LEVEL 4

CVS 1

OUTLINE

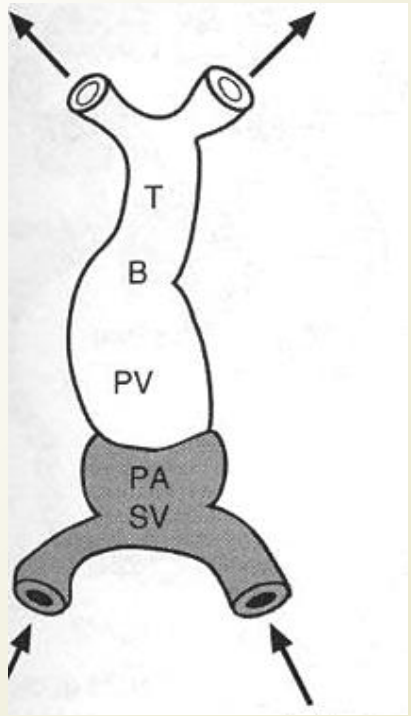
- INTRODUCTION
- EMBRYOLOGY
- IMAGING MODALITIES
- CONGENITAL HEART DISEASES
- SPECIFIC CONDITIONS

INTRODUCTION

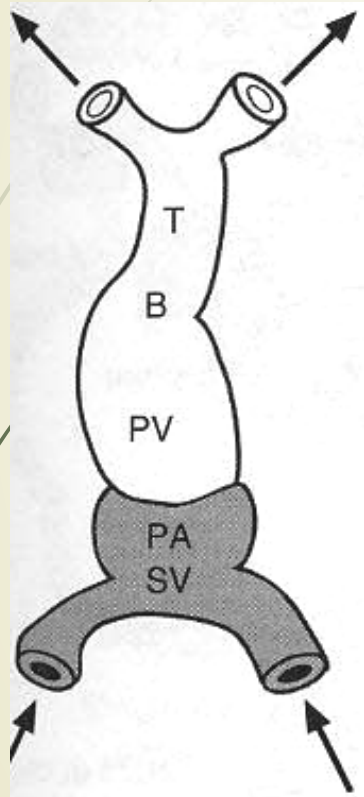
- The fetal heart develops from a series of complex and rapid changes which occur between second and eighth week of intrauterine life.
- During third to fifth weeks cardiac structures develop most actively and are most susceptible to adverse external influence eg. rubella virus or drugs such as thalidomide resulting in CHD.

EMBRYOLOGY

- The heart originates from splanchnic mesoderm
- endocardial tube is the initial basic structure
- development is characterised by rapid and complex changes
- Initially straight longitudinal midline tube.
- 5 dilatations develop along it;
  - common truncus arteriosus
  - bulbous cordis
  - common primitive ventricle
  - common primitive atrium
  - sinus venosus



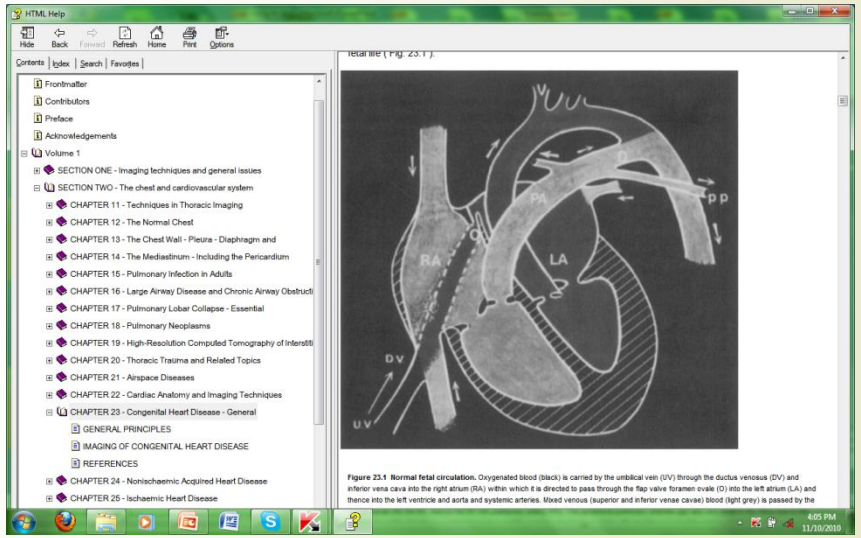
Adult derivatives of the primitive heart



Embryonic Dilatation	Adult Structure
Truncus arteriosus (T)	Aorta Pulmonary trunk
Bulbus cordis (B)	Smooth part of right ventricle **(conus arteriosus)** Smooth part of left ventricle **(aortic vestibule)**
Primitive ventricle (PV)	Trabeculated part of right ventricle Trabeculated part of left ventricle
Primitive atrium (PA)	Trabeculated part of right atrium Trabeculated part of left atrium
Sinus venosus (SV)	Smooth part of right atrium **(sinus venarum)*** Coronary sinus Oblique vein of left atrium

Normal fetal circulation

- Oxygenated blood is carried by umbilical vein (UV) through ductus venosus (DV) and IVC into RA through foramen ovale into LA & LV, aorta & systemic arteries.
- SVC & IVC blood is passed into RA, RV & PA. Most of the RV output passes through PDA into descending aorta to supply lower parts of fetus and umbilical arteries which supply the fetal placental plexus.



CHD IMAGING MODALITIES

- Plain radiography
- Echocardiography- First line to assess anatomy and function.
- Cardiac catheterization- definition of vascular anatomy.
- CT- description of cardiac and vascular anatomy in relation to other structures of the chest·
- MRI- Quantification of cardiac function and vascular flow in vivo.

CHD IMAGING MODALITIES

- Plain radiography
- Echocardiography- First line to assess anatomy and function.
- Cardiac catheterization- definition of vascular anatomy.
- Radionuclide imaging - quantify left-to-right shunts, assess relative pulmonary perfusion.
- CT- description of cardiac and vascular anatomy in relation to other structures of the chest.
- MRI- Quantification of cardiac function and vascular flow in vivo.

Chest radiograph indications

- Suspected congenital heart disease
- Cyanosis
- Cardiac failure
- Failure to thrive
- Difficult in feeding in the neonatal period
- Fainting spells

Chest Radiograph

Standard views - Erect PA/AP chest

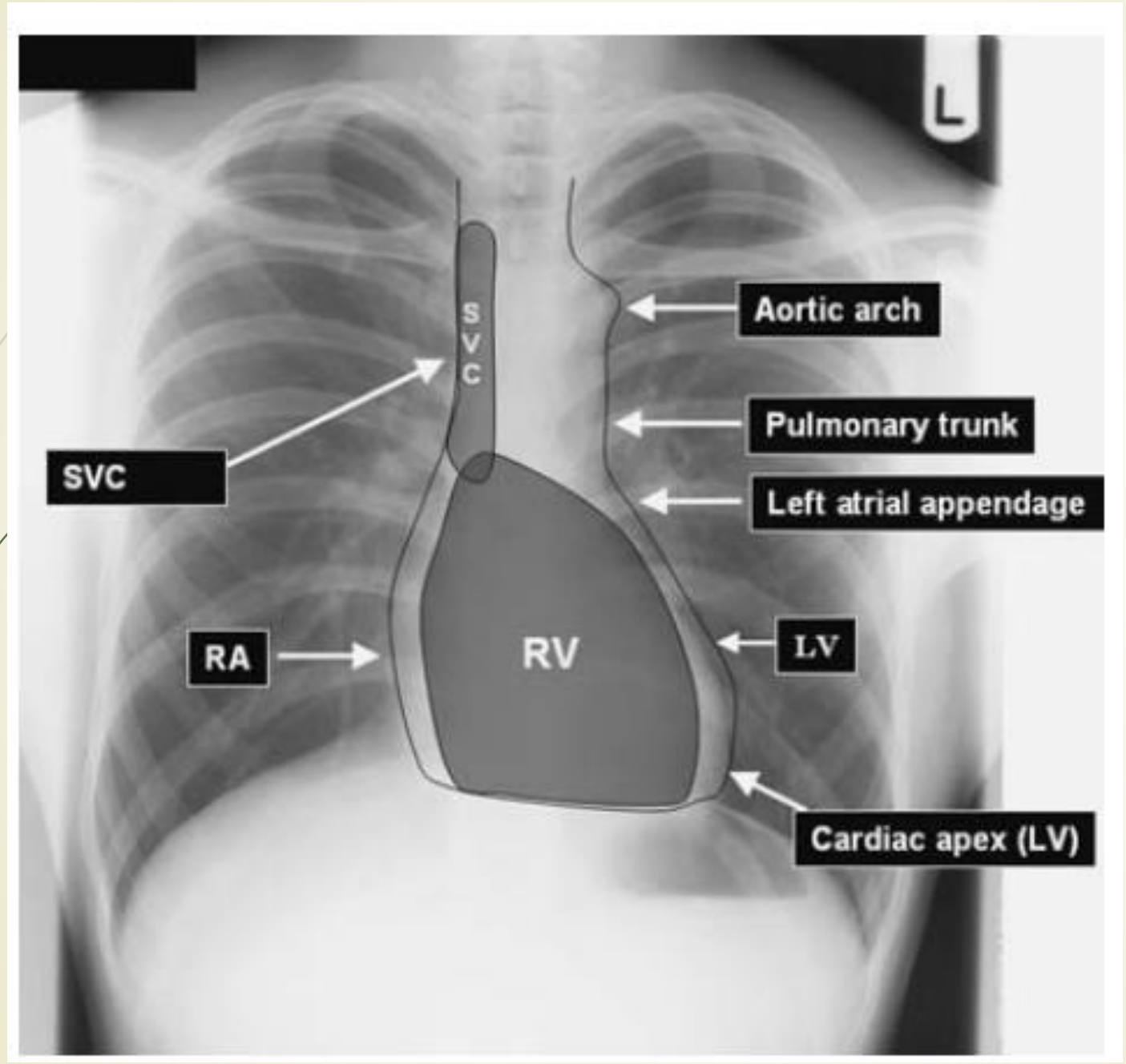
- Left lateral

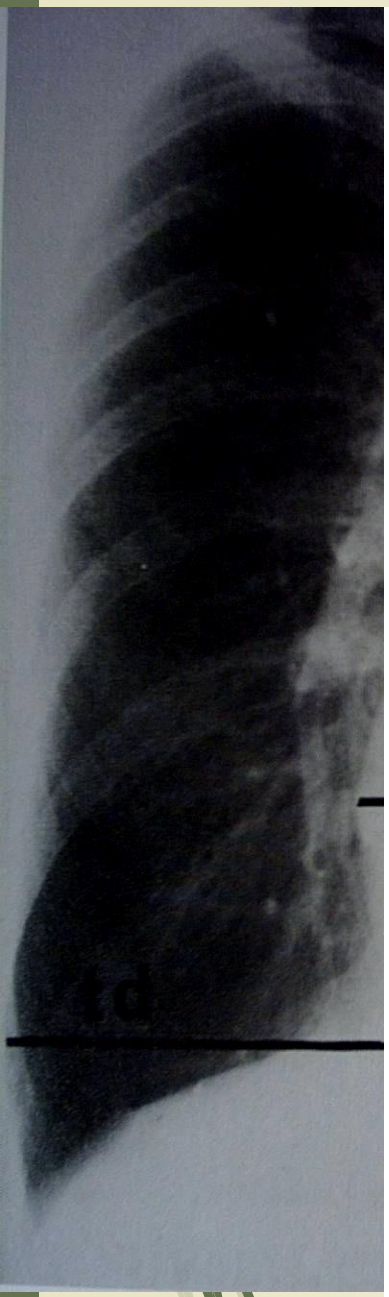
Role- Provides clues to underlying CHD.

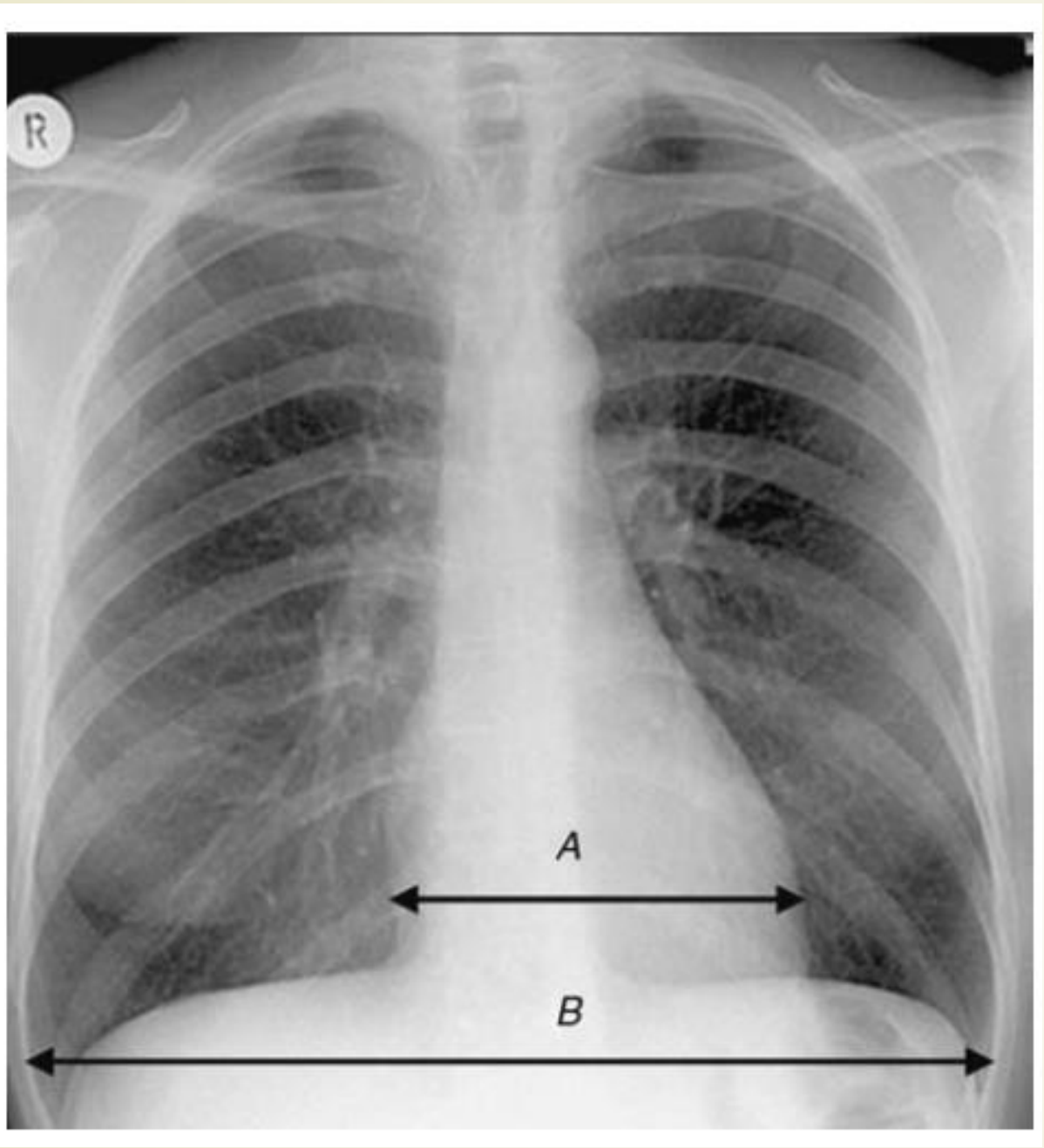
- instrumental in planning further investigations.
- limited use in neonates and young children.
- may show related skeletal anomalies

Assessment:

- Cardiac and visceral situs- situs solitus, inversus, ambiguous
- Cardiac size & shape.
- Mediastinum- Aortic arch, PA, trachea indentation
- Pulmonary vasculature-normal, plethora, oligemia, congestion
- Skeletal features - Rib notching: coarctation of aorta
- post surgical stigmata eg. sternal sutures, implants


L
SVC
SVC
Aortic arch
Pulmonary trunk
Left atrial appendage
RA
RV
LV
Cardiac apex (LV)





CARDIAC SIZE MEASURMENT

r-max. Extension of the heart to the right of the midline

l-max. extension of the heart to the left of the midline

Transverse diameter is of the heart is r+L

td=max. transverse diameter of the thorax

=50-55% upto 60% in children

Echocardiography

- Gives a wealth of anatomical and physiological information without ionising radiation, patient discomfort or significant risk
- 1D (M Mode), 2D most commonly used, 3 D
- Doppler echocardiography ;

  -CFI; ‘non-invasive’ angiogram

  -Pulsed doppler echo (valves)

  -continuous wave doppler
- Contrast echocardiography

Contrast echocardiography

- Use of intravascular contrast effects produced when microbubbles of gas present in solution are injected into a peripheral vein or selected area of the heart via a catheter.

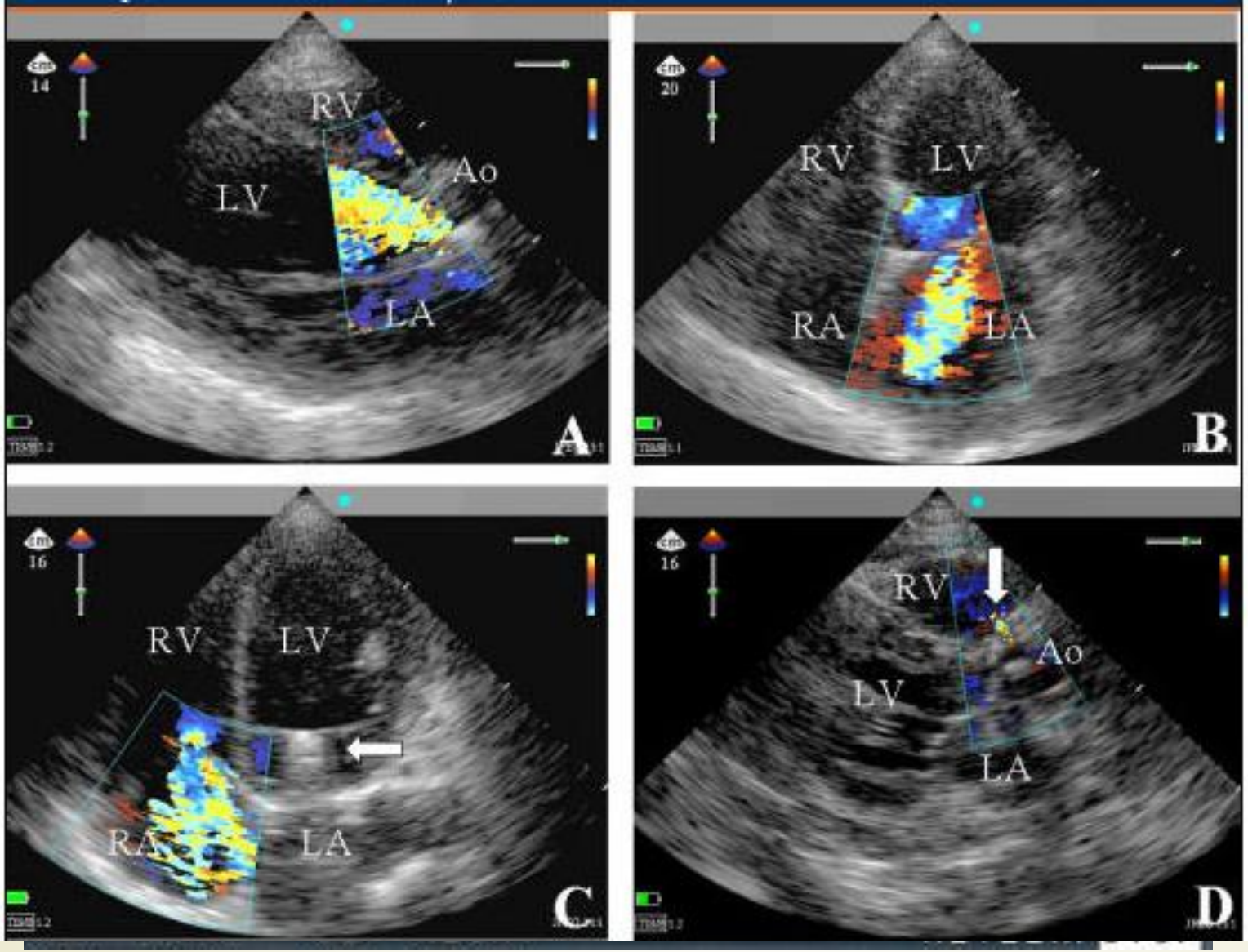
- Microbubbles are generated by rapid injection of saline, blood, indocyanine

Indications

- Myocardial perfusion can be investigated directly/indirectly after intracoronary/ venous contrast injection with/out DSA
- Better define anatomy not shown well on 2D E.g imaging the anomalous venous connections to the heart
- Enhancement of endocardial boundaries increasing sensitivity of wall motion abnormalities e.g.valvular regurgitation,shunts

Doppler echocardiography

- Used to study venous flow from IVC, SVC, pulmonary vessels and flow across the valves
- Measure direction and speed of blood flowing through the heart and blood vessels
- Check for abnormal communication between left and right side of the heart and regurgitation
- Calculate cardiac output/ejection fraction
- Flow towards the transducer- red away from it – blue, and turbulence flow – mixture of red and blue- yellow



A; Parasternal long axis view –shows regurgitant jet consistent with massive aortic insufficiency

B; Apical 4c view-systolic murmer; c:Apical 4c view – severe left vent. Systolic dysfunction.

Tricuspid regurgitation is shown; D;Parasternal long axis – shows aorto-right

Clinical applications of echocardiography

- Congenital heart disease
- Anatomical defects, shunts
- Stress echo
- Pericardial disease
- Coronary artery disease
- Cardiomyopathies
- Valvular disease
- Cardiac masses
- Diseases of the aorta

Angiocardiography

- Important technique for cardiac and great vessel anatomy
- Coronary angiography commonest

  -transfemoral approach using Judkins or Amplatz catheters
- Pulmonary angiography; for possible embolism.
- Adv; Offers high resolution images of the heart and great vessels.
- Disadv ; Requires experienced hands and cardiac monitoring, invasive

Angiocardiography cont'd

- Indications :
- CHD, valvular/myocardial disease, ventricular function
- Technique;

-right sided structures studied by introducing catheter into a peripheral vein, femoral or antecubital or basilic

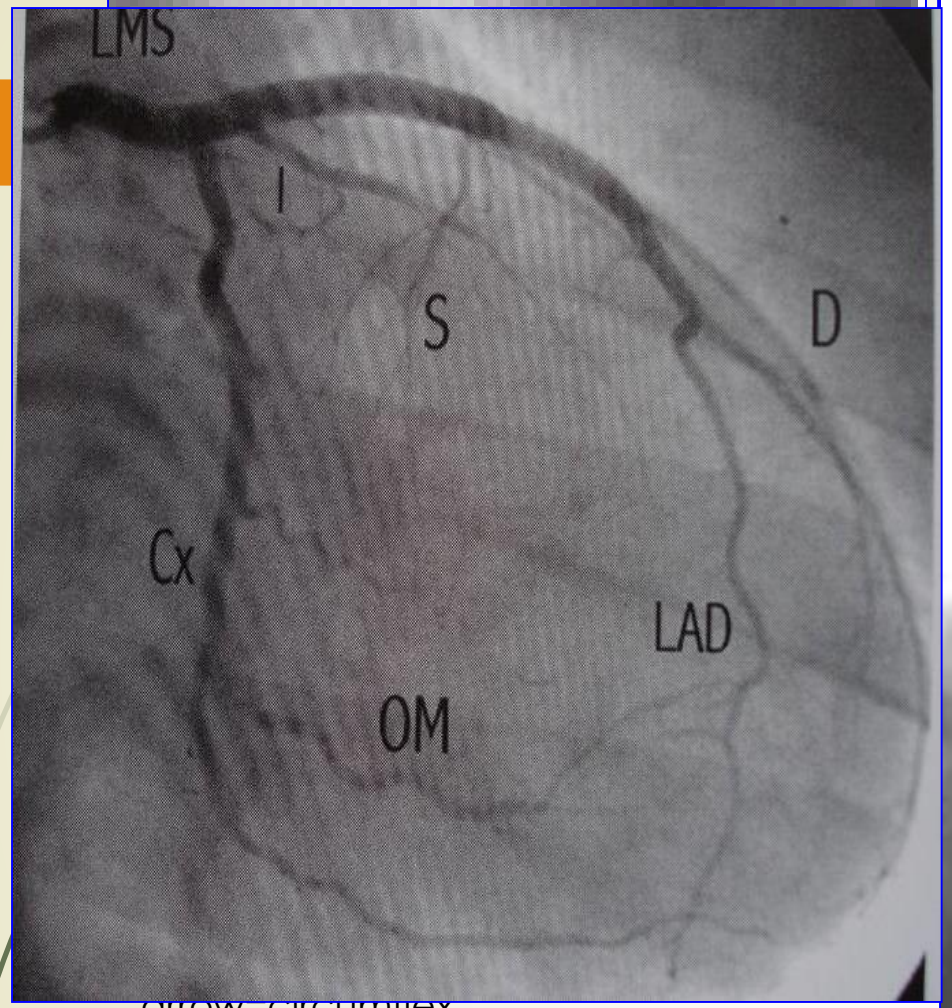
-left sided structures by introducing catheter into femoral artery retrogradely

CM-LOCM 370; 1ml/kg at 18-20mls/s (adults),1.5ml/kg

(0-1yr);1.0ml/kg (>5 yrs)

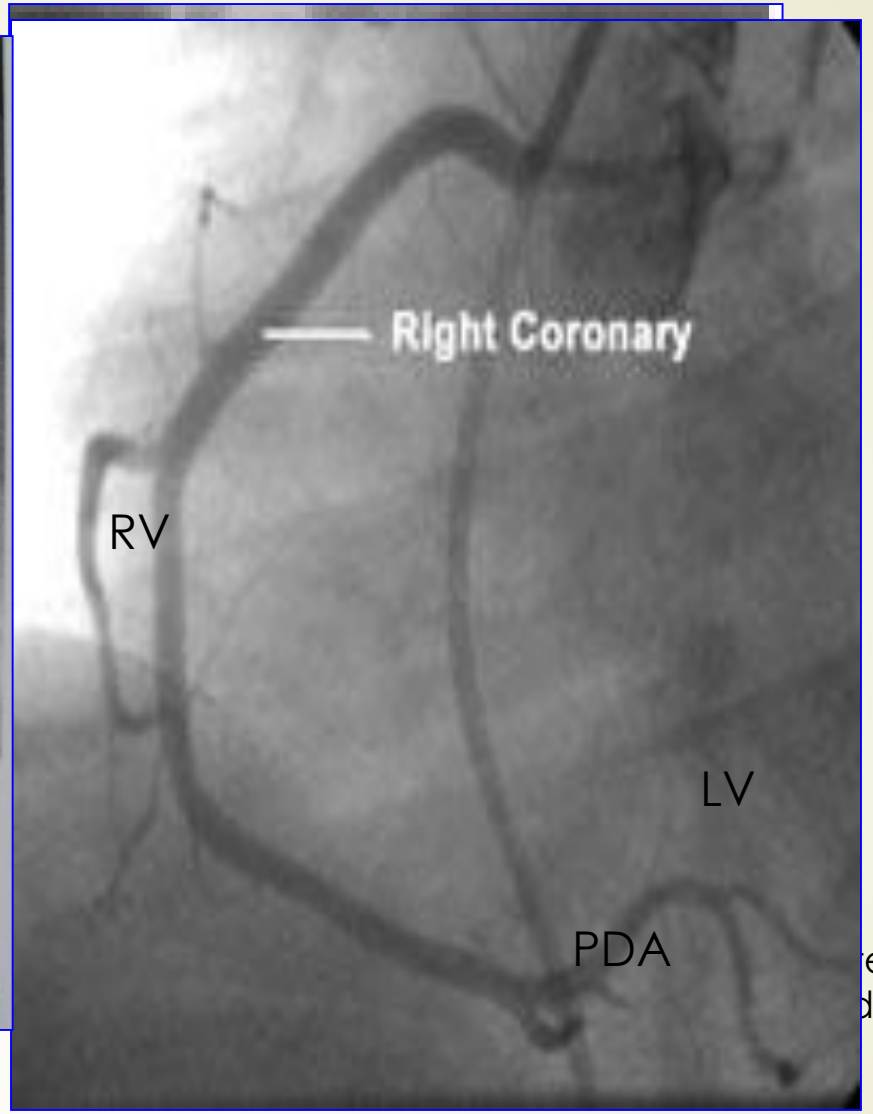
Angiocardiography cont’d

- Equipment- fluoroscopy unit on a C-arm with DSA facilities, pressure recording device, ECG monitor, blood oxygen analyser, catheters (pigtail, NIH), cournand catheter-for pressure measurements
- Angled views which place the pathological lesion at right angles to the Xray beam increase diagnostic accuracy of cardiography
- Complications; sudden death, arrhythmias, MI, C. Media s/effects



arrow=circumflex

RAO=I-intermediate branch,S-septal,D-diagonal
LAD-lt ant.desced,OM-obtuse marginal,Cx-circumflex



erior
de

white arrow =posterolateral left vent. branch

LAO =RV-rt vent. Branch,PDA-post. desced. artery,
LV-inferior lt vent branch

CARDIAC MRI

- Advantages; Provides anatomy, dynamic motion studies of function, flow and chemistry with minimal risk to the patient

- Disadvantage – expensive

  -availability

  - metallic implants

Clinical applications of Cardiac MRI

- ventricular function analysis
- Myocardial function at rest and during stress
- Myocardial perfusion
- Measurement of coronary flow
- Valvular heart disease
- Pericardial pathology
- Cardiac tumors and thrombi
- Cardiomyopathies
- Assessing great vessels- aortic/pulmonary disease

Pulse sequences used

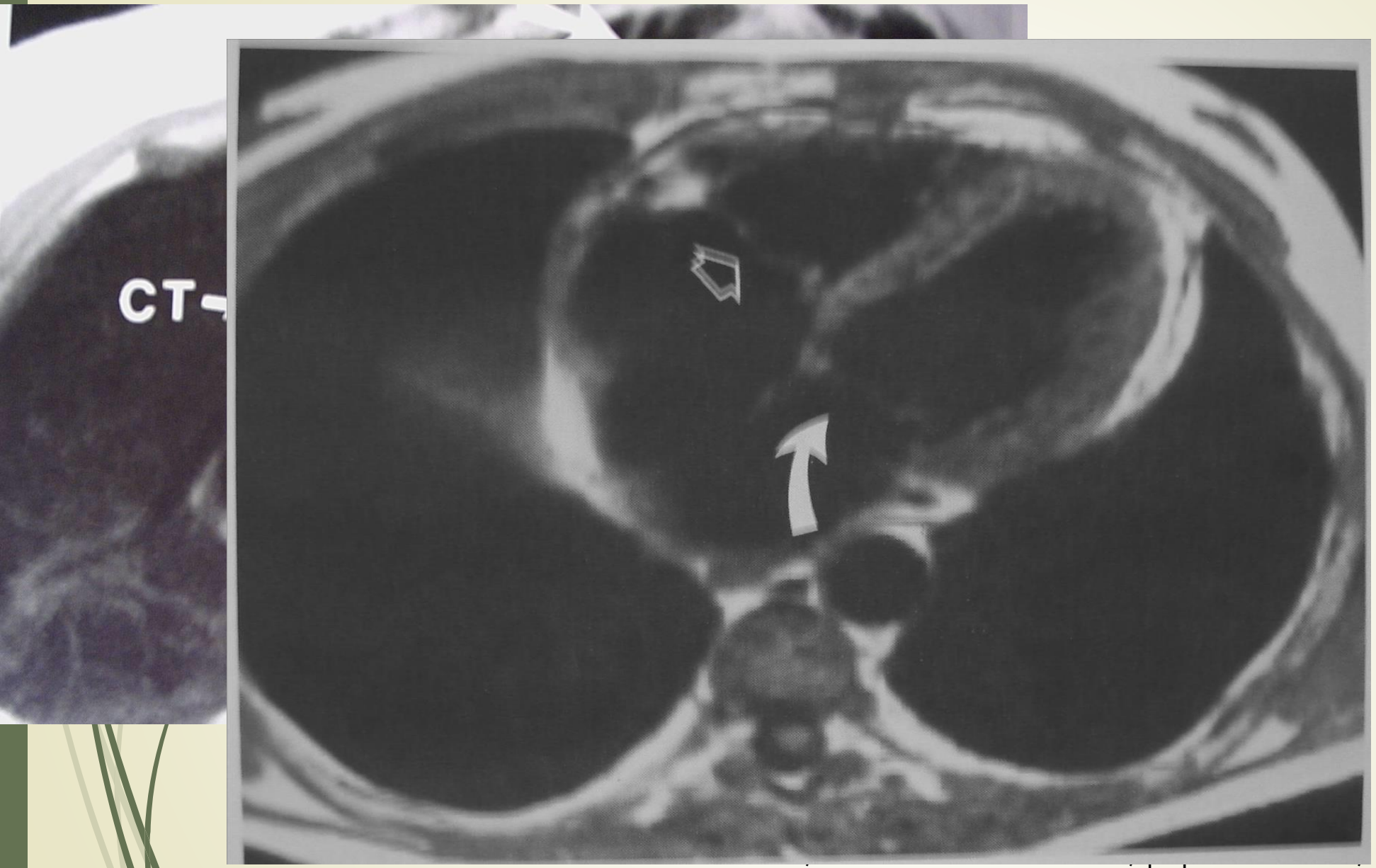
- Tissue characterization-done usingT1W, T2W, contrast enhancement and spectroscopy
- T1W-best anatomic depiction,
  - Blood-show signal void
- Gradient echo-Impart bright signal to coherently flowing blood-white blood
- Fast spin echo- vascular wall bright, blood black

C/MRI –cont'd

- Gradient recalled echo-technique -applied for motion studies,to show flowing blood as well as cardiac motion
- ECG gated MRI of the heart-
- Velocity mapping sequences
- Images-tomographic slices,of any selected plane

MRI Cont'd

- Gadolinium -as contrast used in;
  - myocardial perfusion
  - MR angiography; time of flight MRA, phase contrast MRA, contrast enhanced MRA
  - Myocardial infarction
  - Ischemia



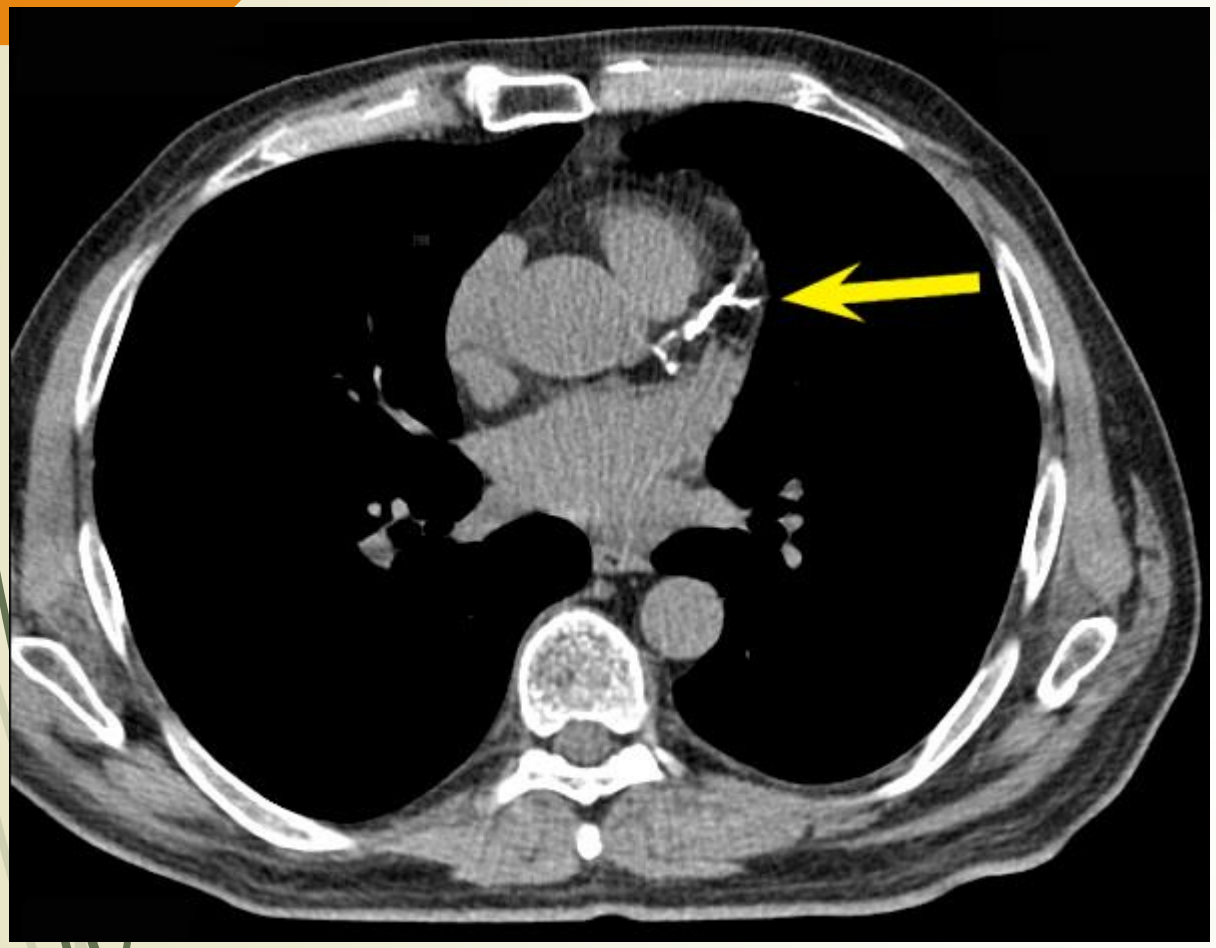
AV-aortic valve

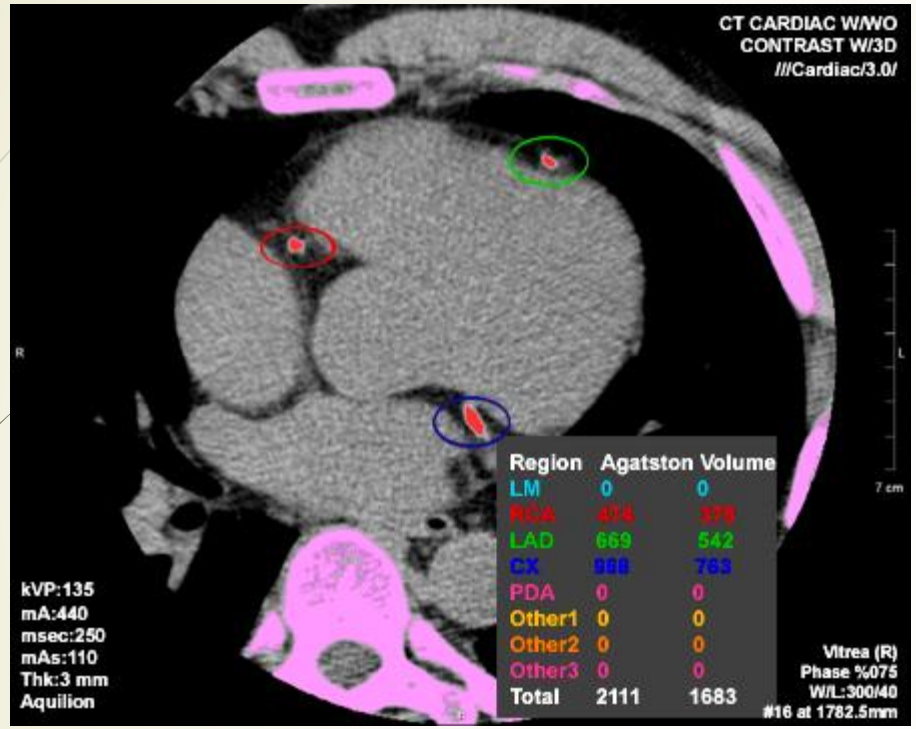
TRANSVERSE IMAGE-through ventricles at AV Valve level

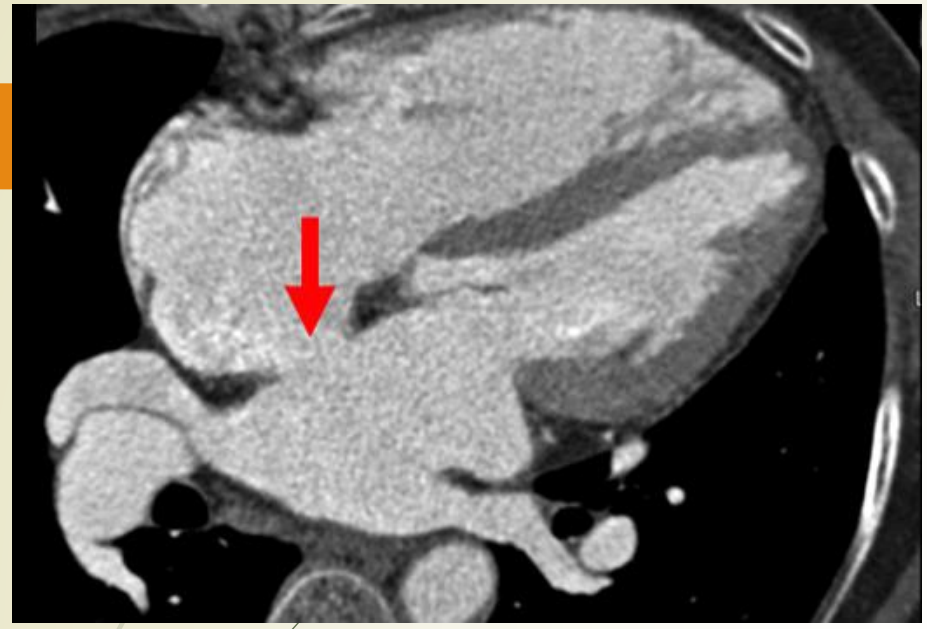
TV-open arrow; MV—curved arrow

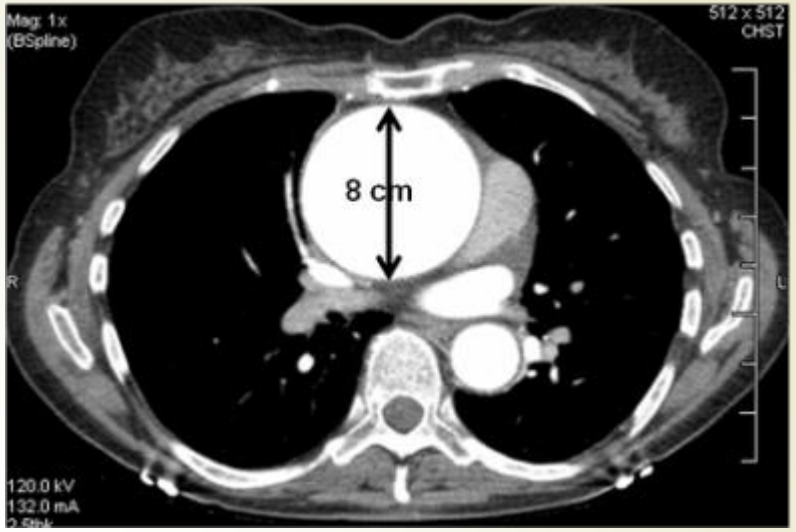
CARDIAC CT

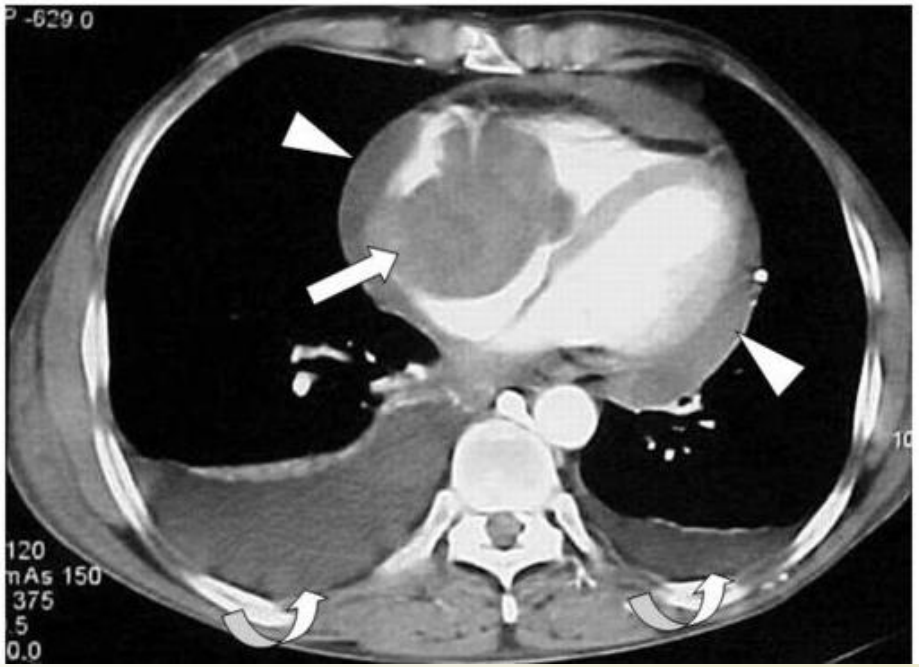
- Good images of thoracic anatomy and major cardiac structures and large vessels
- CECT first line in imaging aortic rupture or dissection
- Calcification on the coronary vessels
- Evaluation of coronary vessels
- Disadvantages; ionizing radiation, motion artifact

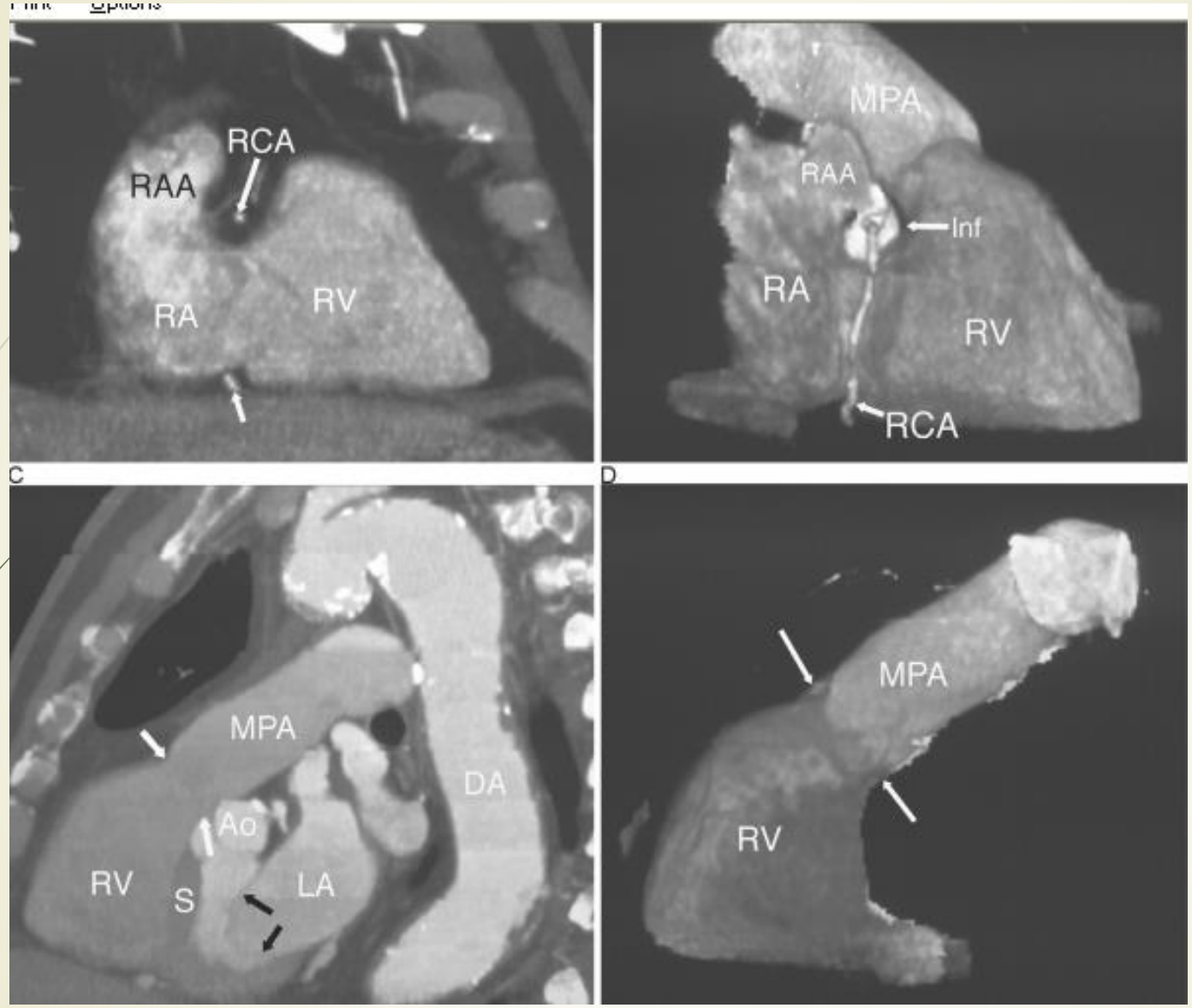



CT CARDIAC W/WO
CONTRAST W/3D
///Cardiac/3.0/
Region Agatston Volume
LM 0 0
RCA 474 378
LAD 669 542
CX 968 763
PDA 0 0
Other1 0 0
Other2 0 0
Other3 0 0
Total 2111 1683
kVP:135
mA:440
msec:250
mAs:110
Thk:3 mm
Aquilion
Vitrea (R)
Phase %075
W/L:300/40
#16 at 1782.5mm
7 cm




Mag: 1x
(BSpline)
512 x 512
CHST
8 cm
120.0 kV
132.0 mA


-629.0
120
mAs 150
375
.5
0.0



Coronal 3D reconstruction of right heart chambers using MDCTA.RAO views of right heart shown by MIP
D-volume rendered technique-RCA running in the low attenuation fat of the ant. Atrioventricular groove
E&F-Sagittal 3D Reconstruction of right chambers using MDCTA by (E) MIP and (F) VRT showing

CARDIAC SCINTIGRAPHY

- Injection of radiophamarceutical to study the heart
- indications;
  - assessment of myocardial perfusion
  - ischaemia,
  - infarction
  - viability.
  - Ventricular function
  - diagnosis and quantification of abnormal passages

Perfusion studies

- Used to diagnose ischemia and m/infarction
- Radiopharmacuticals used include;
- Thallium 201
- technetium 99m labeled tracers;like tetrofosmin

Technique

- scans are taken in;stress and resting phases

Stress induced by:-

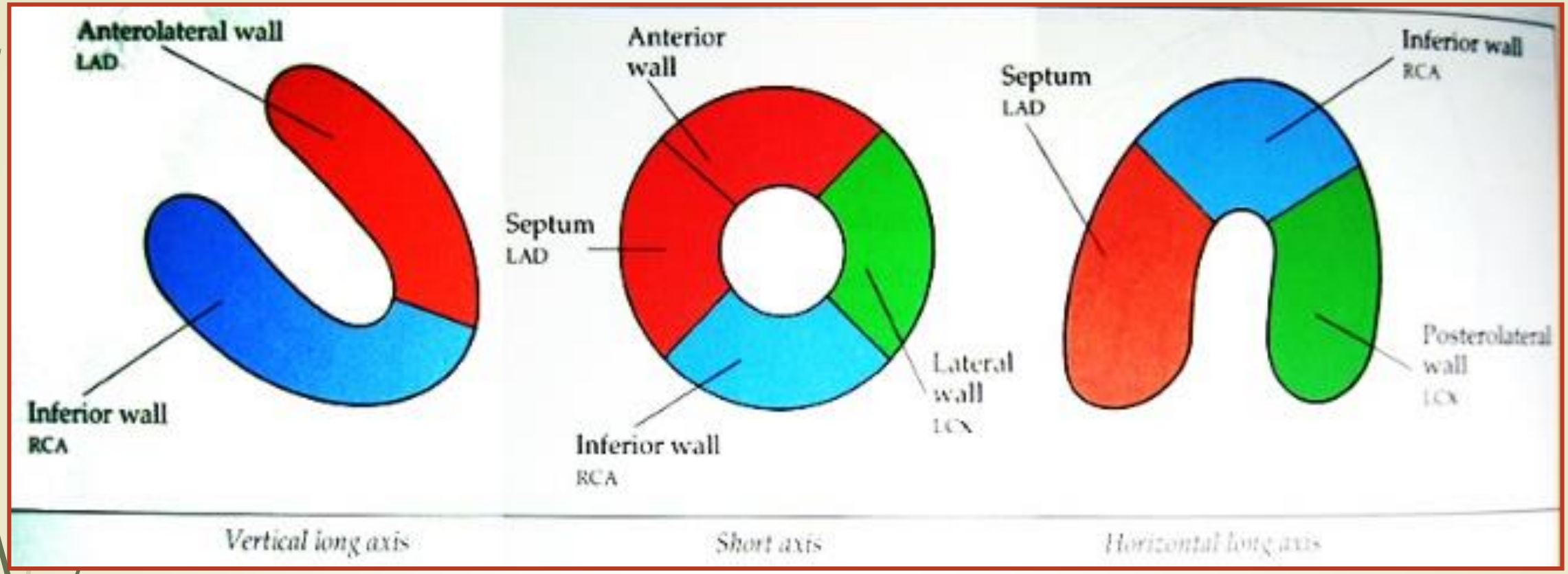
- Physical exercise test: treadmill, bicycling.
- Pharmacologic stress test: dipyridamole, adenosine, dobutamine.

- The heart scanned in longitudinal, vertical and short axes

Normal perfusion scans

- Horse-shoe in vertical and long.axes and doughnut in short axis
- stress and rest images are same in normal myocardial perfusion
- Ischaemia-normal at rest and reduced at stress
- infarcted and scars -reduced in both

Axes

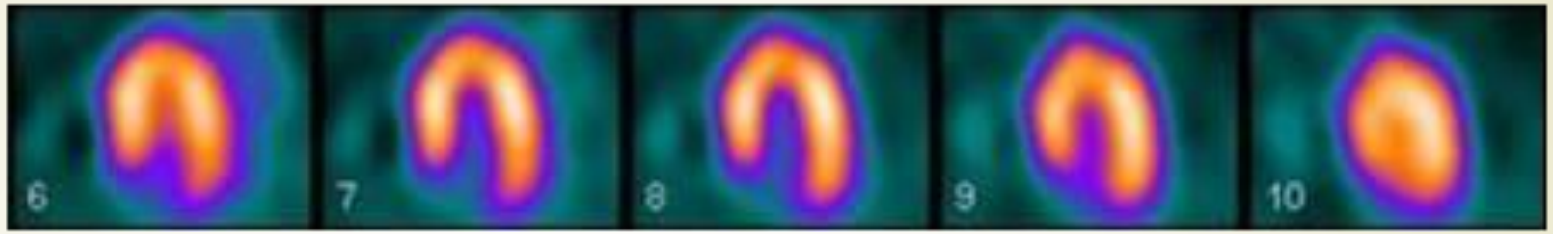


VERTICAL LONG AXIS

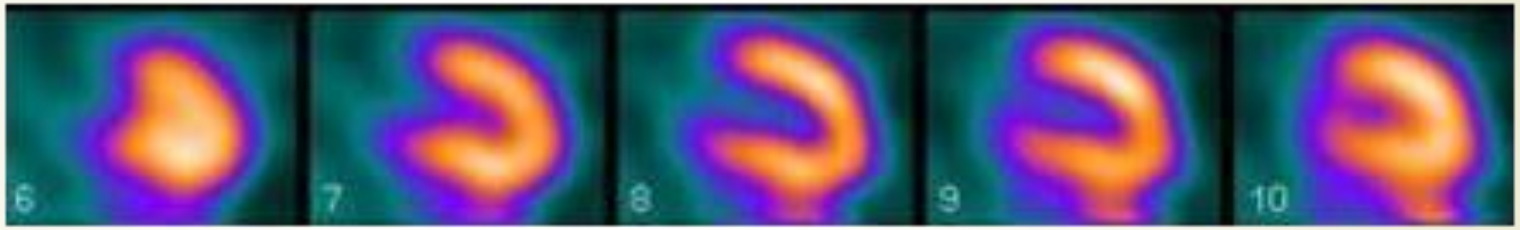
SHORT AXIS

HORIZONTAL LONG AXIS

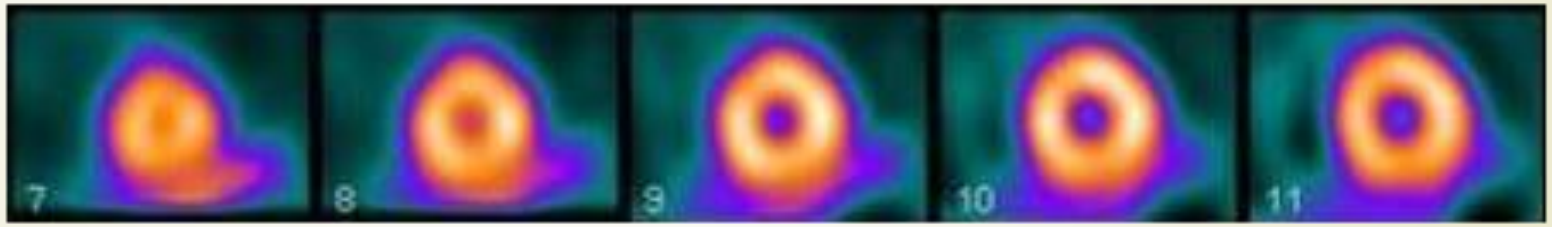
HEART AXES



Horizontal long axis view



Vertical long axis view



Vertical short axis view

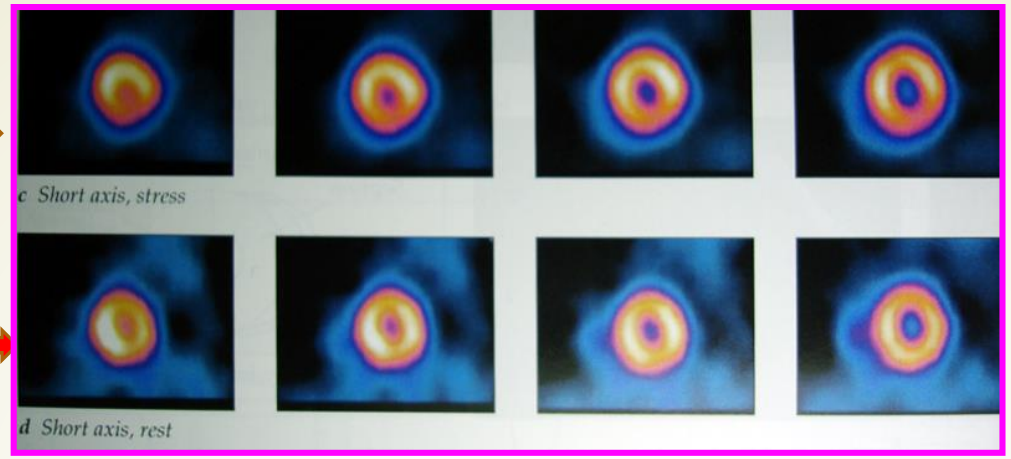
Normal perfusion scan showing cardiac slice information and horizontal long, vertical long and vertical short axis images.

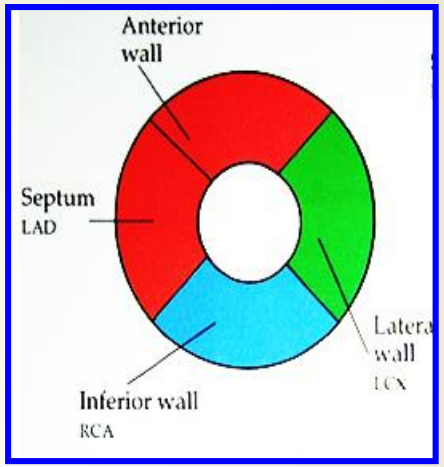
Normal examination

Short axis

-stress

-rest

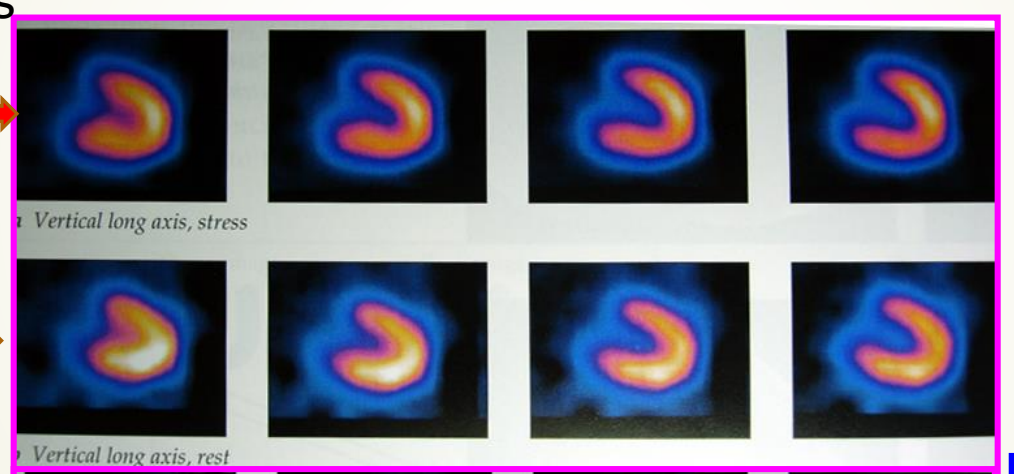


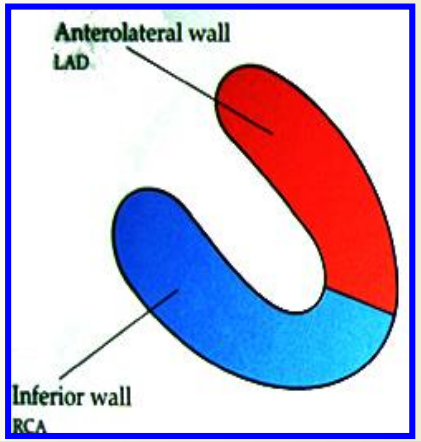


Vertical long axis

-stress

-rest

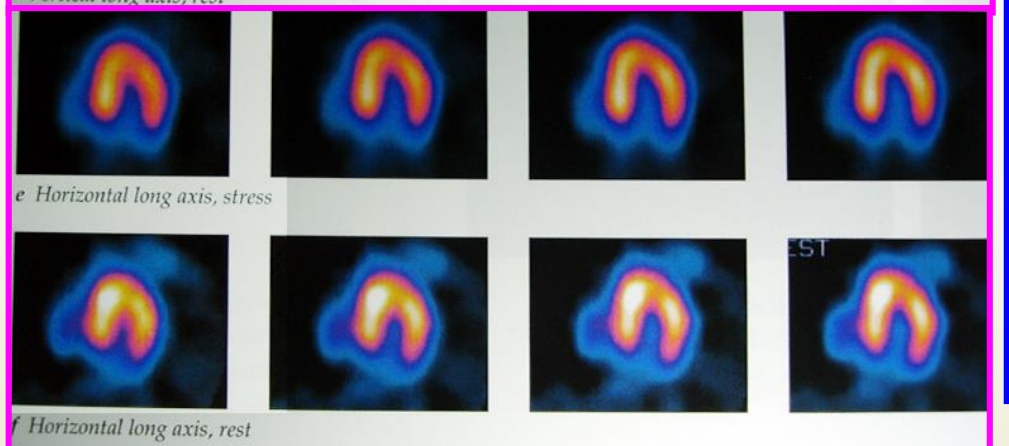


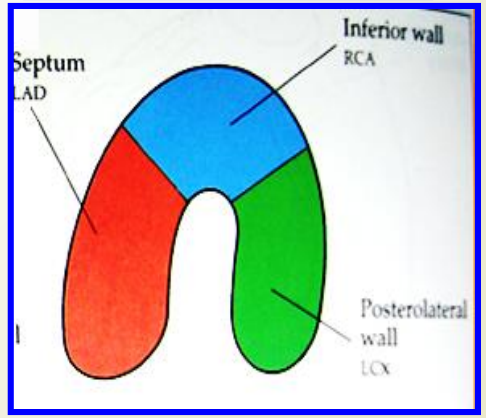


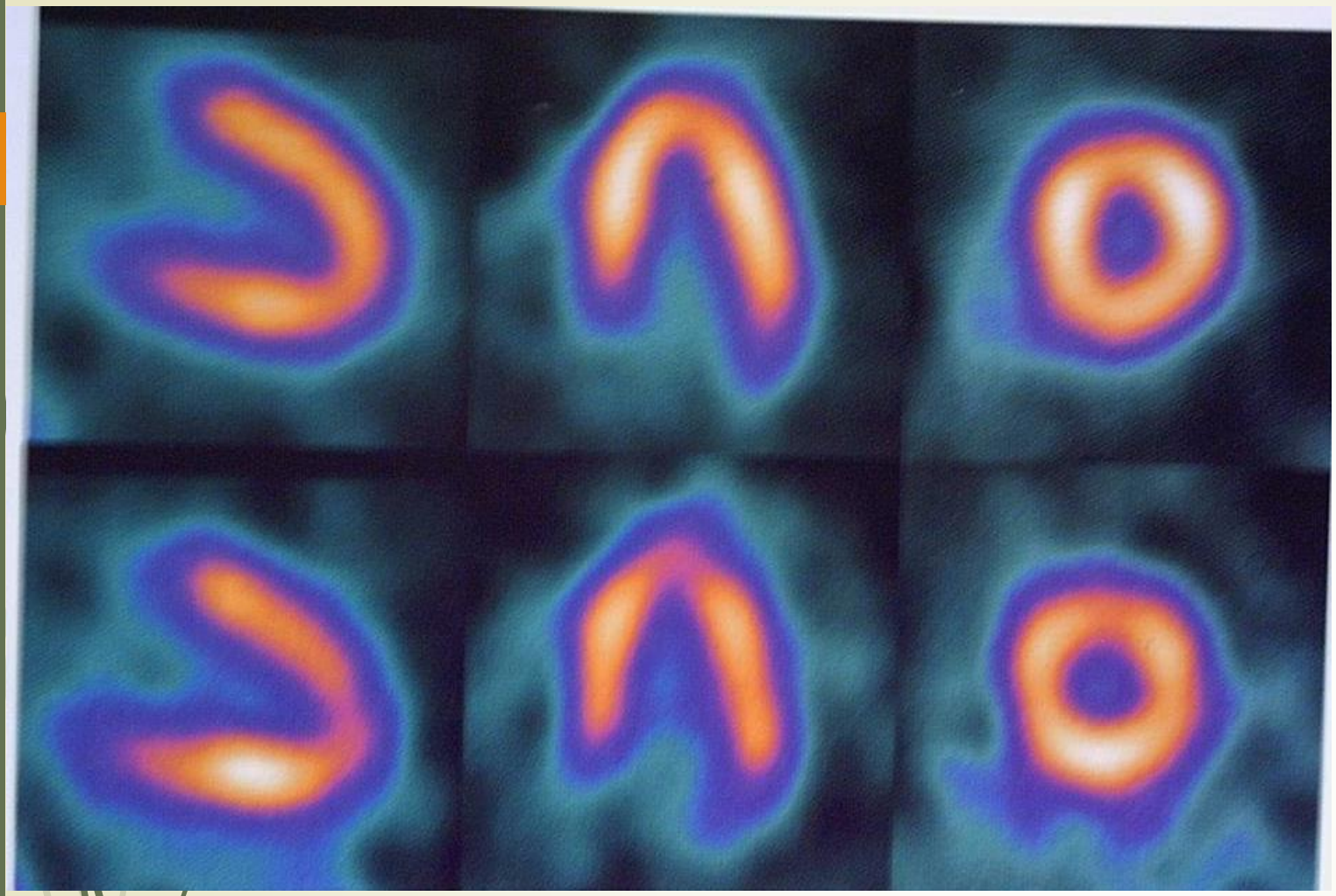
Horizontal long axis

-stress

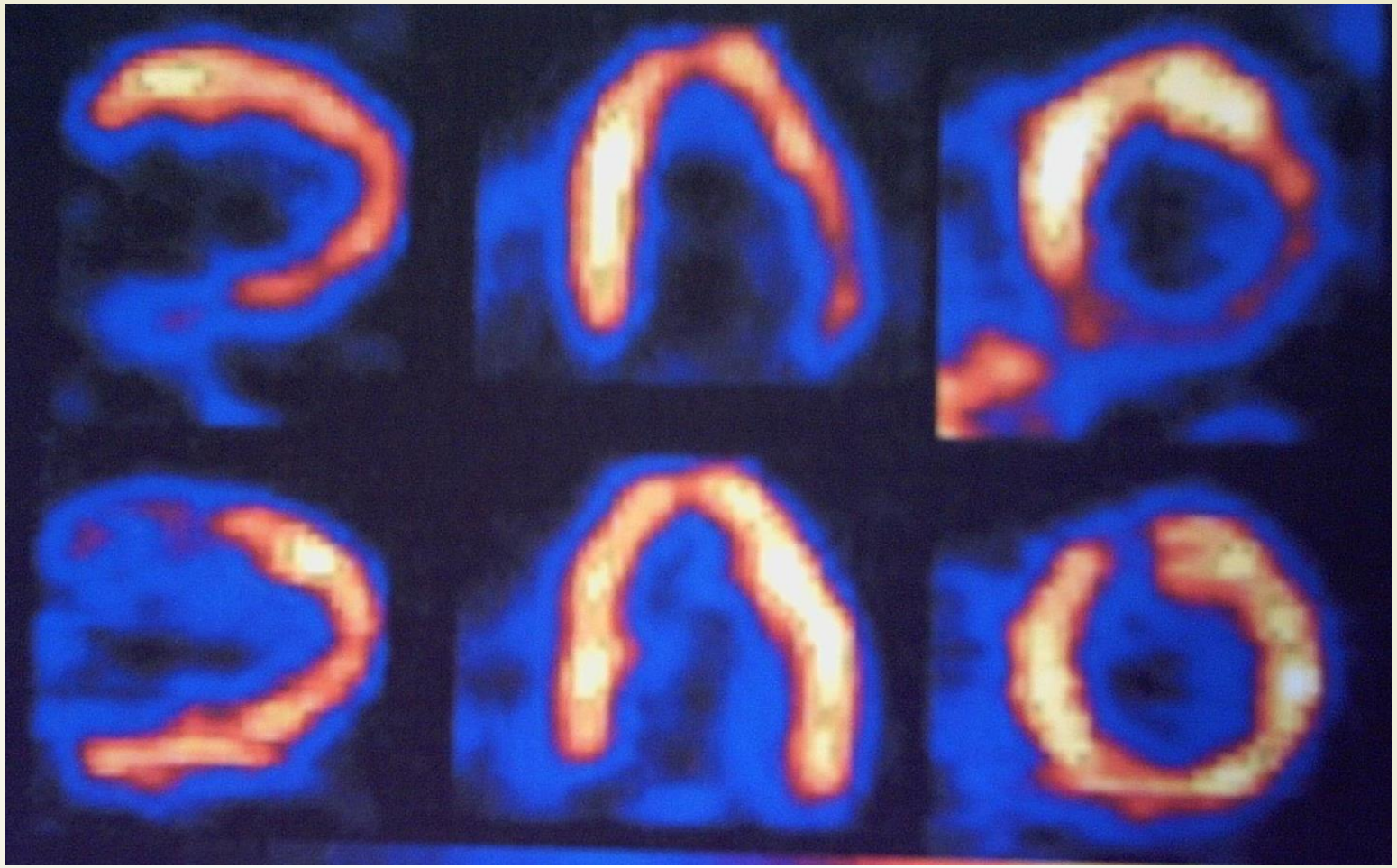
-rest







Normal Thallium myocardial perfusion tomograms in vertical long axis (left), horizontal
long axis (centre), and short axis planes (right). Stress images (top) and redistribution
(bottom). There is homogenous uptake of tracer throughout the myocardium

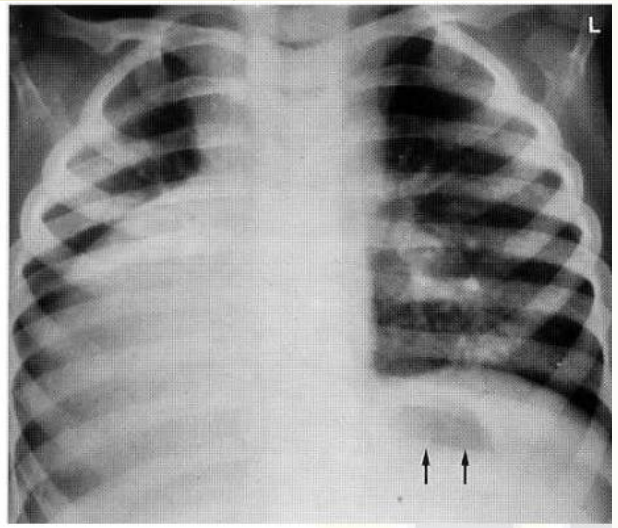


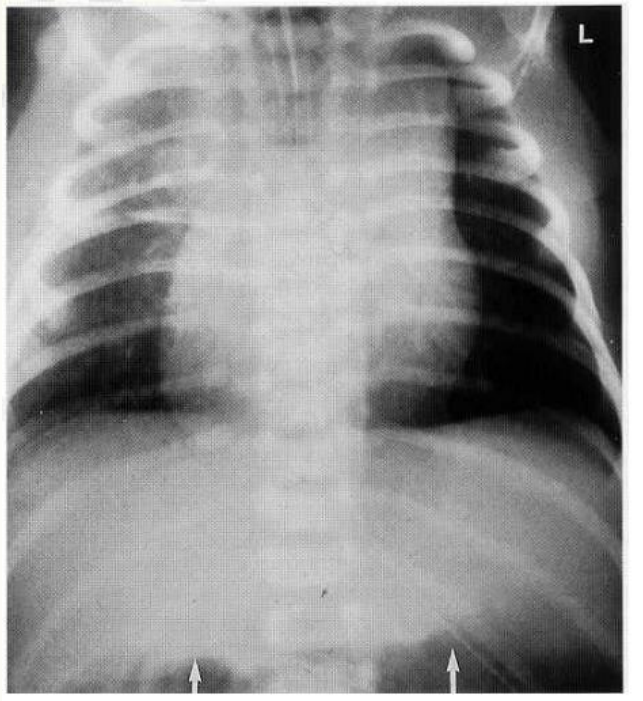
Hibernating myocardium; vertical long axis-(left), horizontal long axis (centre),and short axis(right) PET images. Ammonia perfusion images (top) show decreased uptake in the inferior and basal lateral regions. But the glucose metabolic images (bottom) show increased metabolism in these areas. Typical of hibernating myocardium

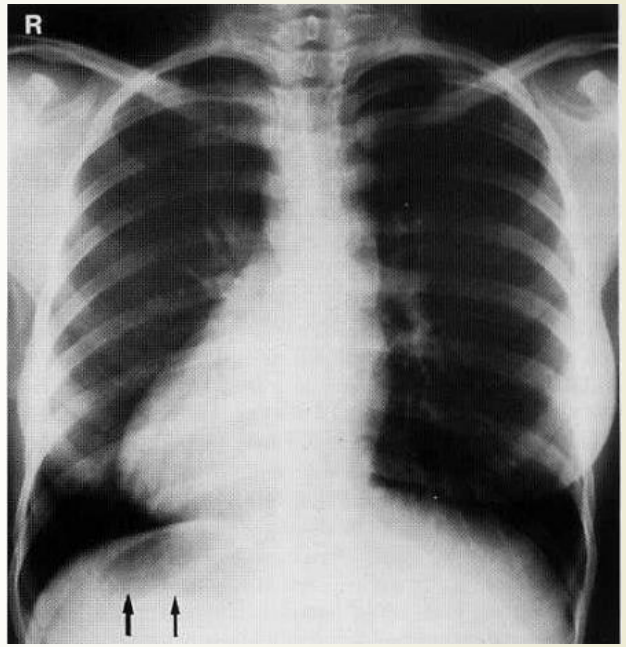
Clinical applications RNI

- Diagnosis of coronary heart disease
- Prognosis of CAD
- Myocardial infarction; infarct detection
- Myocardial viability and hibernation
- Other myocardial disease; alcoholic induced, viral, metabolic
- Valve disease
- arrhythmias

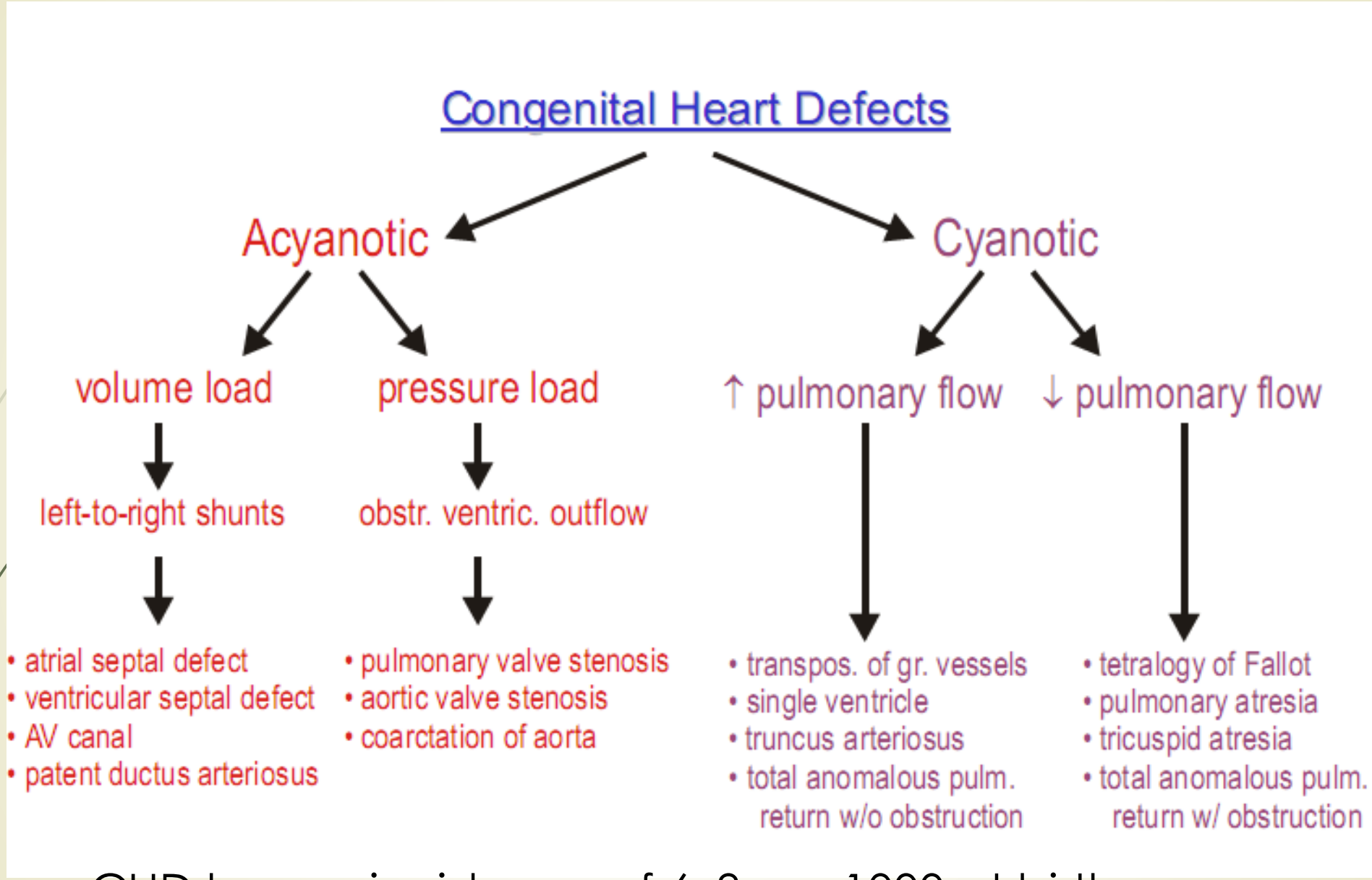
CHD







- Visceroatrial situs solitus, isolated dextrocardia.
- Situs ambiguous- central cardiac apex, transverse liver.
- Situs inversus.

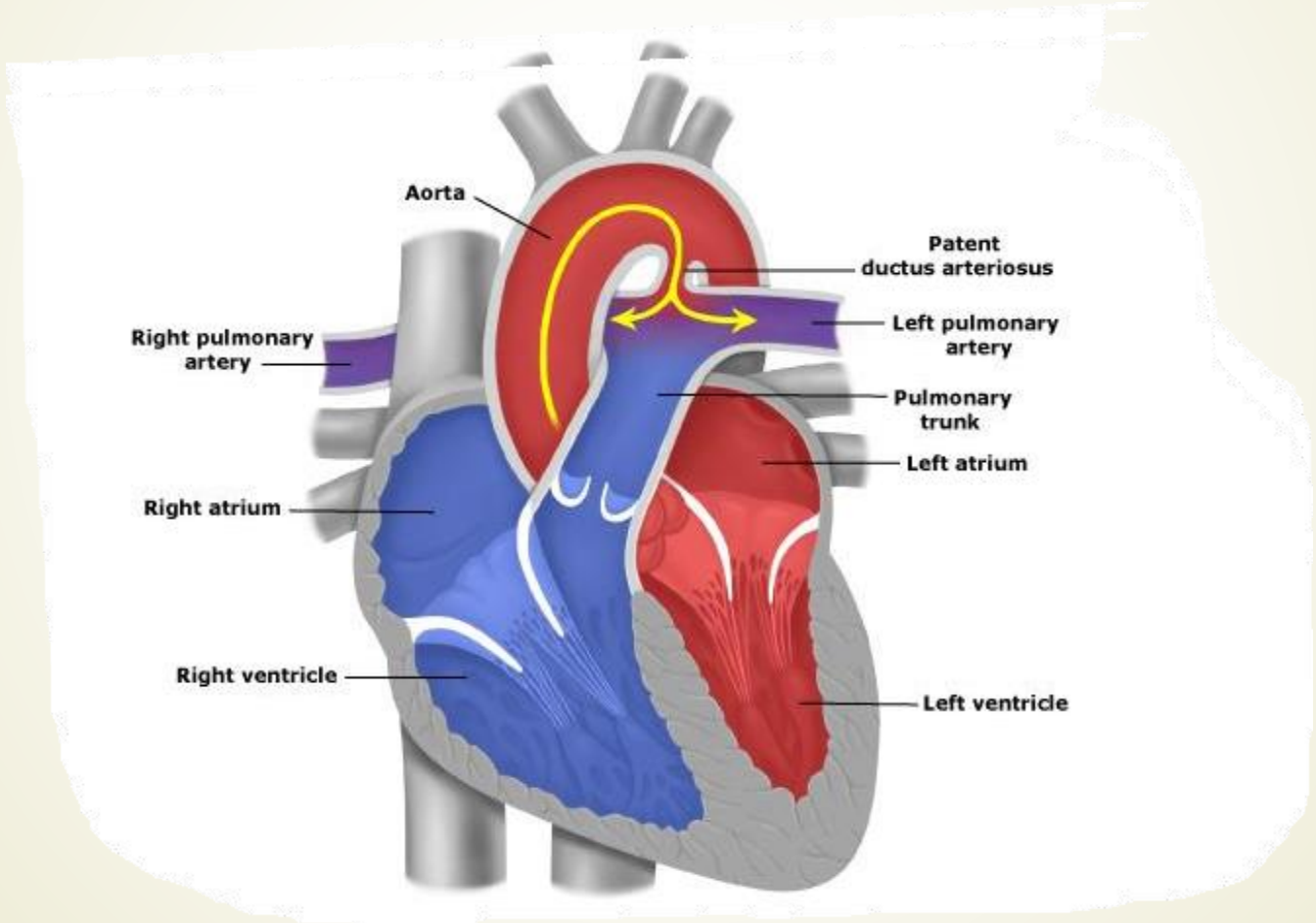


CHD has an incidence of 6–8 per 1000 at birth.

Risk Factors

- Genetic – Downs syndrome-25%
  - Holt-Oram Syndrome
  - Ellis van Creveld Syndrome
  - Turner's, Noonans
- Alcohol.
- Prenatal Rubella infection, Toxoplasmosis.
- Drugs: Thalidomide toxicity---17% have a cardiac lesion.
- Gestational diabetes.

Patent Ductus Arteriosus



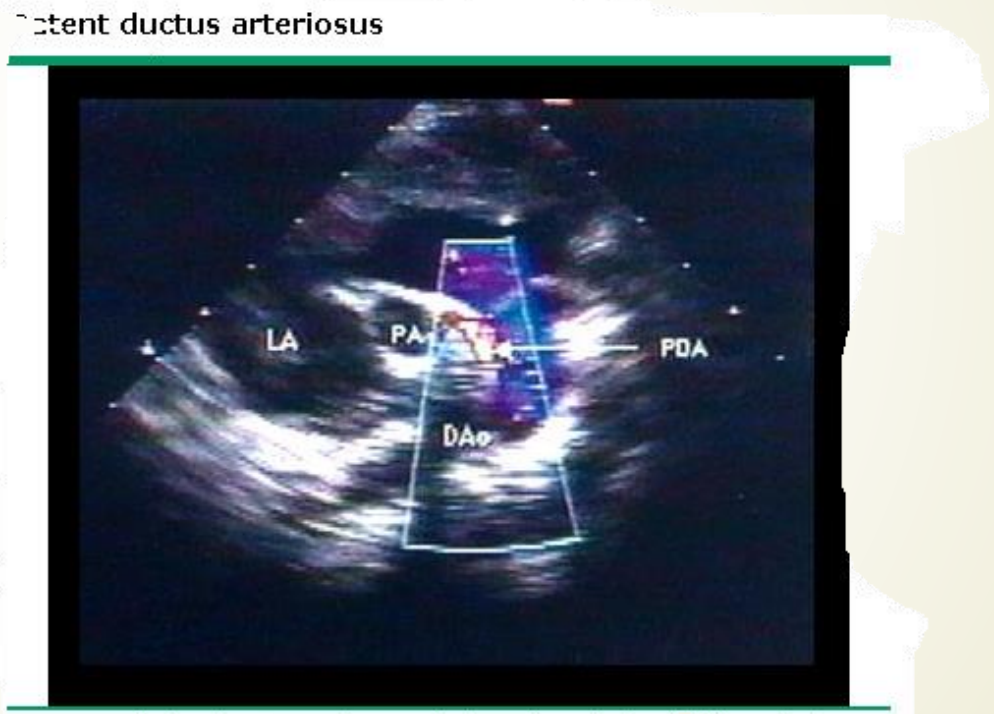
Patent Ductus Arteriosus

- Persistence beyond 10 days is considered abnormal.
- Accounts for 15% of all congenital heart diseases.
- Commoner in females (2-3:1)
- Associated with low birth weight, prematurity, high altitudes.

PDA CXR

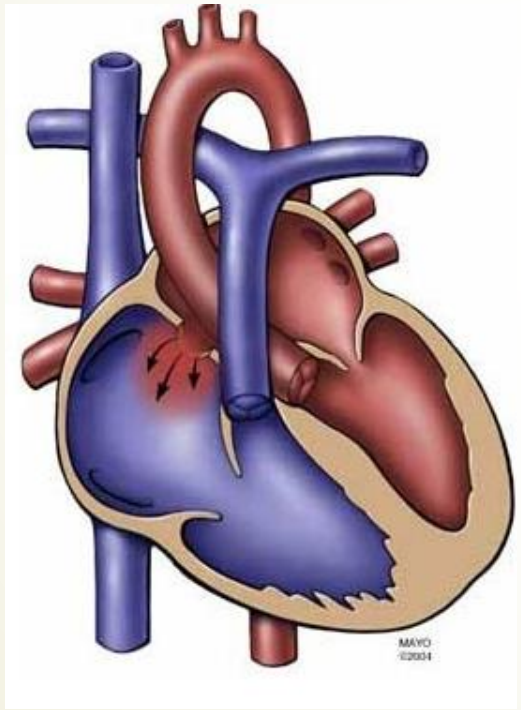
- Prominent central pulmonary arteries.
- Cardiomegally.
- Enlarged left atrial appendage.

2D + Color Doppler Echo

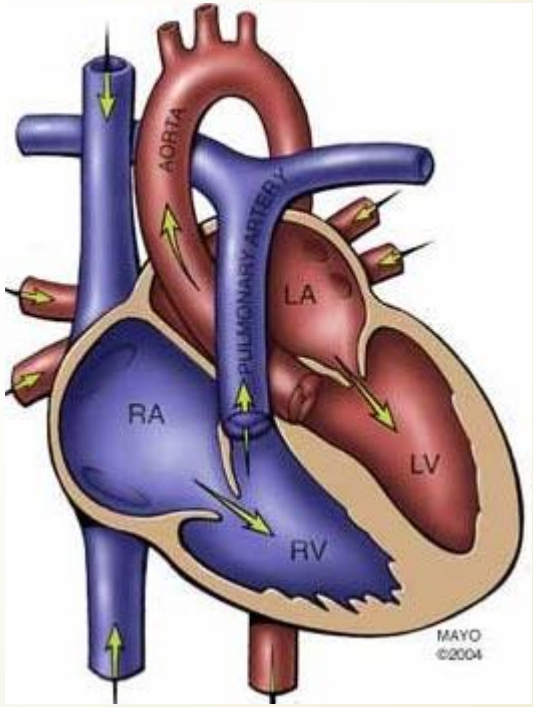


Secundum ASD- accounts for 80%

Defect @ center



Normal Heart for cf.



Secundum ASD

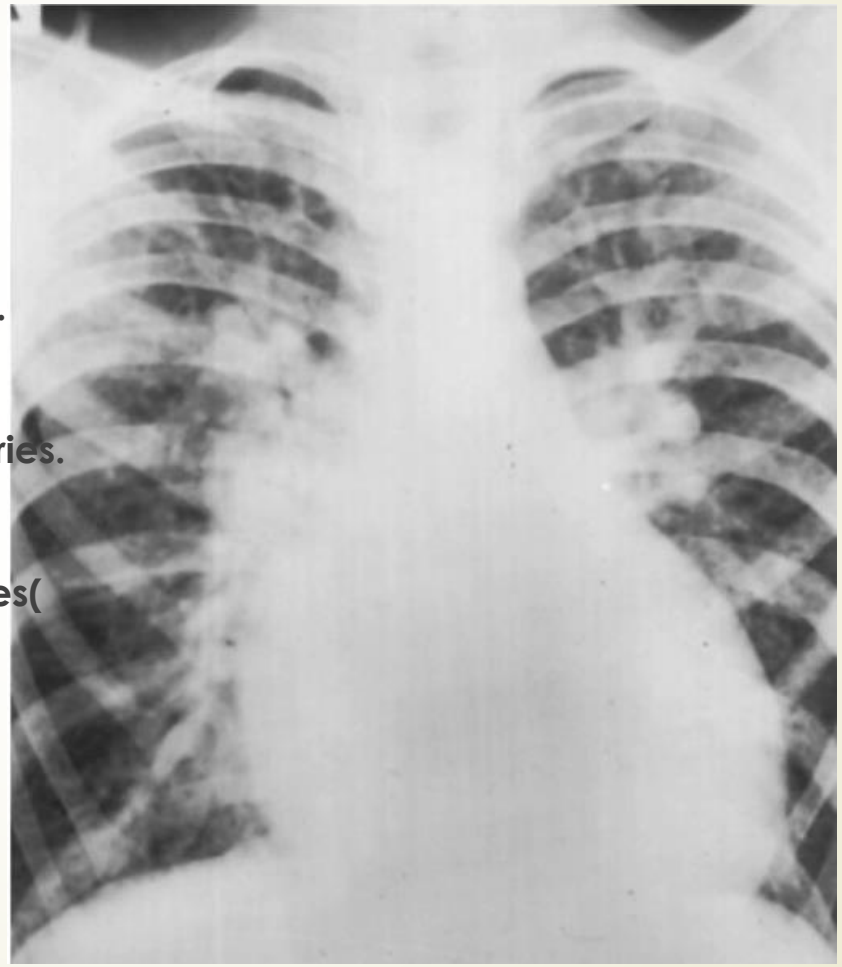
Apical 4 chamber view echo.

Note the enlarged right chambers.

ASD B4 Eisenmenger

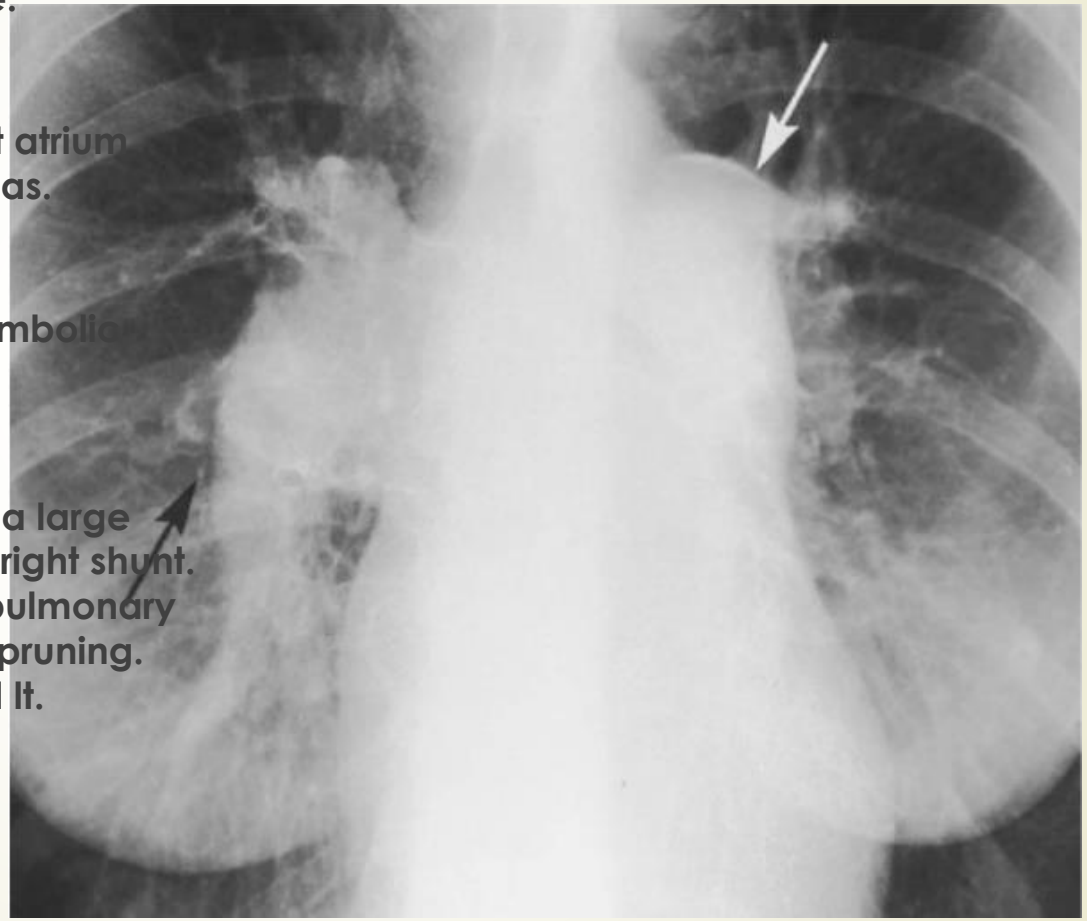
Note the pulmonary plethora:

- Large heart
- Large central pulmonary arteries.
- Large peripheral pulmonary arteries.
- Recruitment of upper zone arteries(Cephalization)
- No pulmonary edema.
- Patient had ASD.



Complications of ASDs.

1. Eisenmenger syndrome.
2. Enlargement of the right atrium causing tachyarrhythmias.
3. Paradoxical thrombo-embolic stroke.
4. This patient initially had a large ASD with a major left to right shunt. Note the large central pulmonary arteries with peripheral pruning. Arrow points @ calcified lt. pulmonary artery.



VSDs.

- One of the commonest anomaly (second only to bicuspid aortic valves). Incidence 3570/1m live births.
- 50% of patients with VSD have associated cardiac anomaly e.g. coarctation of the aorta.
- 80% are perimembranous & the vast majority(75%) of small ones close spontaneously.

VSD

Lets us first understand the anatomy.

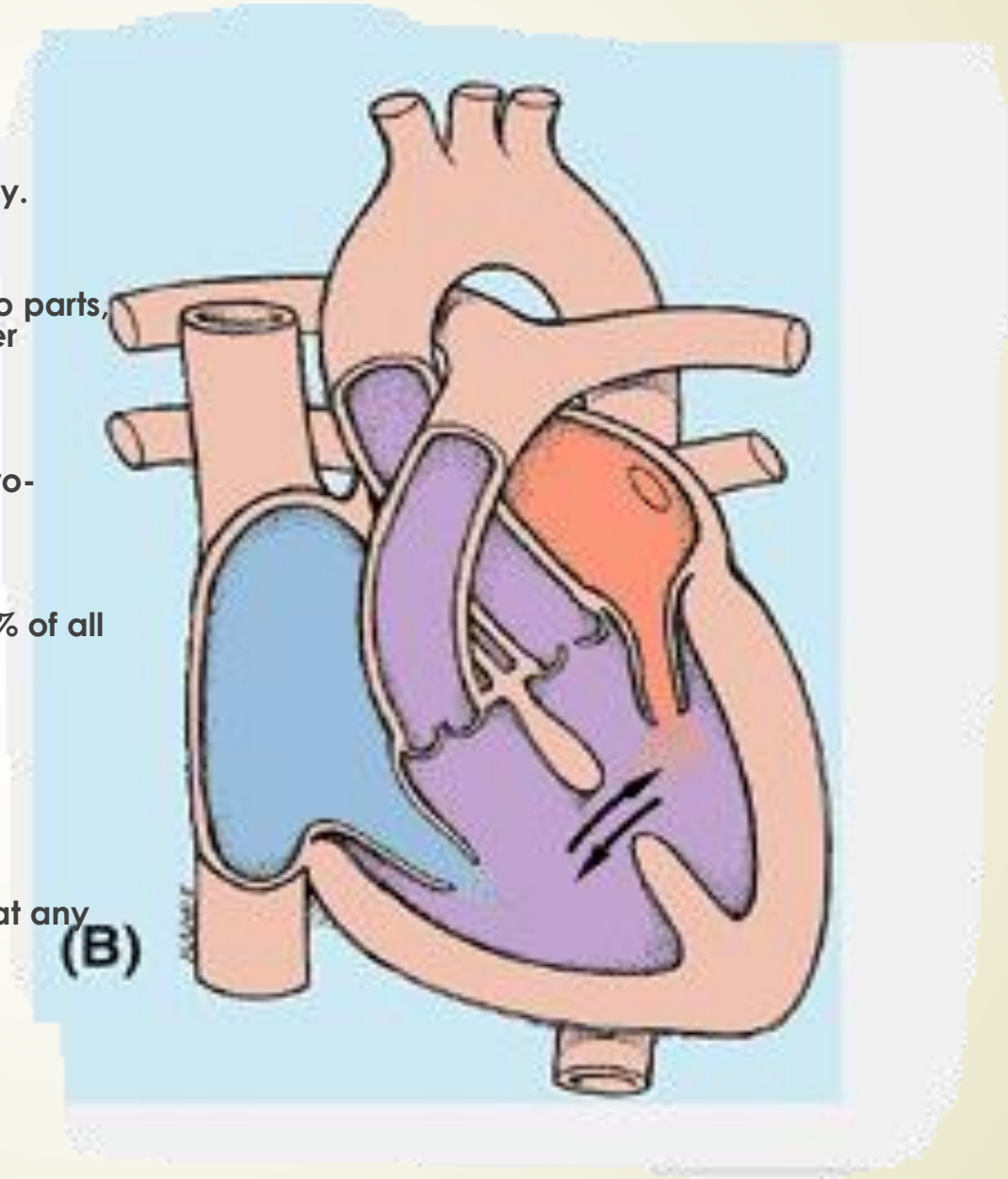
The Interventricular septum has two parts, the larger muscular and the smaller membranous part.

The membranous part is the postero-superior region.

Membranous VSDs account for 80% of all VSDs.

Muscular VSDs tend to close spontaneously.

Its possible to have multiple VSDs at any part of the interventricular septum.



Congenital Cyanotic heart disease

- For clinically detectable central cyanosis, there must be at least 5 g of reduced Hb per 100 ml of aortic blood. Occurs in 3 ways:

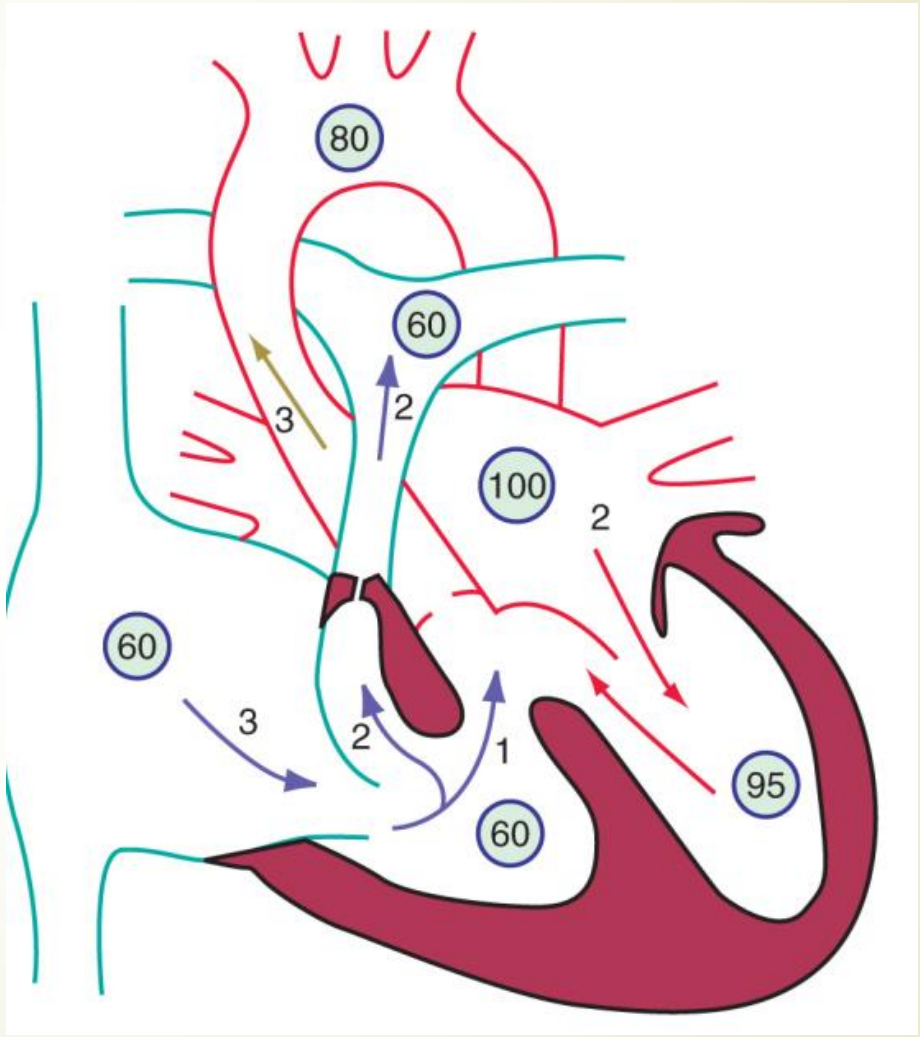
1. Direct right to left shunt eg. TOF
2. Transposition of great arteries.
3. Common chambers eg. Common atria, common ventricle, truncus arteriosus.

TOF

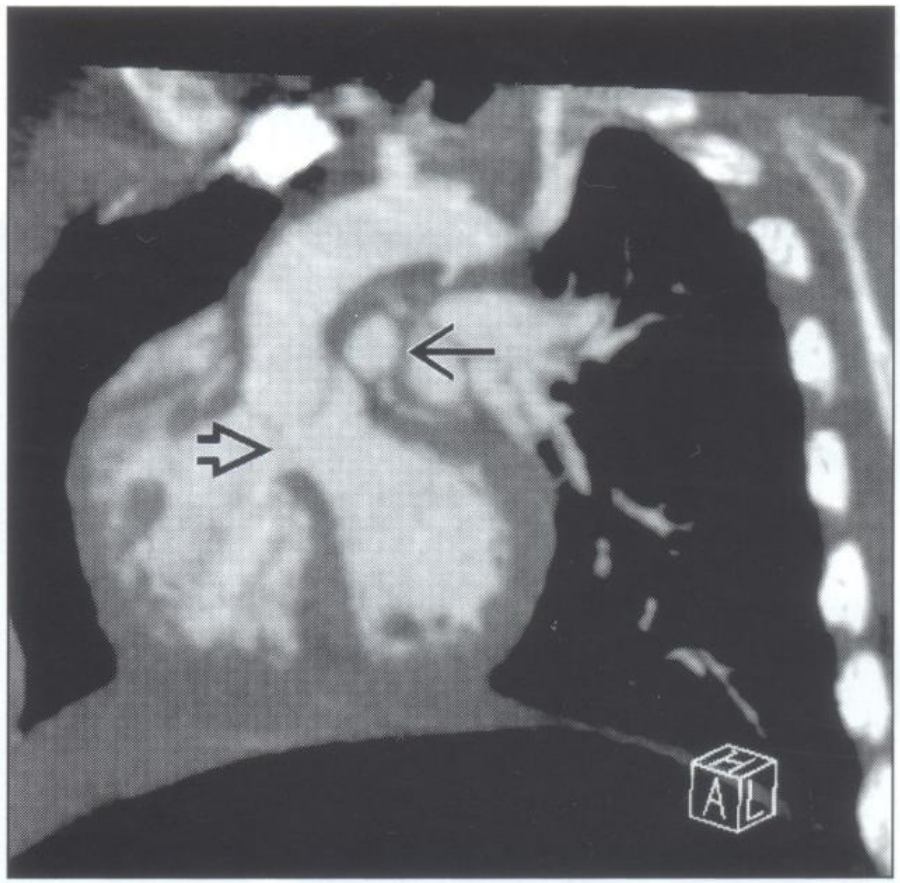
- Most common cyanotic heart lesion.
- Incidence: 3-5 per 10,000 live births.
- Primary hypoplasia of infundibular septum secondary to unequal partitioning of the conotruncus.
- Conotruncal septum displaced anteriorly, and its lowermost component, the Infundibular septum fails to fuse with the top of the IV septum.
- Features include:
- Infundibular right ventricular outflow tract stenosis
- subaortic VSD
- Over-riding aorta
- Right ventricular hypertrophy

TOF

- Desaturated blood enters RA, flows through RVOT into lungs. Some blood shunts right to left through VSD into ascending aorta.



TOF



Graphic- subvalvular(infundibular) stenosis, small pulm valve, large aortic valve, over-riding aorta, VSD, RVH, rt arch.

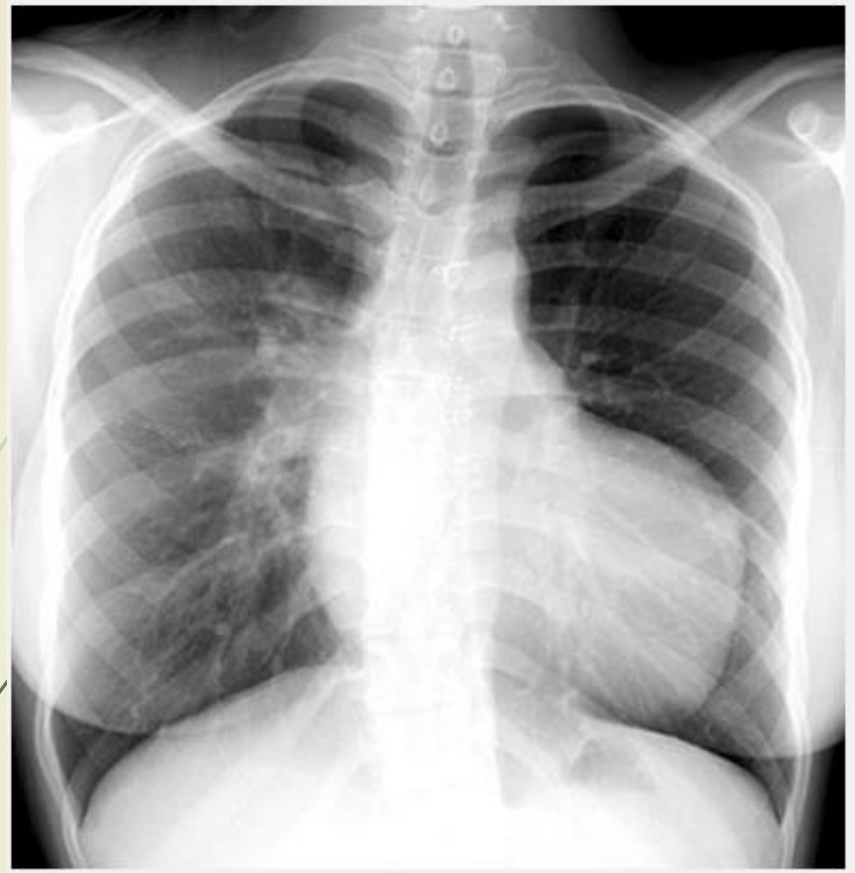
CTA cor- hypoplastic MPA, high VSD, over-riding aorta.

TOF

Radiography:

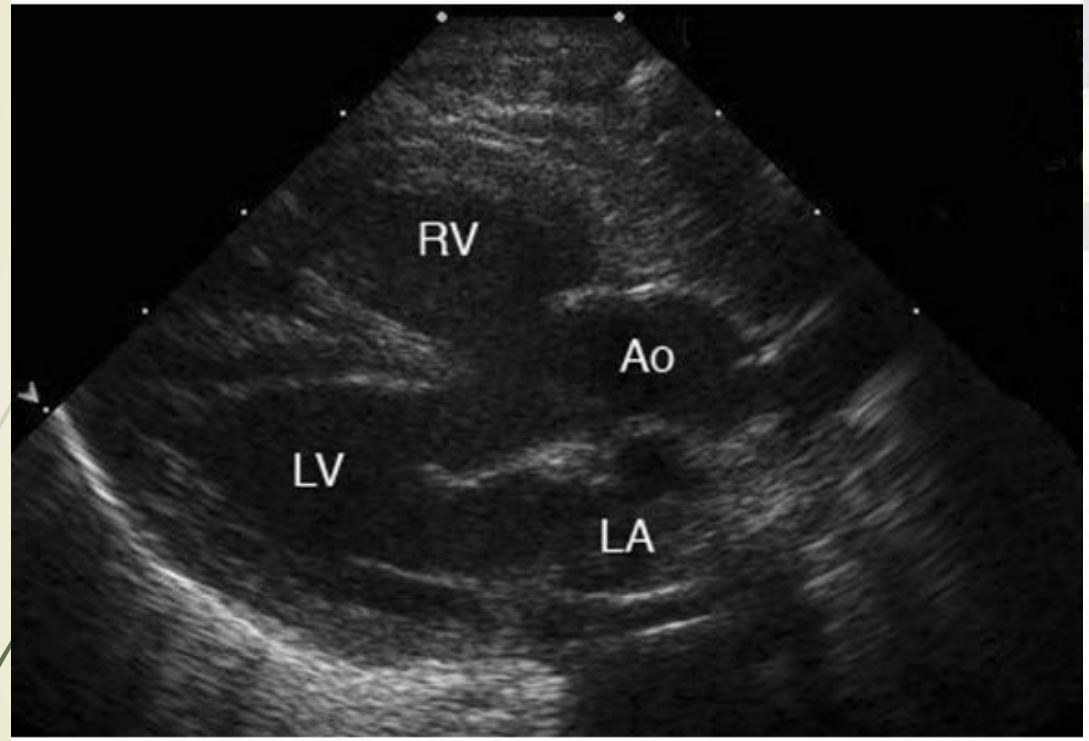
- “Boot-shaped heart” = “coeur en sabot”.
- Right ventricular hypertrophy.
- Concave pulmonary artery segment.
- Decreased pulmonary vascularity(oligaemia).
- Normal heart size at birth.
- Right aortic arch, mirror image branching (25%).

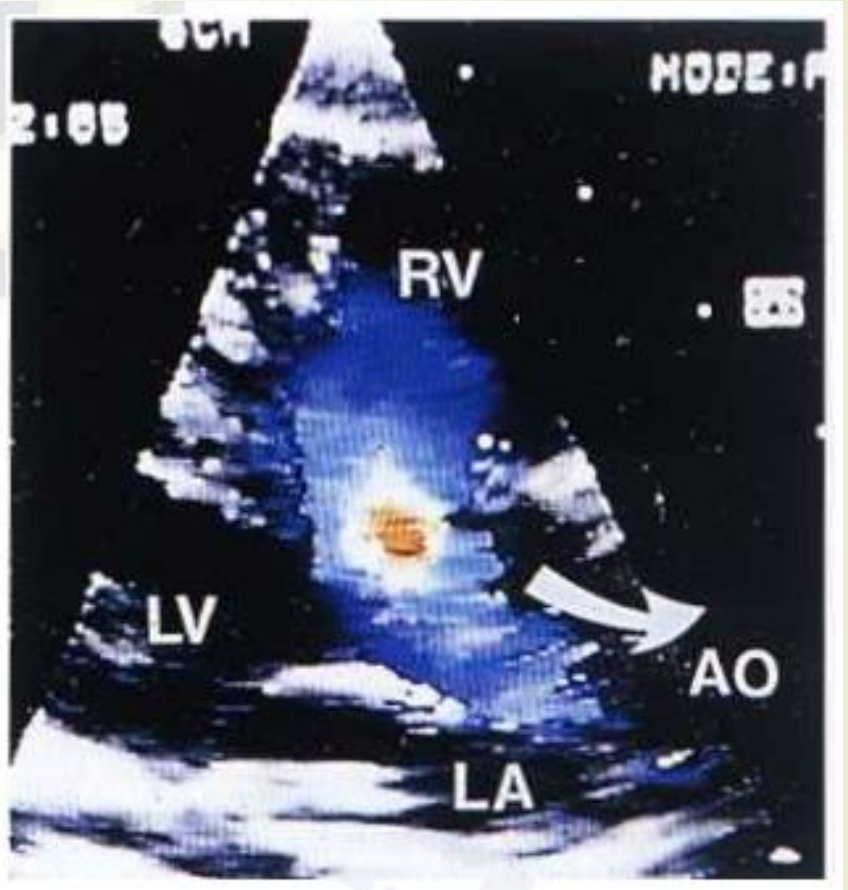
TOF



- boot shaped heart, concave pulmonary artery segment.
- Right aortic arch, elevated cardiac apex, pulmonary oligemia

TOF





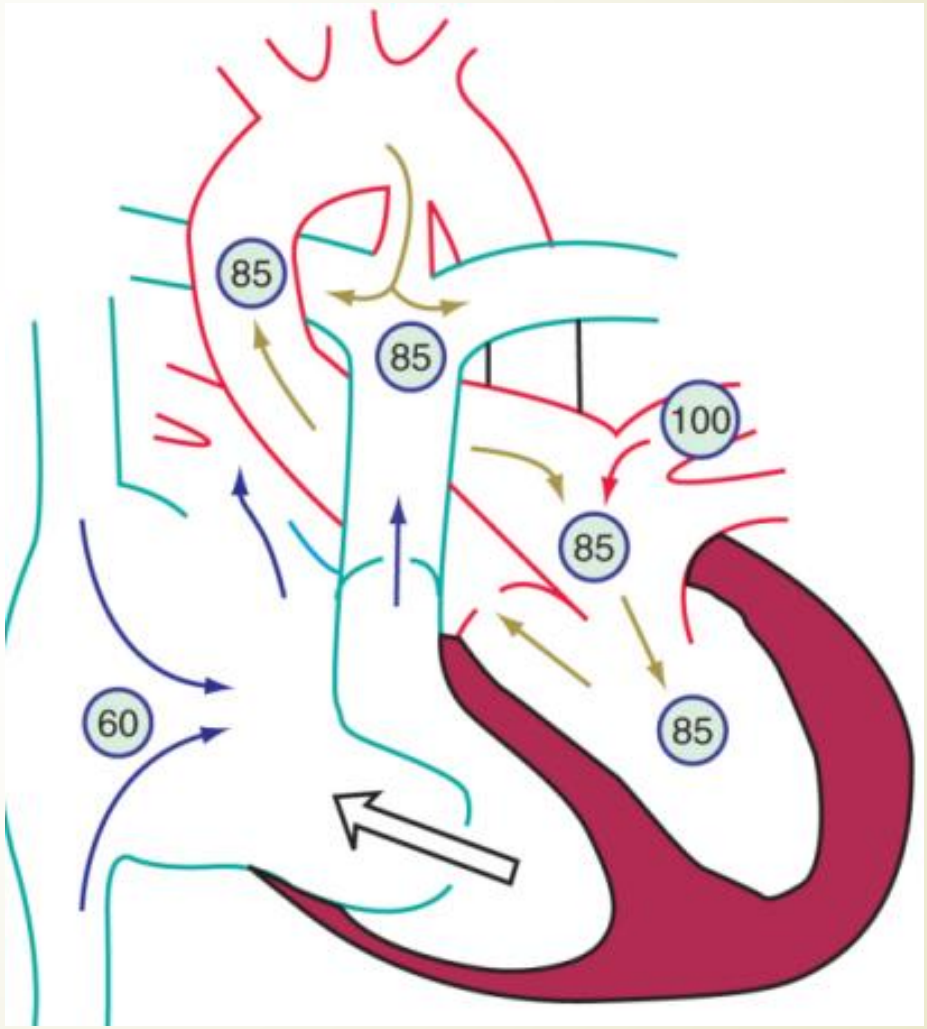
- Parasternal long axis- VSD, over-riding aorta,
- CF doppler- right to left flow from RV to aorta.

Ebstein anomaly

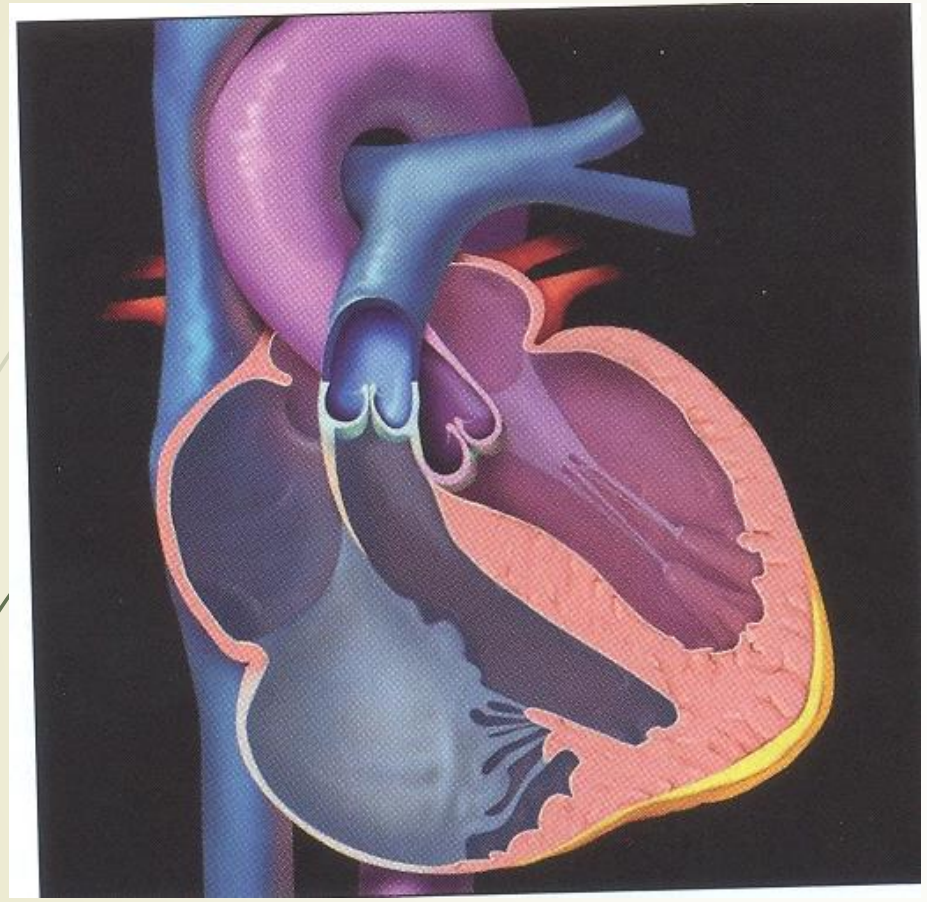
- Downward displacement of septal & posterior leaflets of tricuspid valve.
- Insufficient separation of tricuspid valve leaflets and chordae tendineae from Rt ventricular endocardium.
- Massive tricuspid regurgitation.
- Associated anomalies-PFO, ASD in 90%.
- Normal or decreased vascularity
- Rt. to Lt. shunt thru PFO causes cyanosis.

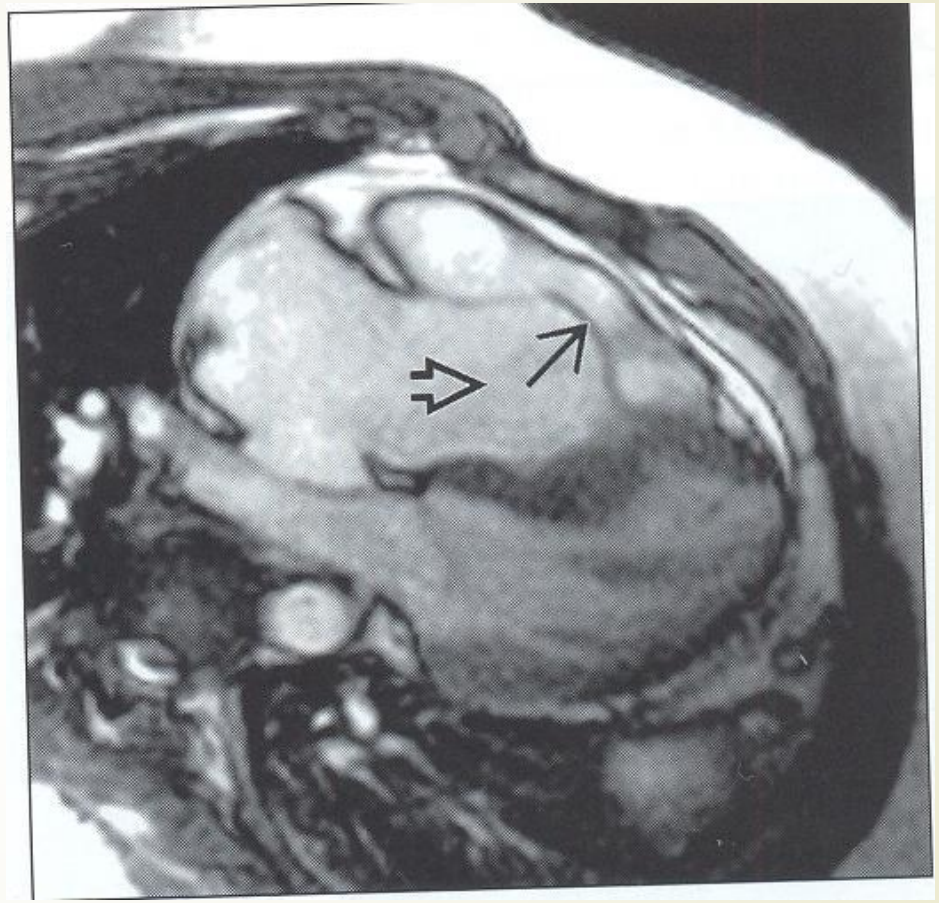
Ebstein anomaly

- Inferior displacement of tricuspid valve leaflets into RV.
- Thin-walled, low-pressure "atrialized" segment of RV.
- tricuspid valve is grossly insufficient.
- RA blood flow is shunted rt. to lt. across an ASD or PFO into LA. Some blood may cross the RVOT and enter PA.
- Severe cyanosis will



Ebstein anomaly





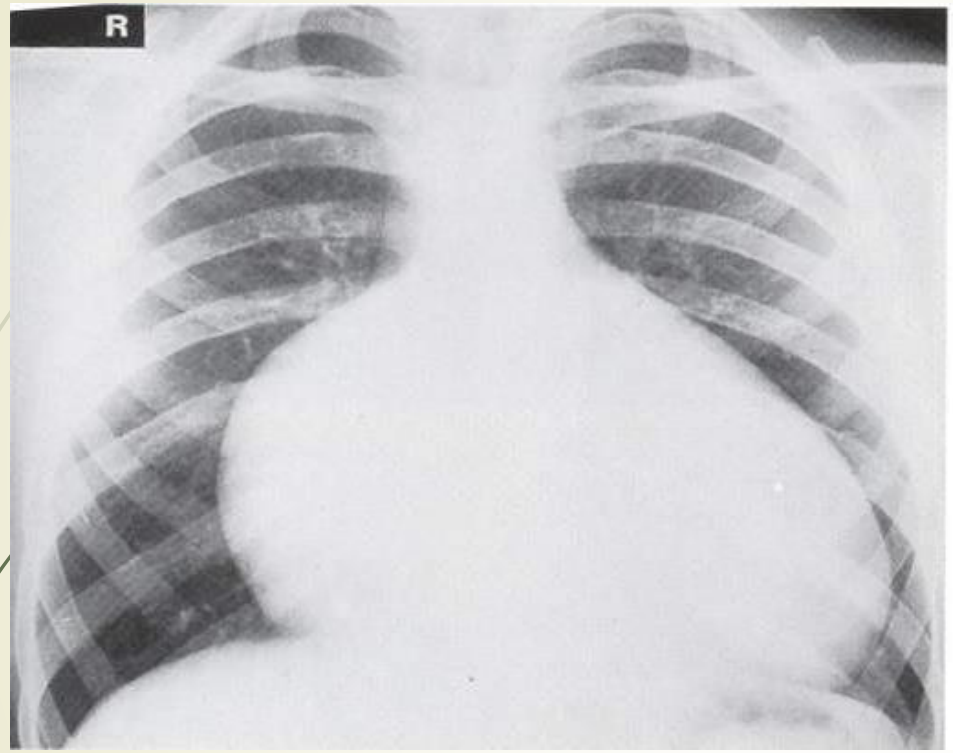
Graphic-Downward displacement of posterior valve leaflet, incorporated into RV.

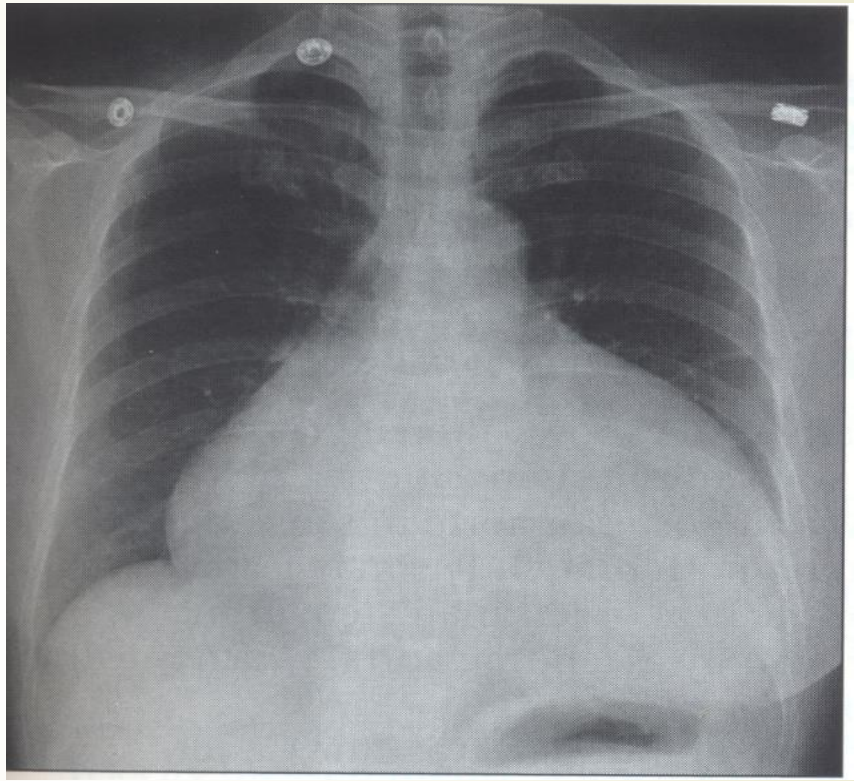
MR cine- Dilated RA, low placement of tricuspid valve, atrialized RV.

Ebstein anomaly

- Radiography: Severe right sided cardiomegally.
- Echo- Rt chamber enlargement, enlarged tricuspid annulus, tricuspid regurgitation, PFO with Rt. to Lt. shunting.
- Treatment- Tricuspid valve replacement and/ or valvuloplasty

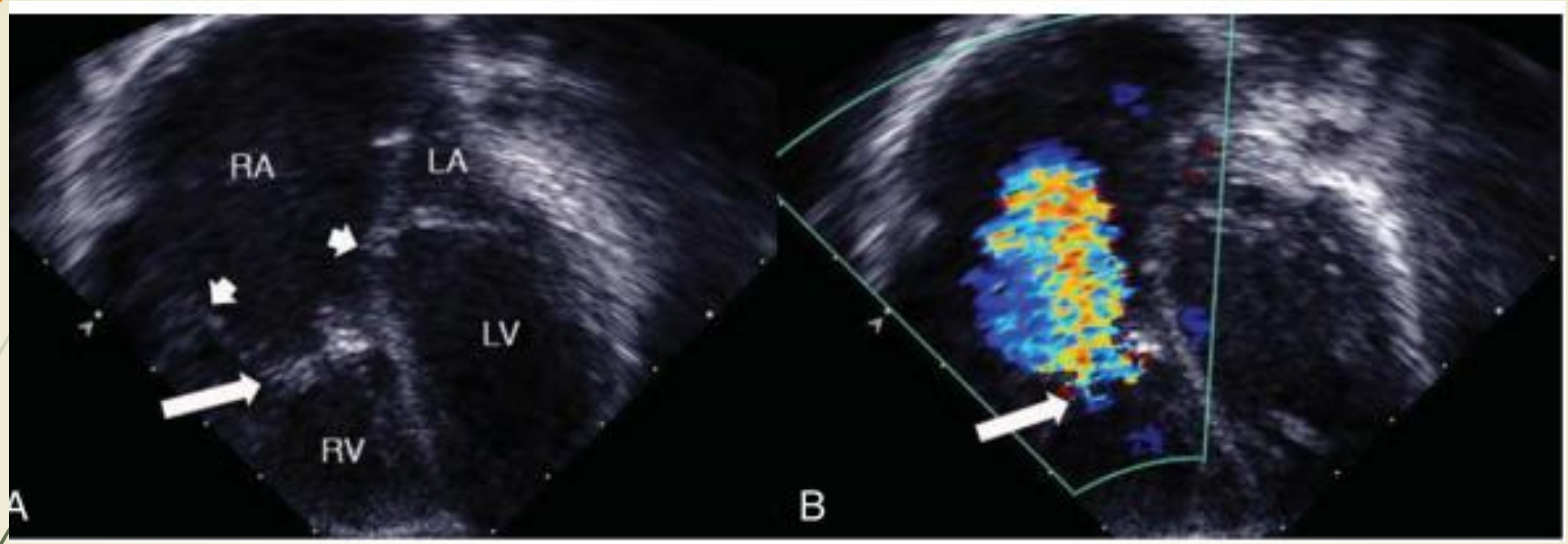
EBSTEIN ANOMALY





- Massive globular cardiomegally (RA dilatation), pulmonary oligemia.
- DDX: pericardial effusion

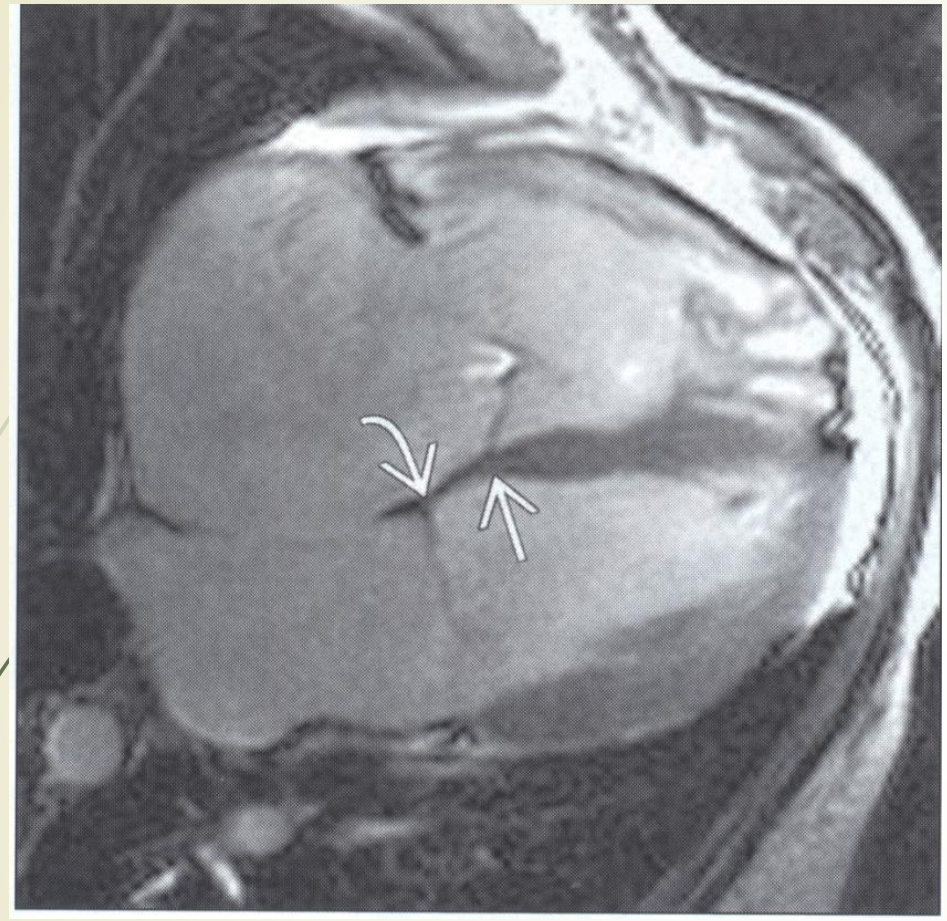
Ebstein anomaly

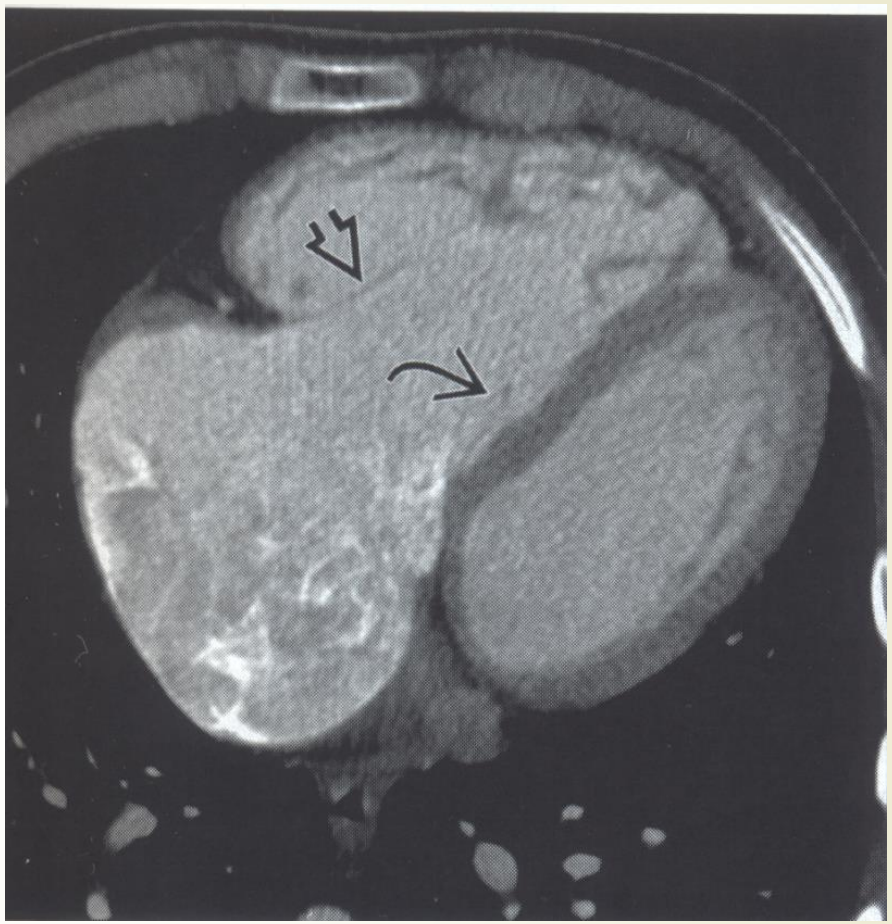


A. Subcostal 4 chamber - displacement of tricuspid valve leaflets inferiorly.

B. Color Doppler- severe regurgitation of the dysplastic tricuspid valve.

Ebstein anomaly





- Mri cine- 16mm distance btn septal leaflets of MV & TV(arrow).
- CECT- RA dilatation, apical displacement of septal tricuspid leaflet.

REFERENCES

1. Grainger&allisons textbook of diagnostic imaging 5th edition
2. Textbook of radiology and imaging. Volume 1. seventh edition. David Sutton