Acyanotic Congenital Heart Diseases

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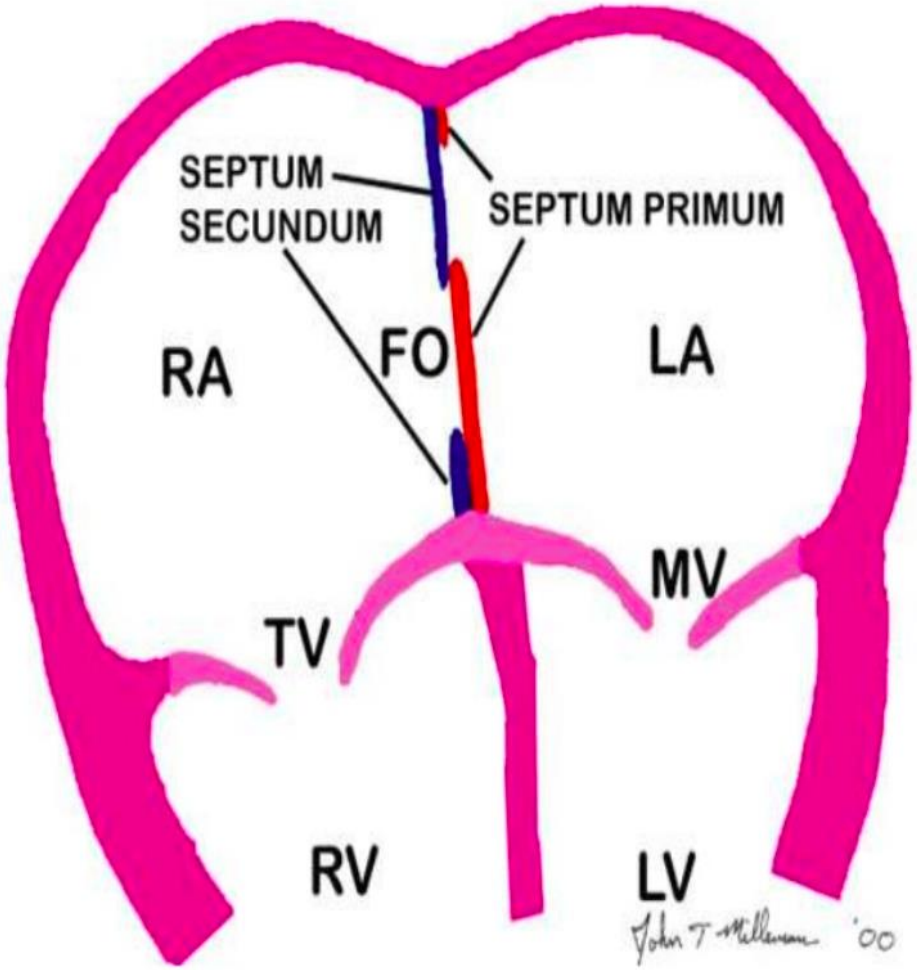
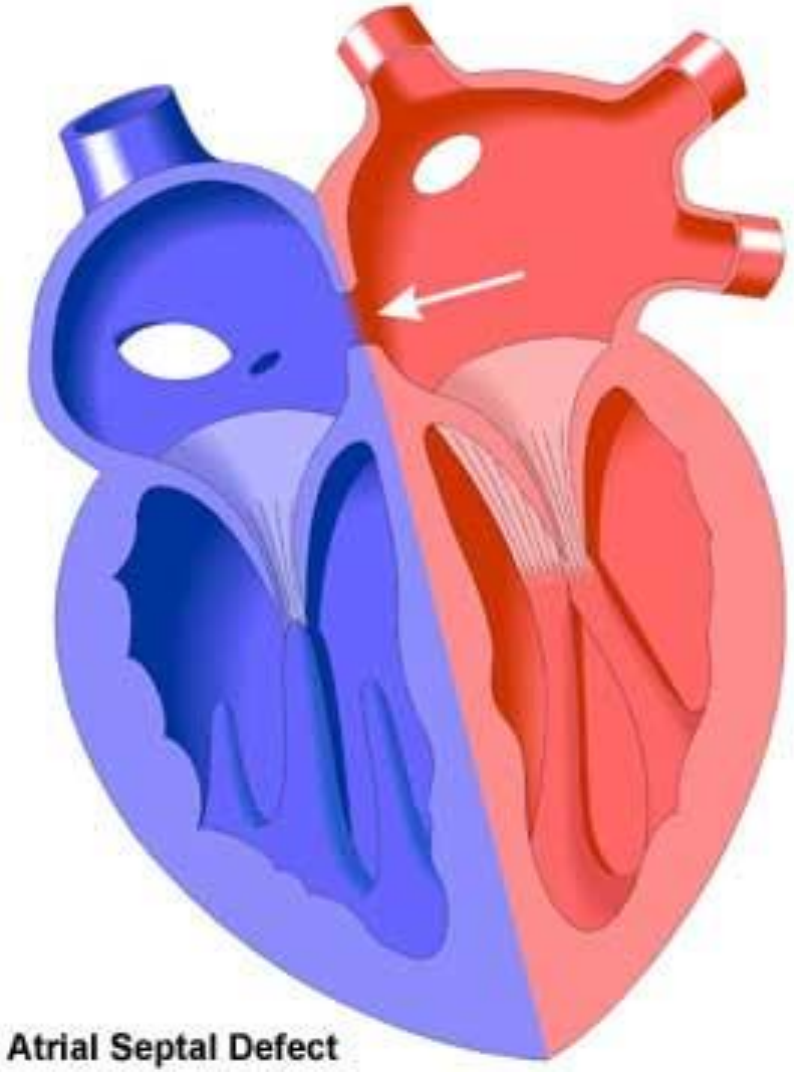
- **Left-to-Right Shunt Lesions :**
  - **Atrial Septal Defect (ASD) .**
  - **Ventricular Septal Defect (VSD).**
  - **Atrioventricular Septal Defect (AV Canal) .**
  - **Patent Ductus Arteriosus (PDA) .**
  - **Partial anomalous pulmonary venous return(PAPVR) .**
  - **Aortopulmonary window(OPW) .**

- **Obstructive lesions:**
  - **Aortic stenosis .**
  - **Pulmonary stenosis .**
  - **Coarctation of aorta.**
  - **Others .**

Atrial Septal Defect





Introduction

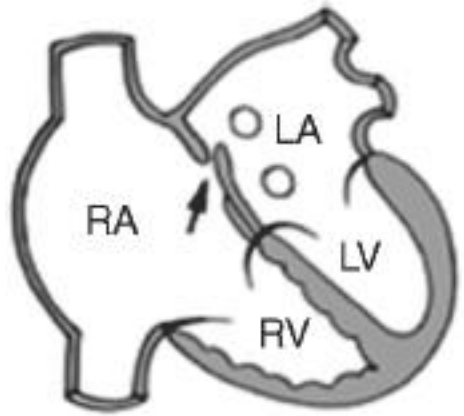
- ASD is an acyanotic CHD characterized by defect in the interatrial septum causing a left to right flow between the atria.
- **Resulting in spectrum from** : **asymptomatic to right sided overload, PAH , even atrial arrhythmias**
- **Pathophysiology :**Shunting occurs during **late ventricular systole and early diastole**

INCIDENCE

- ASD constitutes 8-10% of congenital heart defects in children.
- Incidence = 56 per 100,000 live births
- Recent estimates are much higher (100 per 100,000 live births), likely due to increased recognition in the era of common use of echocardiography.

ETIOLOGY

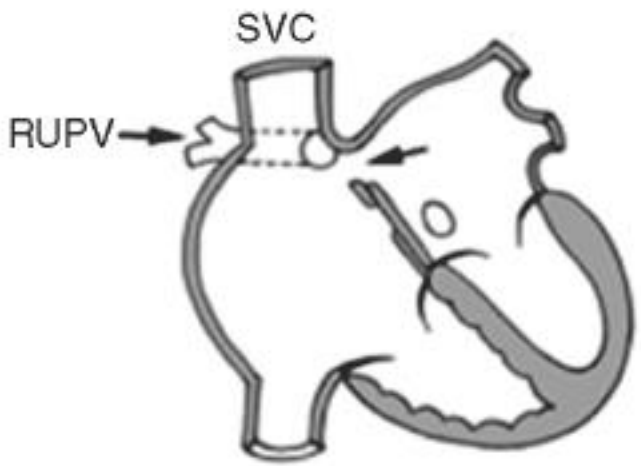
- Actual etiology of this congenital defect is unknown.
- Factors include:
- **Genetic factor:**
  - The genetic basis of ASD is not completely understood.
  - **some homeobox gene defects**..... **familial cases** of ASDs, such as **NKX2- chromosome-5, which has an autosomal dominant inheritence and AV conduction defect**.
  - **HOLT-ORAM Syndrome.....**mutations in the transcription factor **TBX5**, essential in development of **both the heart and upper limbs**.
  - Many other syndromes like
    - **DOWN syndrome**
    - **Noonan syndrome**
    - **Patau's syndrome**
    - **Edward's syndrome**
- **Environmental factor including antenatal use of teratogenic drugs, congenital infection**



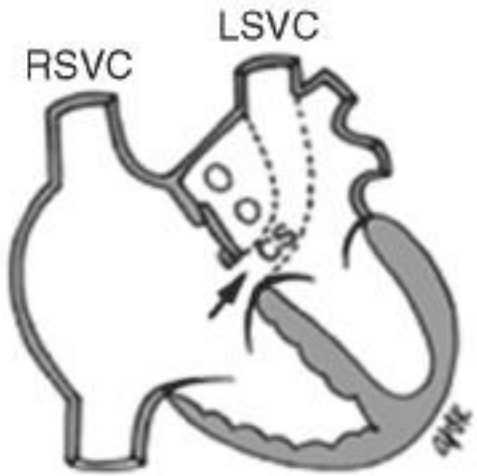
Patent foramen ovale

Fossa ovalis or
secundum defect

Primum defect



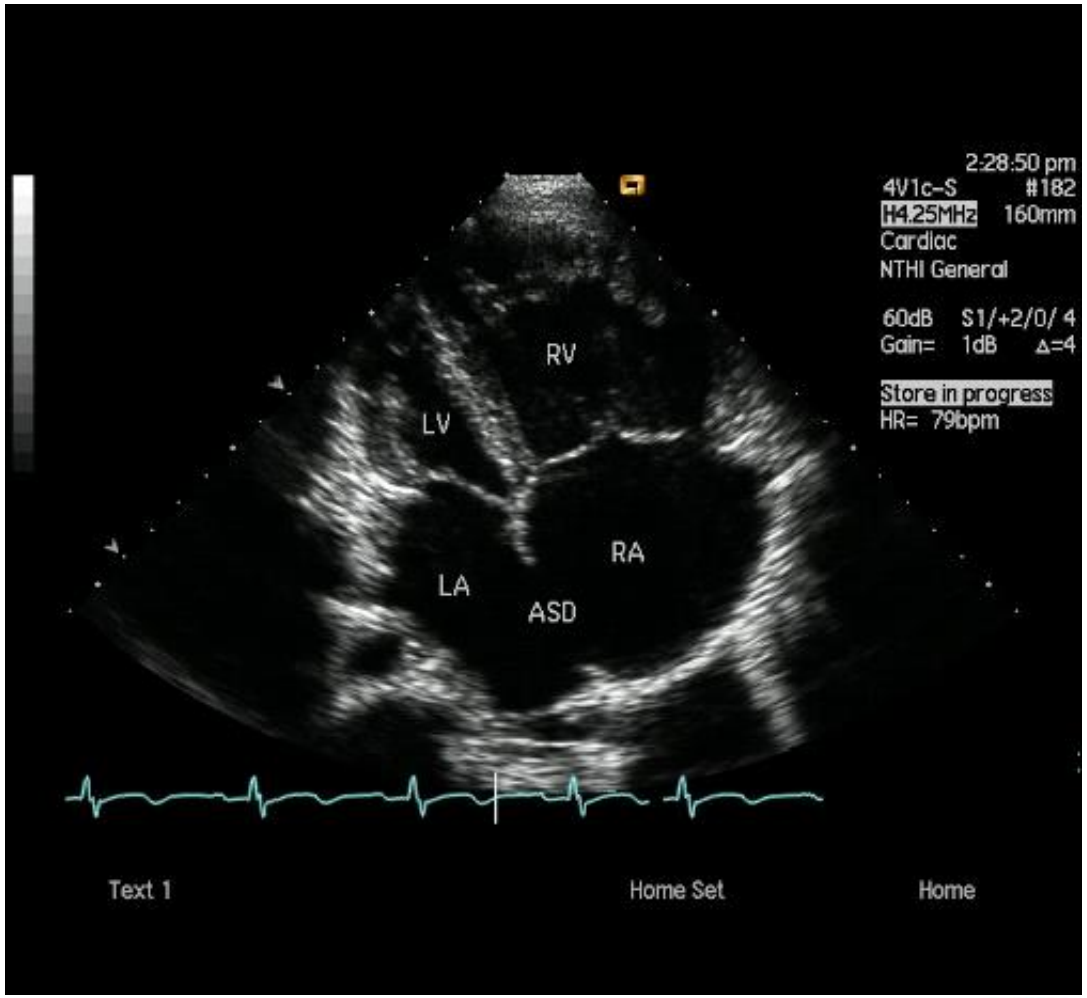
Sinus venosus defect



Coronary sinus defect
(Unroofed coronary sinus)

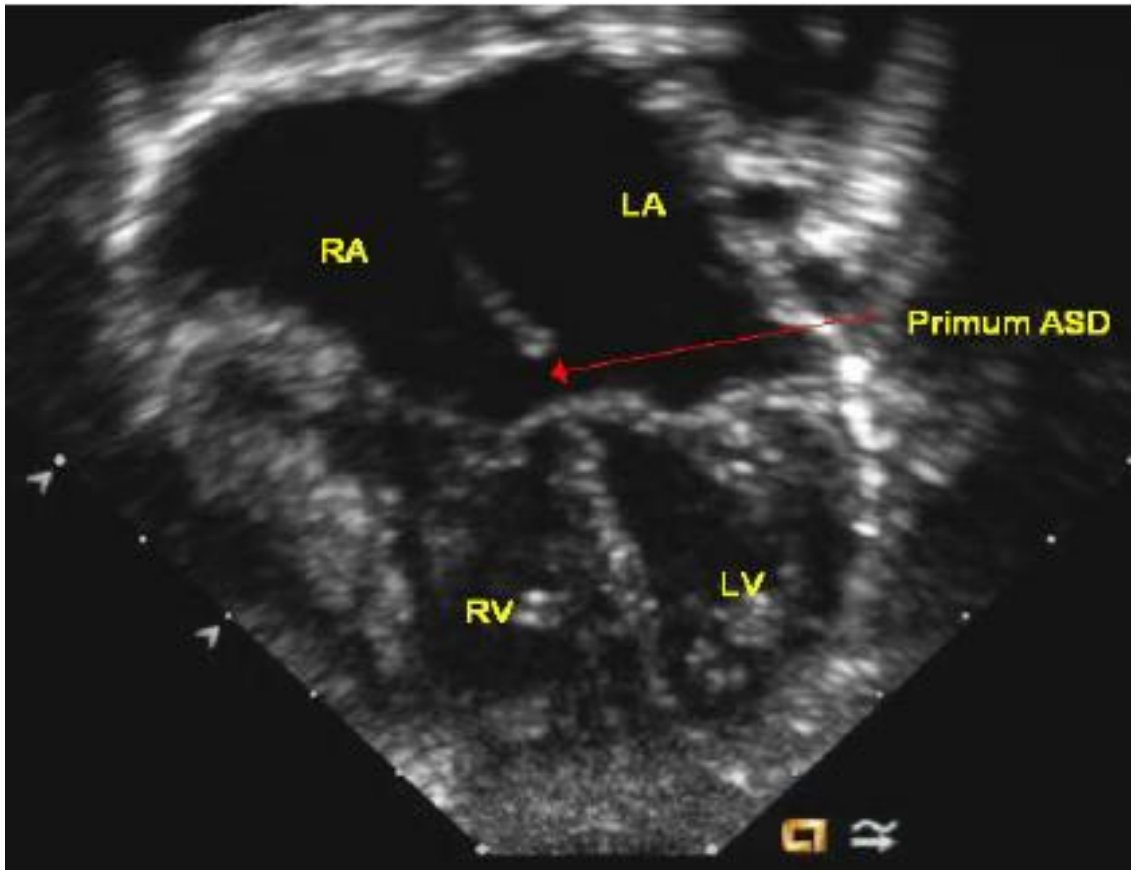
Secundum ASD

- **Secundum ASD (75-85%)** are located in the region of the fossa ovalis.
- **Most common type**.
- **Single or Multiple.**
- **May be associated with other ASDs.**
- Typically located within the area **bordered by the limbus of the fossa ovalis.**
- May be associated with
  - **Partial anomalous venous return most commonly of the right upper pulmonary vein.**
  - **Pulmonic stenosis**
  - **Mitral valve prolapse (**10-20% have a **functional mitral valve prolapse, m**ay be related to changing LV geometry associated with RV volume overload**)**



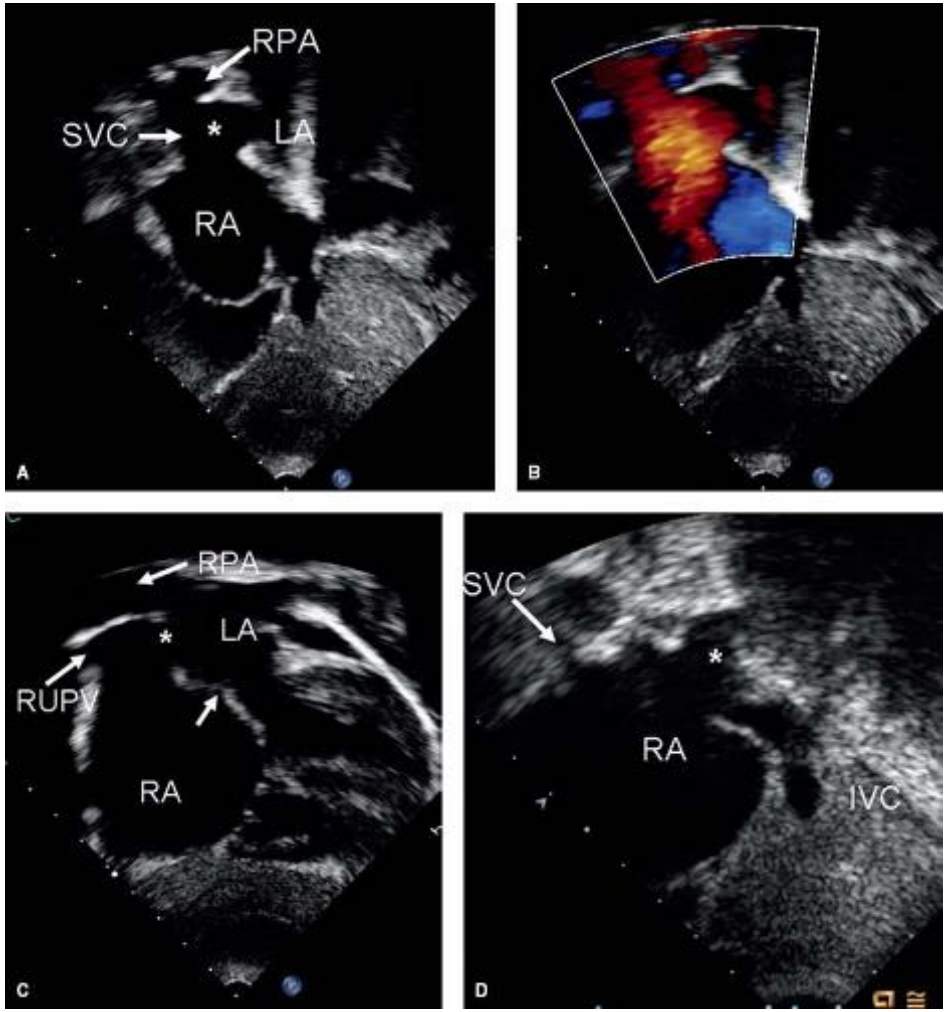
Primum ASD

- **Primum ASD** (10-15%) occur in the lower portion of the atrial septum.
- Occur if the **septum primum does not fuse with the endocardial cushions, leaving a defect at the base of the interatrial septum that is usually large**.
- Usually not isolated – primum ASDs are **typically** associated with
  - anomalies of the AV valves (such as **cleft mitral valve**)
  - and defects of **the ventricular septum (VSDs) or a common AV canal.**
- May associated with **Discrete subaortic stenosis**



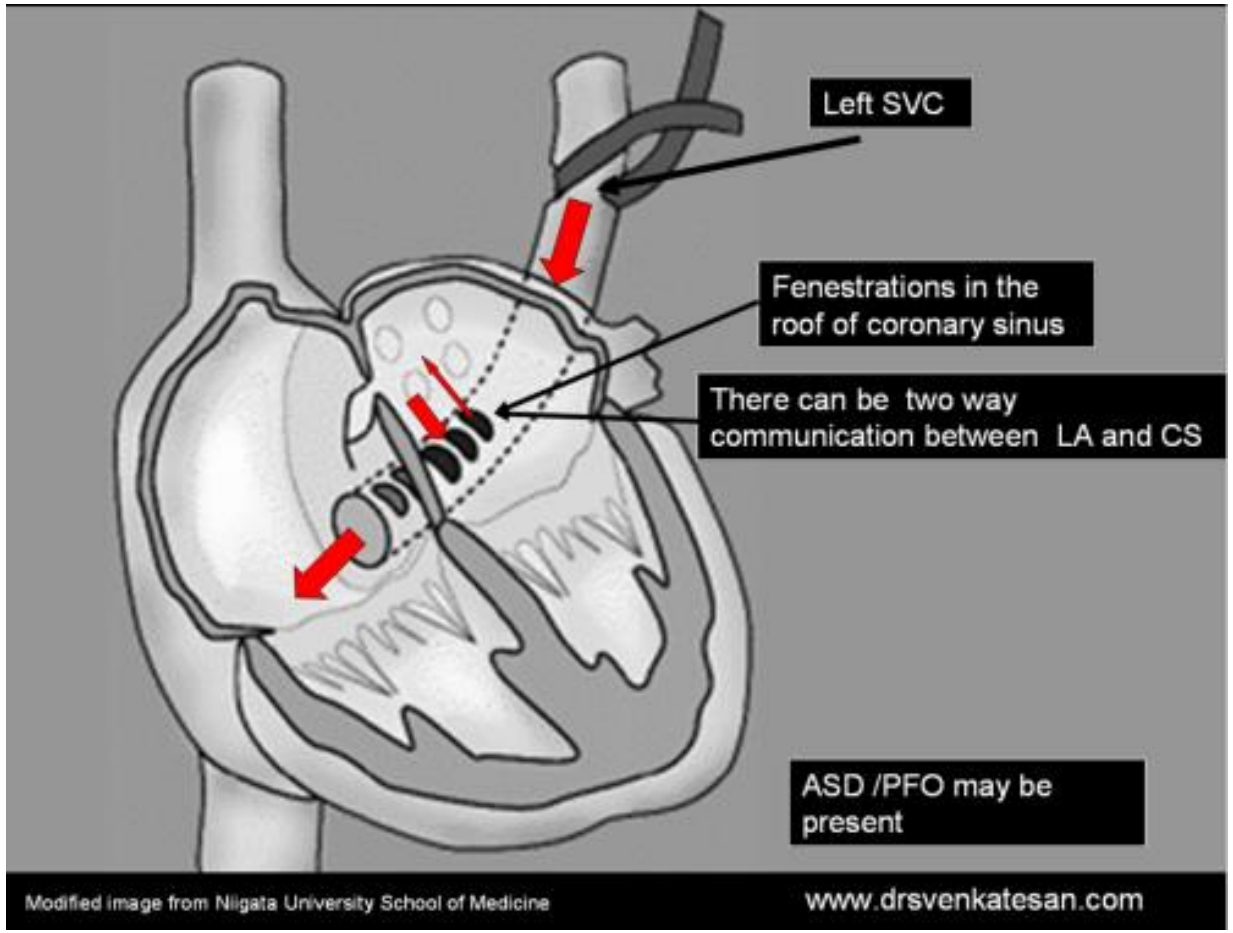
Sinus venosus ASD

- Make up Characterized by malposition of the insertion of the SVC or IVC straddling the atrial septum.
- **Sinus venosus superior defect** (5-10%) are located near the orifice of the superior vena cava.
- **Sinus venosus defects of IVC type** (1%)
- Often associated **with anomalous pulmonary venous return** – the RU/RM pulmonary veins may connect with the junction of the SVC and RA in the setting of a superior sinus venosus ASD.



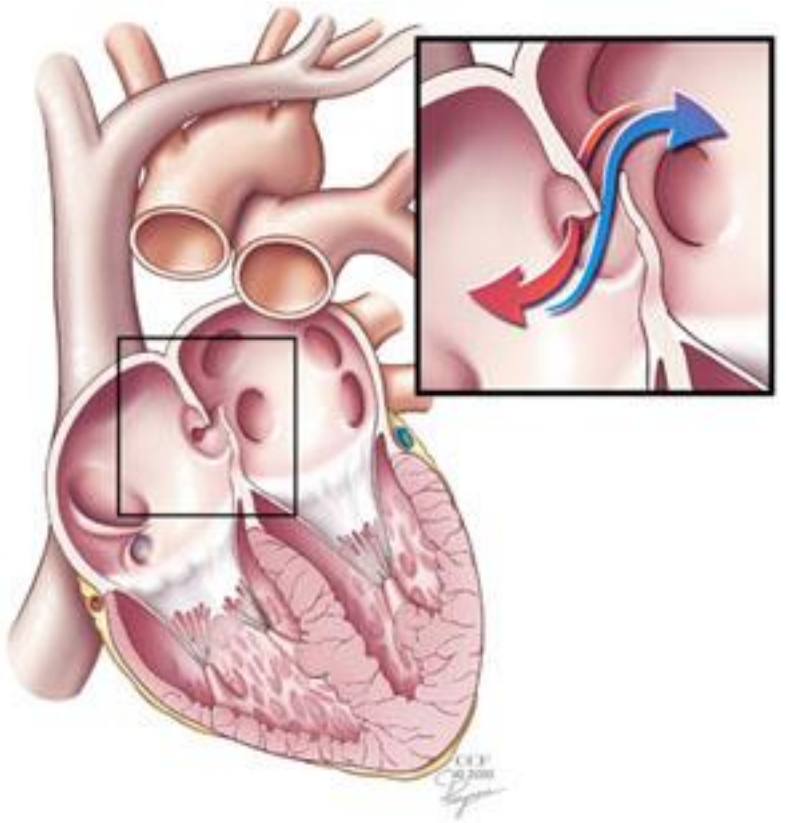
Coronary Sinus Septal Defects

- **Coronary sinus defects**(Less than 1% of ASDs)
- **Defects in the inferior/anterior atrial septum region that includes the coronary sinus orifice.**
- Defect of at least a portion of the common wall separating the coronary sinus and the left atrium – AKA "**unroofed coronary sinus"**
- Can be associated with a
  - **persistent left SVC draining into the coronary sinus**
  - **or Partial and total anomalous pulmonary venous return**



Patent Foramen Ovale

- **Not truly an "ASD" because no septal tissue is missing.**
- Oxygenated blood from the IVC crosses the foramen ovale in utero.
- At birth, the flap normally closes due to
  - Reduced right heart pressure and PVR
  - Elevated LA pressure.
- Flap fusion is complete by age two in 70-75% of children; the remainder have a PFO.

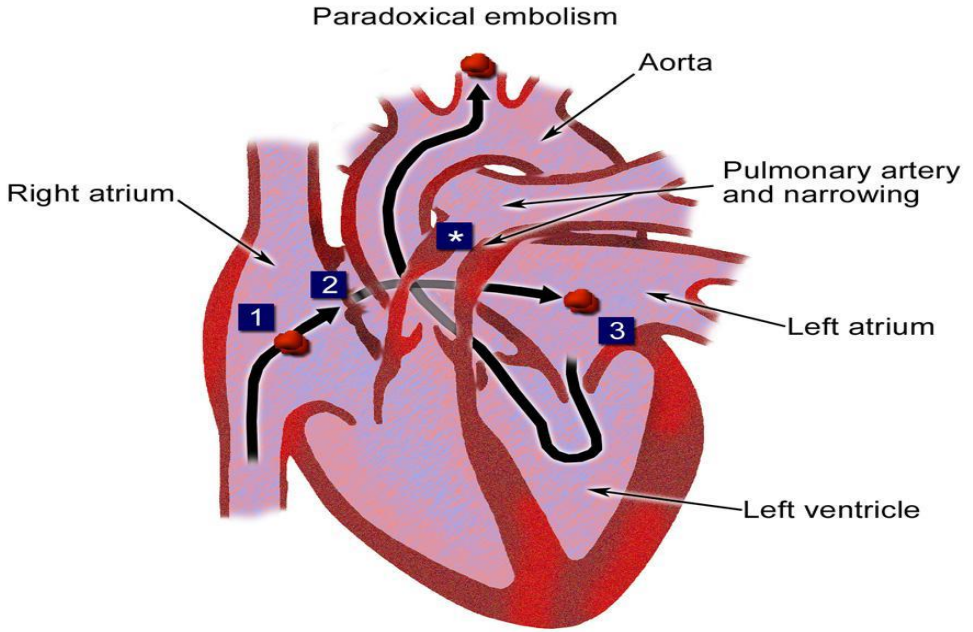


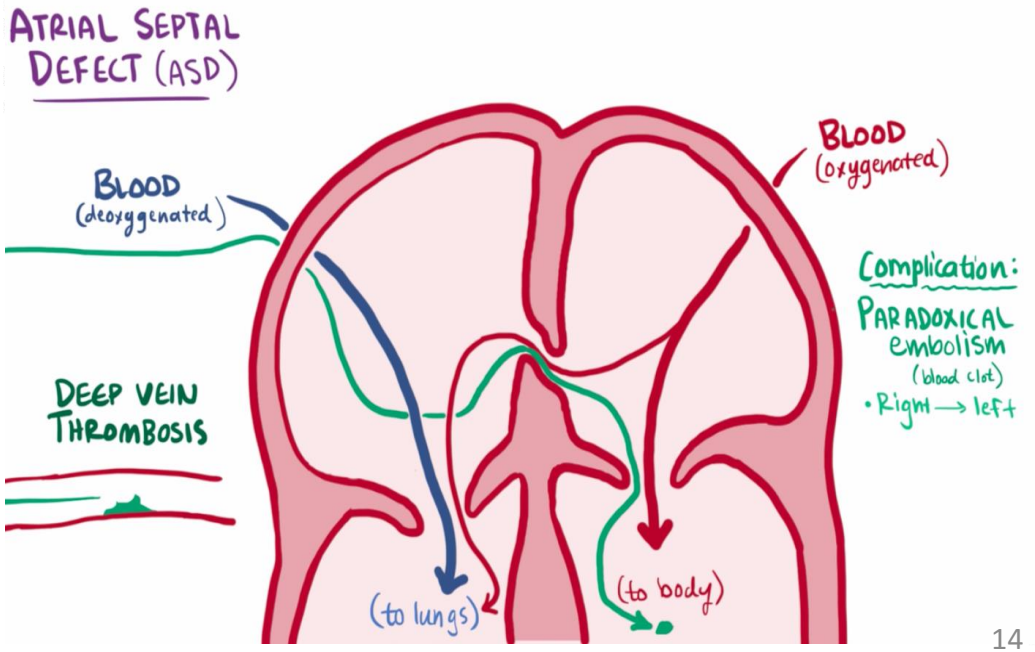
Natural History of ASDs

- **Most small ASDs close spontaneously in infants.**
- Spontaneous closure **is unusual in children and adults**; defects often **become progressively larger.**
- **Advanced pulmonary hypertension seldom occurs before the third decade.**
- **Infective endocarditis does not occur** in patients with isolated ASDs.

Natural History of ASDs

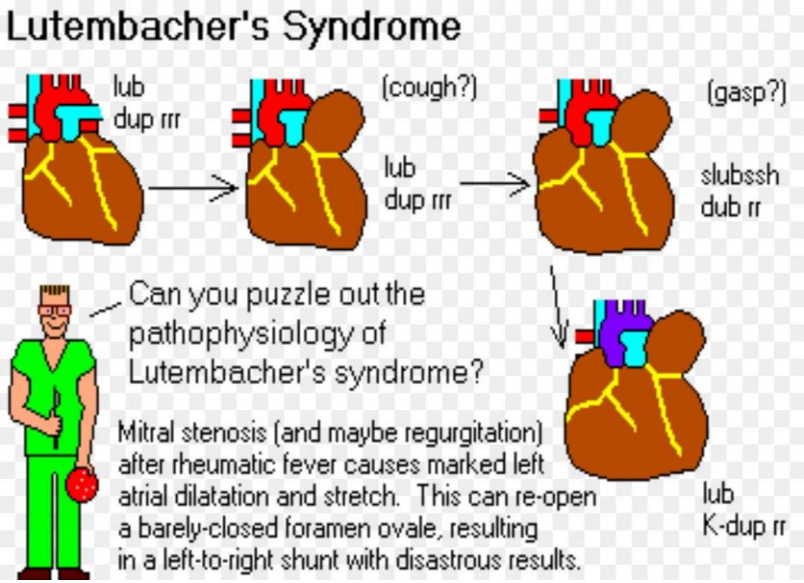
- **Cerebrovascular accident, resulting from paradoxical embolization through an ASD, is <u>a rare complication but important</u>**
- Increased prevalence **of <u>PFO and PFO associated – stroke</u>**.

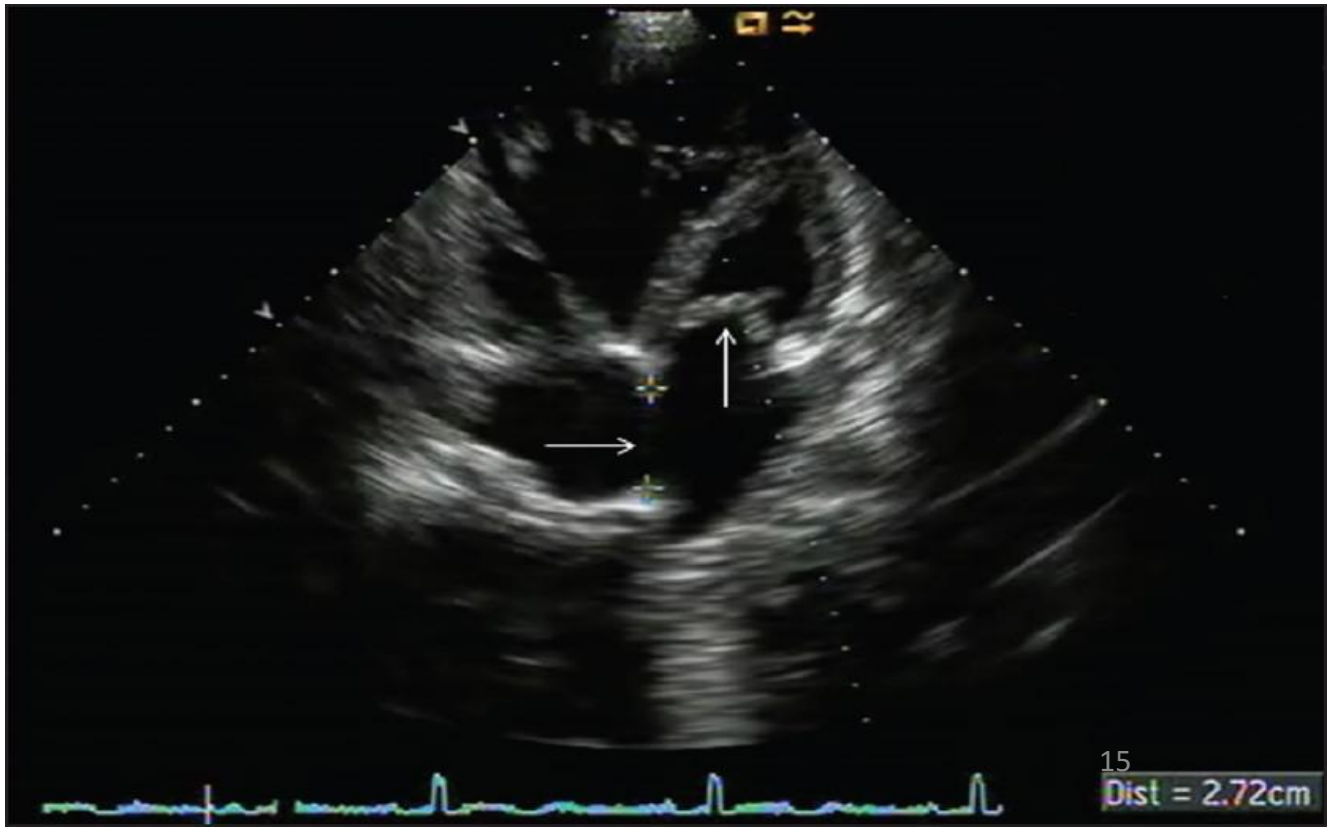




Natural History of ASDs

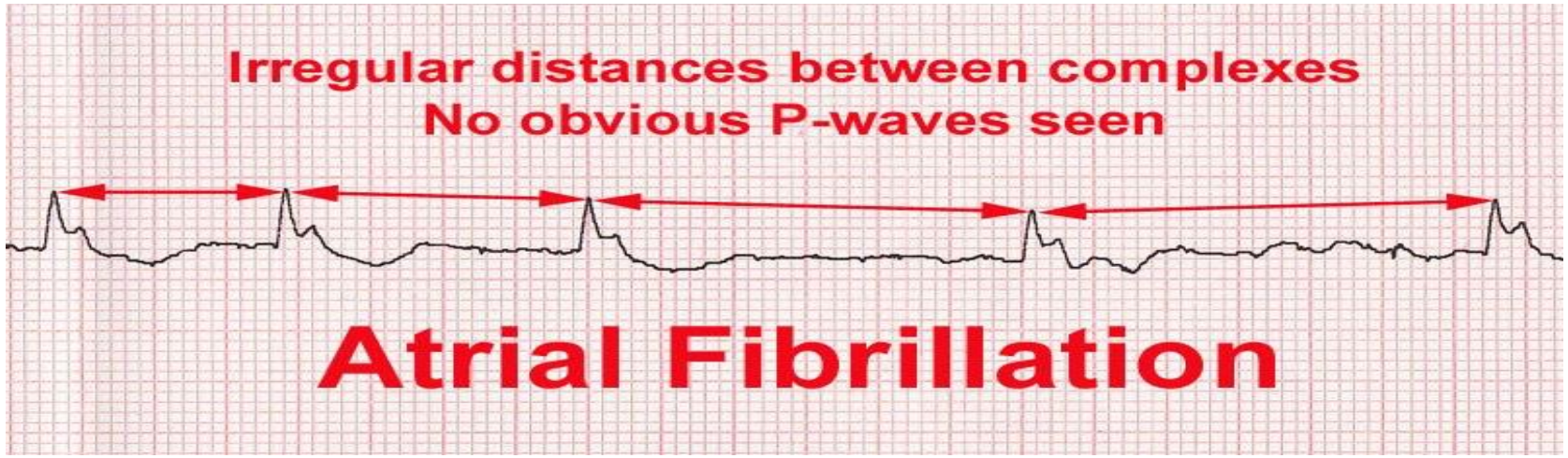
- **Mitral stenosis** may occur as a result of rheumatic fever in a case of ASD (**Lutembacher syndrome**).





Natural History of ASDs

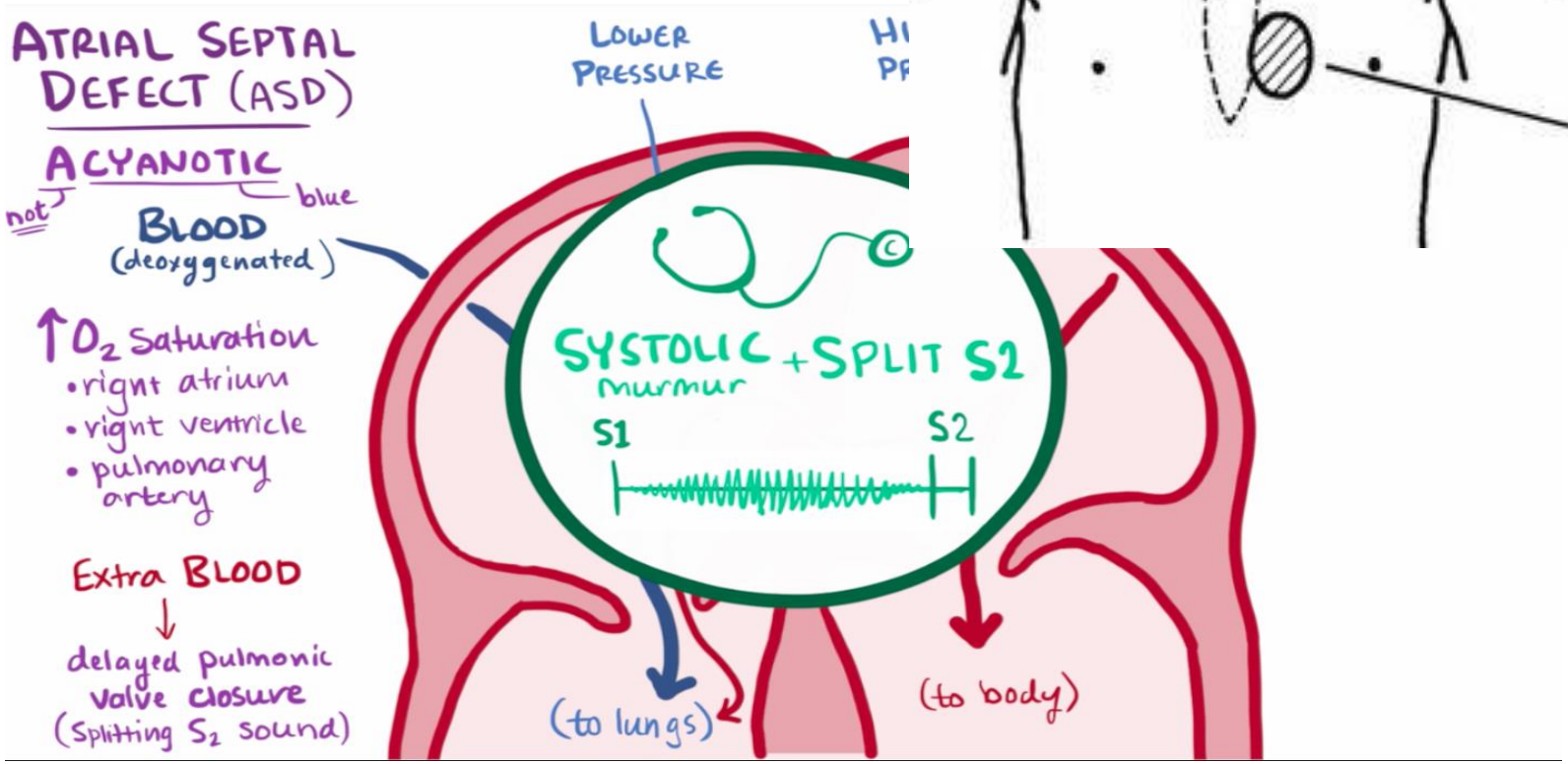
- **Atrial fibrillation and /or Atrial flutter is a <u>late complication</u>**
- Risk of atrial arrhythmias increases with **age and PA pressure.**

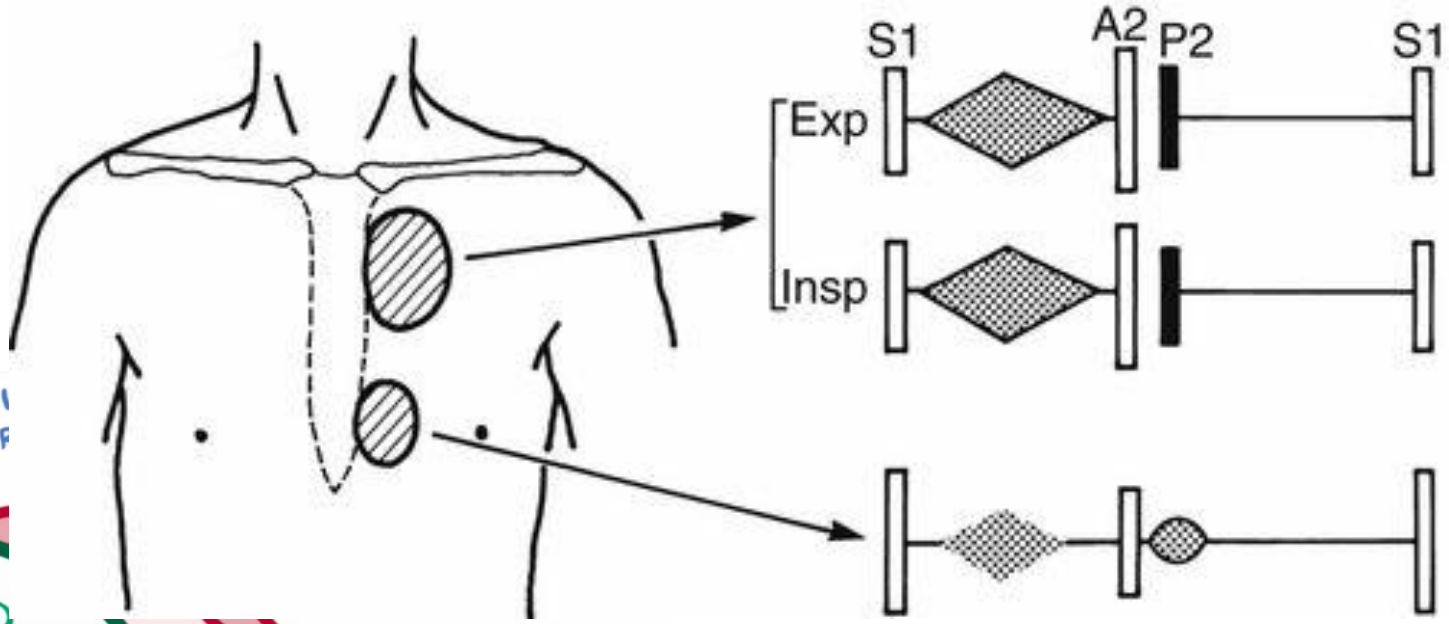


Clinical Manifestations

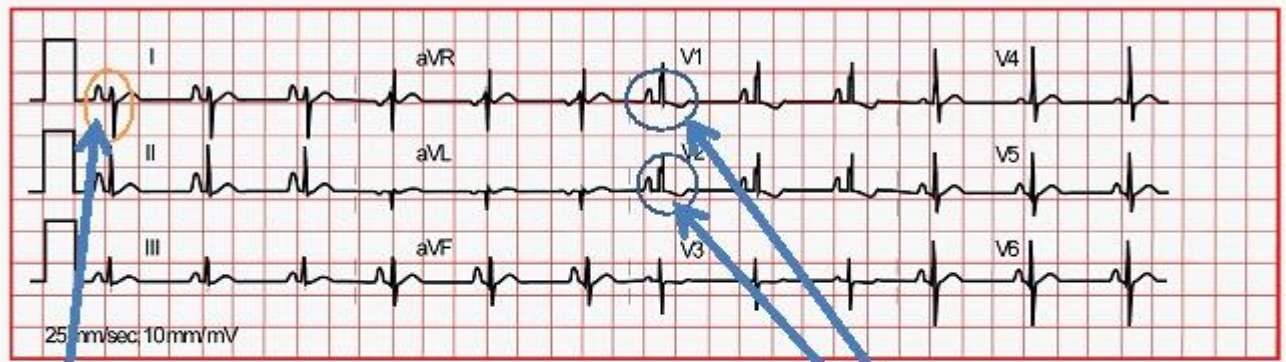
- Vary with the size of defect.
  - **Small defect**: Asymptomatic and is usually diagnosed during a routine health check up.
  - **Large defect:** Symptomatic and patients usually present with Failure to thrive, Easy fatigability. Increased perspiration, Recurrent Pulmonary infections, or Platypnea Orthodeoxia
    - Most patients with a significant shunt flow ratio **(Qp:Qs > 1.5:1)** will **be symptomatic** .

Physical Exam Findings





ECG



Enlarged 'p' wave indicating Right atrial hypertrophy

Also note that the aVF is predominantly upwards as compared to Lead I indicating Right Axis Deviation

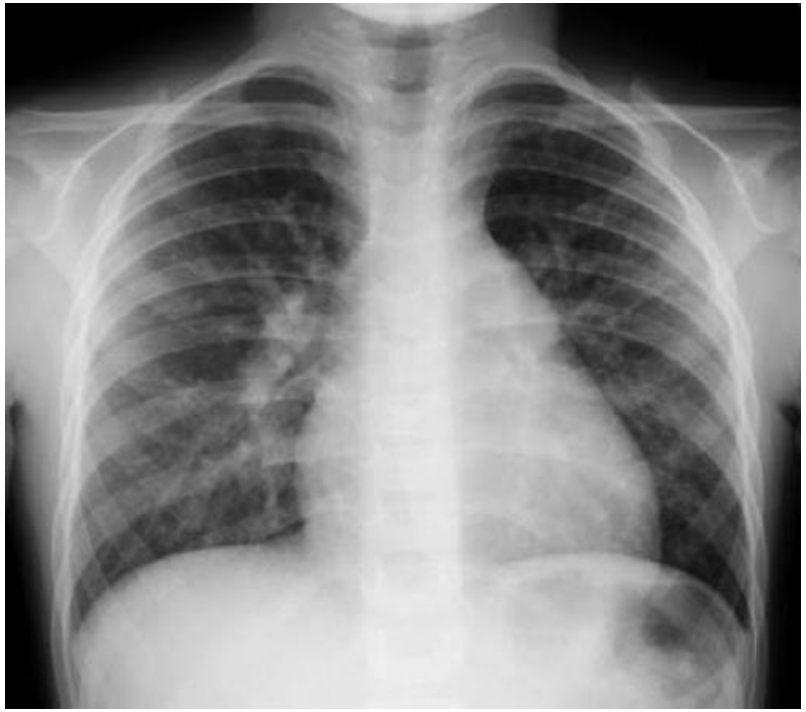
rSR' seen and tall R wave Indicating RBBB and RVH

LAD with rSR' in V1 is suggestive of Ostium primum defect

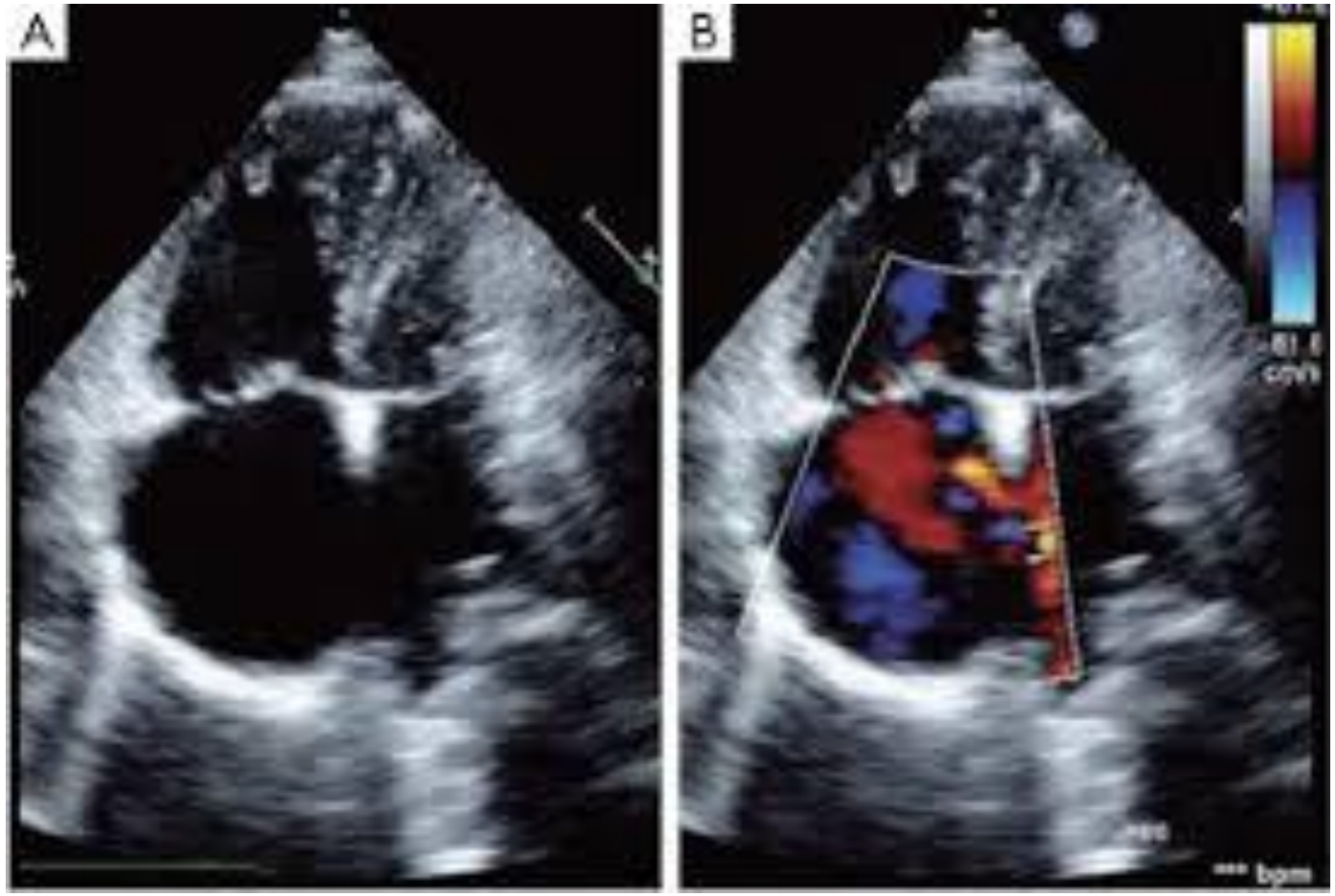
Chest X-Ray

- **Small defect-NL chest x-ray**

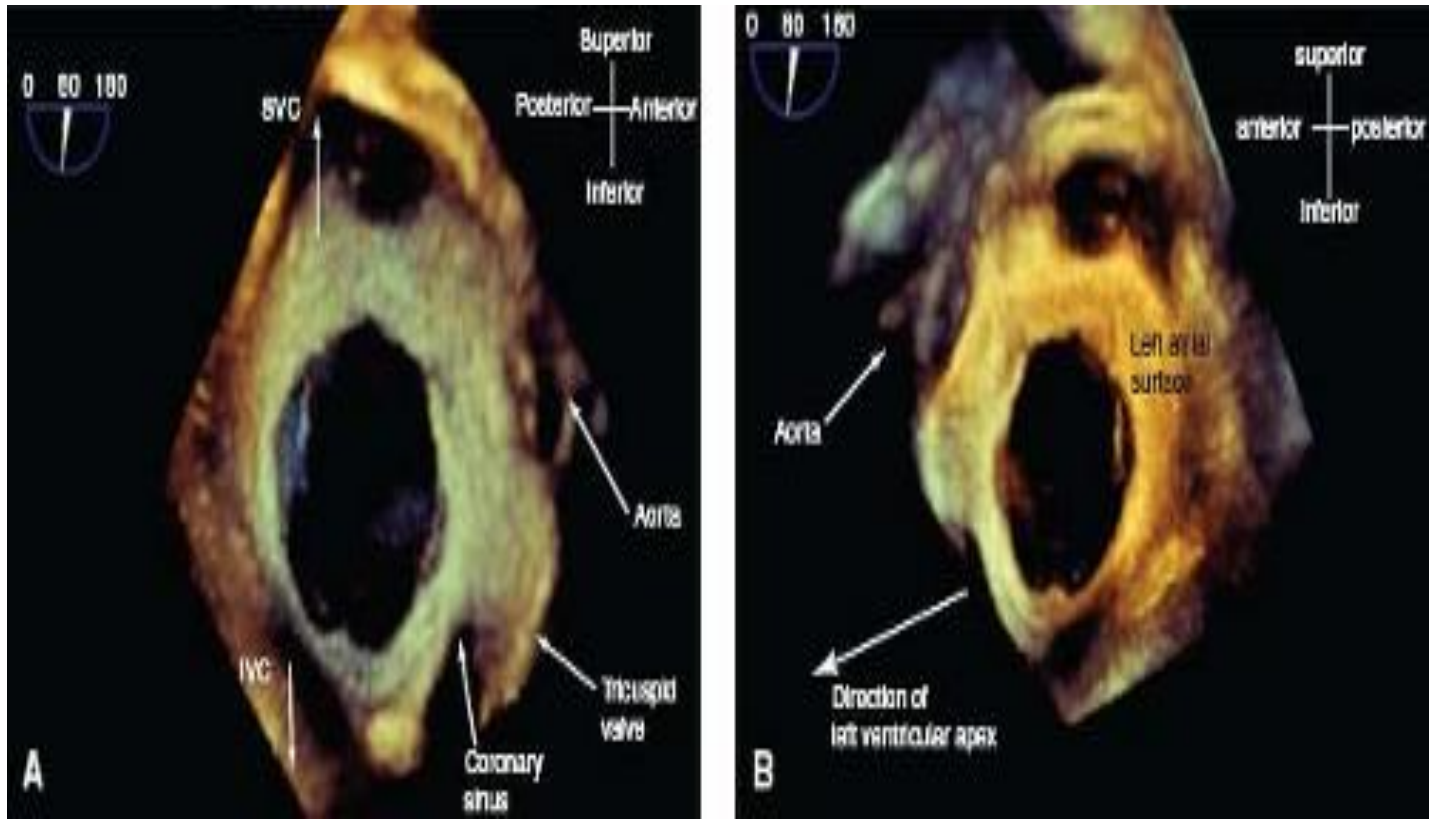
- **Large defect:**
  - **Dilation of RA and RV**
  - **Enlarged main pulmonary arteries and pulmonary vessels**.



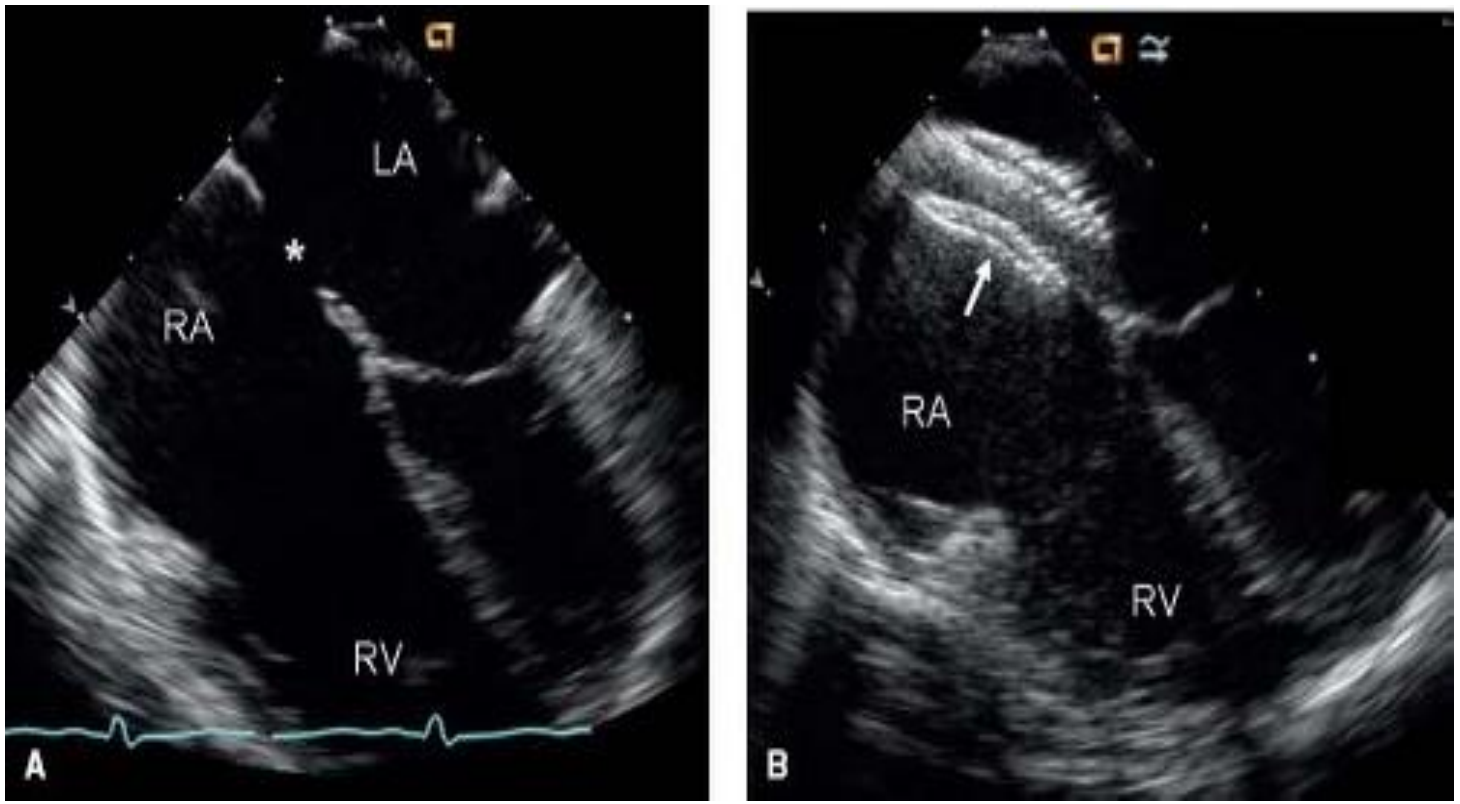
2D Color Doppler TTE



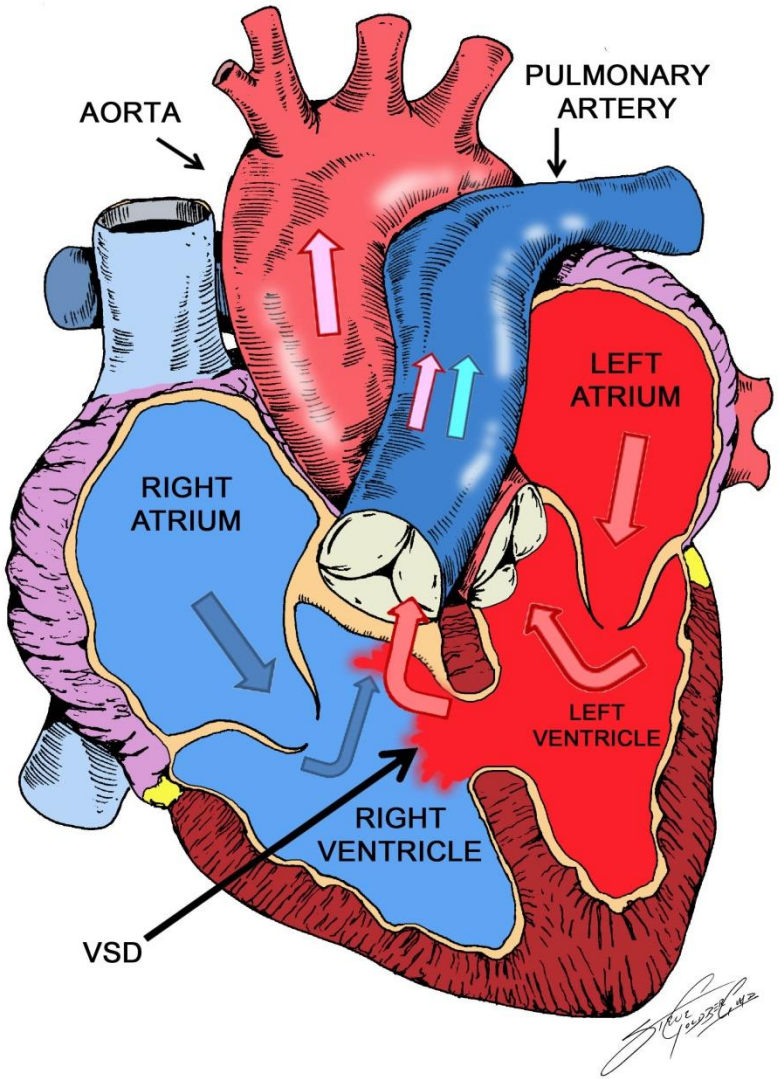
3 D-ECHO


0 60 180
SVC
Superior
Posterior
Anterior
Inferior
Aorta
IVC
Tricuspid valve
Coronary sinus
A
0 60 180
superior
anterior
posterior
inferior
Aorta
Left atrial surface
Direction of left ventricular apex
B

Amplatzer - Occlusion Device



Ventricular Septal Defect (VSD)



Isolated VSD - most commonly recognized CHD .

50 % when associated with other major defects .

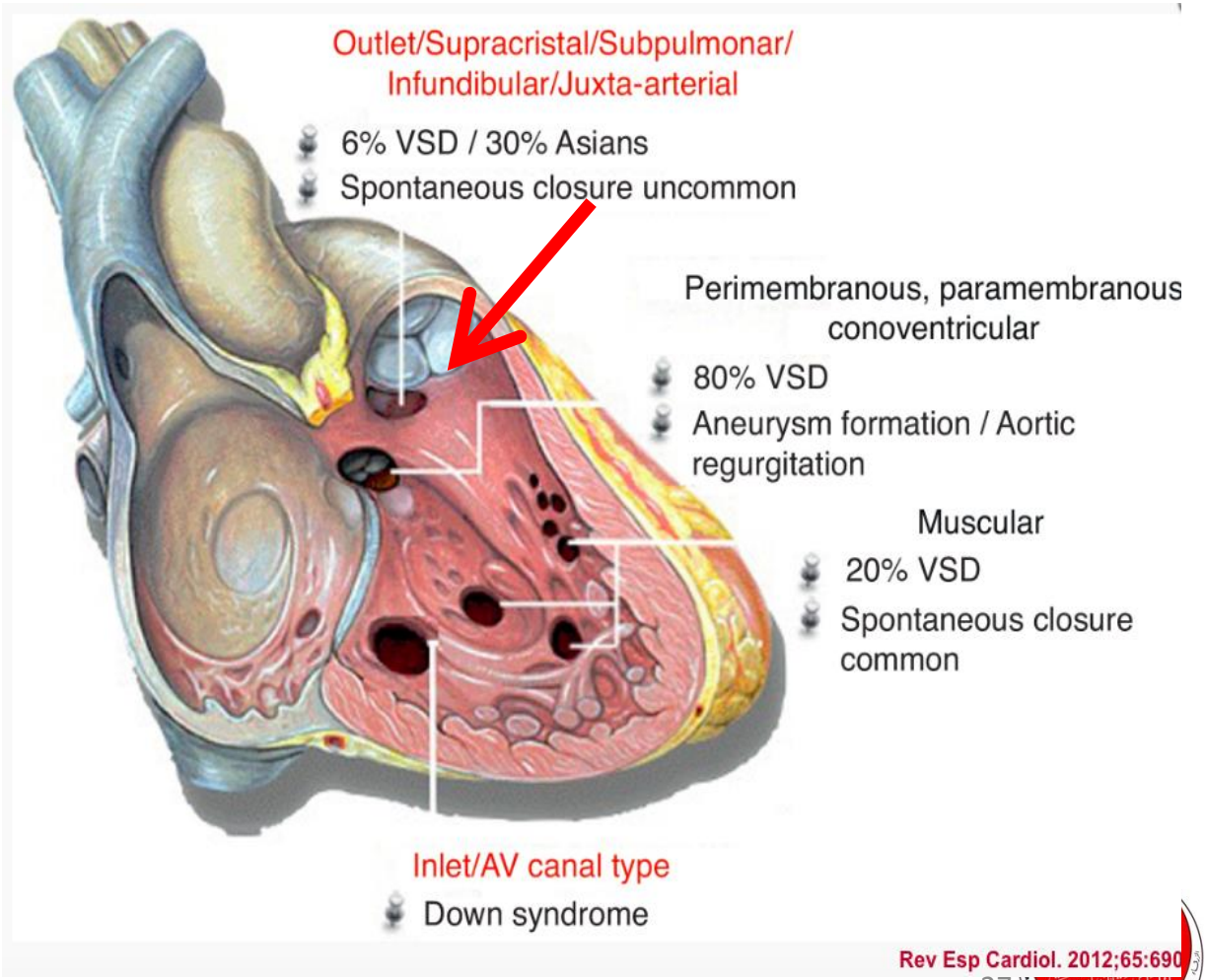
75-80% of small VSD's close spontaneously by late childhood.

Associated syndromes

- **Trisomy 21**: **40%** of T21 will have VSD
- **Trisomy 13, 18**
- **22q11 deletion**
- **Holt-Oram**

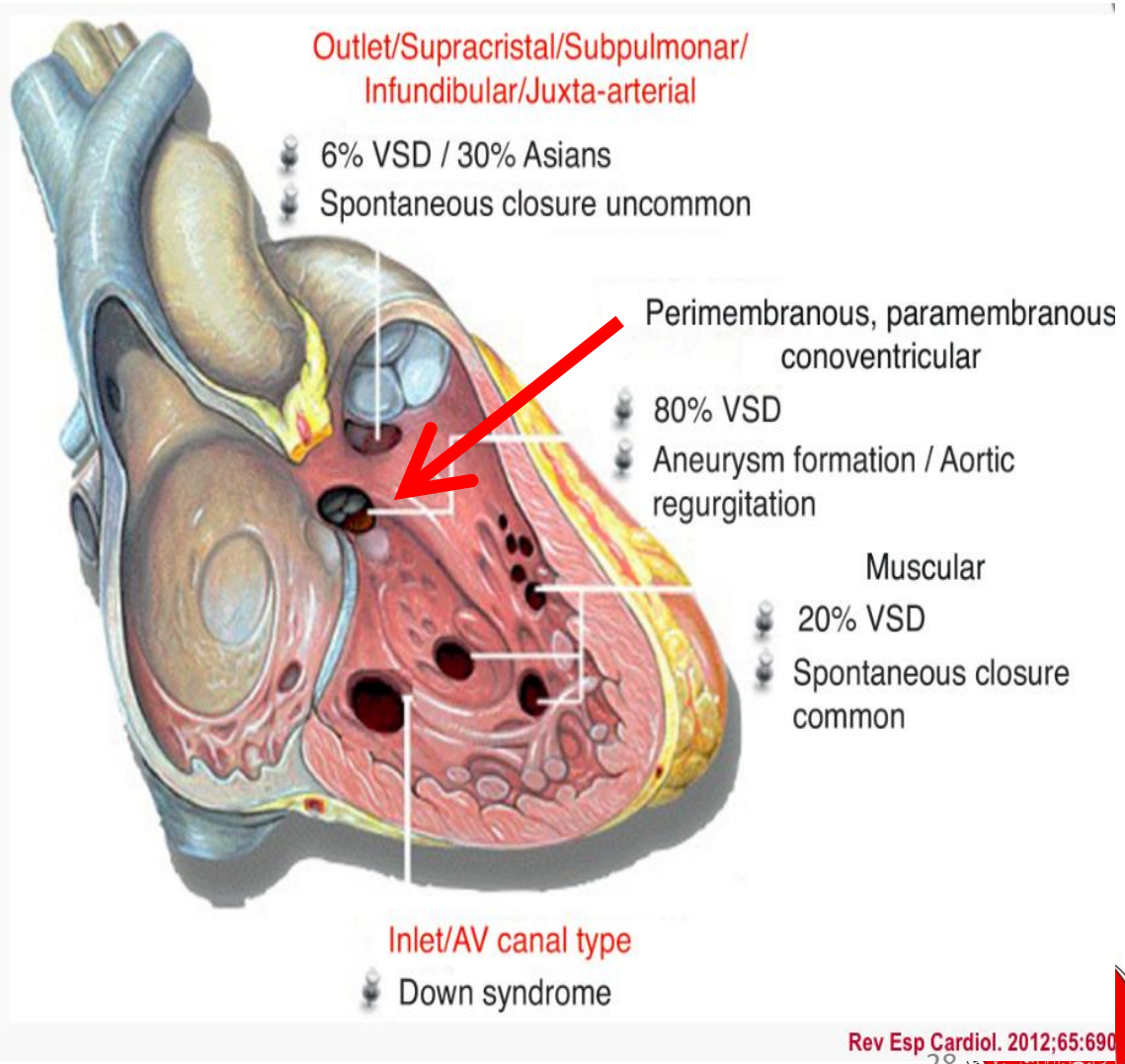
TYPE I (Conal, Supracristal, Infundibular, Subarterial, subpulmonic, doubly committed, outlet) VSDs

- Sub **pulmonary valve**
- Communicate with the RV outflow tract above the supraventricular crest
- Associated with **aortic regurgitation secondary to the prolapse of the right aortic cusp.**
- **Conduction system is <u>not in surgical proximity</u>**

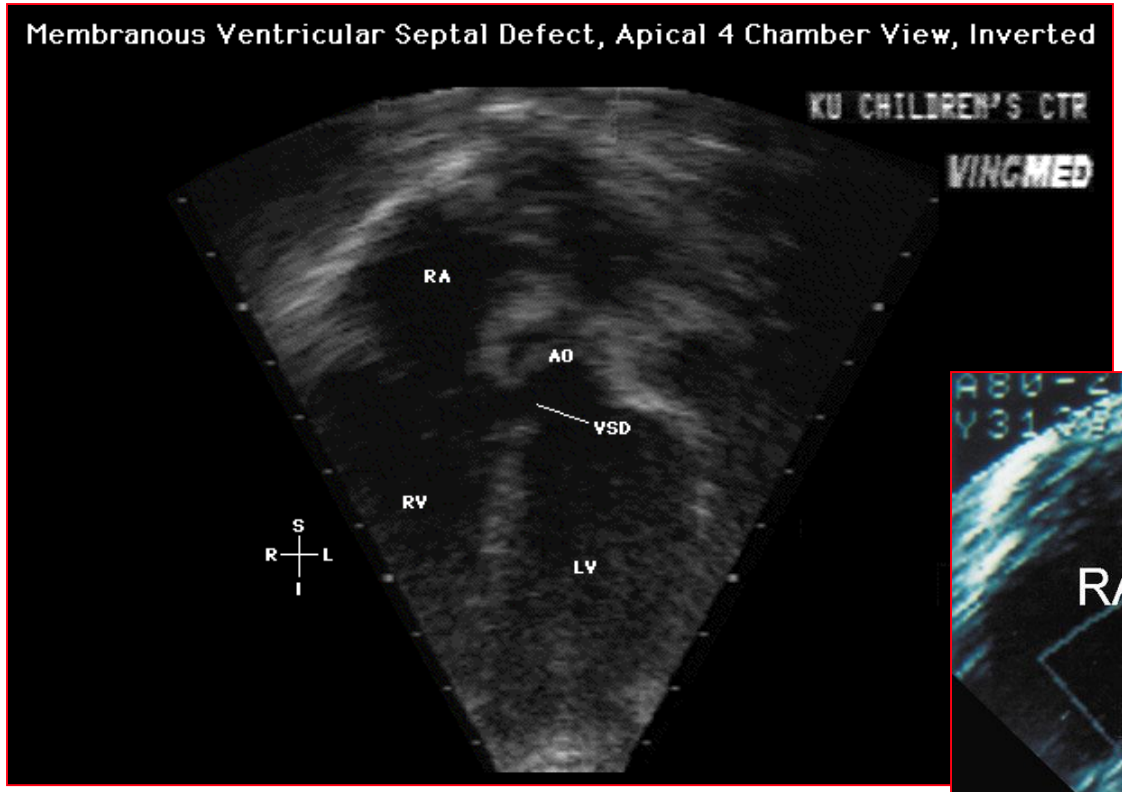


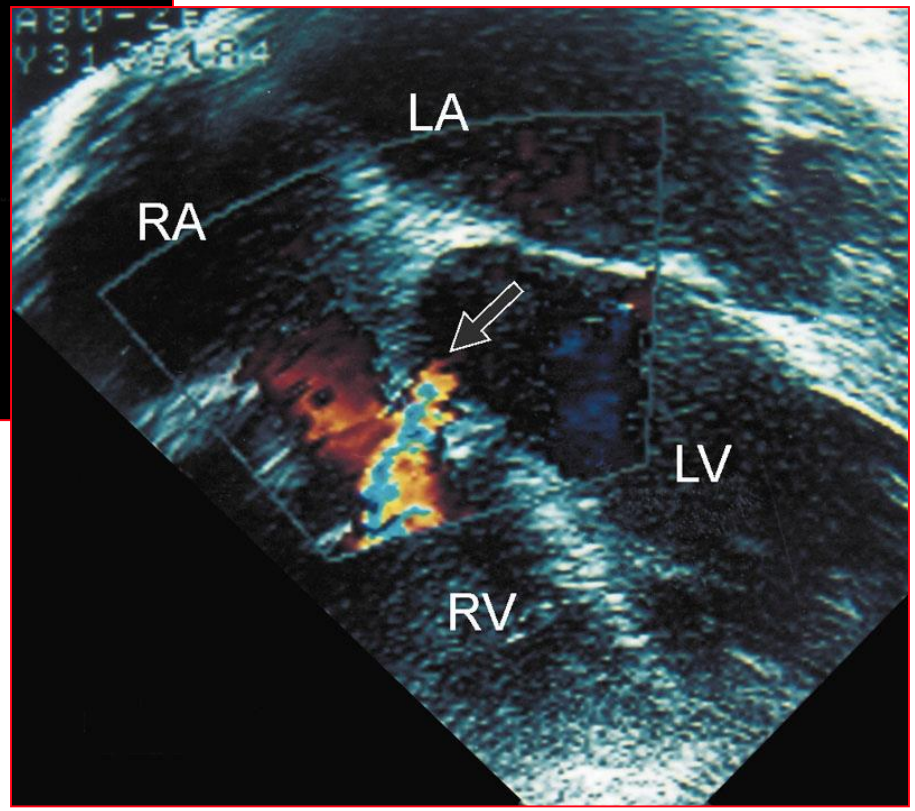
TYPE II – Perimembranous (infracristal,conoventricular) VSD

- **Most common (80%)**
- Sub **aortic valve**
- Associated with **pouches or aneurysms of the septal leaflet of the tricuspid valve**, which can partially or completely close the defect.
- Gerbode shunt :An LV-to-RA shunt may be associated with this defect
- **Danger area- inferior and posterior region of defect.**



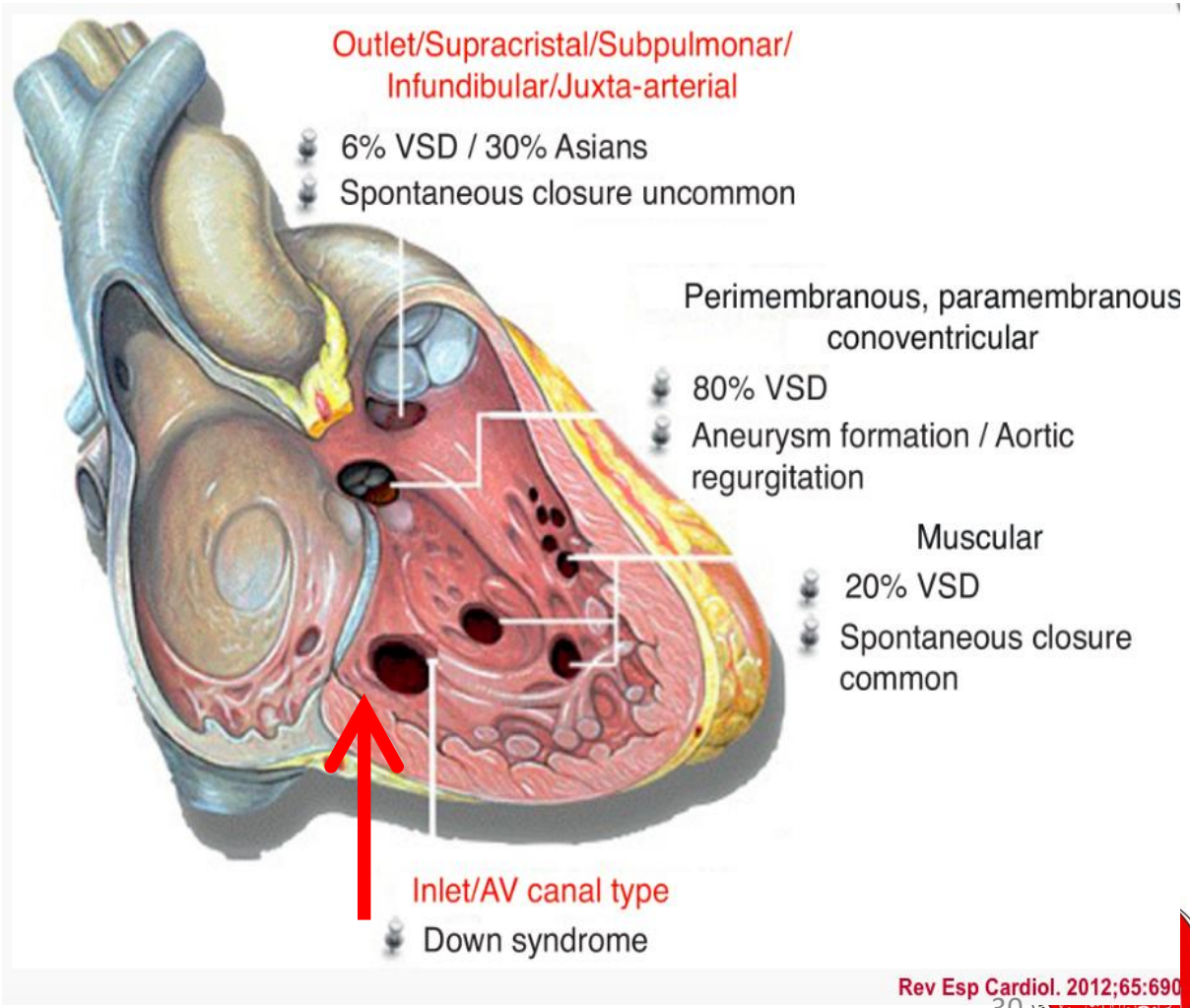
Echocardiogram: Membranous VSD



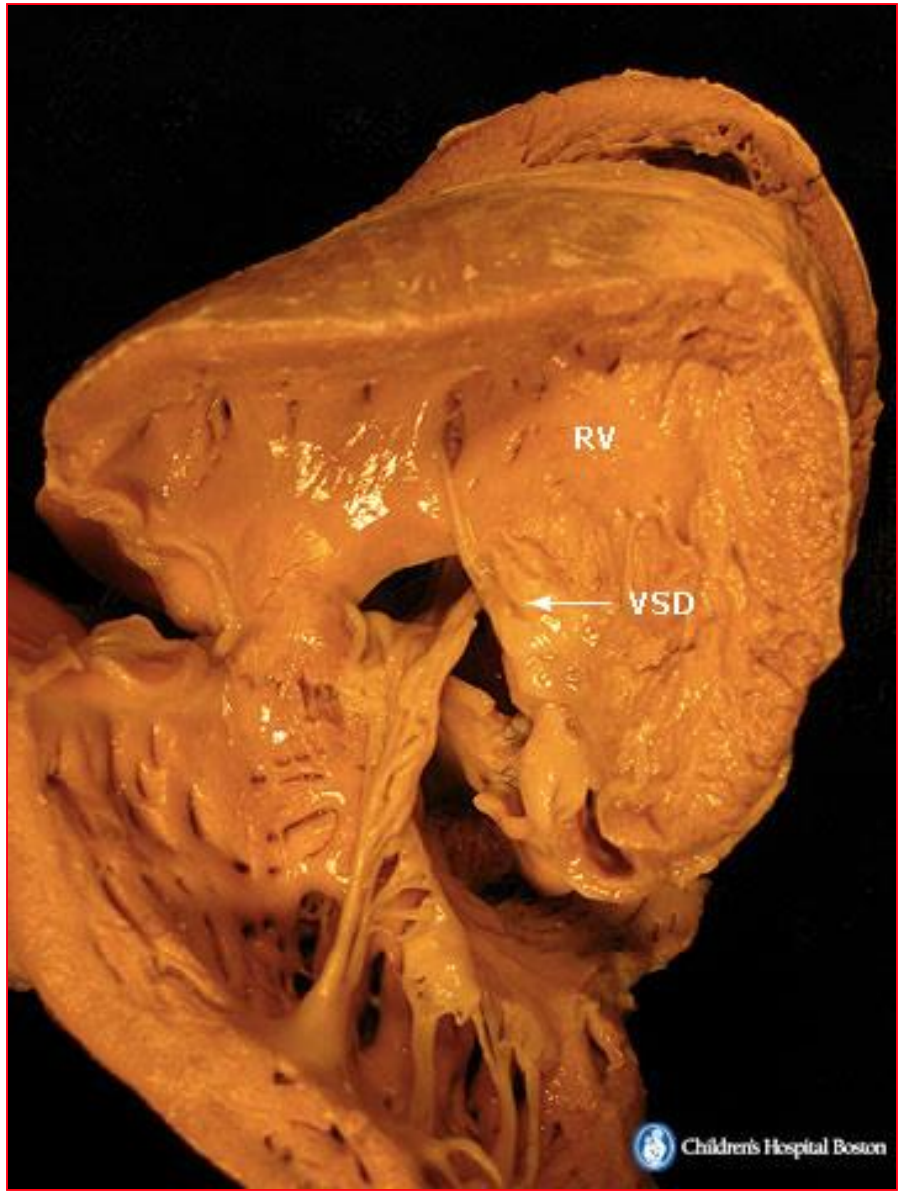


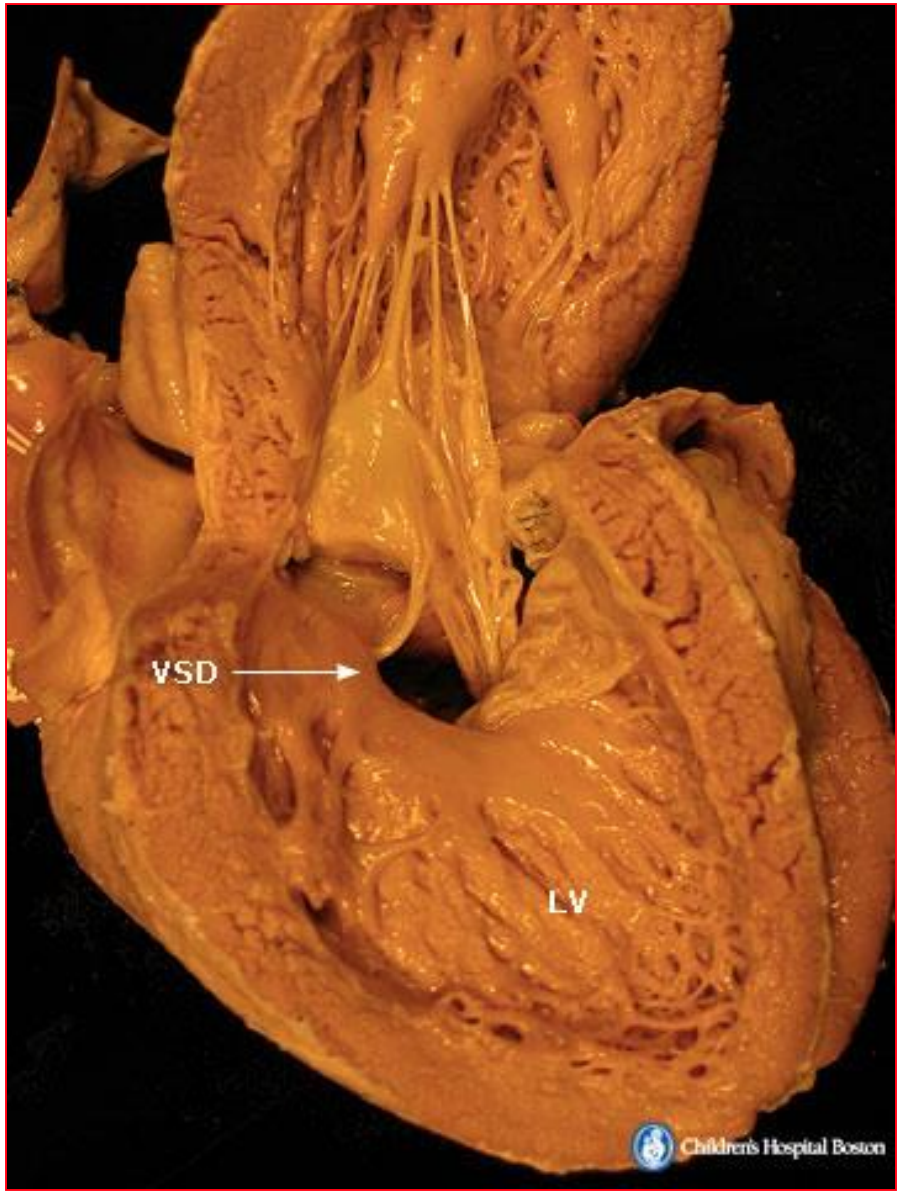
TYPE III AV Canal type / Inlet VSDs

- 5% of all VSDs
- Located posteriorly
- Conduction system at risk – **close proximity to AV node**
- **Common bundle courses around inferior aspect of defect**



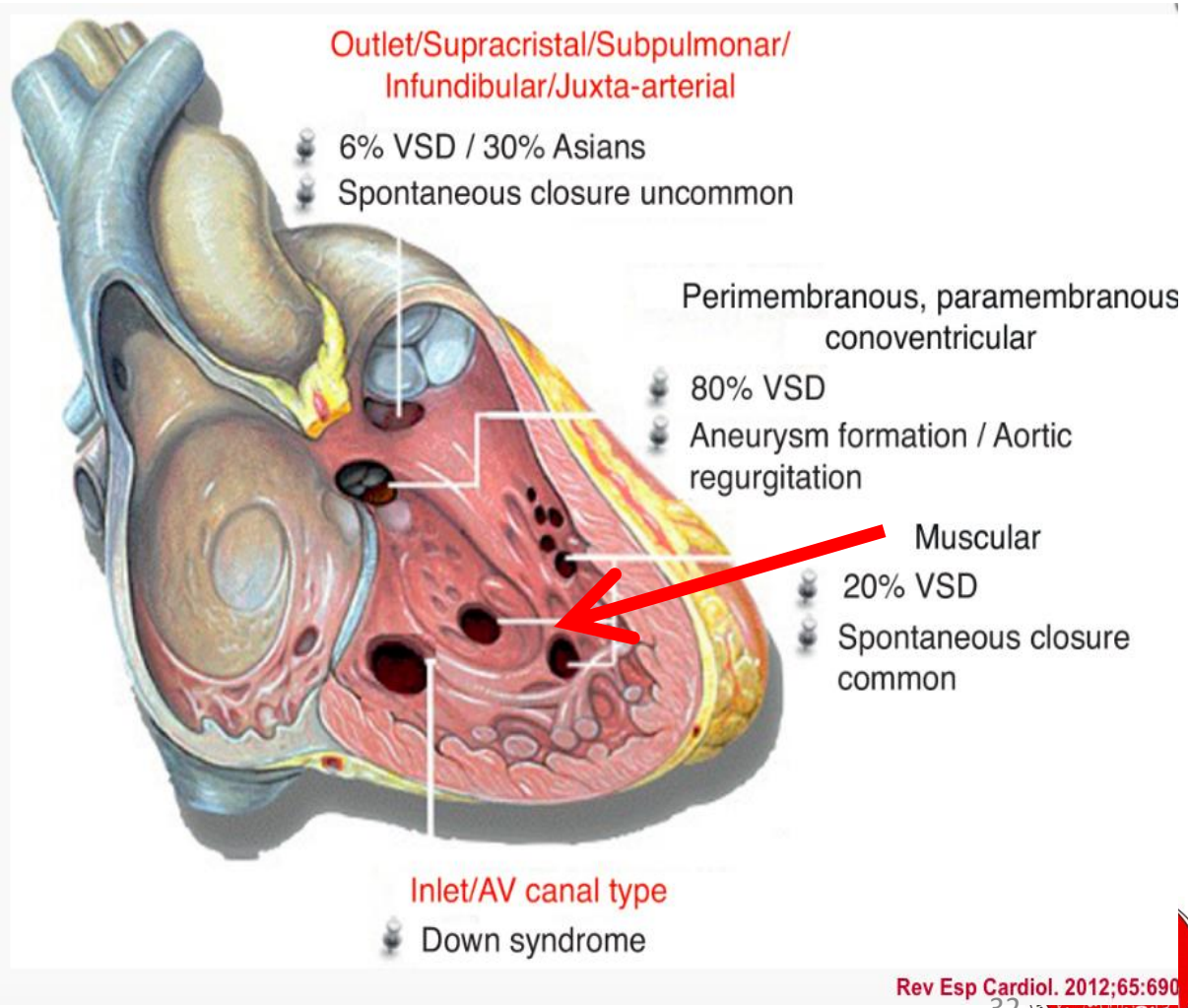
Endocardial Cushion (Inlet VSD)



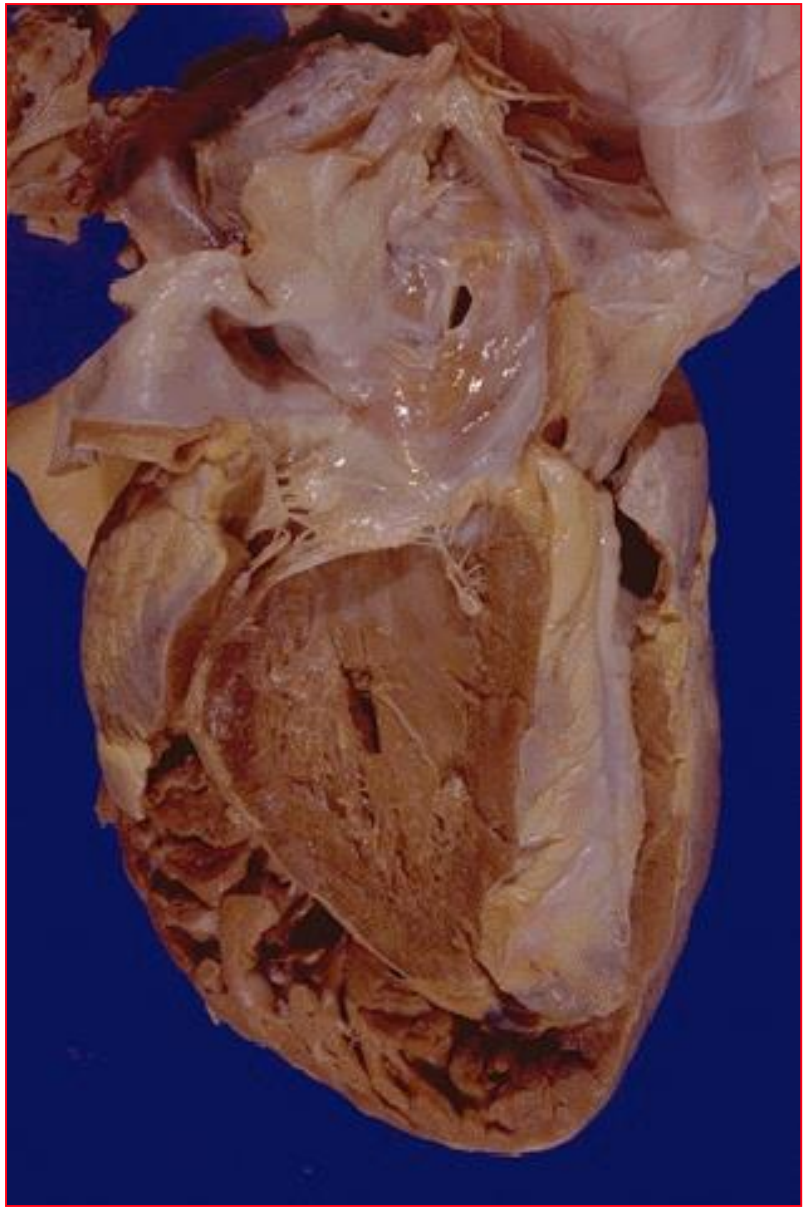


Type IV: muscular VSDs (trabecular)

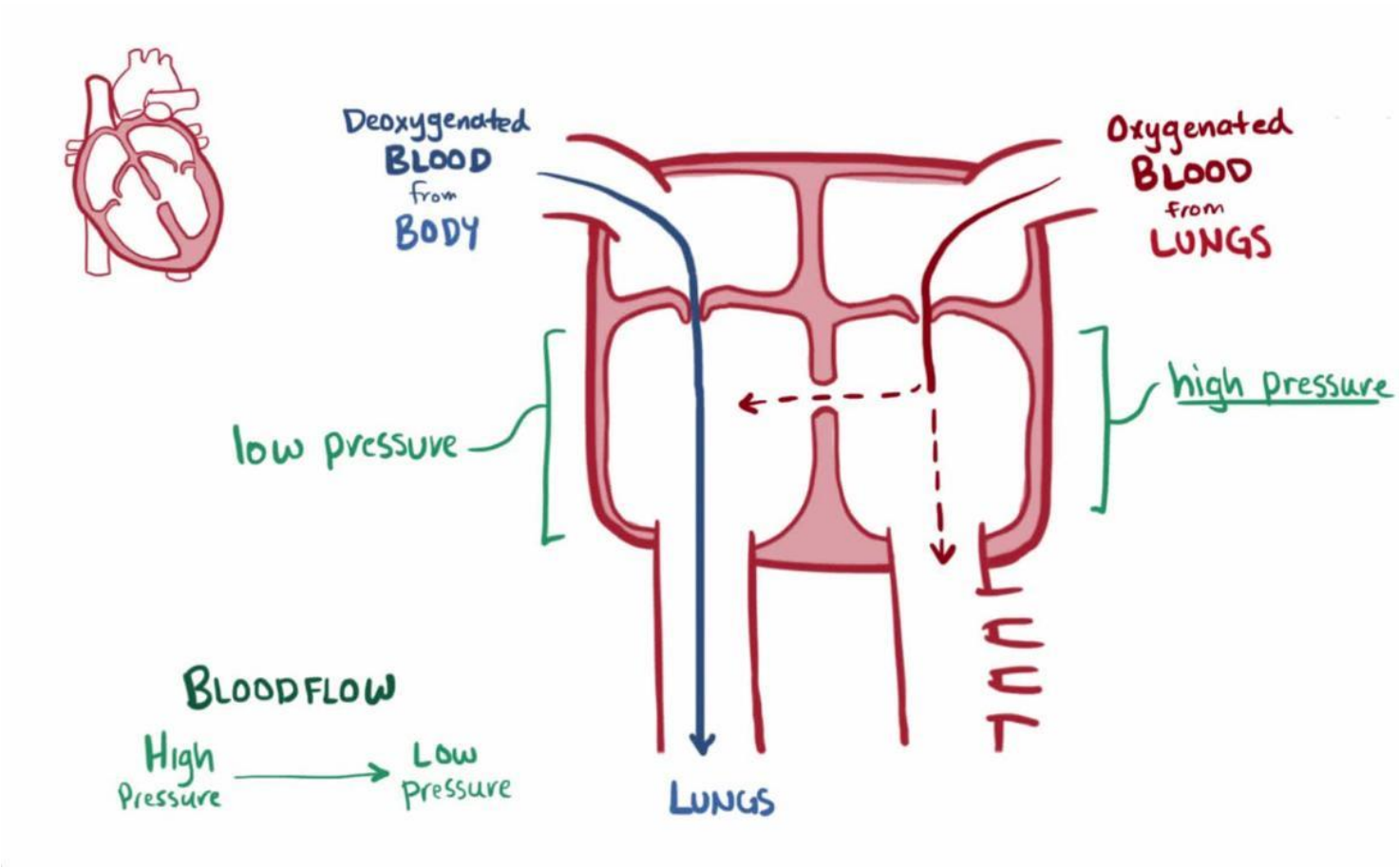
- Muscle tissue all around the defect
- These VSDs account for 5-20% of all defects.
- **May be either**
  - **anterior, in the inlet septum, mid-muscular or apical**



Muscular VSD



CLINICAL FINDINGS



- Precordium is hyperkinetic with a systolic thrill at LSB
- S1&S2 are masked by a **PSM** **at Lt.sternal border**
- **Max. intensity** of the murmur is best heard at **3rd,4th&5th Lt interspace**.
- **Delayed diastolic murmur at the apex &S3**

Pathophysiology and clinical manifestation

- **After birth ,** PVR falls (2-4 weeks until 8 weeks) ------ Large flow across shunt if large VSD Causes increased PA pressure (initially flow related), increased PV return, hence LA enlargement and LV overload
- **PH initially flow related and reversible**

Pathophysiology and clinical manifestation

- **Later**, ----- Intimal proliferation and medial hypertrophy leads to fixed irreversible PH
- Flow through the lungs decreases as PVR increases, hence shunt volume decreases
- Eventually PVR > SVR, hence R to L shunt across VSD ,Cyanosis, Eisenmenger

Pathophysiology and clinical manifestation

- A small restrictive VSD
  1. **75% spontaneously close < 2yrs**
  2. **Rarly produce PH**
  3. **Patient is asymptomatic.**
  4. **Murmur can be present since a few days after birth.**

A moderately restrictive VSD

1. **Variable increased PVR in less than 2 years**
2. **Frequent** respiratory tract infections. CHF **(rare)**. Cyanosis is **absent**
3. **Functional aerobic capacity is usually moderately reduced**

- A large or non-restrictive VSD
  1. **Rarely** close spontaneously
  2. Produce PH in less **than 2 years**
  3. **More frequent** respiratory tract infections. **Defective growth**. **Moderate cyanosis at times with exertion** Congestive heart failure **frequent in the first years of life**
  4. **Functional capacity markedly reduced**

Pathophysiology and clinical manifistation

- An **Eisenmenger** VSD
  1. **Net right-to-left shunt.**
  2. Infants with Eisenmenger may become
     - easily fatigued, especially during crying spells and at feeding time
     - Low tolerance for extra exertion
     - Shortness of Breath (dyspnea) and/or rapid breathing
     - Fainting (syncope)
     - Difficulty eating, breathing or sucking ,Poor weight gain, Slow growth or other physical retardation

Natural History

Spontaneous Closure

- **Spontaneous** closure is known, primarily with **perimembranous and muscular VSDs** with **restrictive VSD**
- (**Subarterial, doubly committed)** VSDS **and inlet** VSDs are **rarely** close

Progressive AR

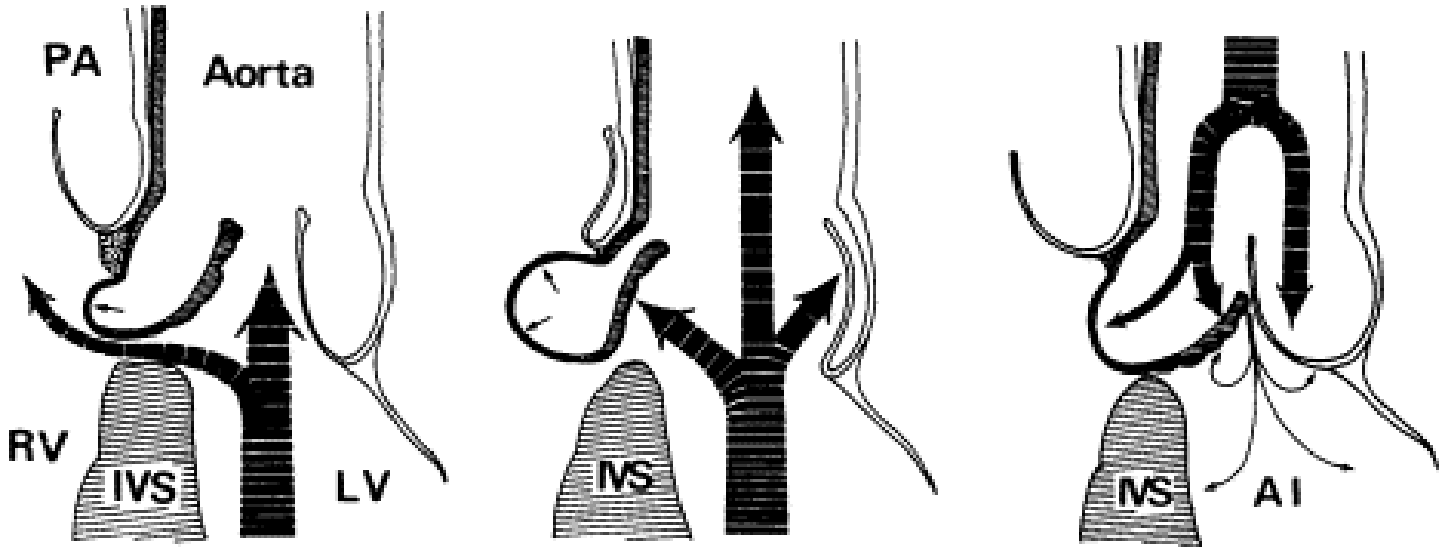
A perimembranous VSDs and (Subarterial, doubly committed) VSDs

- **(vuenturi effect)**

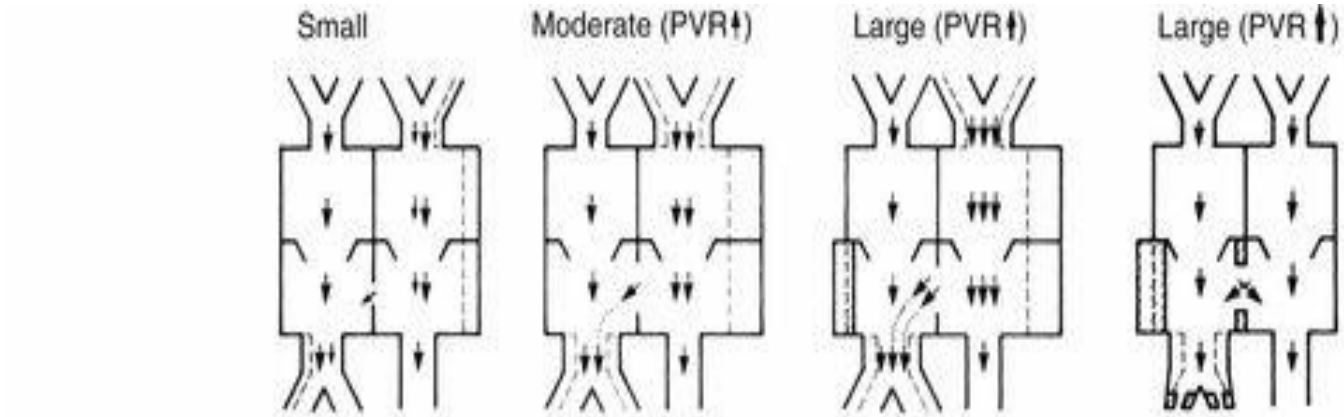
Gerbode shunt

(some P.M VSDs) - Left ventricular to right atrial shunt

VUENTURI EFFECT



ECG

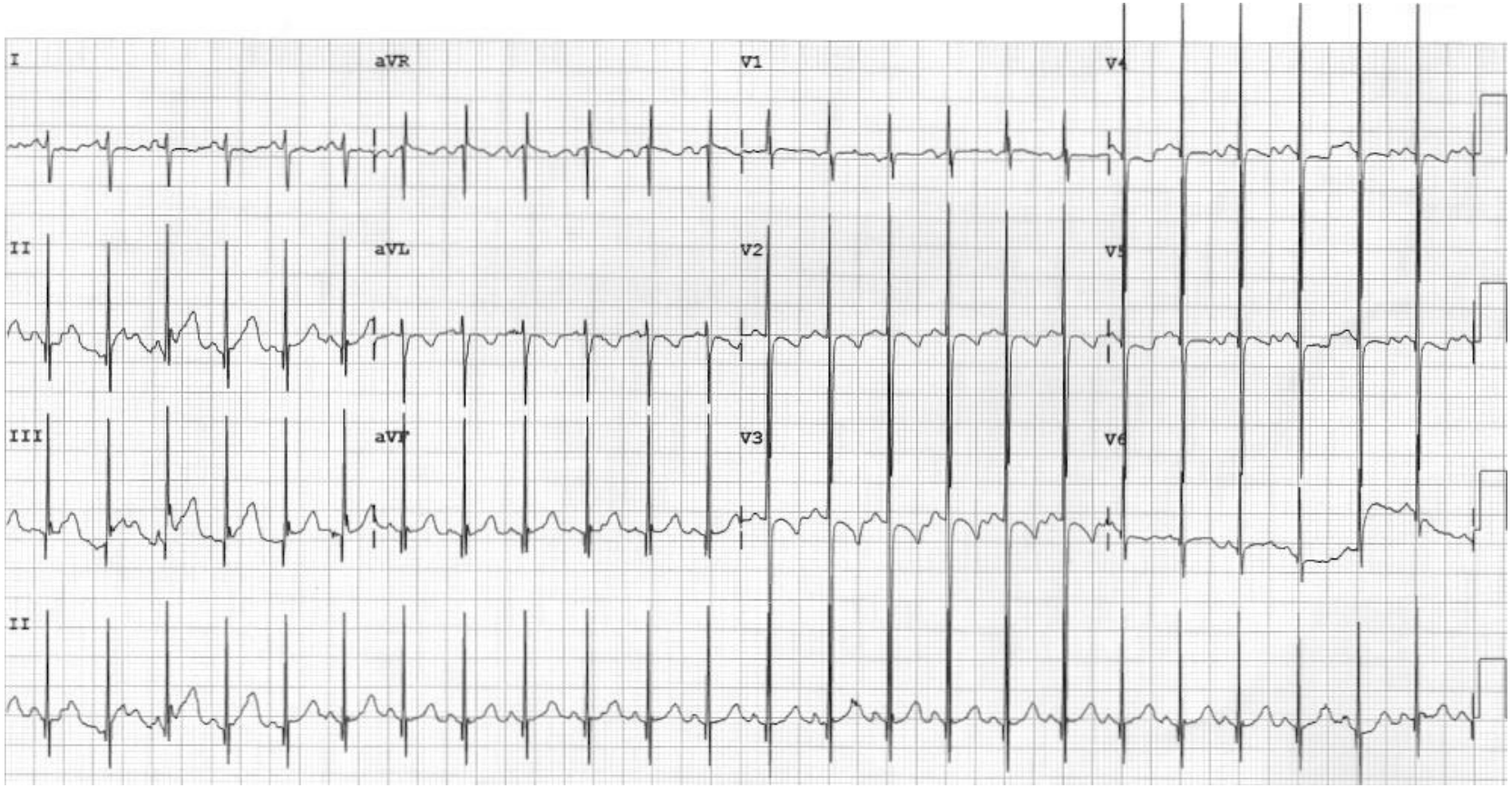


Cardiomegaly on x-rays	No or minimal	Moderate	Marked	No
RVP (%LVP)	25%–30%	30%–50%	60%–80%	100%
ECG	Normal	LVH LAH (±)	LVH RVH LAH	Pure RVH

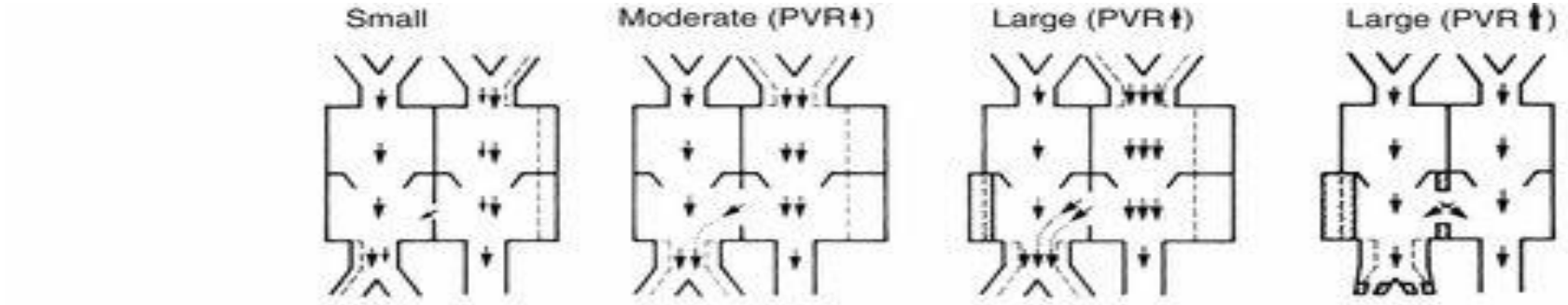
- Small restrictive VSDs…. **Normal tracing**
- Medium-sized VSDs
  - Broad, notched P wave characteristic of **left atrial overload**
  - Signs of **LV volume overload** — deep Q and tall R waves with tall T waves in leads V5 and V6
- With further progression, the ECG shows **biventricular hypertrophy; P waves may be notched or peaked.**

KATZ WATCHTEL SIGN

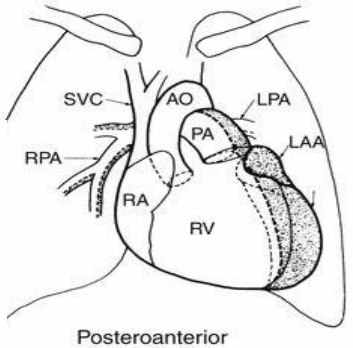
- The *Katz Wachtel phenomenon* — large biphasic QRS complexes in V2-4. This is the classic ECG pattern of **BVH**, most commonly seen in children with large ventriculo-septal defect (VSD).



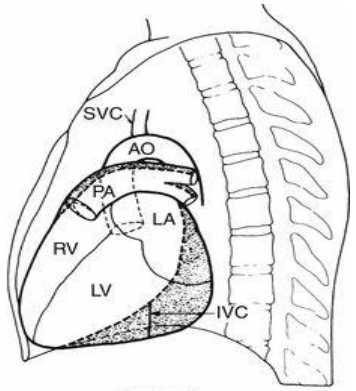
CHEST X-RAY



Cardiomegaly on x-rays	No or minimal	Moderate	Marked	No
RVP (%LVP)	25%–30%	30%–50%	60%–80%	100%
ECG	Normal	LVH LAH (±)	LVH RVH LAH	Pure RVH



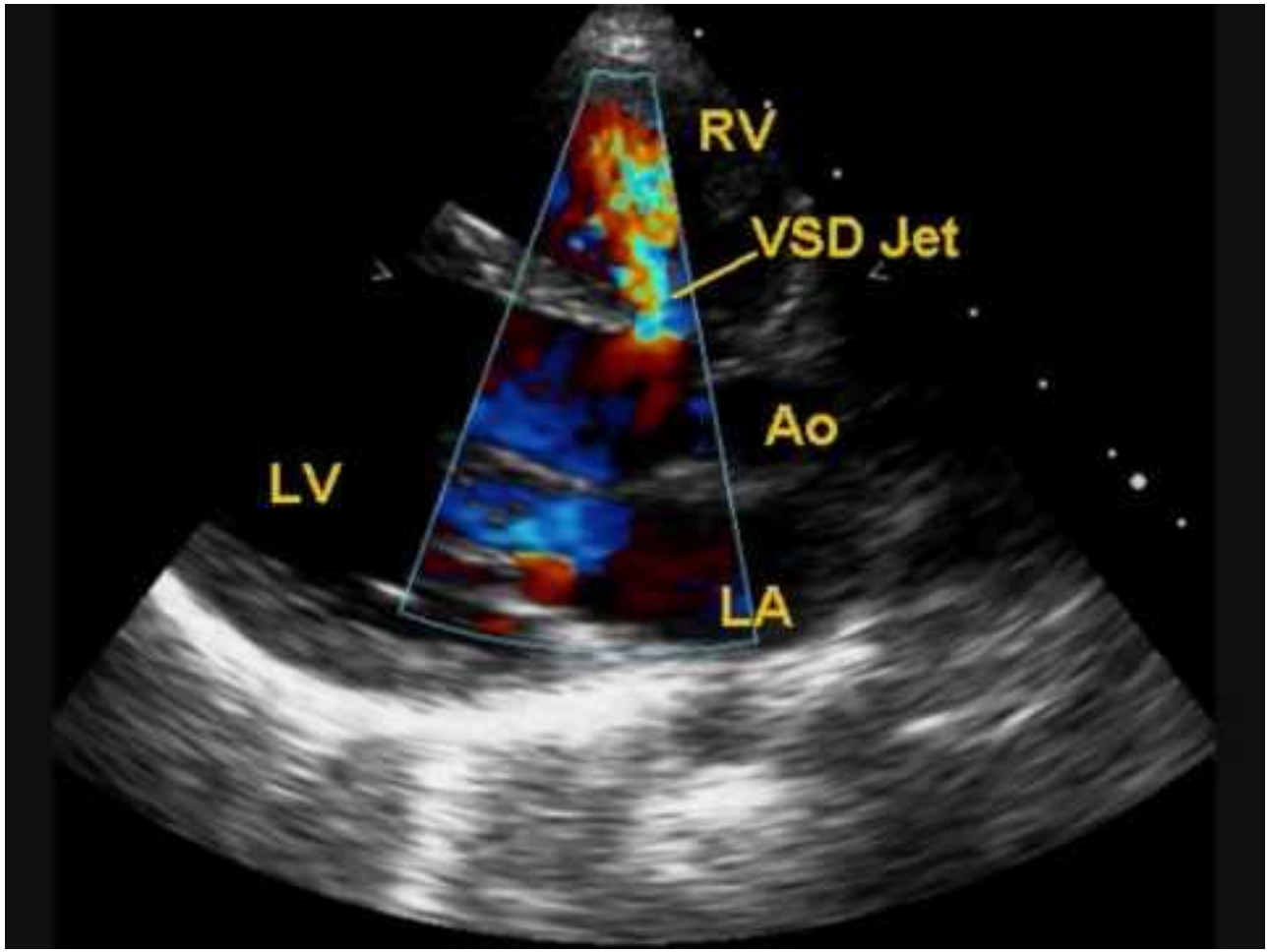
Posteroanterior

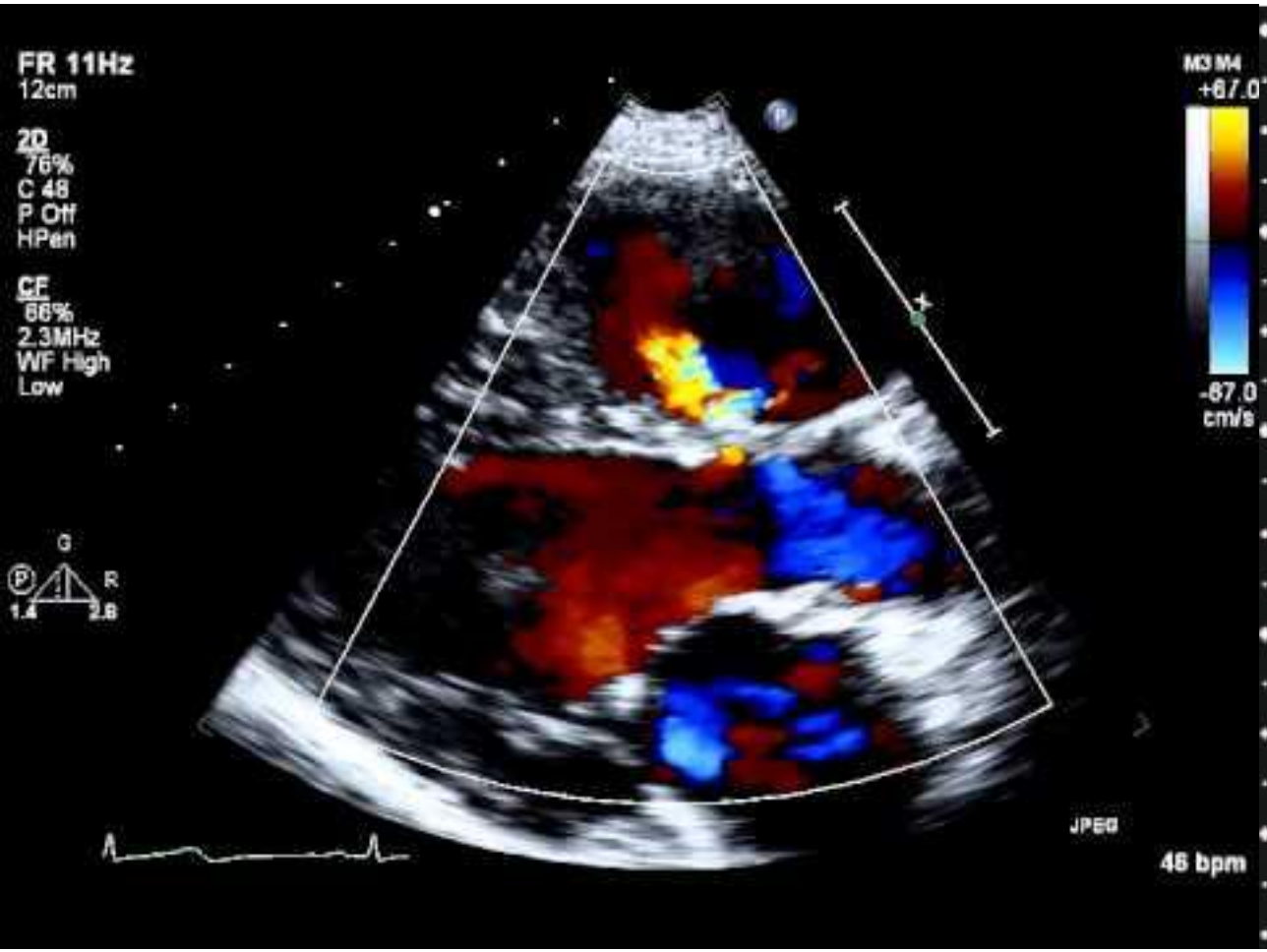


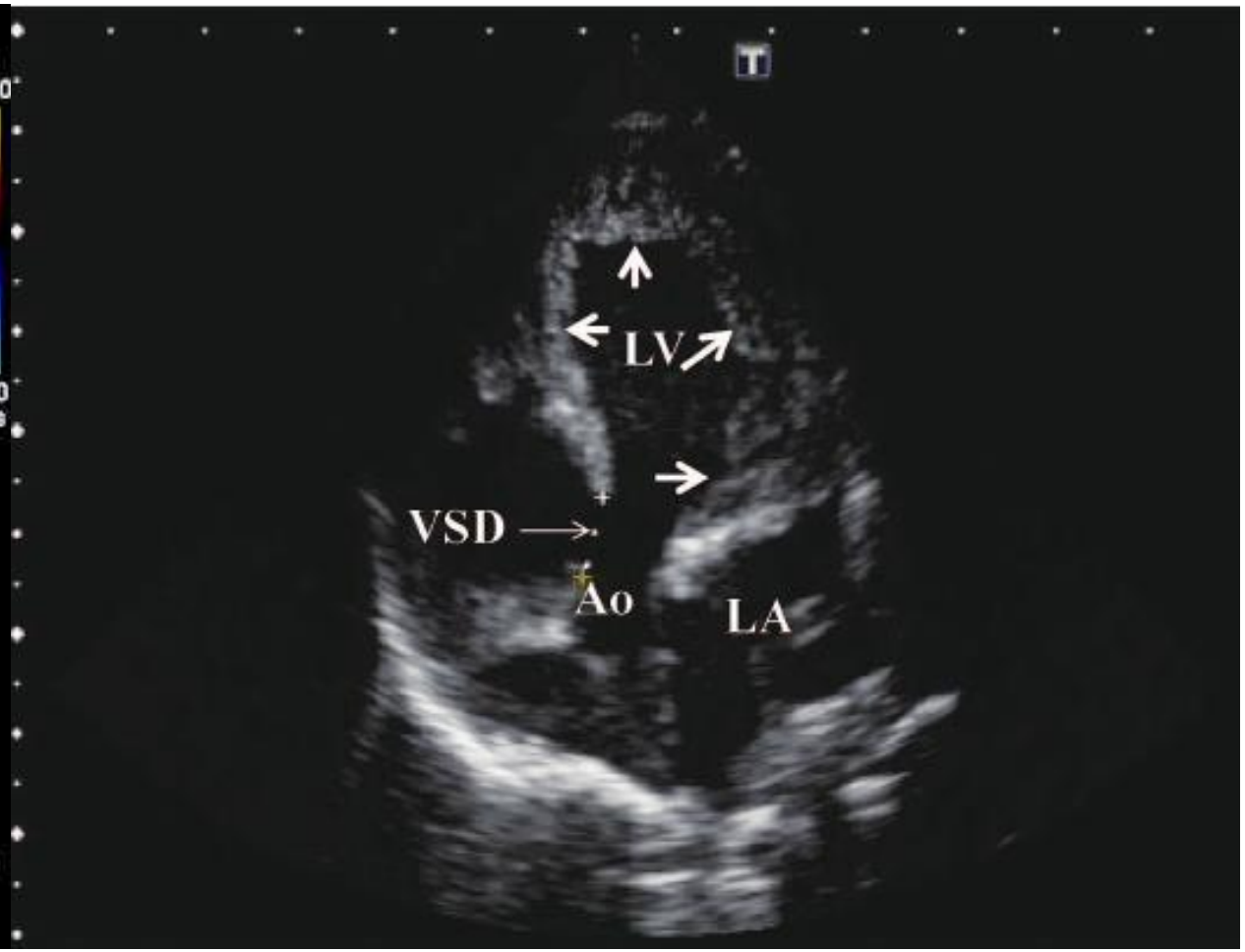
Lateral

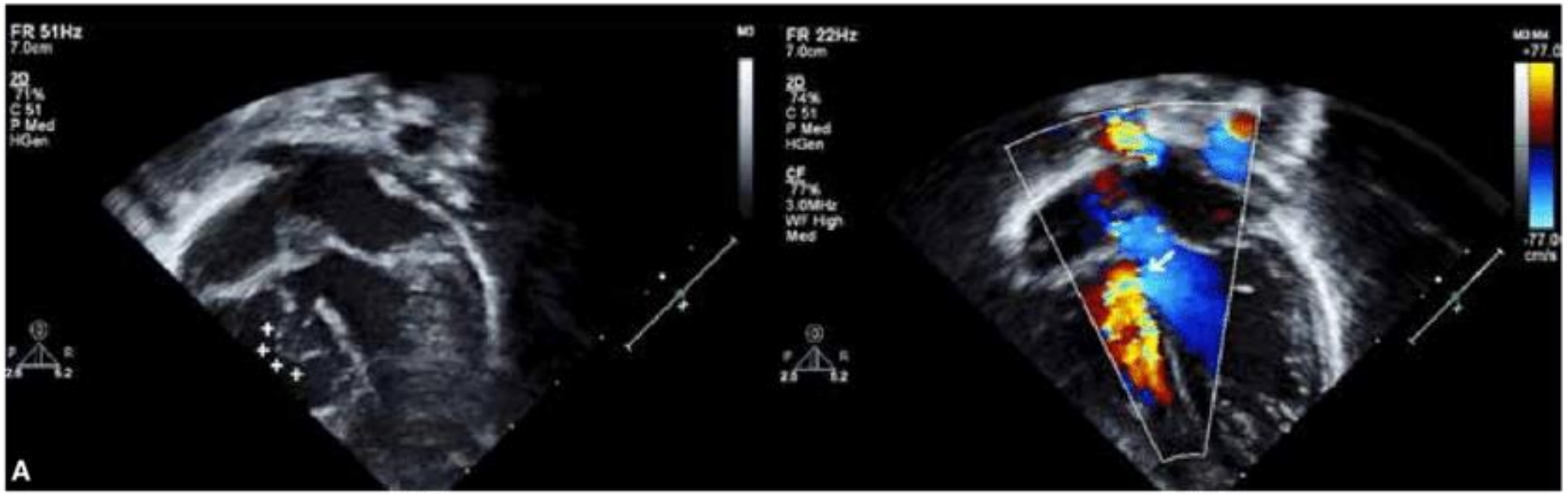
- Cardiomegaly : proportional to the volume overload. Mainly LV, LA and RV enlargement.
- Increased pulmonary blood flow.
- RV may not be as enlarged as anticipated as it receives the shunt into its outflow tract.

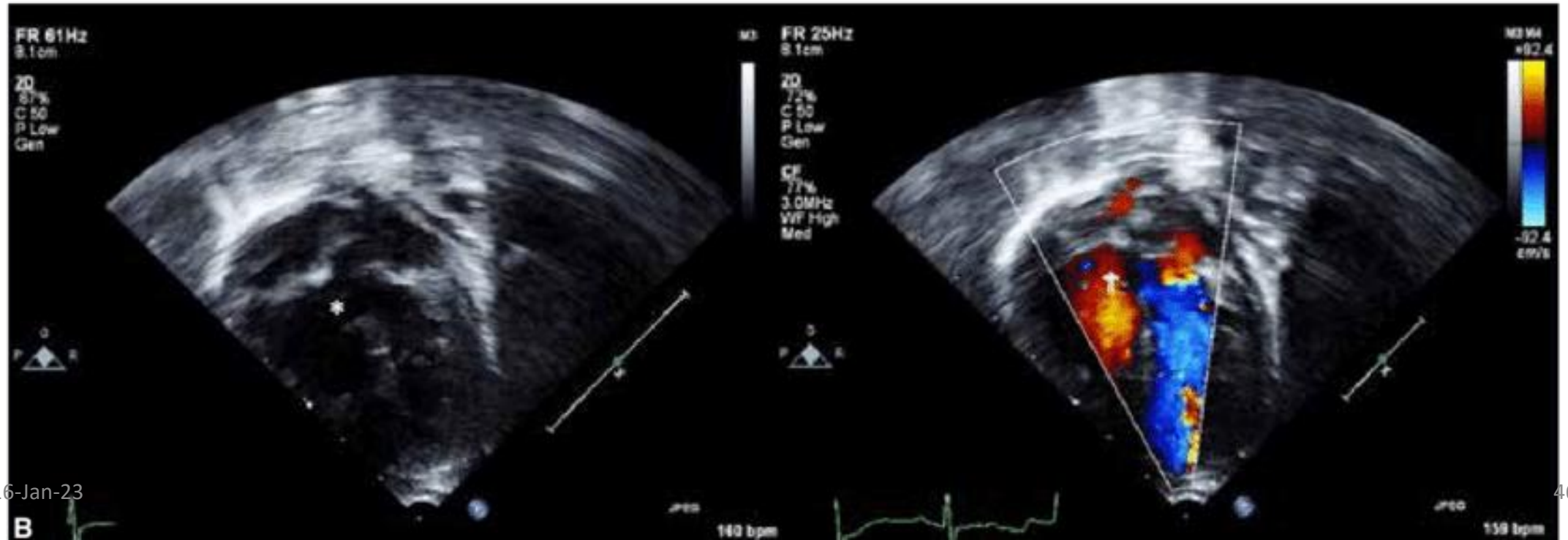
Doppler Echo



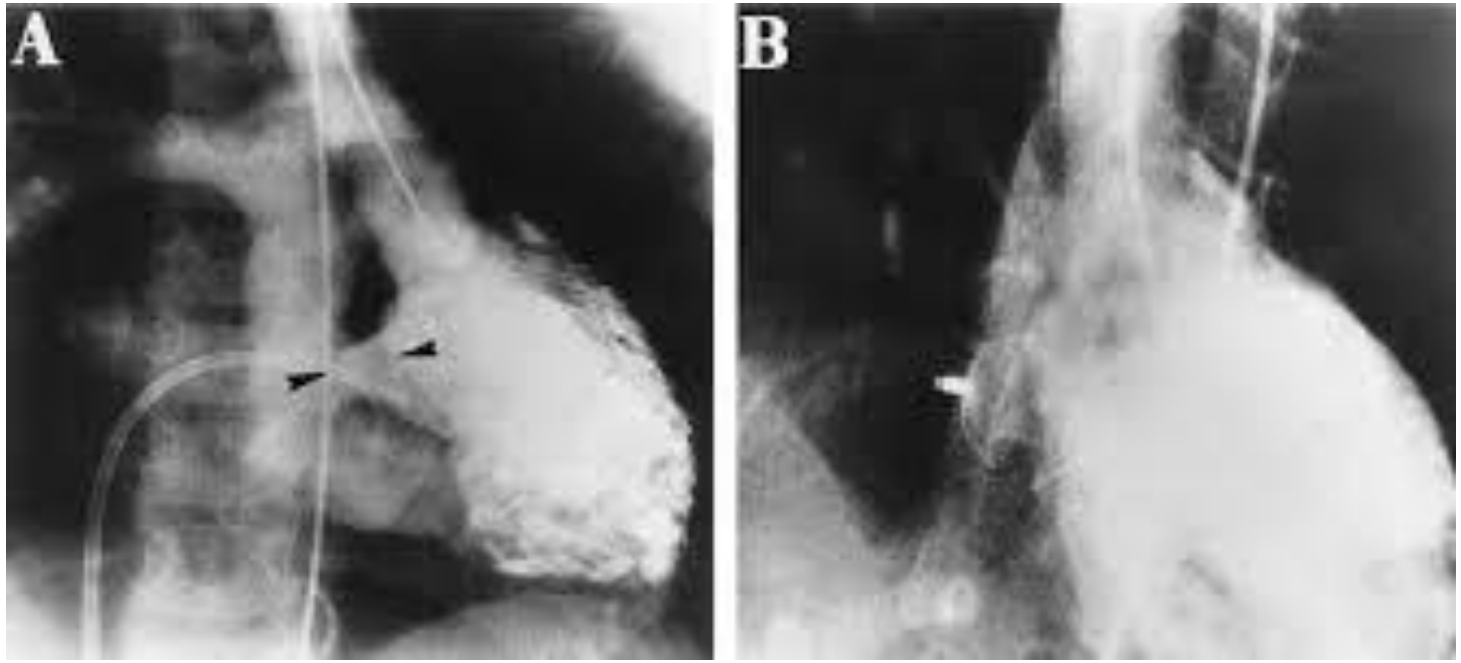

FR 11Hz
12cm
2D
76%
C 48
P Off
HPen
CF
66%
2.3MHz
WF High
Low
M3 M4
+67.0
-67.0
cm/s
G
R
1.4
2.8
JPEG
46 bpm


LV
VSD
Ao
LA


FR 51Hz
7.0cm
2D
71%
C 51
P Med
HGen
FR 22Hz
7.0cm
2D
74%
C 51
P Med
HGen
CF
77%
3.0MHz
WF High
Med
A

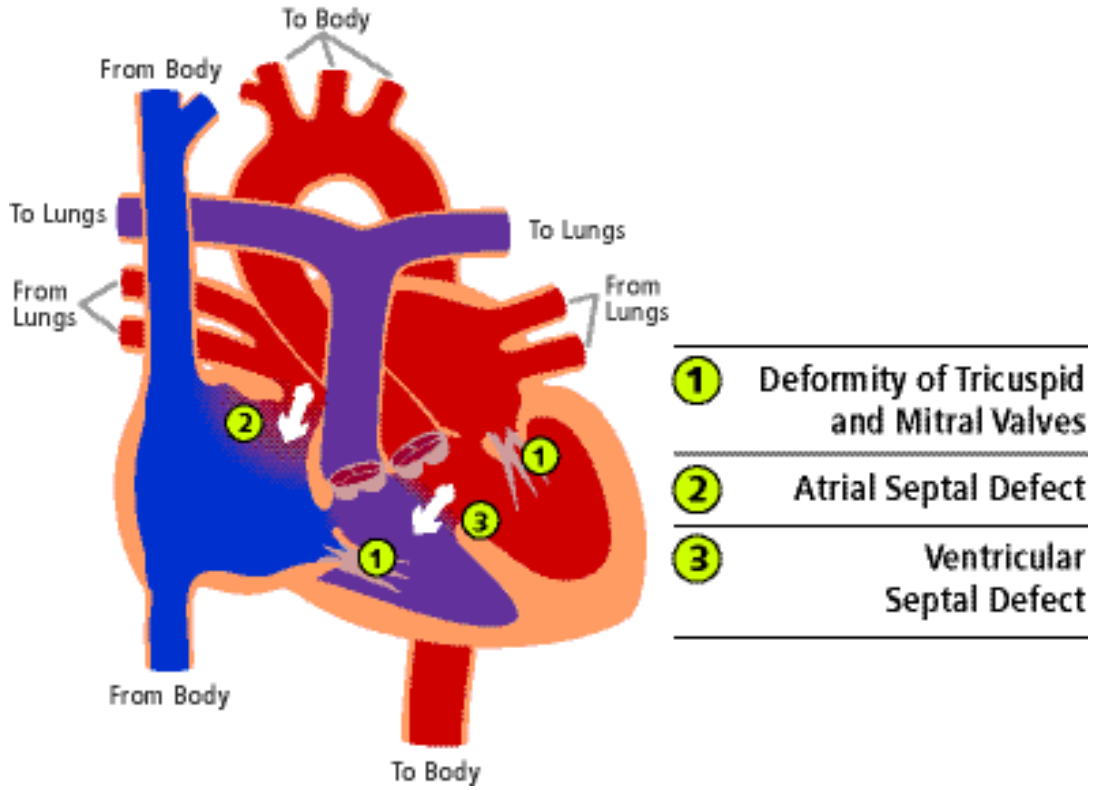

FR 61Hz
8.1cm
2D
87%
C 50
P Low
Gen
FR 25Hz
8.1cm
2D
72%
C 50
P Low
Gen
CF
77%
3.0MHz
WF High
Med
B
140 bpm
159 bpm

Cardiac catheterization



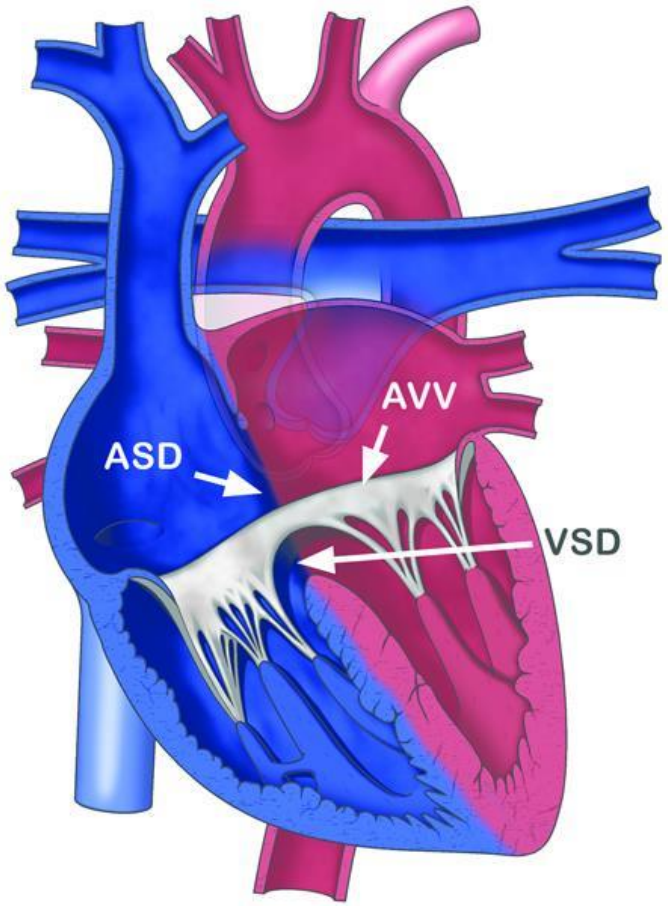
Complete Atrioventricular Septal Defect

- **AVSD** results from incomplete fusion the the endocardial cushions, which help to form the lower portion of the atrial septum, the membranous portion of the ventricular septum and the septal leaflets of the triscupid and mitral valves.
- **<u>Complete AVSD</u>**
  - Most common in Trisomy 21 (Down syndrome) patients



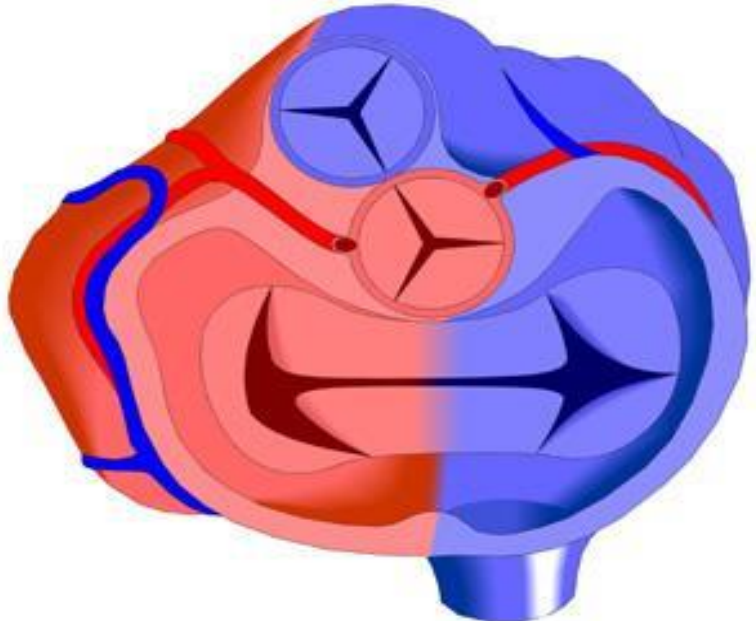
Abnormal development of the atrioventricular septum

- Involves the primum atrial septum and the inlet ventricular septum



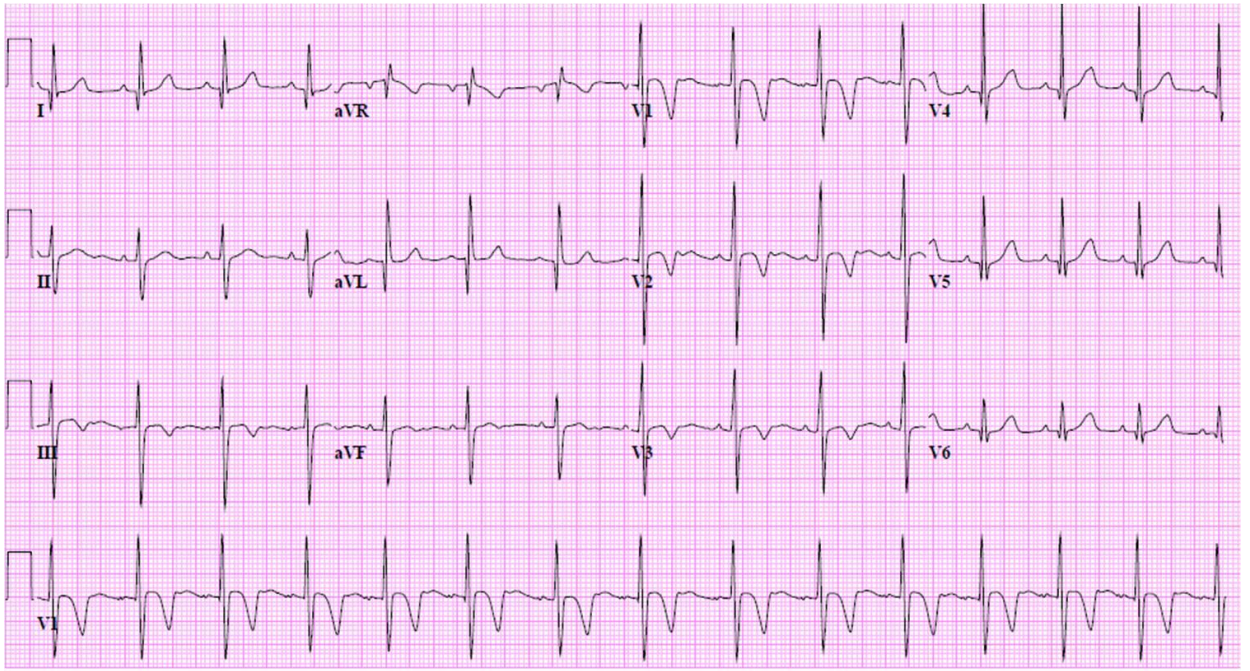
Abnormal development of the AV valves

- Involve septal portions of the mitral and tricuspid valve
- Lower attachment on AV septum creates large primum ASD
- Higher attachment on AV septum results in larger VSD
- Common AV valve
- No or abnormal septal attachments of the AV valves
- Single anterior and single posterior leaflets bridging the septal orifice



Posterior displacement of atrioventricular node

- Results in changes in electrocardiogram (In approximately 50% of patients)
  - **Prolonged PR interval**
  - **“Superior” QRS axis (left axis deviation)**
- Increases risk of surgically induced heart block

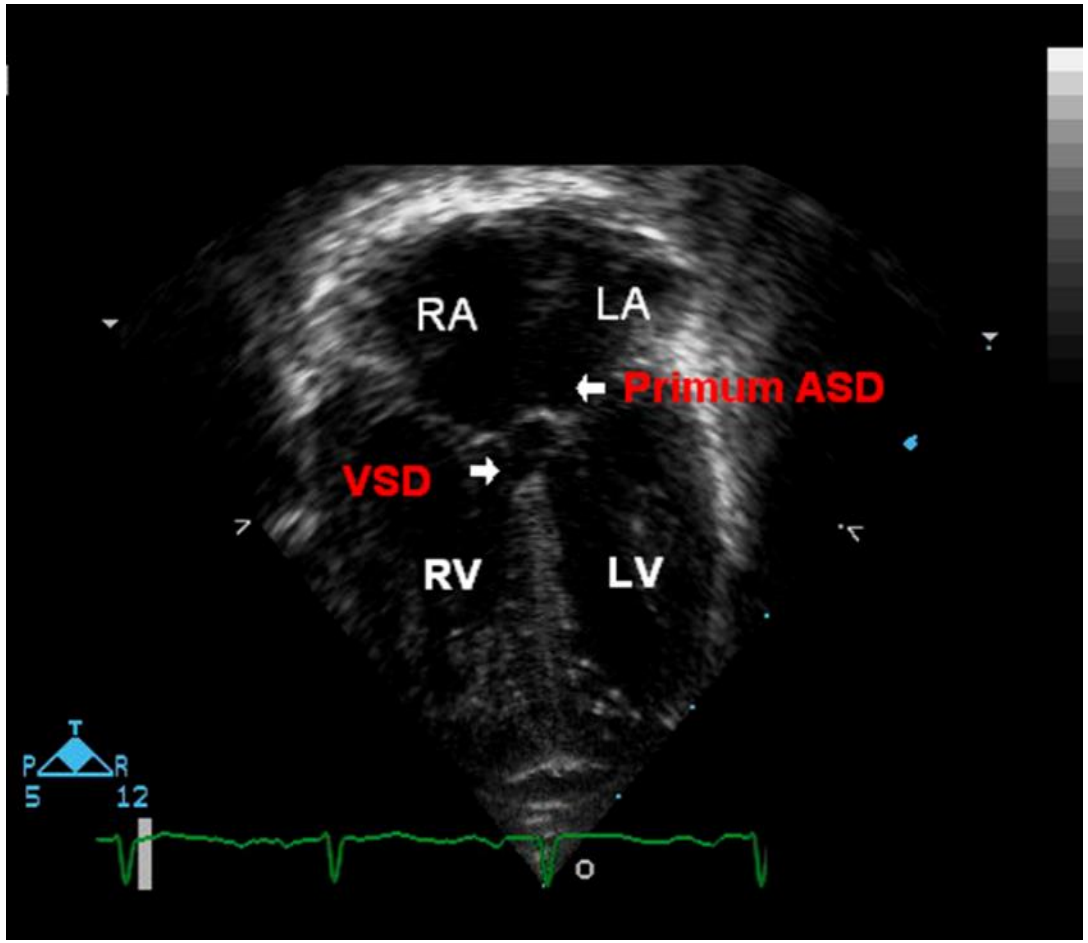


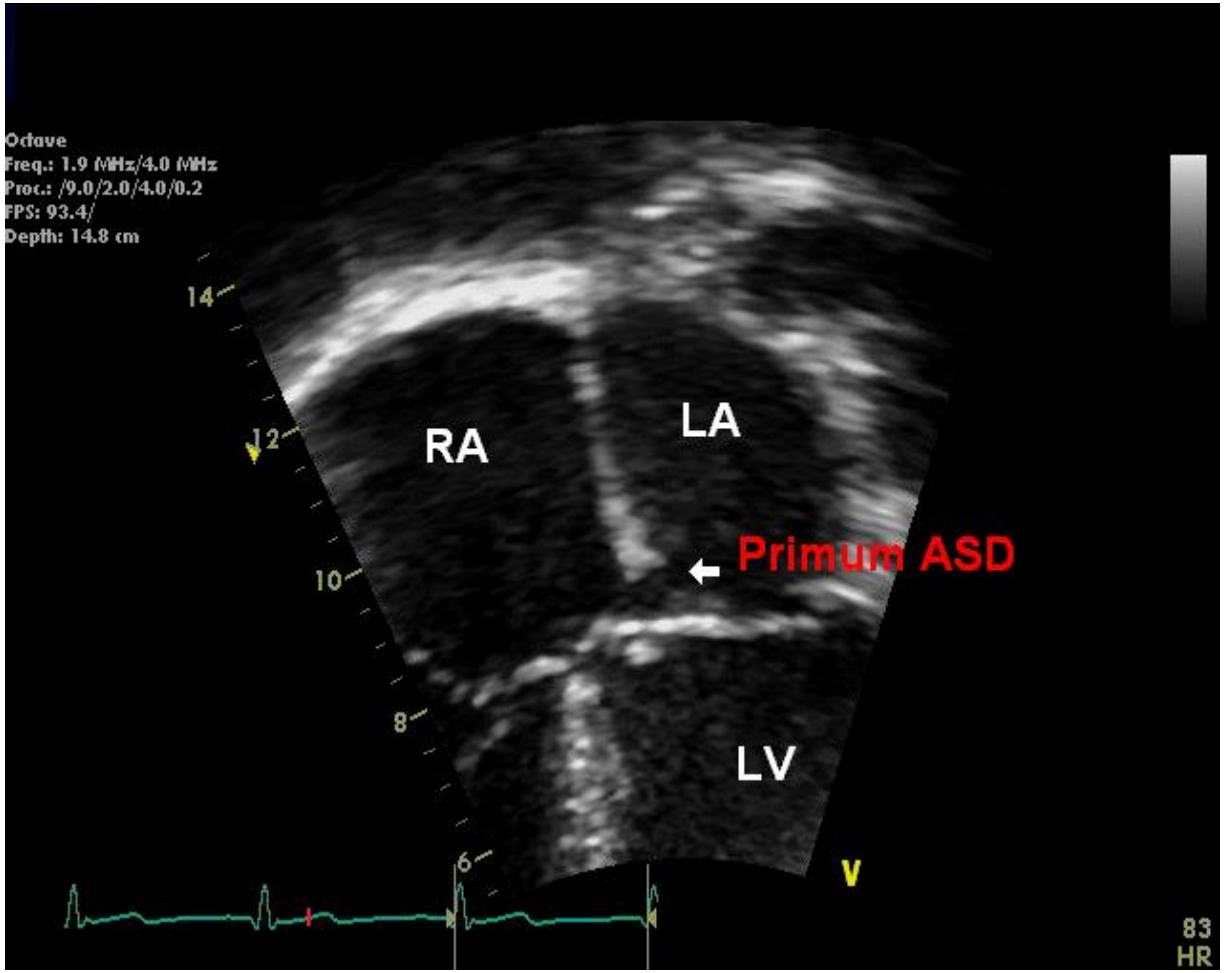
Clinical Presentation

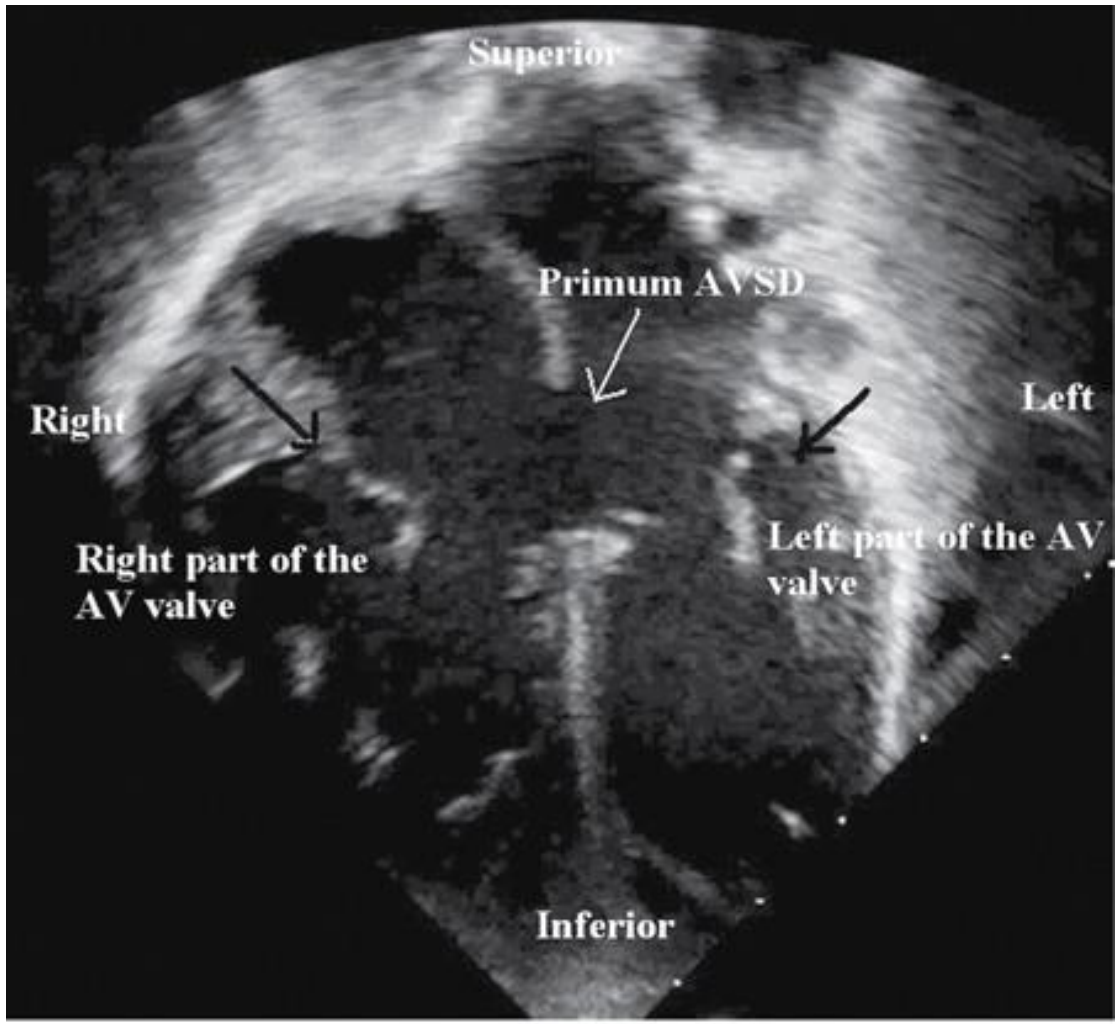
- **Congestive heart failure** in infancy.
- **Recurrent pulmonary infections.**
- **Failure to thrive.**
- **Late cyanosis** from pulmonary vascular disease w/ R to L shunt.

Clinical Signs:

- Hyperactive precordium with a systolic thrill at LLSB
- **Narrow split of S2**
- **P2 increased in intensity**
- **A grade 3-4/6 holosystolic murmur--- VSD**
- **Pulmonary systolic ejection murmur w/thrill—P. overflow**
- **Holosystolic murmur at apex w/radiation to axilla--MR**
- **Mid-diastolic rumbling murmur at Left Sternal border—Large L-R shunt**


RA
LA
Primum ASD
VSD
RV
LV
T
P
R
5
12


Octave
Freq.: 1.9 MHz/4.0 MHz
Proc.: /9.0/2.0/4.0/0.2
FPS: 93.4/
Depth: 14.8 cm
14
12
10
8
6
RA
LA
Primum ASD
LV
V
83
HR

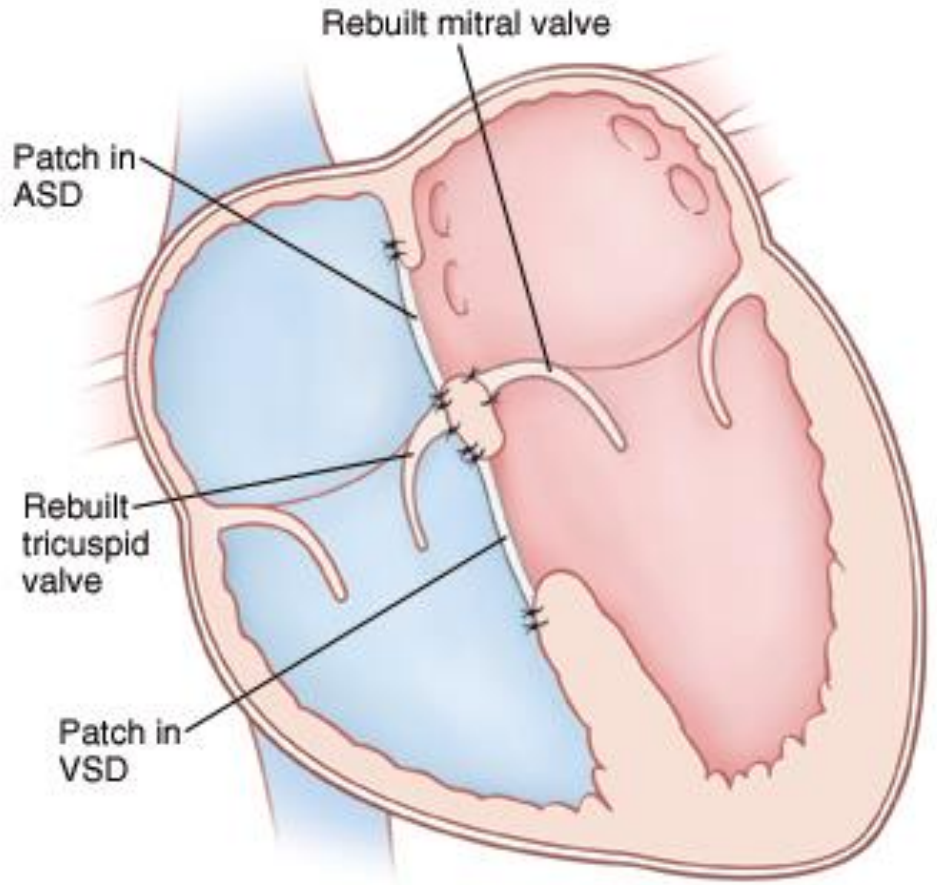


A

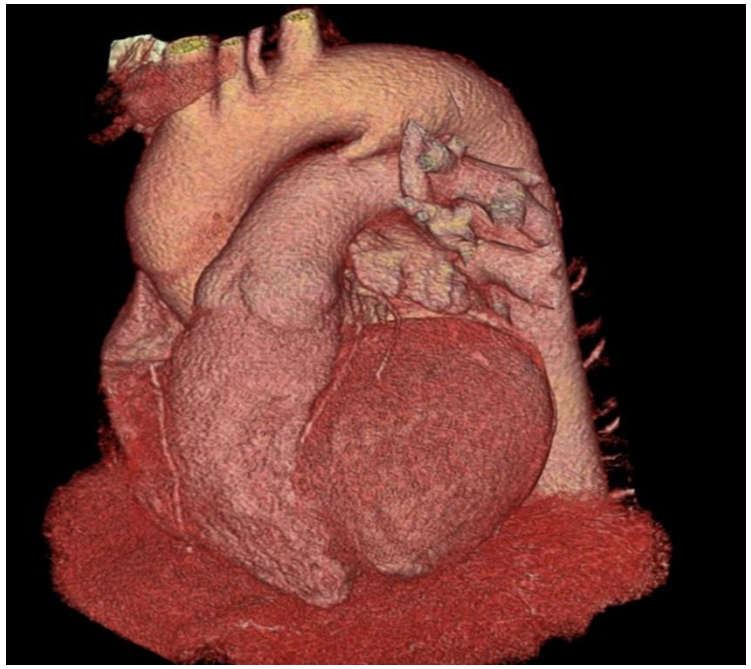
Source: Pahlm O, Wagner GS: *Multimodal Cardiovascular Imaging: Principles and Clinical Applications*: www.accessmedicine.com

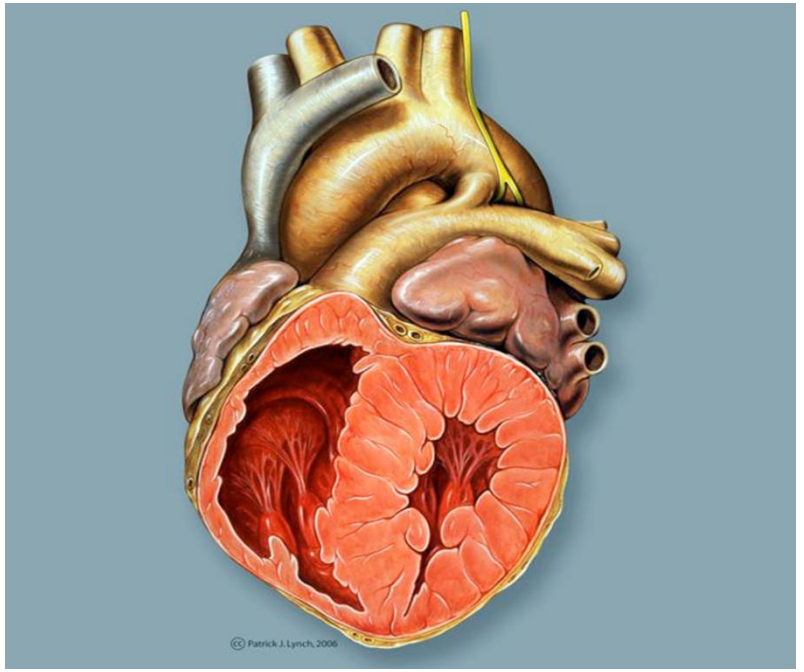
Treatment

- Surgery is always required.
- Treat congestive symptoms.
- Pulmonary banding maybe required in premature infants or infants < 5 kg.
- Correction is done **during infancy to avoid irreversible pulmonary vascular disease.**
- Mortality low w/incomplete 1-2% & as high as 5% with complete AVSD.



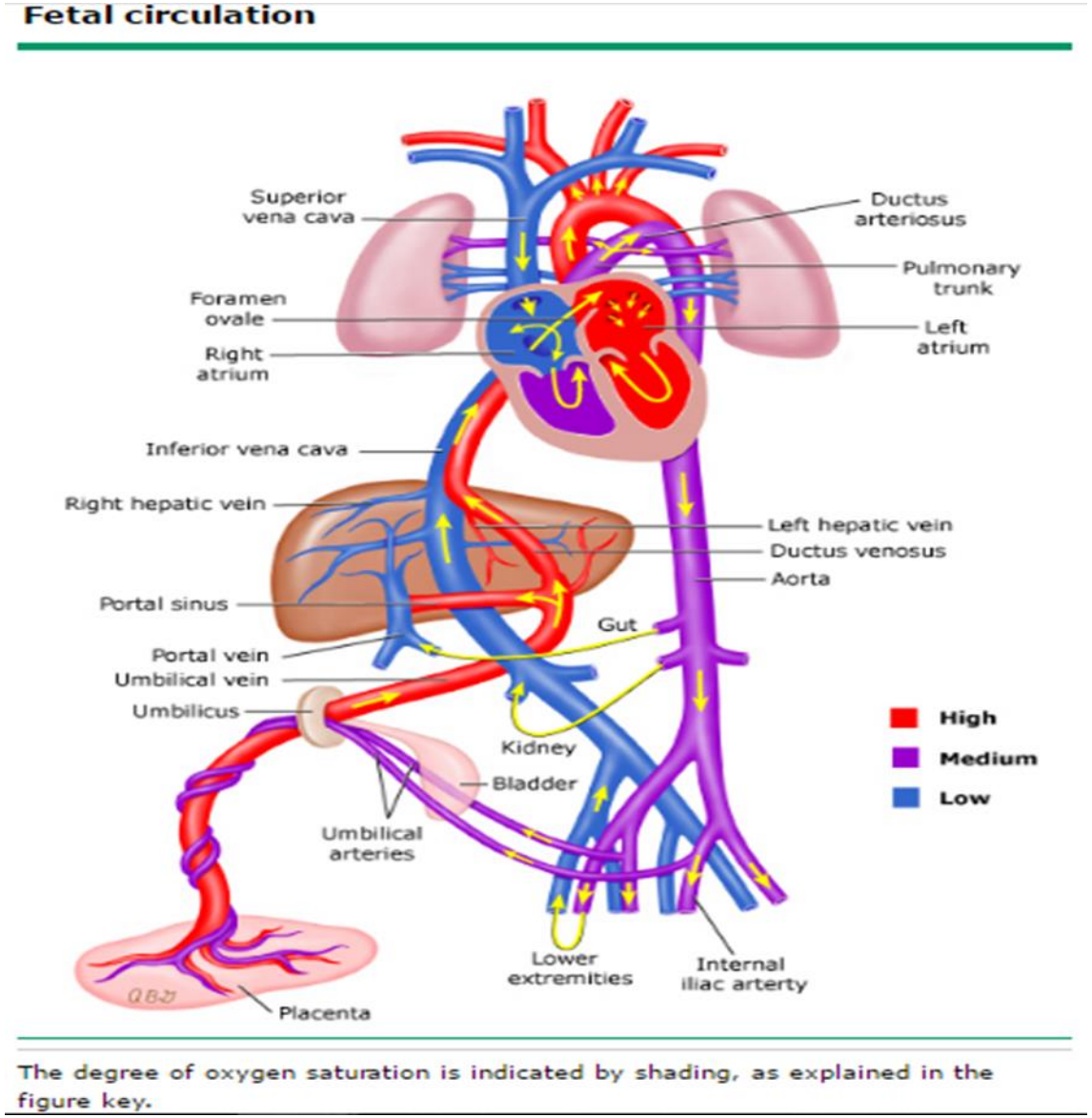
Patent Ductus Arteriosus





Anatomy

- During fetal life, the ductus arteriosus is a normal structure that allows most of the blood leaving the right ventricle to bypass the pulmonary circulation and pass into the descending aorta.
- Typically, **only about 10% of the right ventricular output passes through the pulmonary vascular bed.**
- The ductus arteriosus is a **remnant of the distal sixth aortic arch**
- **Connects the pulmonary artery at the junction of the main pulmonary artery and the origin of the left pulmonary artery to the proximal descending aorta just after the origin of the left subclavian artery.**

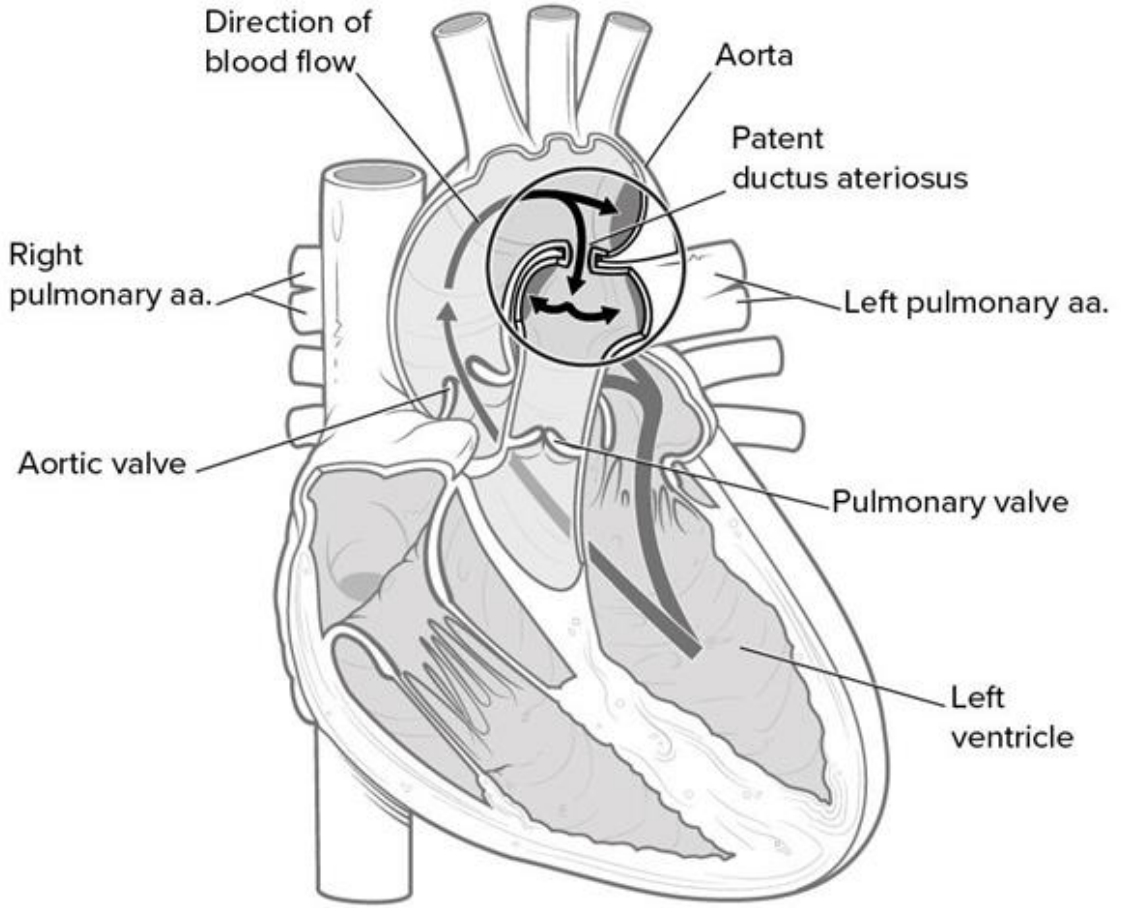


Functional closure

- In the fetus,
  - **the oxygen tension is relatively low**, because the pulmonary system is nonfunctional.
  - Coupled with **high levels of circulating prostaglandins** due to:
    - **The little amount of pulmonary circulation**
    - **The high levels of production in the placenta.**
- At birth,
  - **the placenta is removed**, eliminating a major source of prostaglandin production,
  - **the lungs expand**, activating the organ in which most prostaglandins are metabolized.
  - **oxygen tension in the blood markedly increases**. Pulmonary vascular resistance decreases with this activity.

- Normally, **functional closure** of the ductus arteriosus occurs by **abrupt contraction of the muscular wall by about 15 hours of life in healthy infants born at term.**
- A **second stage of closure(Anatomical) related to fibrous proliferation of the intima is complete in 2-3 weeks.**

- Communication between the pulmonary artery and the aorta
- Location – distal to left subclavian



Etiology ….**Prematurity**

- IN **preterm neonates** has been suggested to be due to
  1. **Poor prostaglandin metabolism because of immature lungs.**
  2. **high reactivity to prostaglandin**
  3. **reduced calcium sensitivity to oxygen in vascular smooth muscle cells due to immaturity of the smooth muscle within the structure**

Etiology ….Implicated teratogens

- **Congenital rubella infection,**
- **Fetal alcohol syndrome**
- **Maternal amphetamine use**
- **Maternal phenytoin use**

Etiology….**Genetics**

- **Familial cases** of patent ductus arteriosus (PDA) have been recorded, but a **genetic cause has not been determined**.

Clinical presentation

- ❑ **Irritable, feed poorly, fail to gain weight and sweat excessively**
- ❑ **Increased respiratory effort and respiratory rates**
- ❑ **prone to develop recurrent pneumonia**

Clinical Signs:

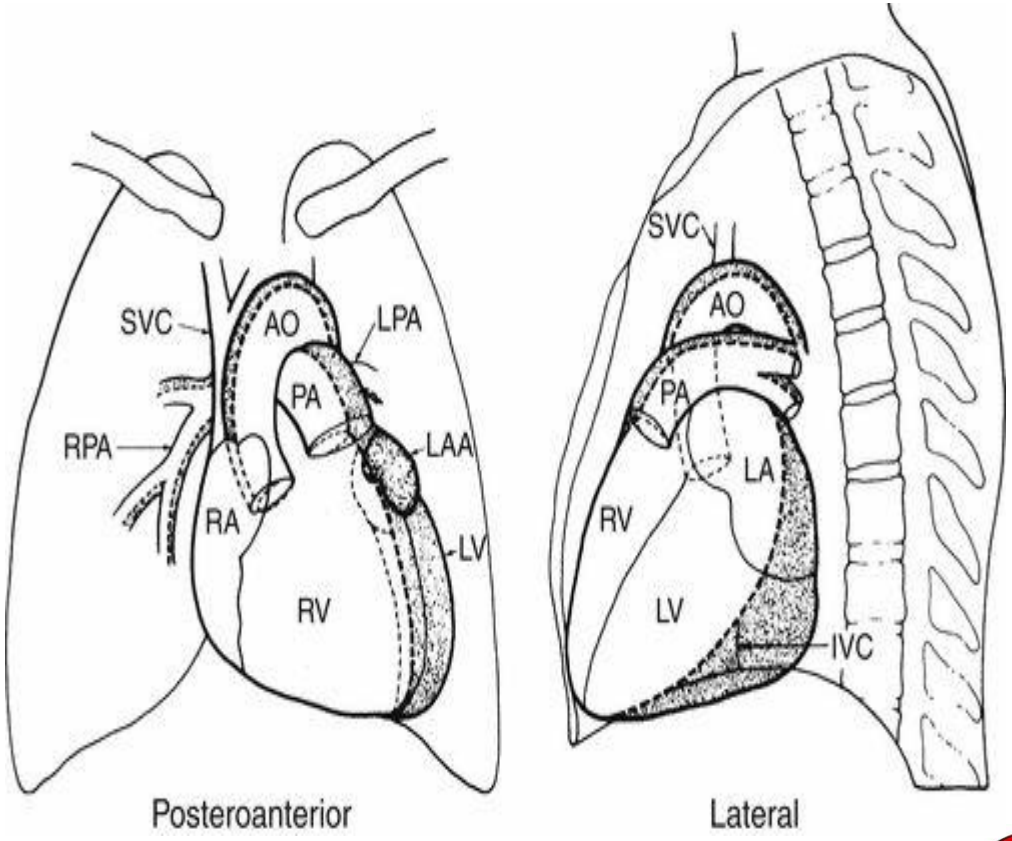
- **Murmur:** systolic at LUSB/Left Infraclavicular, may progress to **continuous (machinery**) murmur.
- **Mitral delayed diastolic murmur <u>related to</u>** increased flow through left atrium and ventricle.
- **Narrowly or paradoxically split S2_** (large shunts) **<u>??</u>**
- **<u>Wide pulse pressure</u>** – <u>bounding peripheral pulses??</u>
- **<u>Hyperkinetic apex releated to</u>** the large left ventricular stroke volume
- **The apical impulse is laterally displaced**
- **Dilatation of the ascending aorta , Aortic ejection click** – preceding the continuous murmur, **Aortic ejection systolic murmur** – drowned by the loud continuous murmur

ECG

- ✓Small PDA: nl ECG.
- ✓Large PDA: Left ventricular hypertrophy and Left atrial enlargement may be present
- ✓**In the presence of significant pulmonary hypertension**, there may be evidence of <u>right ventricular hypertrophy.</u>

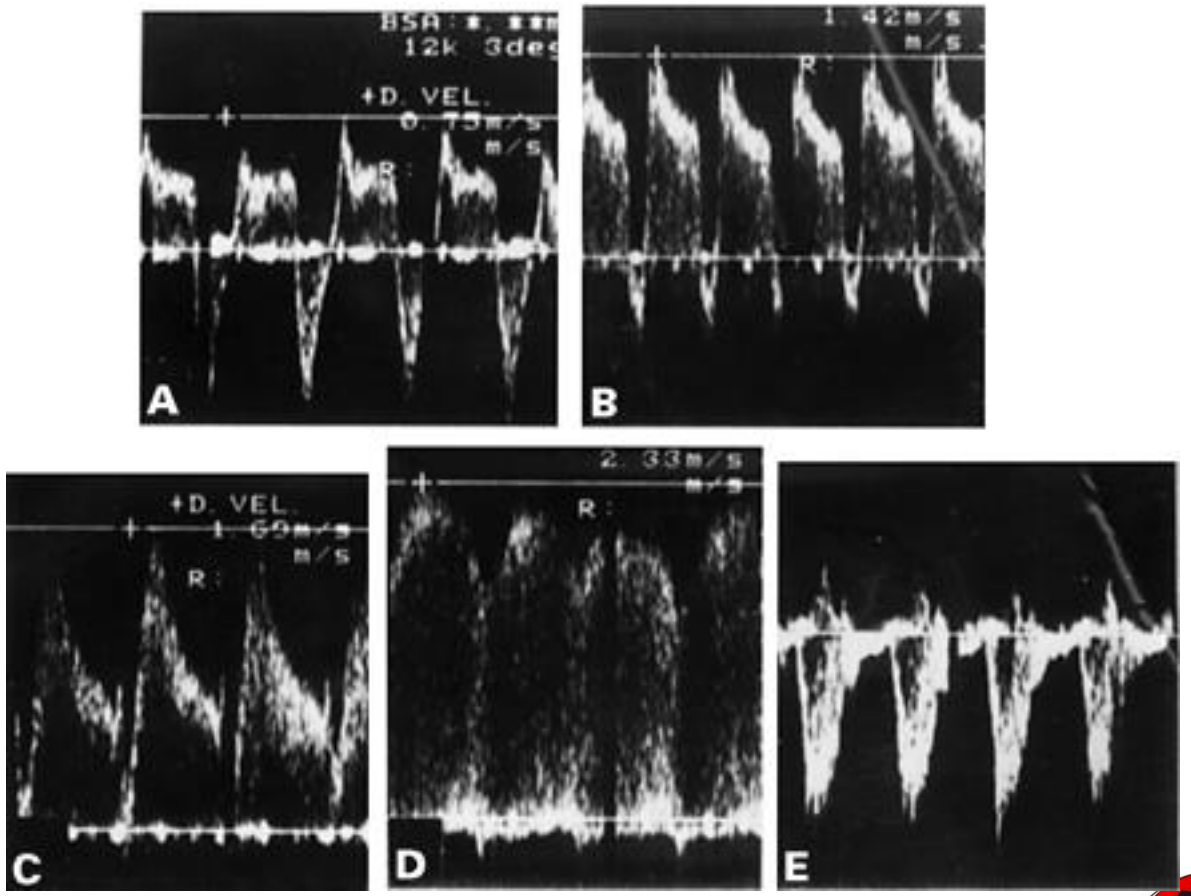
CXR

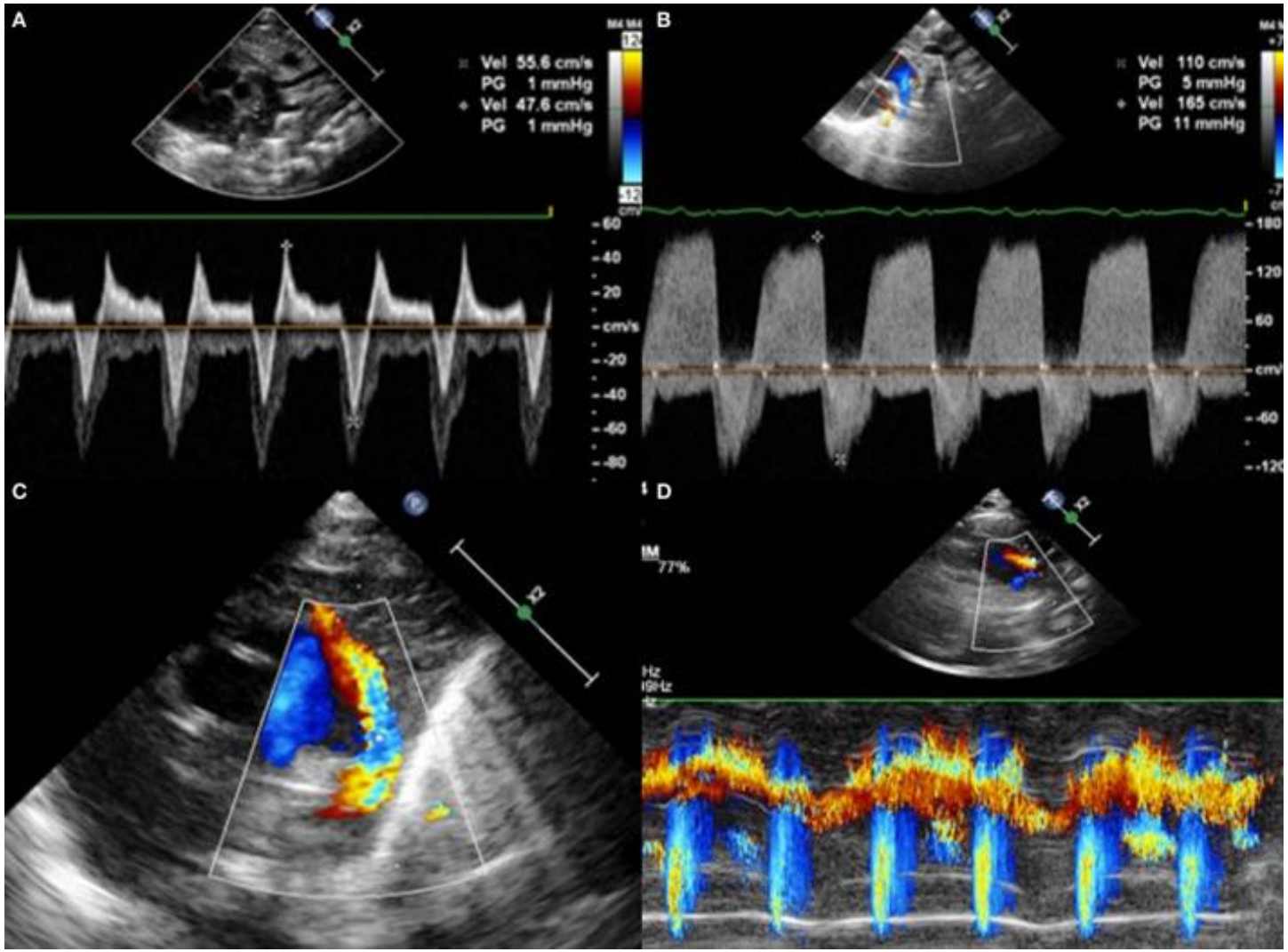
- Small PDA: NL ECG
- Large PDA With marked pulmonary over circulation, pulmonary edema occurs. Accentuated peripheral pulmonary vascular markings and increased pulmonary venous markings are noted.



Doppler Echocardiography

- Gold standard for diagnosing PDA




A
Vel 55.6 cm/s
PG 1 mmHg
Vel 47.6 cm/s
PG 1 mmHg
B
Vel 110 cm/s
PG 5 mmHg
Vel 165 cm/s
PG 11 mmHg
C
D

Complications: hemodynamic significant PDA

- Congestive Heart failure
- Premature n.b is at risk for:
  - **NIC**
  - **IVH**
  - **Renal impairment**
- Infective endarteritis
- Eisenmenger syndrome

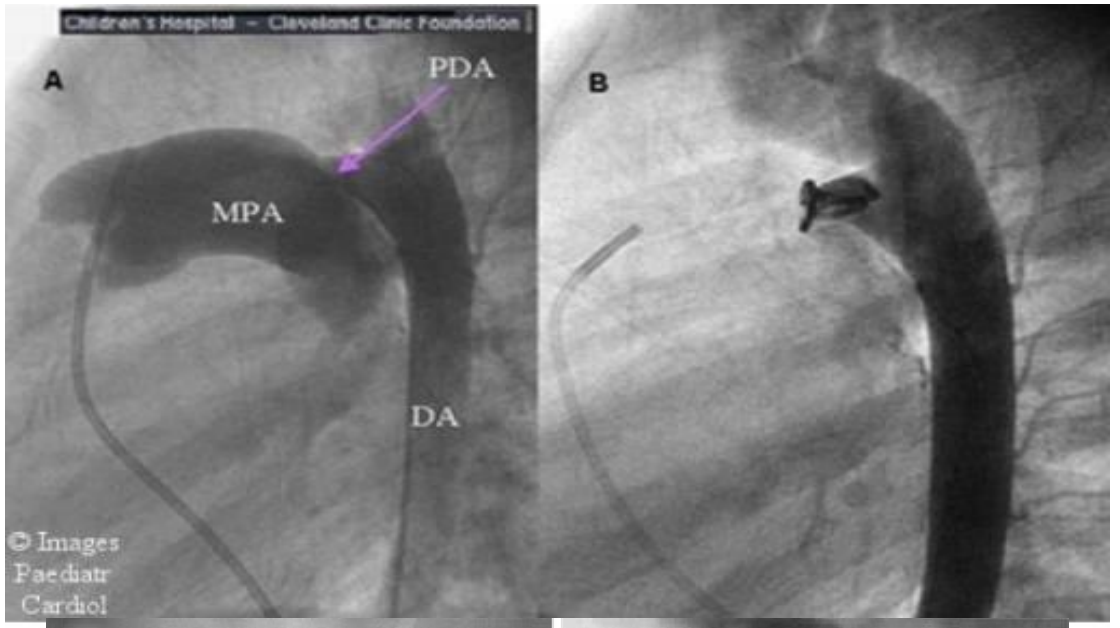
Treatment :

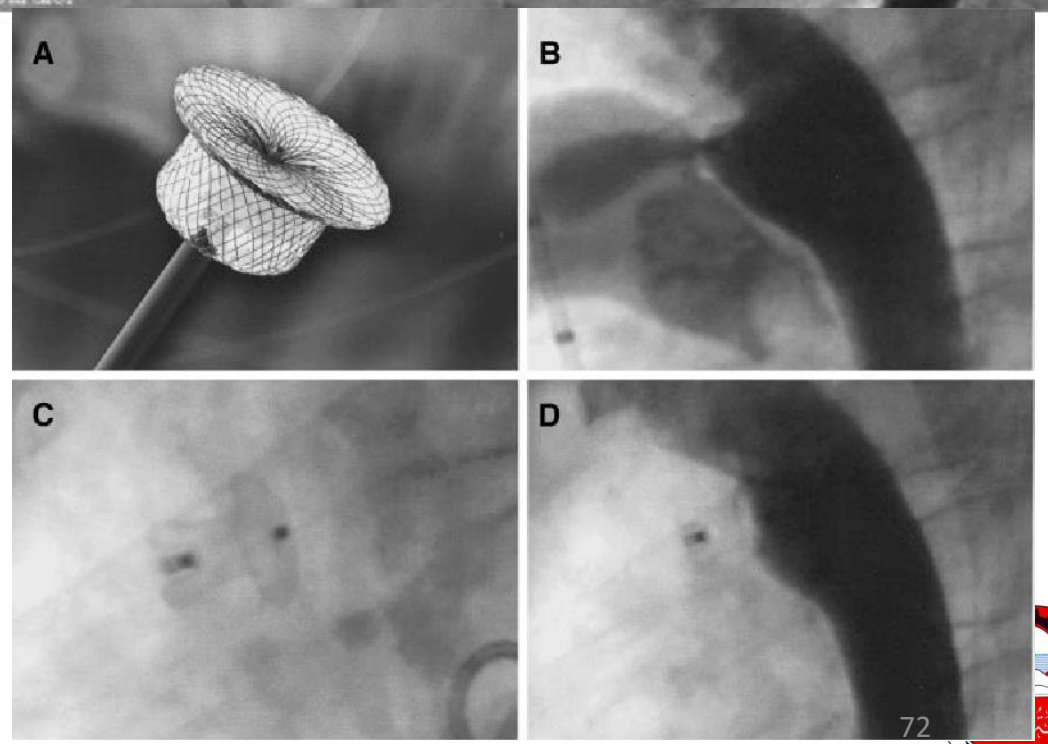
- **Fluid restriction and watchful waiting: In a premature N.B, a PDA often closes spontaneously**
- **If premature newborn is symptomatic and has a hemodynamic significant PDA after few days (3 days of life!) :**
    - **Fluid restriction**
    - **Mechanical ventilation?**
    - **Closure trial with medications(Pharmacologic Management) (mainly for preterm births)**
        - **Ibuprofen?**
        - **Paracetamol ?**
    - **Anti-failure medications (lasix ,digoxin)**

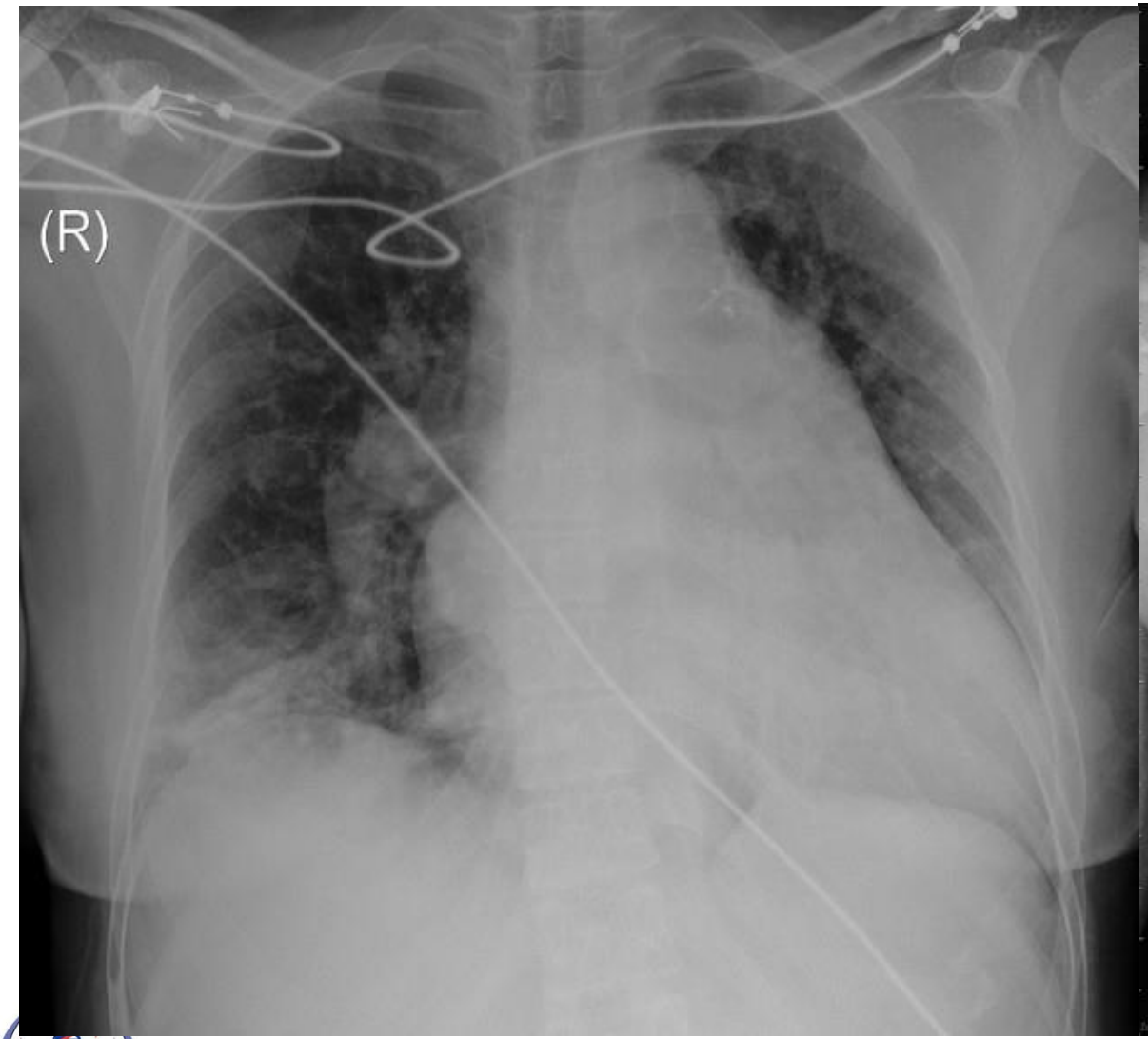
- **In full-term babies, children and adults who have small PDAs without any medical symptom or problem, follow up might be enough.**
    - **If non-significant asymptomatic PDA(mild L-R shunt) and silent (no murmur): needs f/u**
    - **If non-significant asymptomatic PDA(mild L-R shunt) and murmur: needs f/u, and better to be closed**

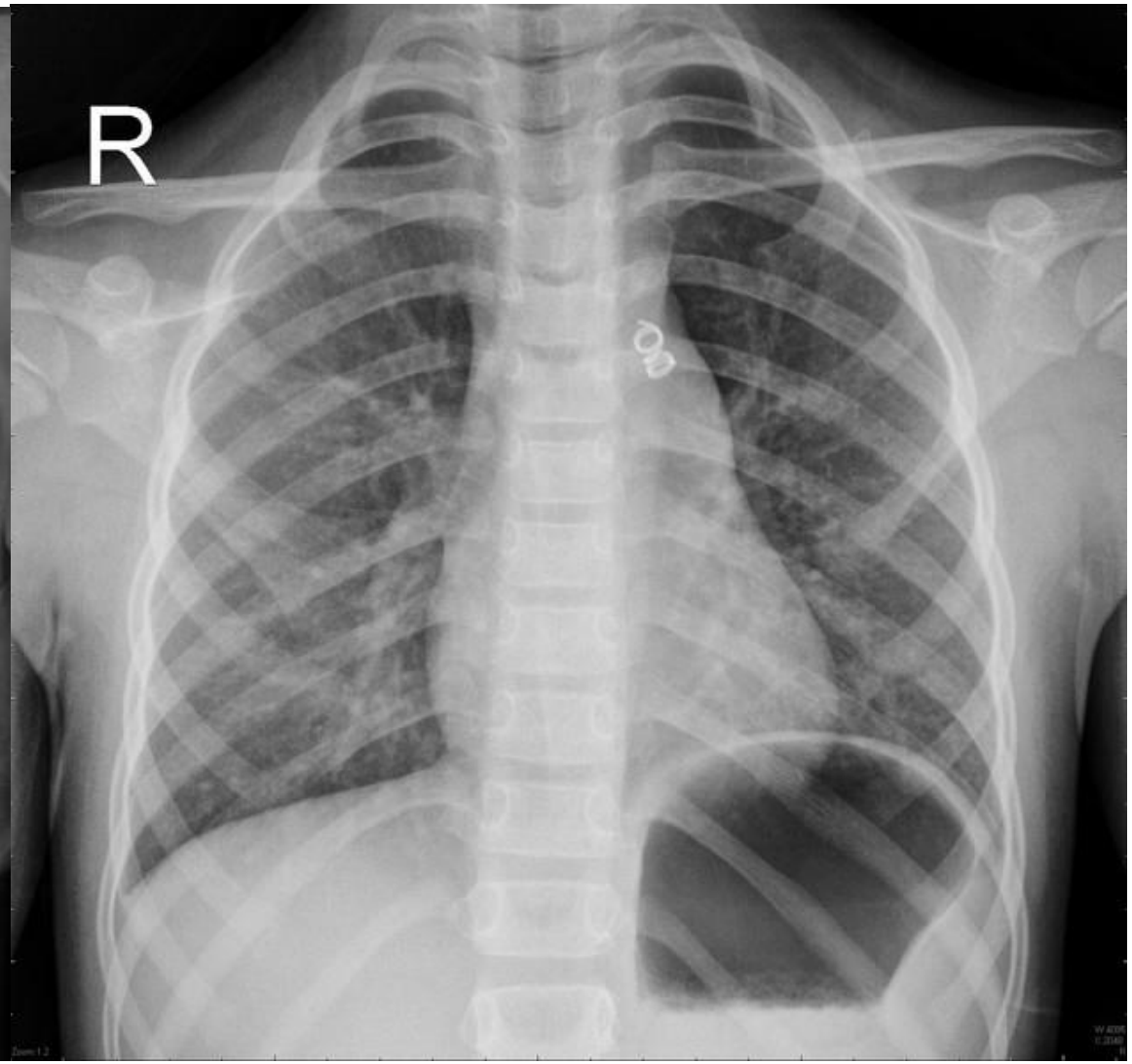
Interventional closure of PDA:

- **Trans-catheter closure**
  - Intravascular coils
  - Amplatzer patch
- **Surgical ligation --Left thoracotomy**



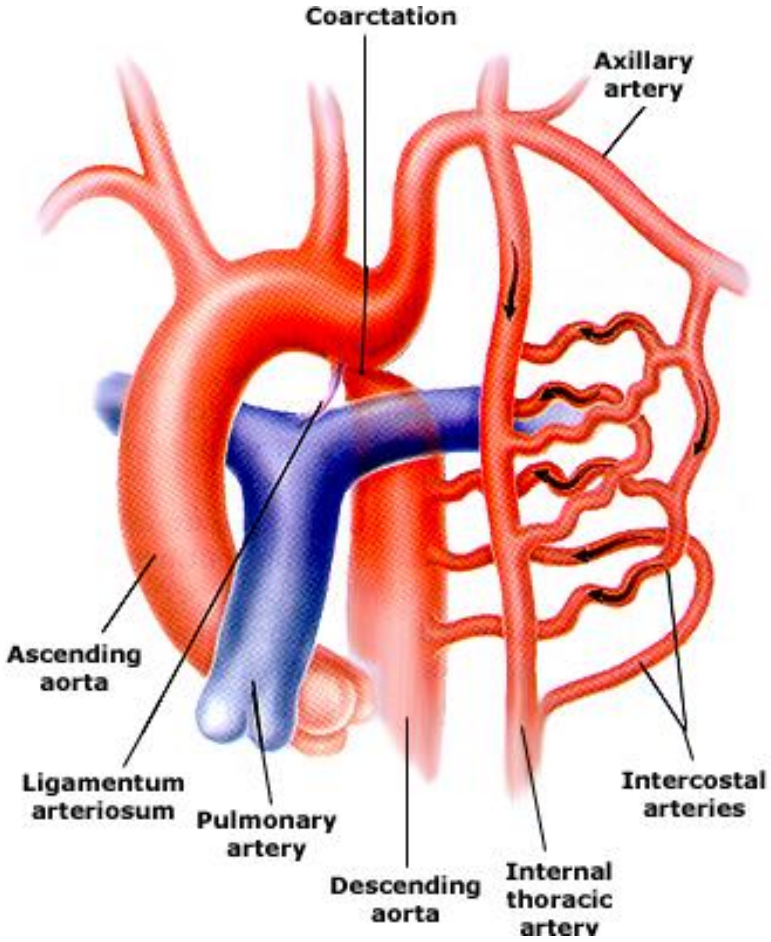



(R)


R

COA

- Discrete narrowing of the thoracic aorta
  - Distal to left subclavian artery
    - At ductus arteriosis
  - Proximal to left subclavian artery
- Rarely long segment or tubular hypoplasia



Epidemiology/ Pathogenesis

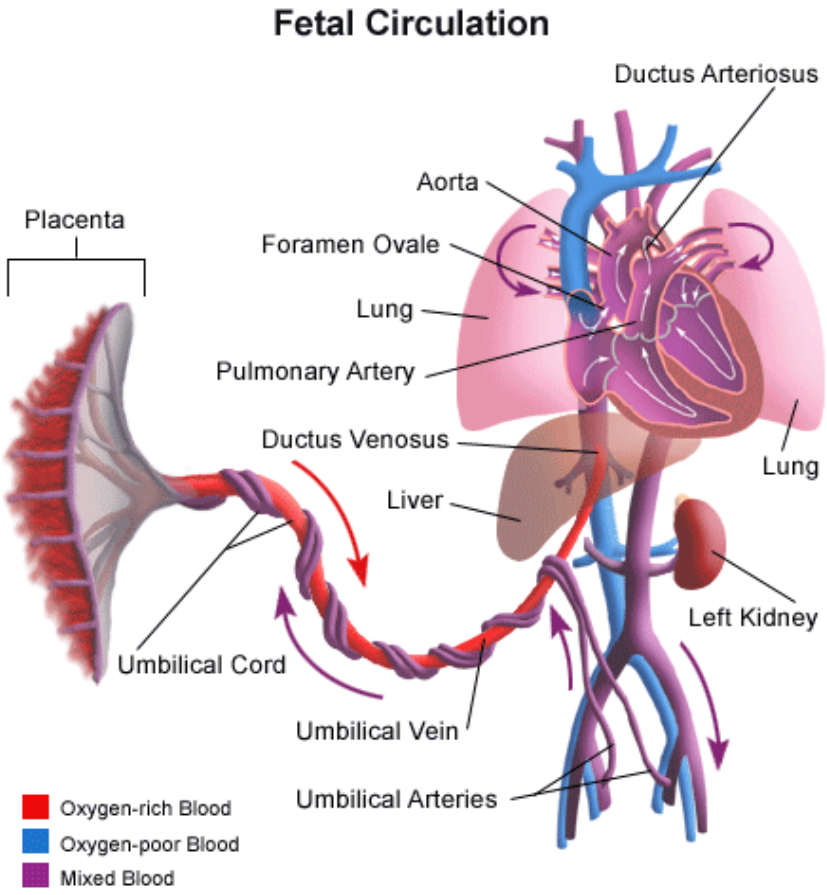
- 6-8% of all congenital heart defects
- Male: Female 2-5:1
- Sporadic; rare familial
- **Congenital**
  - ❖**Most common**
- **Acquired**
  - ❖**Inflammation/Arteritis, eg, Takayasu**
    - ❖**Mid-thoracic, abdominal aorta**
  - ❖**Severe atherosclerosis**

Pathogenesis/Pathology

- Mechanism unknown
  - Genetic defects?
  - Intrauterine defects, eg impaired blood flow → altered endothelial development?
- Medial thickening + intimal hyperplasia → posterolateral ridge encircling lumen
- Surgical specimens:
  - ↑ collagen
  - ↓ smooth muscle mass in pre vs poststenotic areas
  - Cystic medial necrosis: disarray of elastic tissue

Being born can be a problem

- **In utero:**
  - High PVR, low SVR
  - 90% cardiac output: → PDA → descending ao
- **At birth:**
  - ↑ SVR
  - ↓ PVR
  - PFO and PDA closure
  - CO through ascending aorta

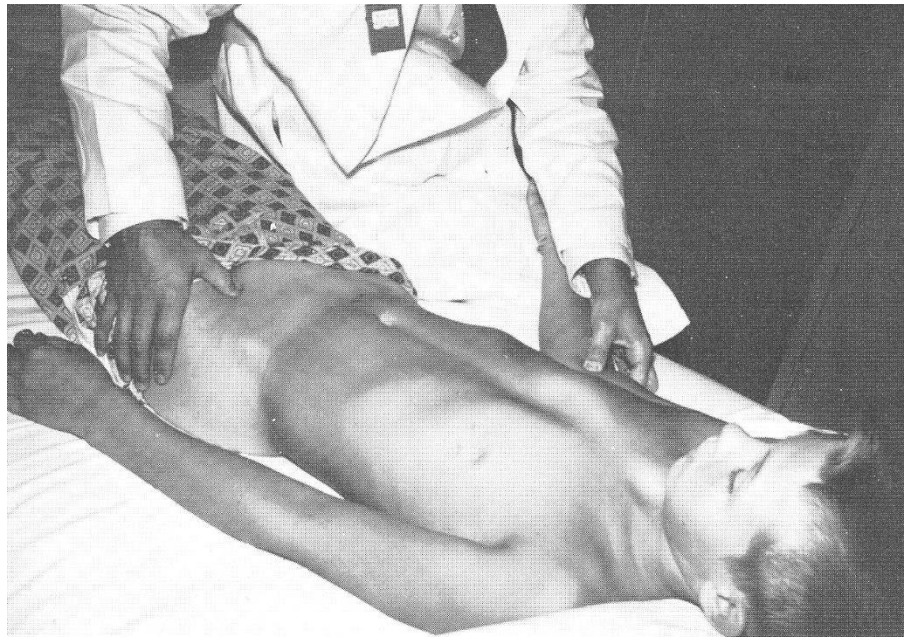


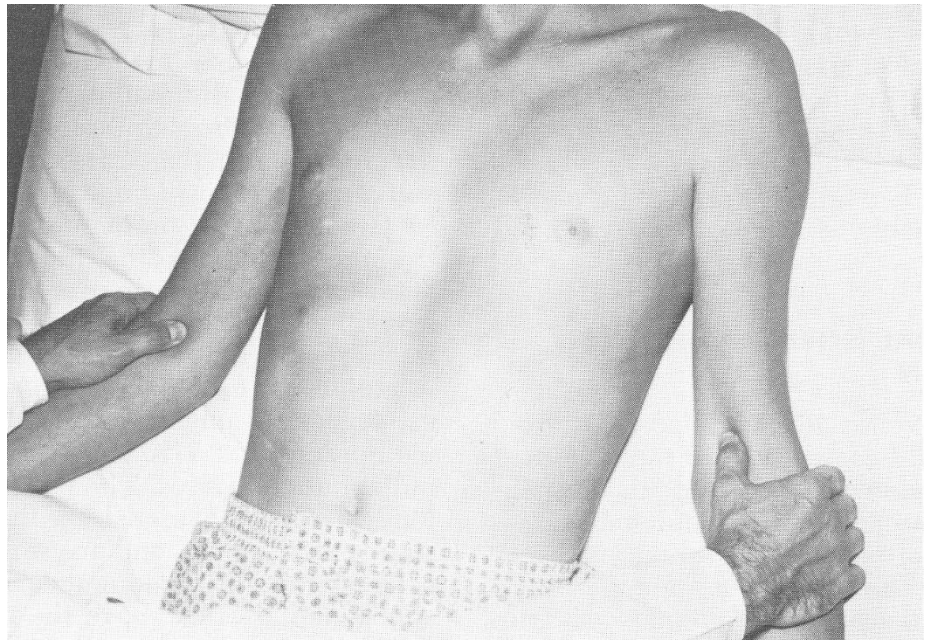
Clinical Manifestations

- **Neonates**
  - Absent/delayed femoral pulse
  - Differential cyanosis if severe and large PDA R→L shunt
  - Heart failure/ shock in first days of life
- **Children**
  - Delayed diagnosis in mild coarctation
  - Chest pain with exercise, cold extremities, claudication
- **Adults**
  - Hypertension
  - Claudication

Differential Blood Pressure

- **Classic findings**
  - Hypertension in upper extremities
    - **Mechanical obstruction**
    - **↑ renin secretion → volume expansion**
    - **Less sensitive baroreceptor reflex**
    - **Reduced arterial compliance**
  - Decreased/ delayed femoral pulse
  - Low blood pressure in lower extremities





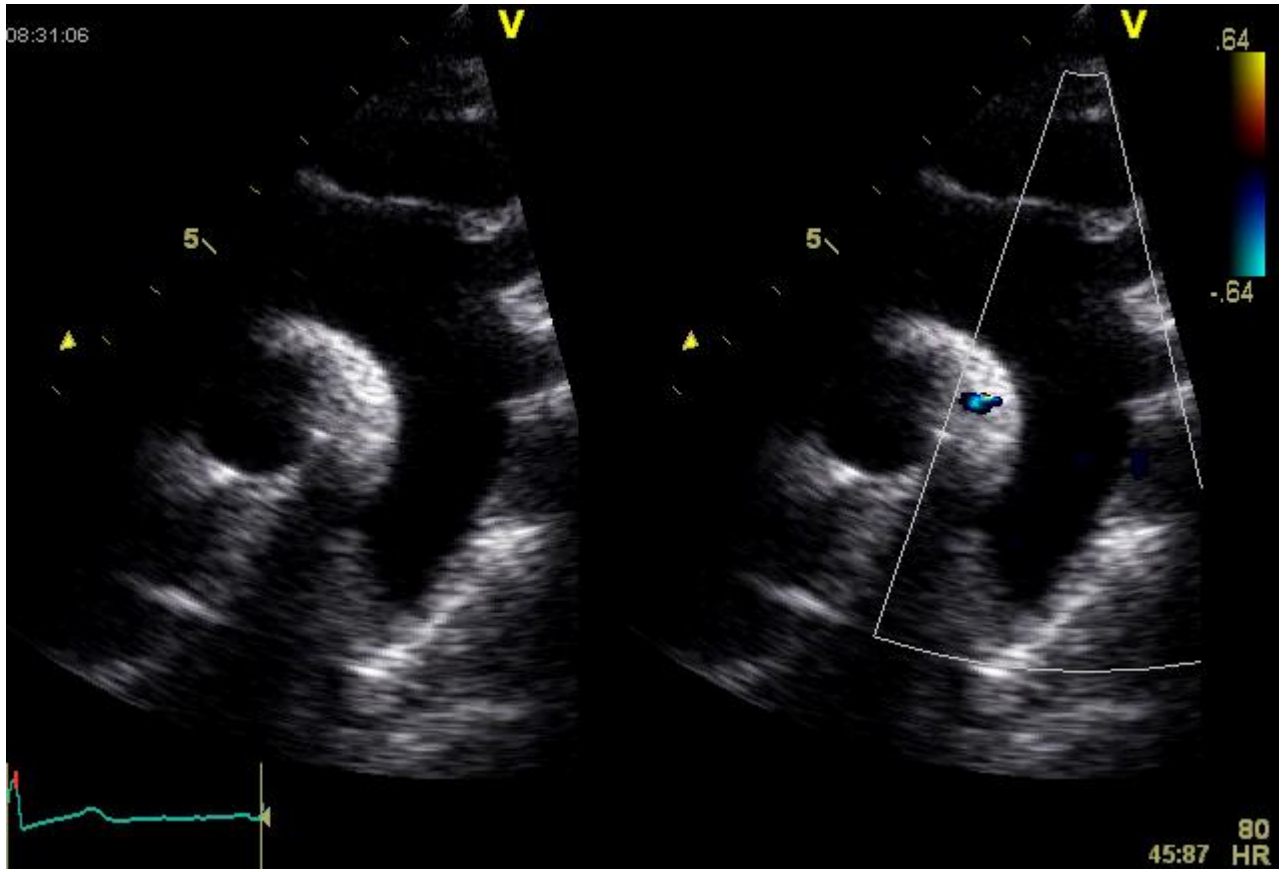
Neurologic Comorbidities

- Increased frequency of intracranial aneurysms
- Dilation of collateral spinal arteries → compress spinal cord

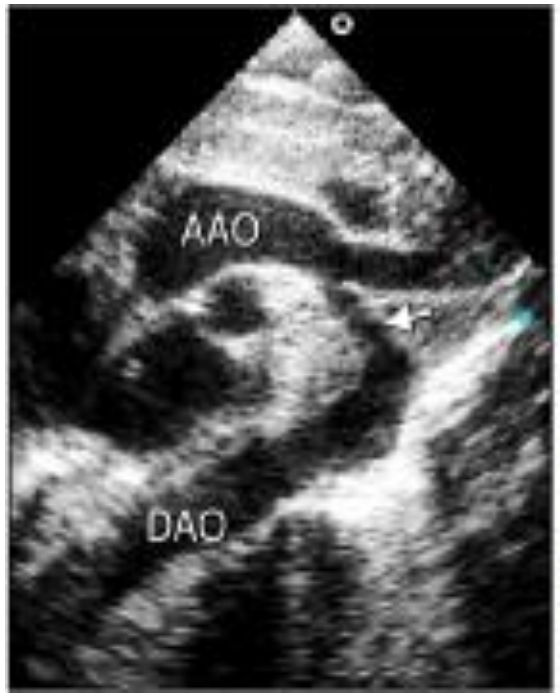
Cardiac Exam

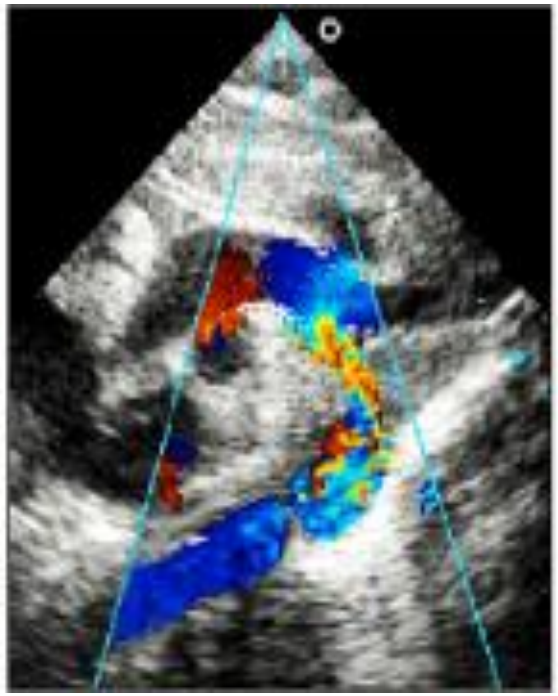
- Often normal without co-existing defects.
- Continuous murmur if large collateral vessels.
- Systolic ejection click and/or murmur if bicuspid aortic valve.
- Vascular murmur from flow across Coarctation itself.

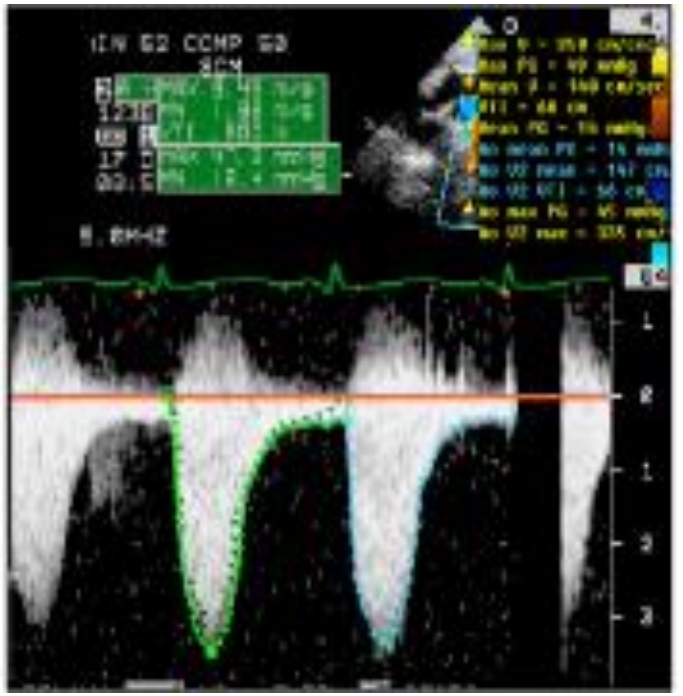
ECHO
Normal Aortic Arch



COARCTATION







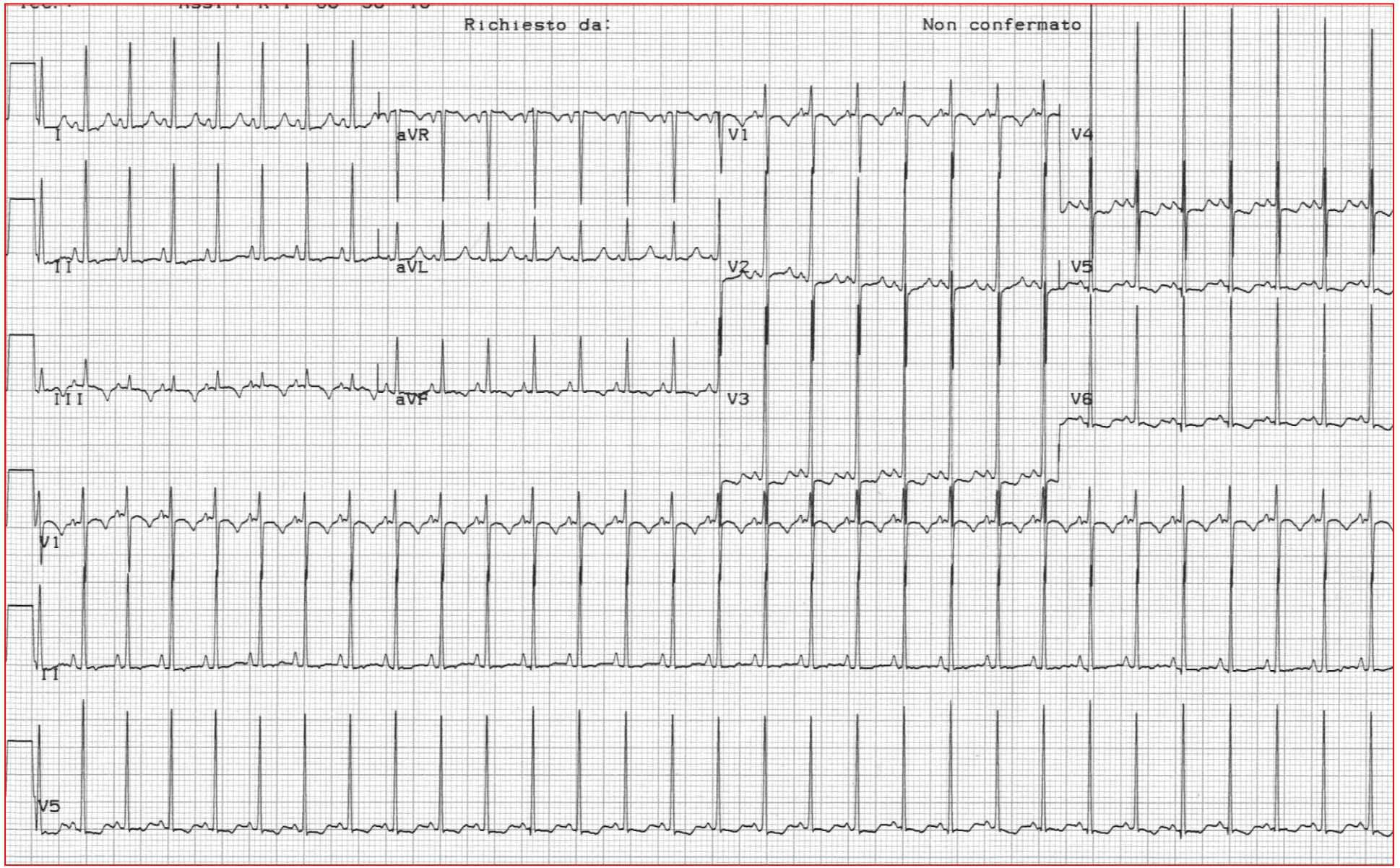
ECG

Neonates

- Sinus tachycardia
- Right ventricular hypertrophy
- Right axis deviation
- Left ventricular hypertrophy (more rare and more late and in the cases with assotiated lesions)

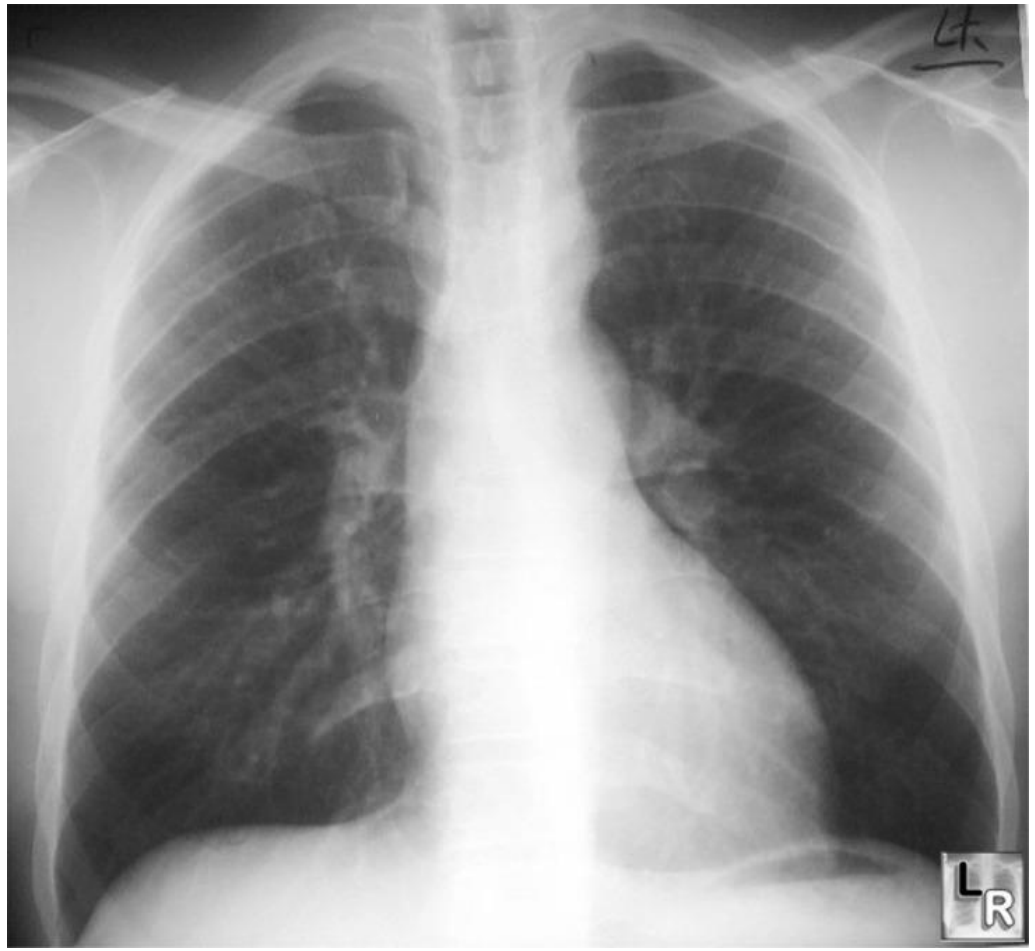
Children

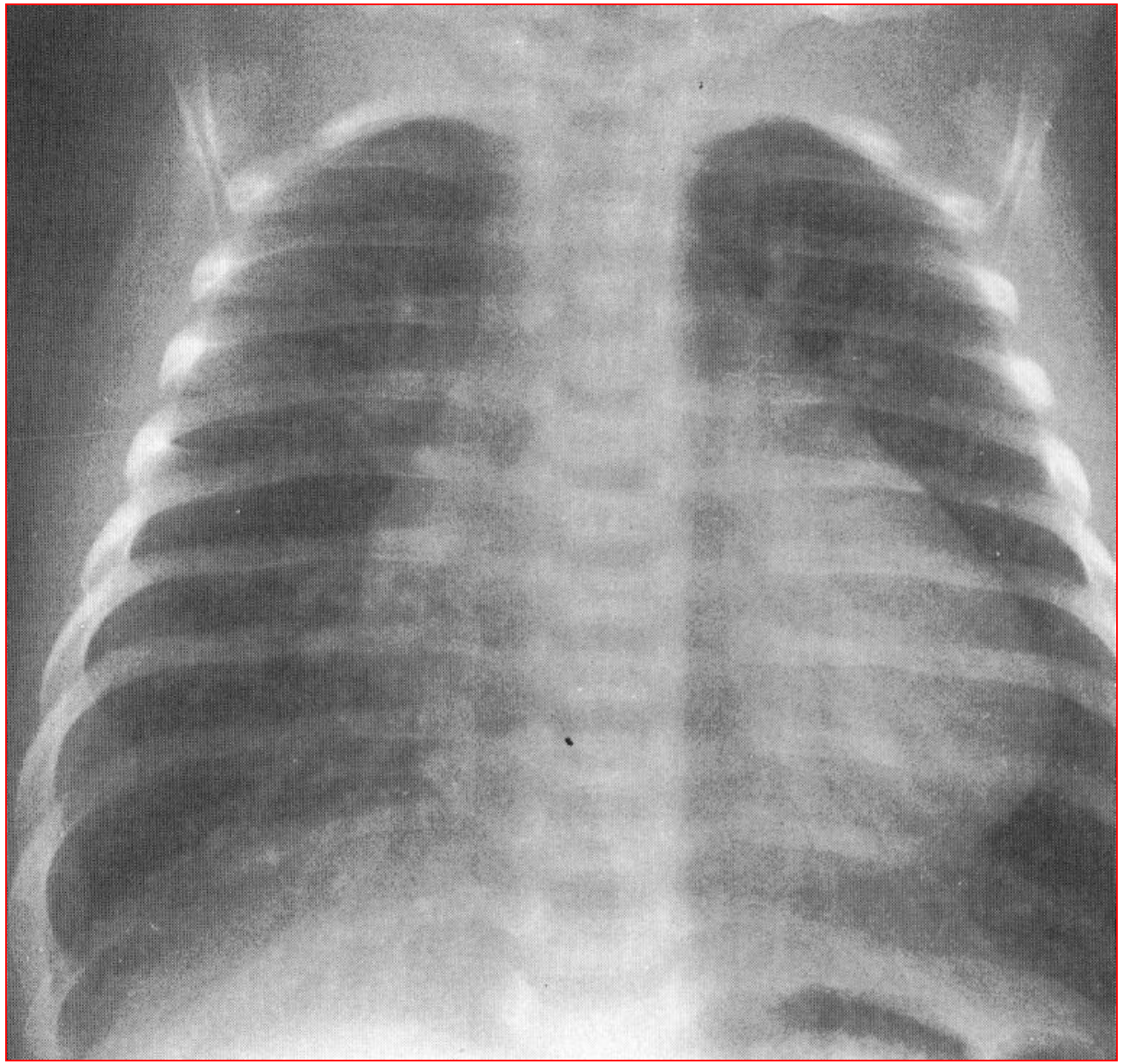
- Left ventricular hypertrophy
- Left axis deviation (think of AVC, DORV)
- Right ventricular hypertrophy if pulmonary hypertention

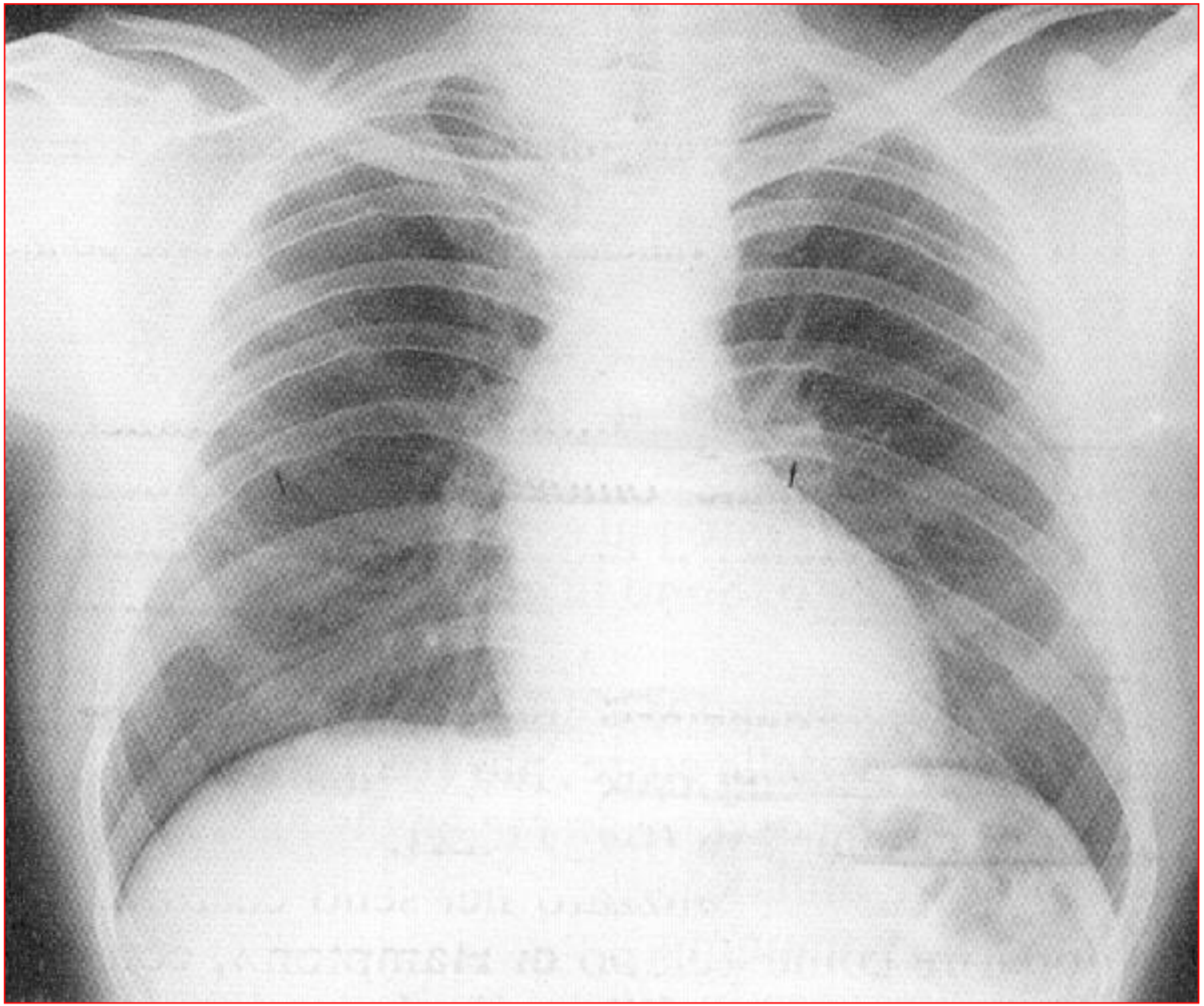

Richiesto da:
Non confermato
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
V1
II
V5

CXR

- Infants with severe disease: cardiomegaly, heart failure
- Notching posterior ribs: erosion by collaterals
- “3” Sign: Indentation of aortic wall with pre and poststenotic dilatation



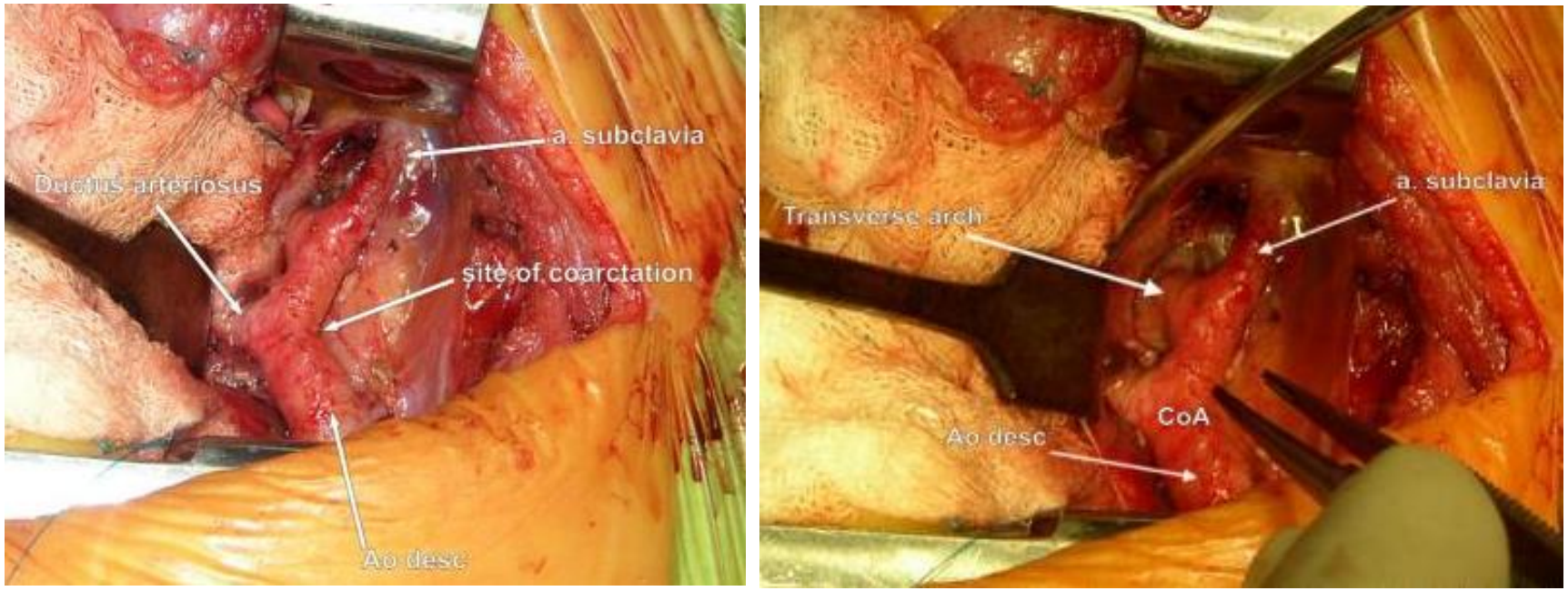


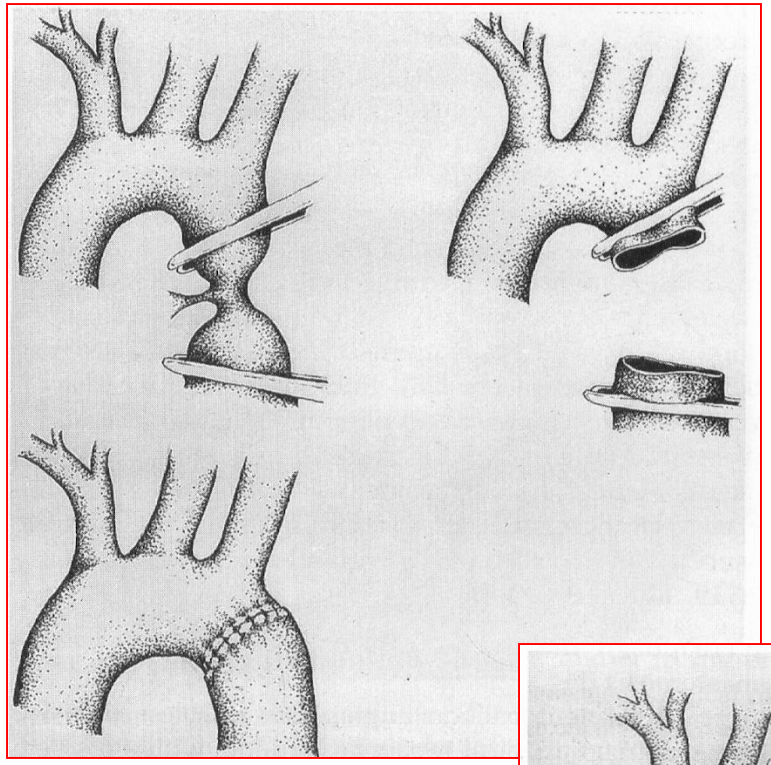


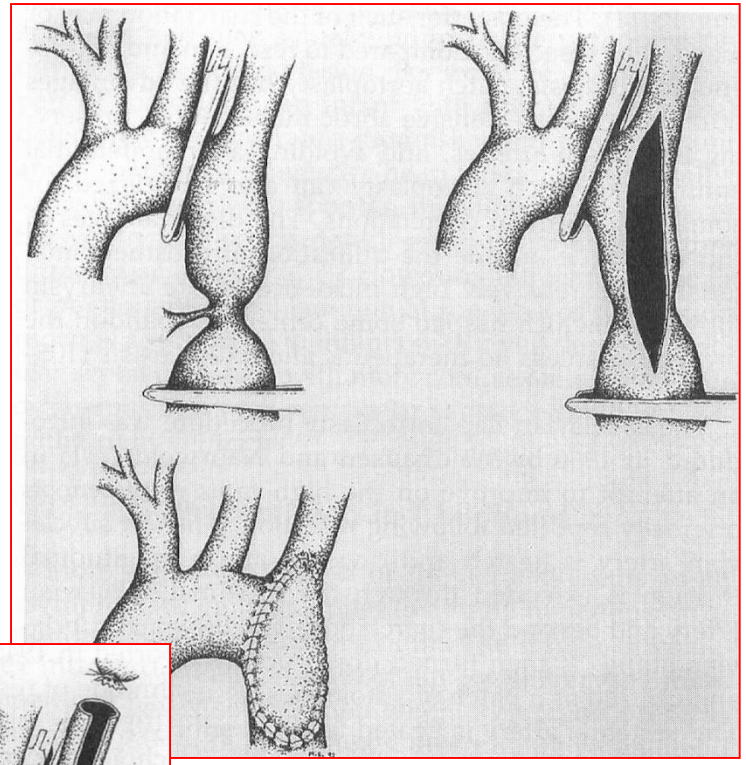
COA: Early neonatal presentation management

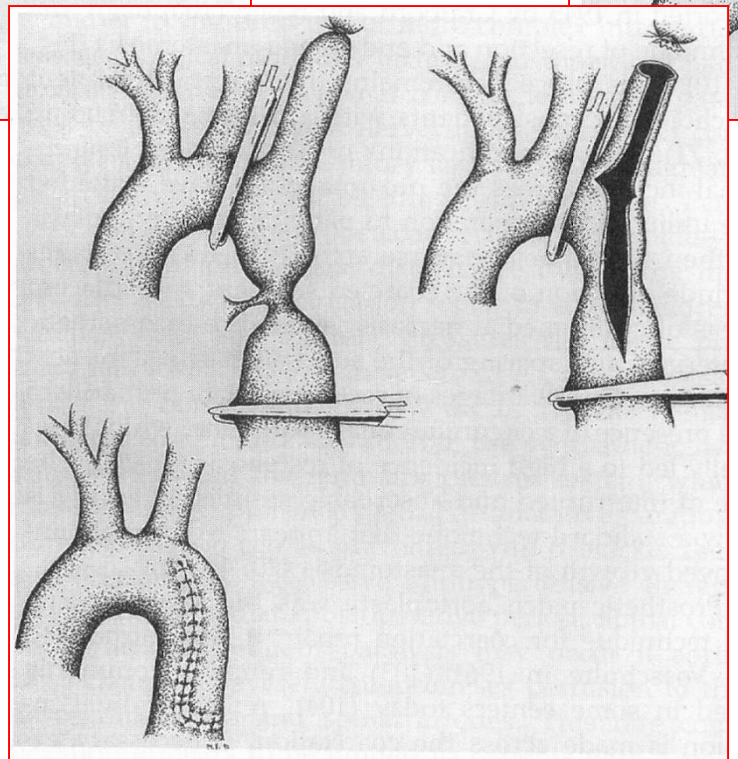
- Prostaglandin E1 (0.05-0.1 mcg/kg/min) infusion to open DA.
- Anti-failure medication for congestive heart failure (CHF) treatment.
- Inotropic medications (as dopamine, dobutamine, or epinephrine) may be used in case of lv dysfunction assotiated with hypotention.
- Renal function and urine output monitoring.
- ABGs and metabolic acidosis monitoring.
- After stabilization with medication , N.B will be ready for surgical intervention.

Surgery


a. subclavia
Ductus arteriosus
site of coarctation
Ao desc
Transverse arch
a. subclavia
CoA
Ao desc



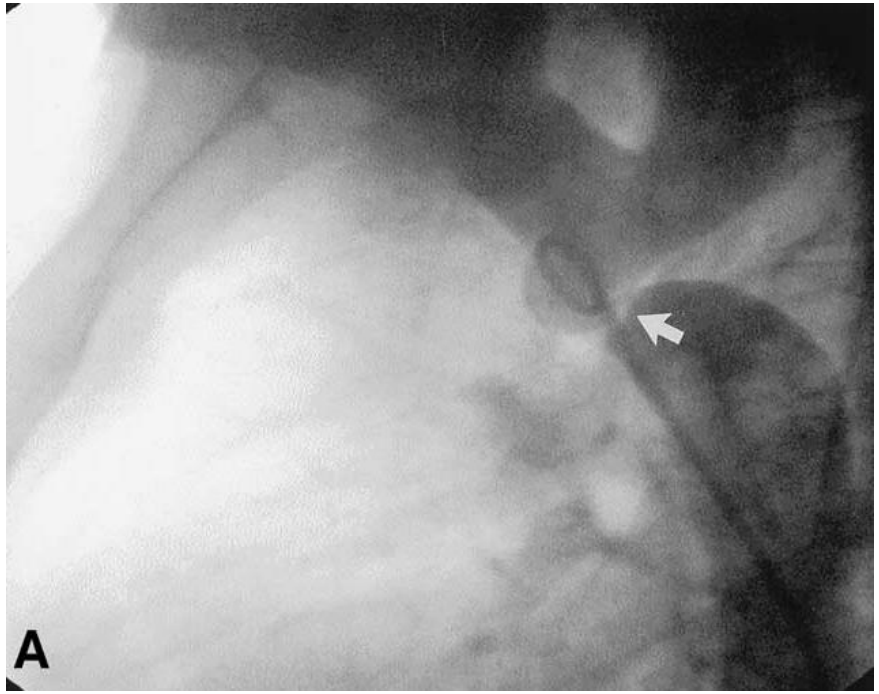


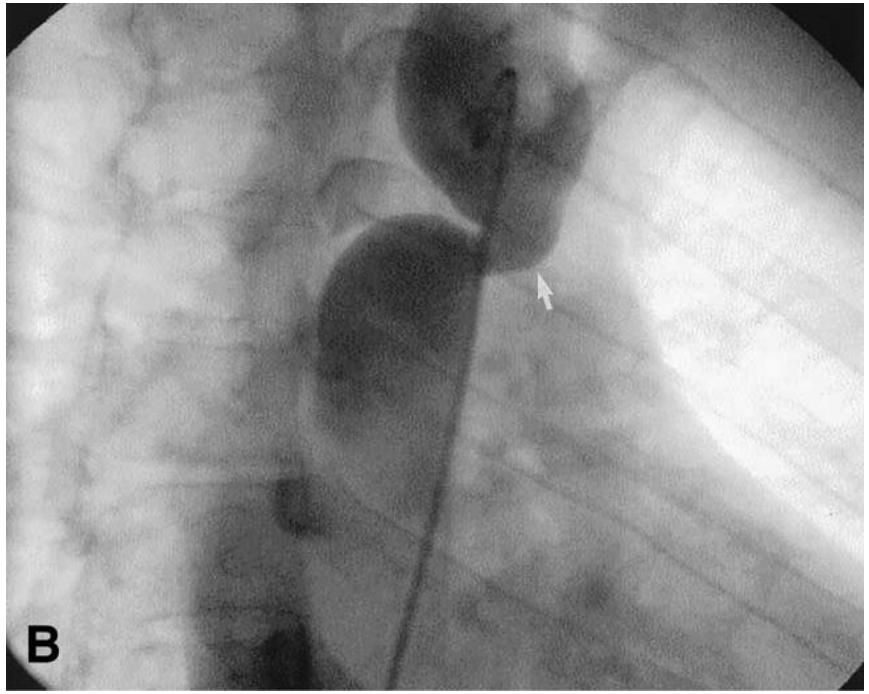


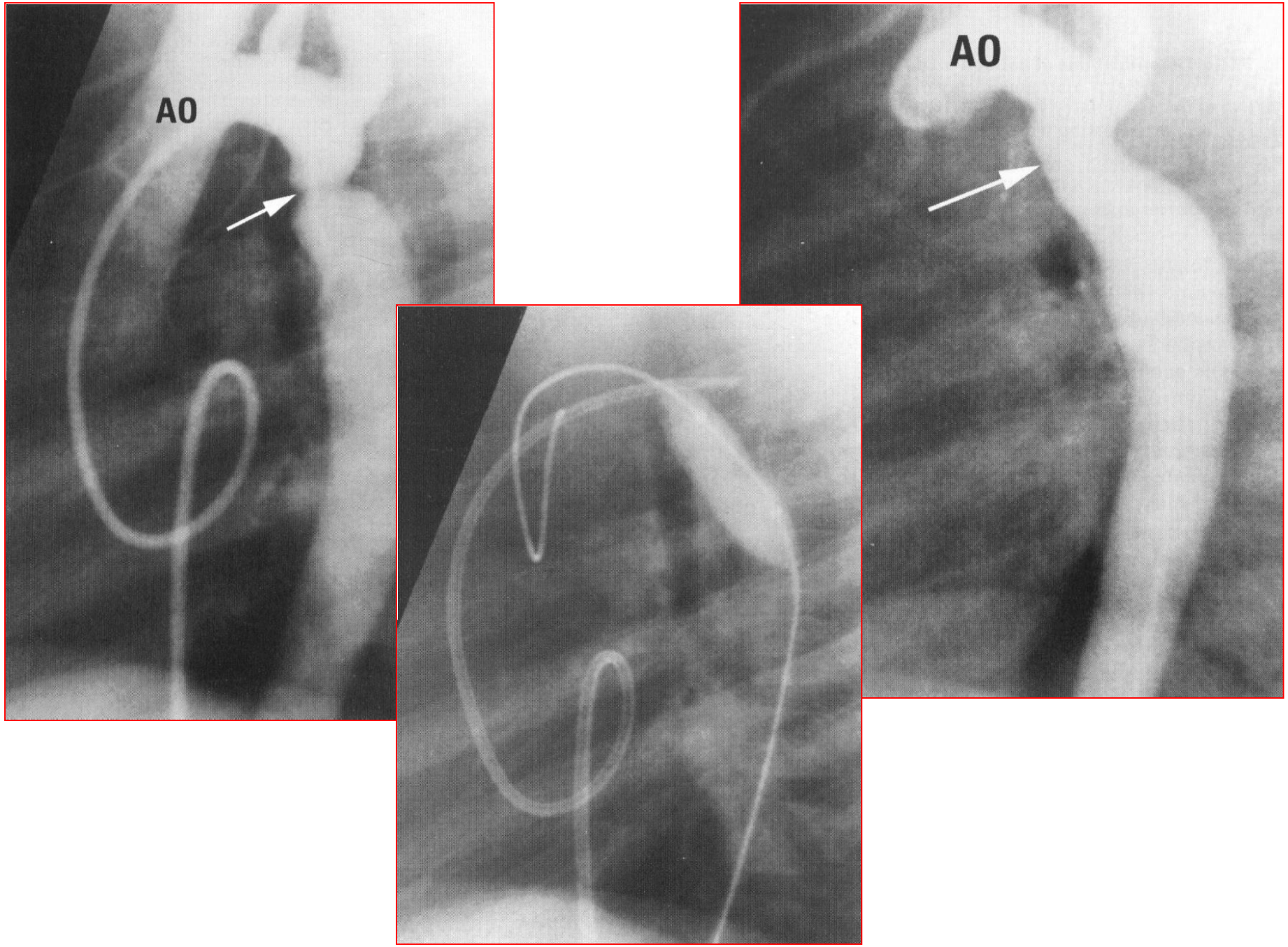
COA:Late presentation of coarctation of the aorta

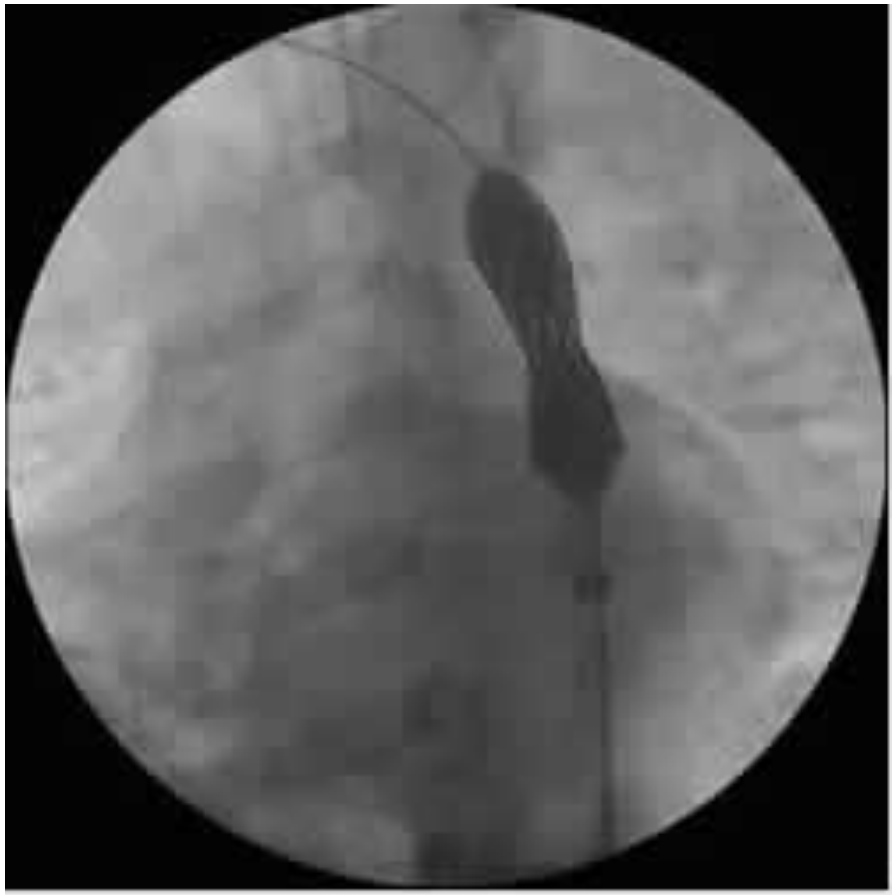
- If there is Significant hypertension or congestive heart failure (CHF), Surgical relief of the aortic obstruction or hemodynamic interventional techniques (as balloon angioplasty with stent) is indicated
- Proper imaging with MRA or Aortic arch MRI may be needed for proper evaluation to select the best way of coarctation intervention

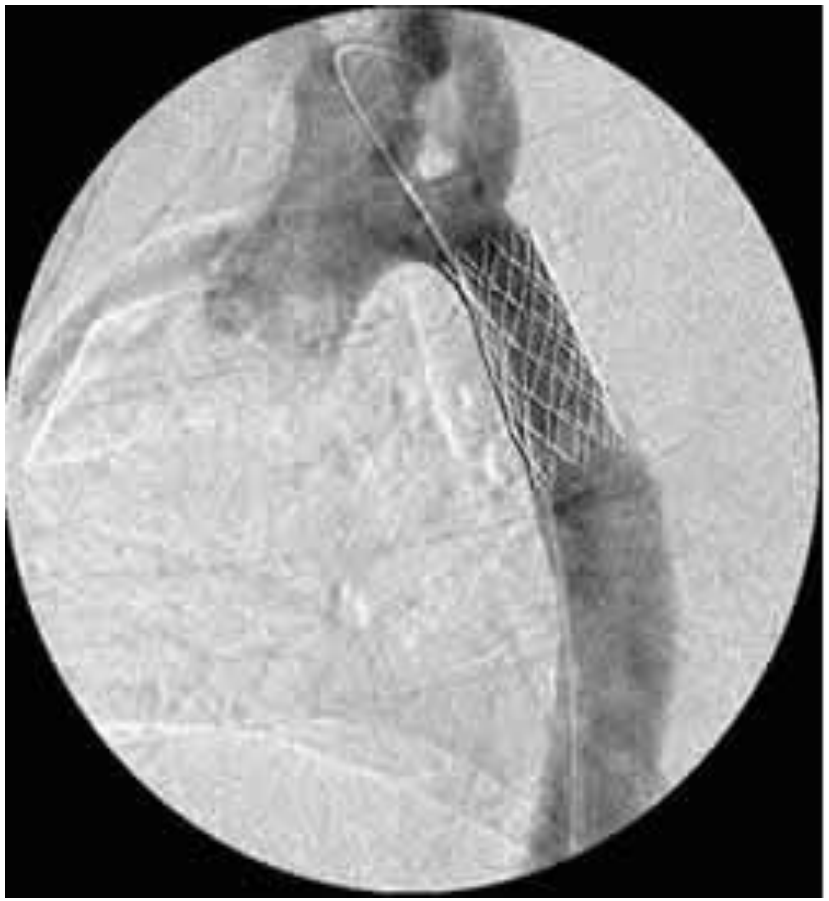
Cardiac Catheterization



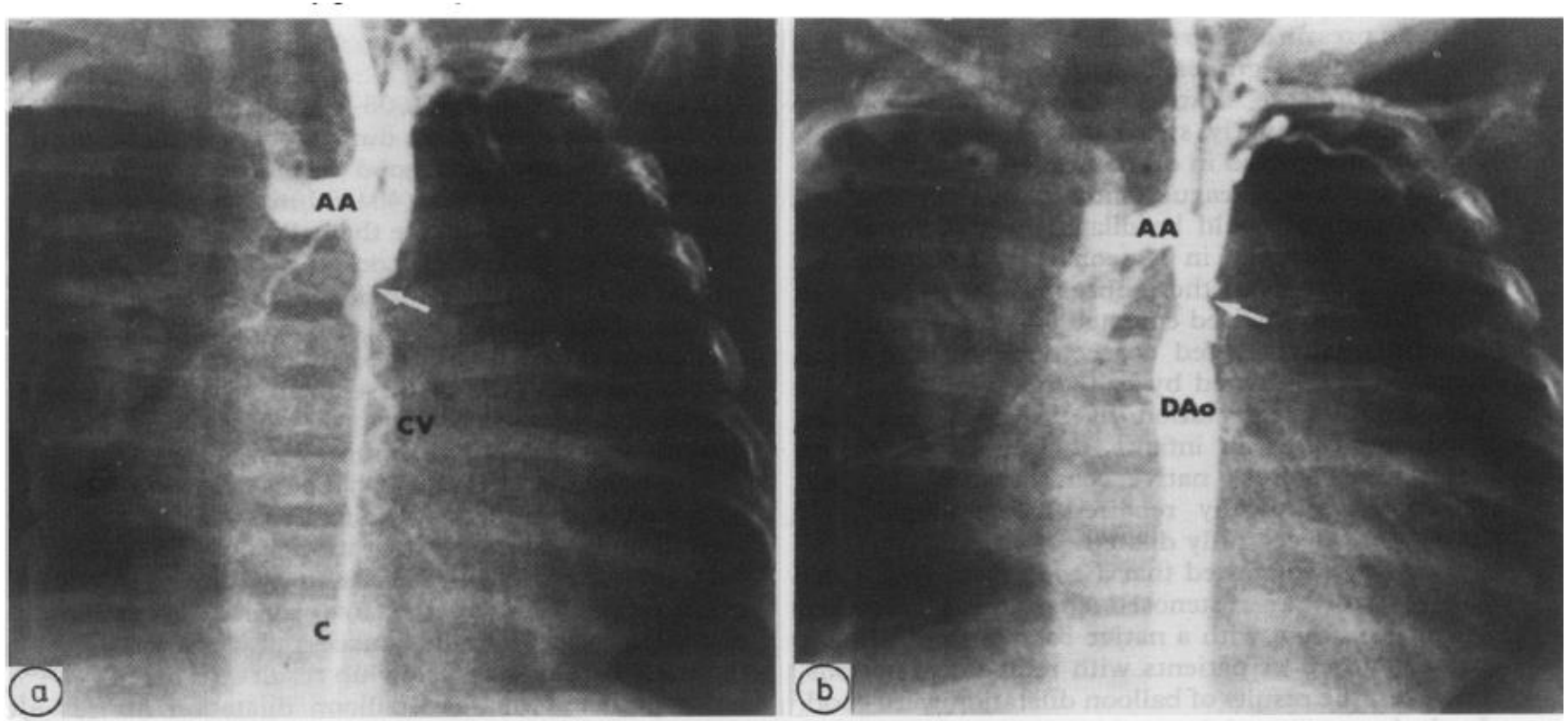



AO
AO





Balloon Angioplasty (BA)


AA
CV
C
a
AA
DAo
b

THANK YOU FOR YOUR ATTENTION