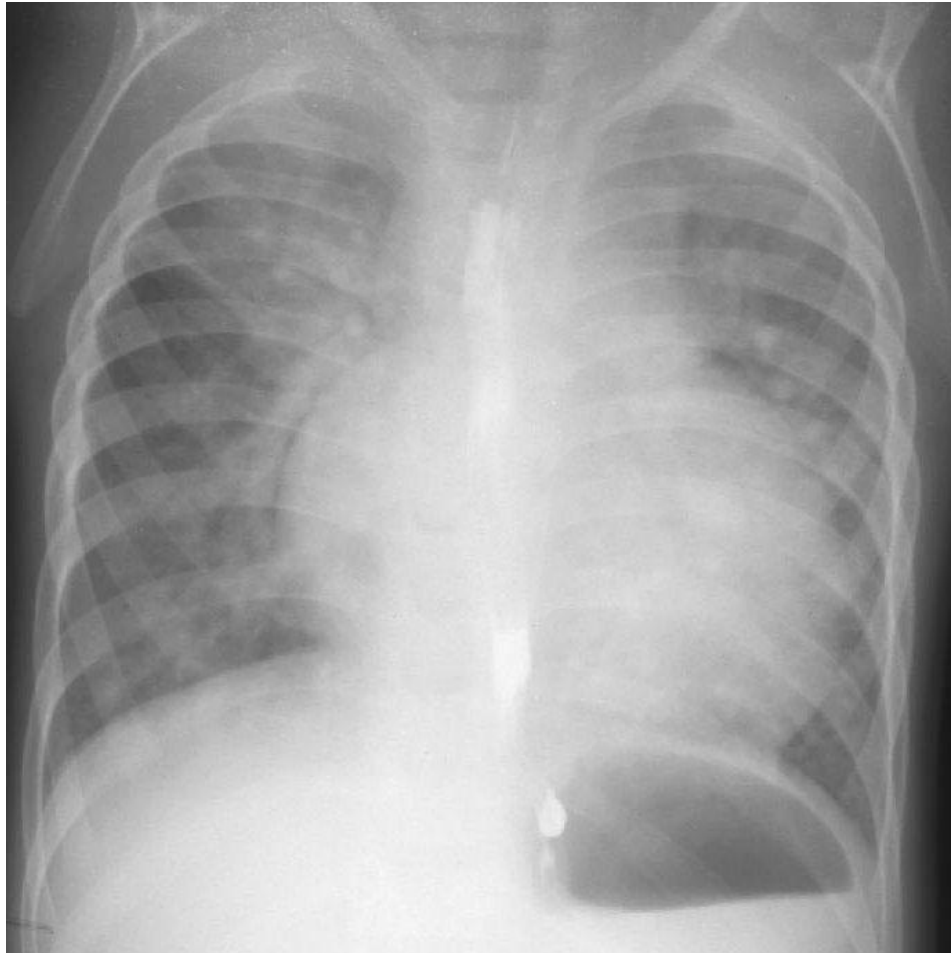
VSD – Lateral CXR



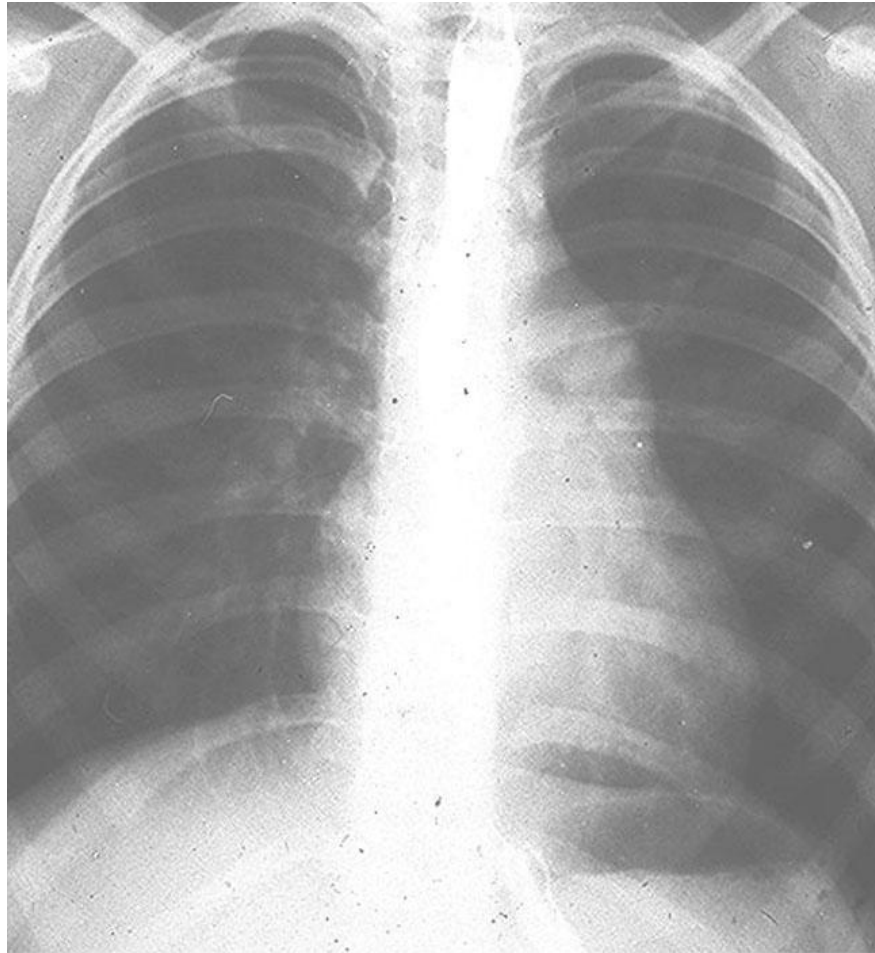
VSD – Large Shunt



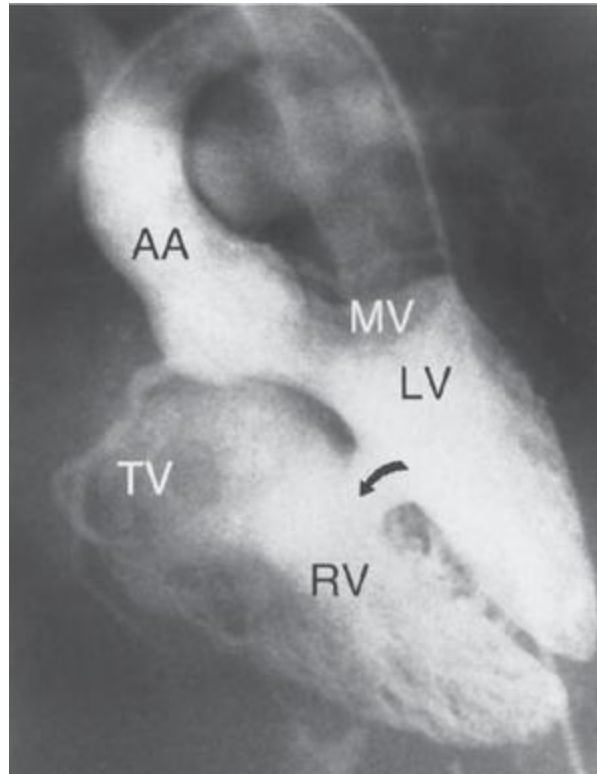
VSD – Large Shunt

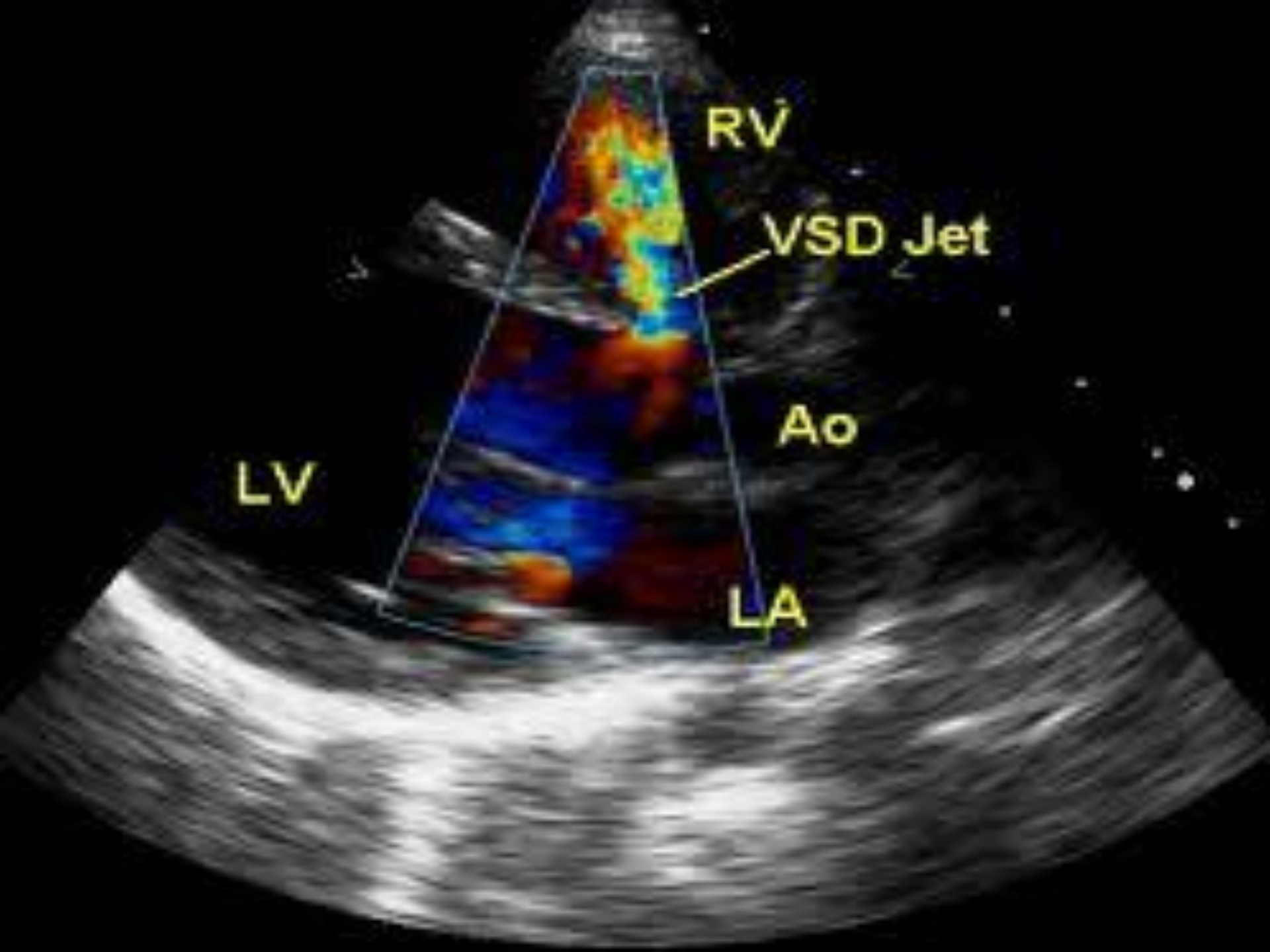


VSD - Eisenmenger



VSD - Ventriculogram



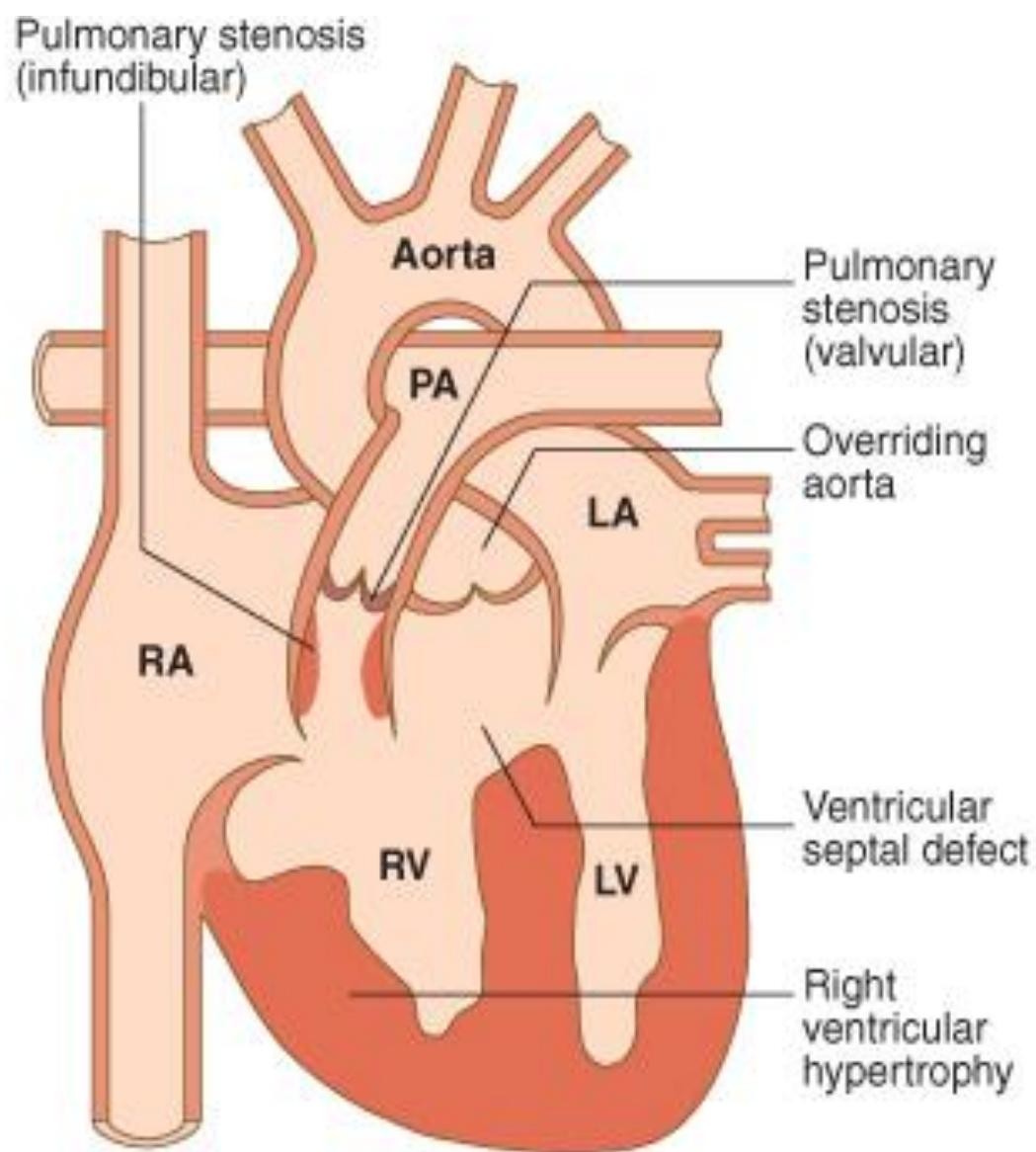


VSD - Management

- Intervention is not needed in small VSD
- Transcatheter closure or surgery are needed for symptomatic patient or those with large shunt.
- Closure is contraindicated in R to L shunt

Tetralogy of Fallot (TOF)

1. VSD
2. RVH
3. Pulmonary Stenosis
4. Overriding aorta.



TOF – Clinical Features

Symptoms :- cyanosis appears few months after birth.. Clubbing..

Squatting after exertion..polycythemia

Cyanosis, unconsciousness and apnea with feeding & crying (Fallot's spells)..

Signs :- Cyanosis, clubbing, systolic murmurs of PS & VSD , diminished P2

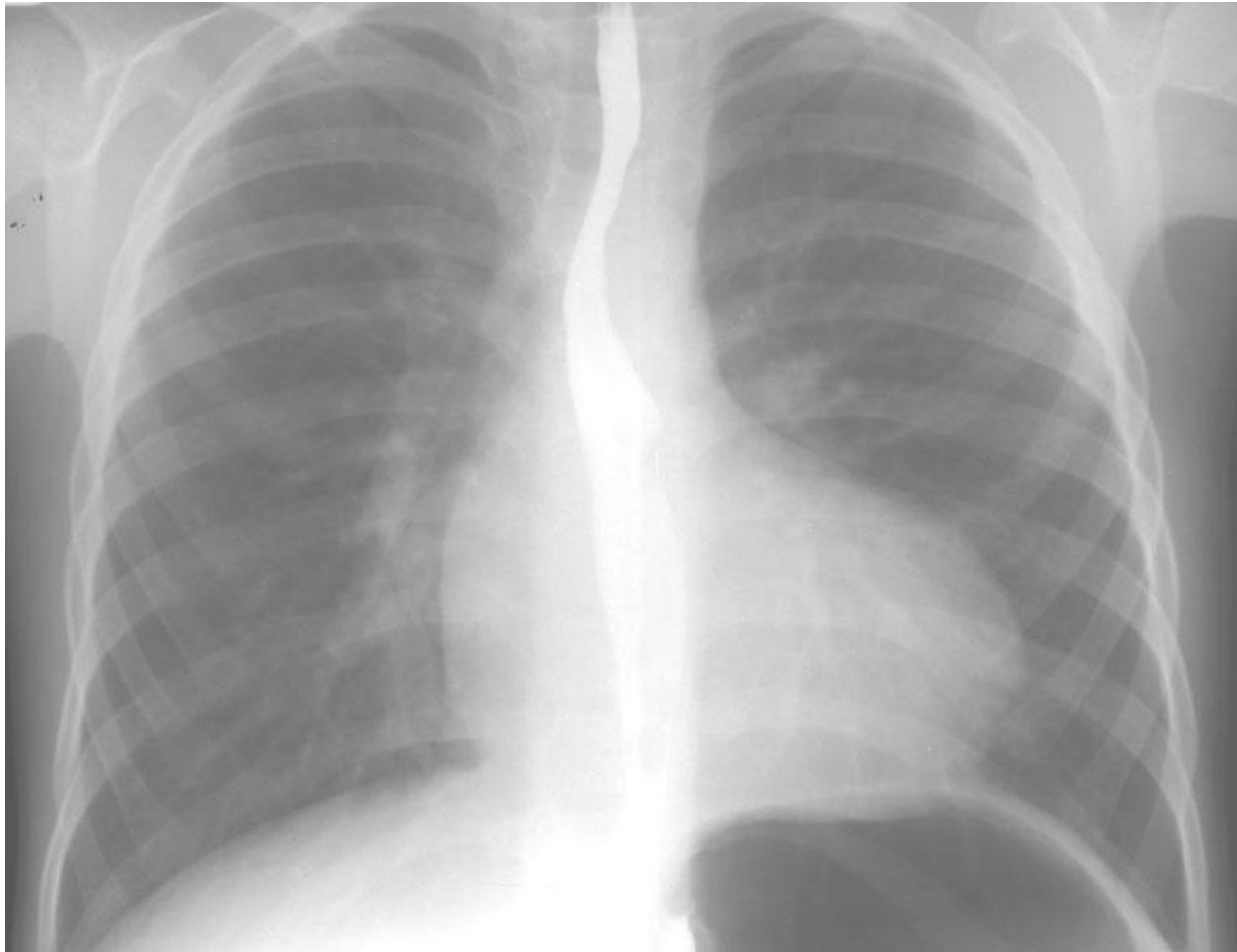
TOF – Investigations

ECG :- RVH

CXR :- Boot shaped heart &
diminished pul. Vascularity

2 D echo – Doppler.... Angiography

TOF - CXR



TOF – Treatment

- Surgery before the age of 5 by complete correction...
- If Pulmonary arteries are small Blalock-Taussig shunt (subclavian to pulmonary art.)

Pregnancy in women with cong. Heart Disease

- **Obstructive lesion like AS is not well tolerated.**
- **Pregnancy should be avoided in cyanotic conditions.**
- **Incidence of congenital HD is higher in children born to mothers with congenital HD.**

OTHER CAUSES OF CYANOTIC CONGENITAL

Defect

Tricuspid atresia

Transposition of the great vessels

Pulmonary atresia

Ebstein's anomaly

Features

Absent tricuspid orifice, hypoplastic RV, RA to LA shunt, VSD shunt, other anomalies
Surgical correction *may* be possible

Aorta arises from the morphological RV,
pulmonary artery from LV

Shunt via atria, ductus and possibly VSD

Palliation by balloon atrial

septostomy/enlargement

Surgical correction possible

Pulmonary valve atretic and pulmonary artery
hypoplastic

RA to LA shunt, pulmonary flow via ductus

Palliation by balloon atrial septostomy

Surgical correction may be possible

Tricuspid valve is dysplastic and displaced into
RV, right ventricle 'atrialised'

Tricuspid regurgitation and RA to LA shunt

Wide spectrum of severity. Arrhythmias. Surgical
repair possible, High mortality.

