Chapter

13 Role of Radiography in Congenital Heart Diseases

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INTRODUCTION

The role of radiography especially chest X-ray (roentgenogram) has decreased as a tool of cardiovascular thoracic imaging, because of the widespread reach of antenatal (fetal echocardiography) and postnatal transthoracic 2D echocardiography (TTE). The early detection of congenital heart disease (CHD) by TTE and its management, has in turn led to lessened 'classical' appearances on radiography. Still it continues to play an important role in evaluation of both pediatric and grown up congenital heart diseases (GUCH).

Today, chest X-ray (CXR) is not a main diagnostic tool for detection of CHD, however it is complimentary to clinical assessment and TTE. With its decreased importance over the last two-three decades, the ability of interpreting X-ray has also declined across the medical fraternity. Although diagnosis of CHD on CXR can be attempted, it is unfortunately less accurate. Intricate anatomical and physiological phenomena make the interpretation of CXR a complex process compared to echocardiography. Echocardiography allows direct visualization of the cardiac structures and their functional profile, whereas CXR provides mere summation of these complex anatomic and pathophysiological processes. In most cases, cardiac pathology is reflected in an abnormal CXR image. In few situations, it is easier to get and store data in terms of roentgenogram rather than echocardiography, e.g. calcification of the patent ductus arteriosus (PDA) and aortic knuckle, pulmonary arteriovenous (AV) malformation, situs, pulmonary vasculature and laterality of the aortic arch.

Chest X-ray needs to be regarded as a necessary tool with limitations, with a specific role in the diagnosis of CHD. The gallery of images showing the few 'current' clinical scenarios, where CXR has a vital role is given in this chapter. Evaluation of the quality of X-ray requires some understanding of the technical factors involved in the production of an X-ray image. Without such understanding, the risk of making an interpretive error is increased.

BASIC INTERROGATIONS ON CHEST X-RAY

1. Cardiac position.
2. Situs.
3. Bony cage.
4. Rotation.
5. Cardiothoracic ratio.
6. Thymic shadow.
7. Underexposed or overexposed?
8. Degree of inspiration.
9. Pulmonary vascularity.
10. Cardiac silhouette.
11. Lung fields.
12. Cardiophrenic (CP) angles.

Cardiac Position

Normally the major portion of the heart (2/3rd) lies to the left of the midline. Any position other than left is considered as malposition. The left sided heart with left ventricular apex on the left side is called 'levocardia'. When most of the heart is in the right hemithorax and the base to apex axis points to the right, it is called 'dextrocardia'. When the heart is in midline it is termed as 'mesocardia'.

Situs

Situs is the site or position of the viscera. Situs solitus is the normal position, in which the liver is to the right and the fundal gas shadow is to the left. Dextrocardia with situs solitus is called isolated dextrocardia (Figure 1). In mirror image dextrocardia there is situs inversus (Figure 2).

Visceroatrial Situs

Visceroatrial situs means that the right atrium (RA), liver, inferior vena cava (IVC) and trilobed right lung are on one

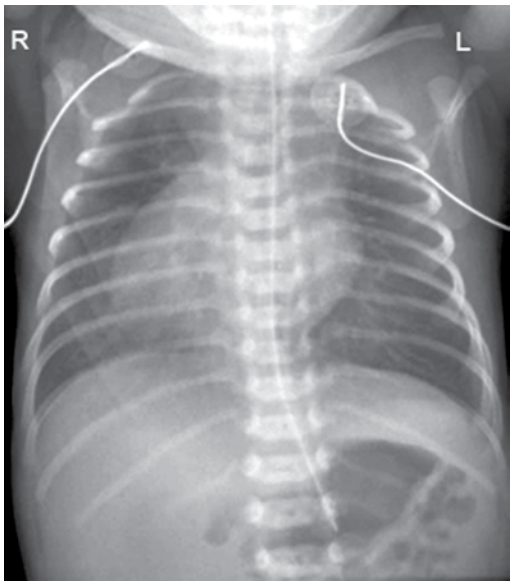


Figure 1: CXR image shows isolated dextrocardia with liver shadow to the right and fundal gas shadow to the left (situs solitus, dextrocardia)

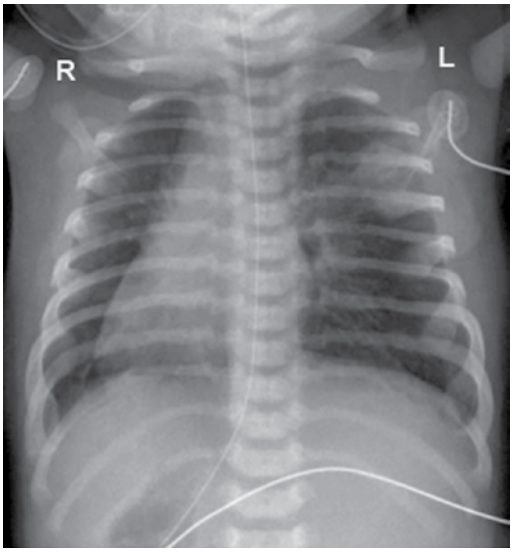


Figure 2: CXR image shows mirror image dextrocardia with liver shadow to the left and fundal gas shadow to the right (situs inversus dextrocardia)

side and the left atrium (LA), stomach gas bubble, spleen and bilobed left lung are on other side. The position of the stomach gas bubble helps in identifying the visceroatrial situs.

Bronchial Morphology

The bronchial morphology is important in isomerism (Figure 3). CXR image gives valuable clues to the diagnosis, as shown in Figures 4 and 5.

i. Tracheal bifurcation is best seen in an oblique view.
ii. Right bronchus is short, wide and straight.

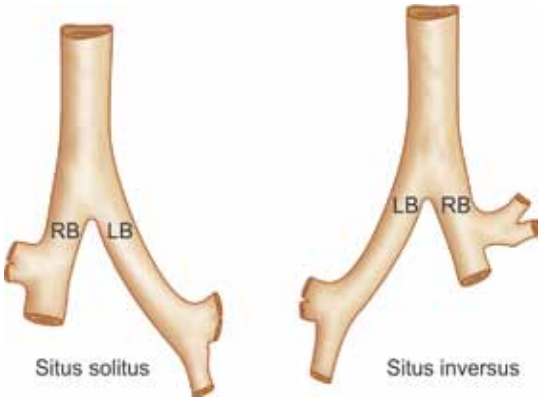


Figure 3: Schematic representation of bronchial morphology (RB = Right bronchus; LB = Left bronchus)

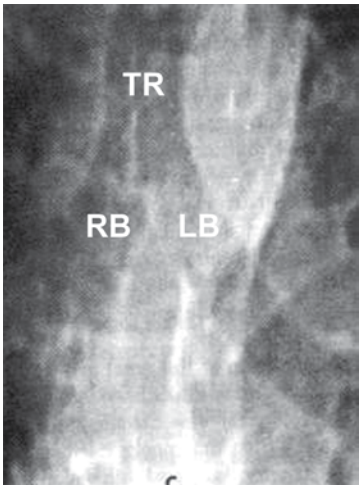


Figure 4: CXR image shows in oblique view shows normal tracheal bifurcation (TR = Trachea; RB = Right bronchus; LB = Left bronchus

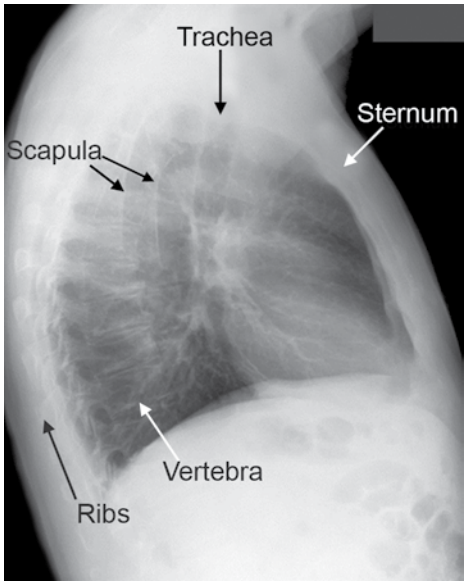


Figure 5: CXR image in right lateral view shows how to identify the normal structures

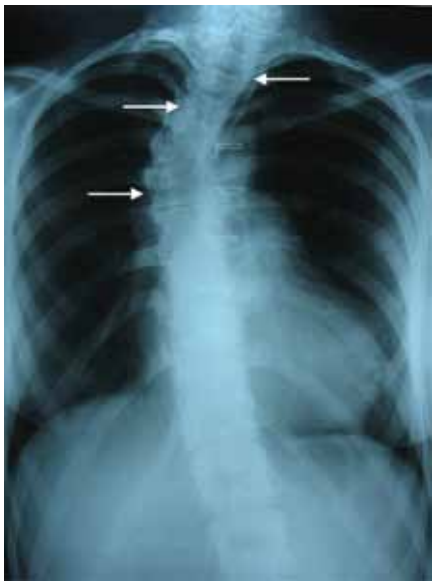


Figure 6: CXR image shows scoliosis

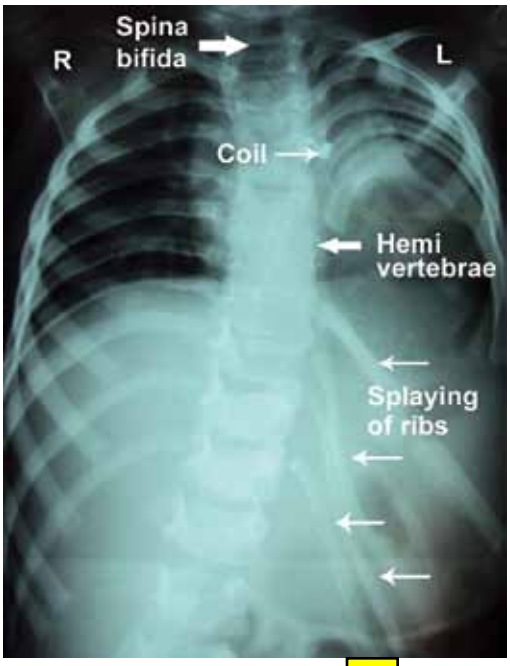


Figure 7: Chest X-ray in a 2 years old child o with meningocoele, shows spina bifida, hemivertebrae (arrow) with splaying of left lower ribs and the PDA coil is in situ

iii. Left bronchus is long, thin and curved.
iv. Measure the bronchus from the bifurcation, the longer of the two is the left bronchus.
v. If the ratio between the left and right bronchus is greater than or equal to 2 then it is situs solitus (longer bronchus is on left) or situs inversus (longer bronchus is on right). If the ratio is less than 1.5, then it indicates isomerism.

Bony Cage

All the CXR should be carefully observed systematically for skeletal abnormalities from the cervical spine, scapula, clavicle, thoracic spine and ribs in both frontal and lateral view (Figure 5). In the bony cage one should look for scoliosis (Figures 6 and 7) in posteroanterior (PA) view and for kyphosis in lateral film. The incidence of scoliosis in acyanotic CHD is 20 percent and in cyanotic CHD it is 66 percent. As the skeletal deformity can pose problems during surgery, chest X-ray is extremely useful in detecting the problems in the bony cage. Rare conditions like hemivertebrae can be easily detected by simple X-ray. The children with Down syndrome often have only 11 pairs of ribs. Premature fusion of sternal segment is usual in cyanotic CHD. Bilateral rib notching is seen in coarctation of aorta (COA) and unilateral rib notching is seen in subclavian pulmonary artery anastomosis as in Blalock-Taussig shunt.

Is there Significant Rotation?

Rotation means that the baby was not positioned flat on the X-ray film and one plane of the chest is rotated in comparison to the plane of the film. The centering of the CXR has to

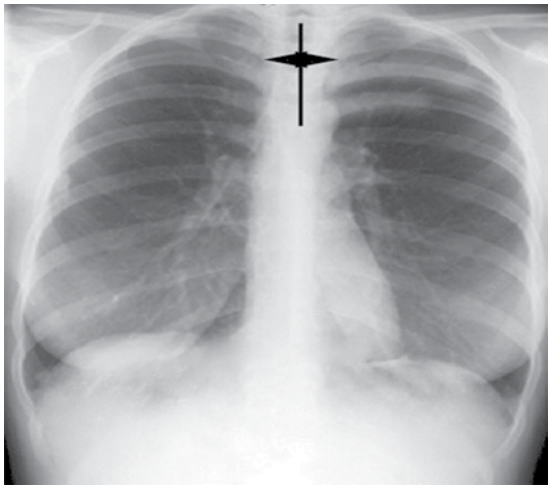


Figure 8: CXR image with correct centering. There is equal distance between the medical end of clavicle and midline

be checked before reading it. The centering is checked by clavicular symmetry. There should be equal distance between the medial end of the clavicle and midline (Figure 8). If the distance between medial end of the clavicle and center line is unequal then centering is not correct (Figure 9). If the centering is not correct then the interpretation can go wrong due to rotation. Rotation can make the lungs look asymmetrical and it can change the orientation of the cardiac silhouette. If there is significant rotation, the side, which has been lifted

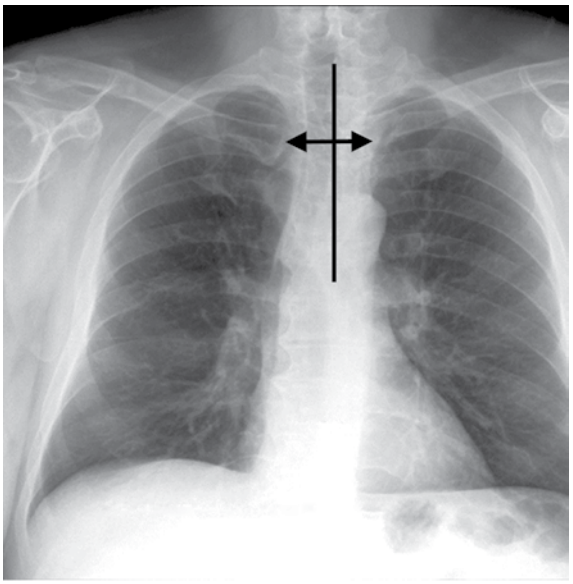


Figure 9: CXR image with incorrect centering—unequal distance between the medial end of clavicle and central line

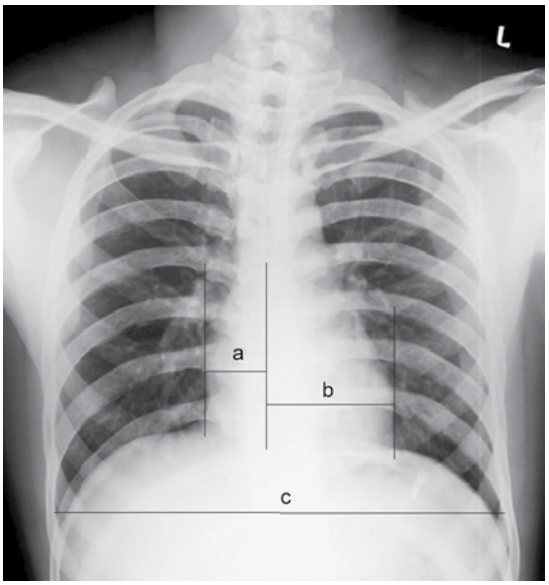


Figure 10: The correct method of measuring the cardiothoracic ratio (CTR) CTR = $\frac{\text{TCD (a + b)}}{\text{TTD (c)}}$

may appear narrower and more dense (white) and the cardiac silhouette appears more in the opposite lung field.

Cardiothoracic Ratio

Before commenting on cardiomegaly the *cardiothoracic ratio* (CTR) must be measured meticulously (Figure 10). The CTR is equal to the transverse cardiac diameter (TCD) divided by the transthoracic diameter (TTD) measured at the inner border of the 9th rib (CTR = TCD/TTD) as shown in Figure 10. Irrespective of CTR an increase of > 2 cm of TCD is significant if previous CXR is available. The normal values of CTR ratio in an adult is 0.41 to 0.5. The upper limit of normal CTR ratio in infants is 0.55 and 0.60 in neonates. A TCD of greater than 15 cm is significant irrespective of normal CTR. An expiratory roentgenogram can lead to pseudocardiomegaly and a prominent aorta. Epicardial fat in the cardiophrenic angle can mimic as cardiomegaly as shown in Figure 11.

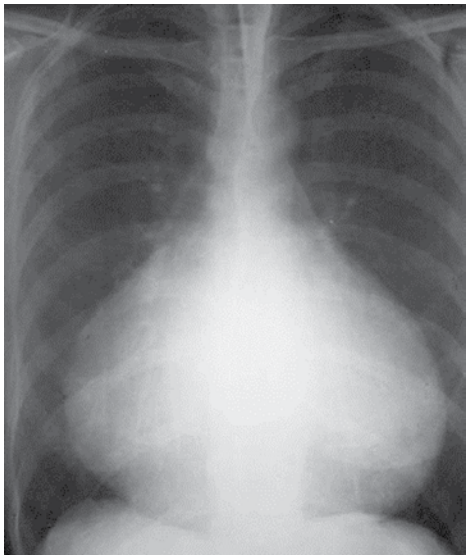


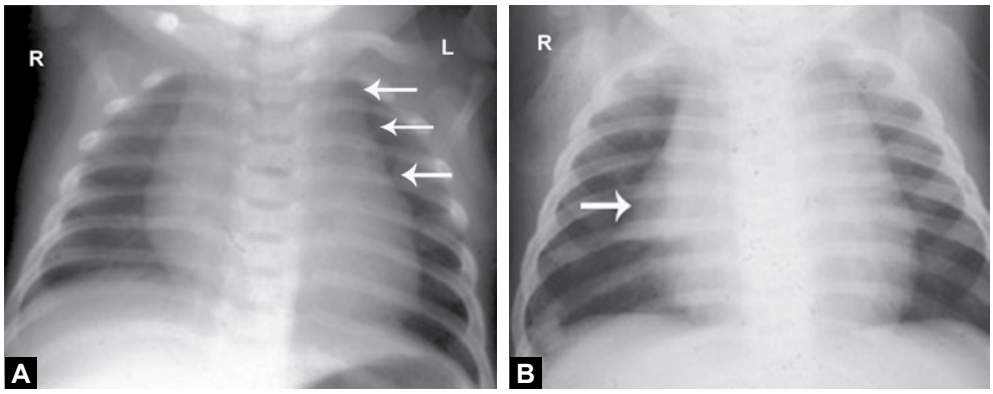
Figure 11: CXR image in a patient epicardial fat in cardiophrenic angle mimicking cardiomegaly

Thymic Shadow

The thymic shadow is usually prominent in the first few years of life. Thymic shadow should not be mistaken for mediastinal widening. The shadow of the thymus lies anteriorly in relationship to the heart and great vessels. The relative size of the thymus increases with expiration and decreases with inspiration. The thymus decreases in size during periods of stress, such as during sepsis. Because the thymus is a soft organ, its lateral margin can have indentations caused due to the overlying costal cartilages, causing a 'wave' sign (Figure 12A). The right lobe of the thymus can insinuate into the minor fissure, causing a 'sail' sign (Figure 12B). The awareness of various apperances of thymus on CXR can bail one out of a situation wherein thymus mimics a pericardial mass on echocardiography.

Is the X-ray Underexposed or Overexposed?

A properly penetrated chest radiograph is one, in which the intervertebral bodies can be seen. Normally only the first four



Figures 12A and B: A. CXR image shows the ribs causing indentation on the soft thymus, causing 'wave' sign (arrows); B. Right lobe of the thymus can insinuate into the minor fissure, causing 'sail' sign (arrow)

vertebral bodies are visible. In *overpenetrated* or *overexposed* CXR, the vertebral bodies are visible clearly through the cardiac shadow. In *underpenetrated or underexposed* film the vertebral bodies are not visible at all. An underpenetrated chest CXR does not differentiate the vertebral bodies from the intervertebral spaces. An overpenetrated film shows the intervertebral spaces very distinctly. An overpenetrated CXR will be darker and the subtleties will be harder to see. An underpenetrated CXR will emphasize normal lung and make it appear as if there are infiltrates. Hence it is important to know whether the CXR is *overexposed* or *underexposed*. Overexposure causes the image to be dark. Under these circumstances, the thoracic spine, mediastinal structures, retrocardiac areas, nasogastric and endotracheal tubes are well seen, but small nodules and the fine structures in the lung cannot be seen. However, the widespread availabilty of digital imaging (computed radiography, direct digital radiography) has reduced the importance of 'optimal' exposure as images can be manipulted on viewing monitors.

Degree of Inspiration

Normally, the right hemidiaphragm should be at the level of 6th rib anteriorly and the 9th rib posteriorly. If diaphragm is below this level, it is a 'hyperinflated' film. Expiratory films produce apparent cardiomegaly and pseudotracheal deviation. The heart appears larger on anteroposterior (AP) than in PA view. Expiratory film, simulates pulmonary edema and the heart appears larger.

Pulmonary Vascularity

Pulmonary vascularity can be nicely made out on CXR and one can know whether the patient has decreased pulmonary flow (oligemia) or increased pulmonary flow (plethora). Abnormal pulmonary vascularity can be generally divided into four types:

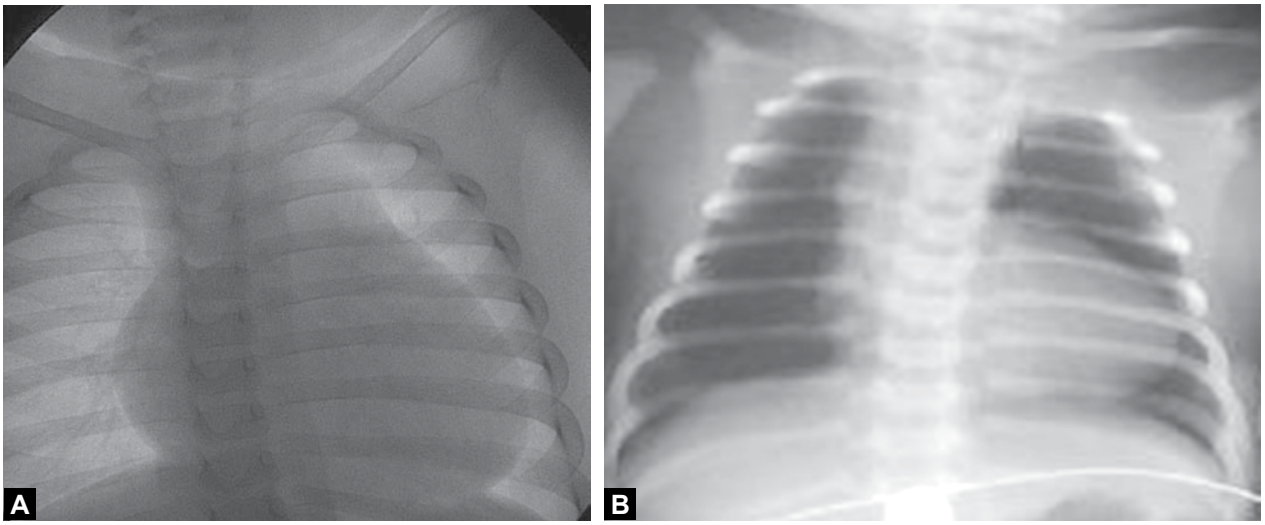
i. Oligemia.
ii. Plethora.
iii. Pulmonary venous hypertension (PVH) (ground glass appearance).
iv. Pulmonary edema (bat-wing appearance).

Oligemia

In oligemia, vascular shadows are reduced. They are not seen even in the intermediate lung zones. Main pulmonary artery (MPA), left pulmonary artery (LPA) and right descending pulmonary artery (RDPA) are of small size (normally RDPA is of the same size as the right lower lobe bronchus). Oligemia occurs in critical pulmonary stenosis (PS) and in tetralogy of Fallot (TOF). In pulmonary valvular stenosis along with oligemia there is poststenotic dilation of the MPA and the right ventricle (RV) and RA are enlarged (Figure 13A). In TOF pulmonary bay is empty (MPA is small) and severe RVH is seen without RA enlargement (as RV pumps into the over-riding aorta and the pressure is not transmitted to RA). Nearly 25 percent of TOF can have right sided aortic arch (Figure 13B). The left or right aortic arch can be made out by the ipsilateral indentation of the trachea.

Plethora

In plethora (Figure 14) the vascular shadows are numerous and vessels can be traced in the lateral one-third of the lung fields also. The MPA, LPA and RDPA are large. End on vessels are more in number (≥ 3 in one lung field or ≥ 5 in both lung fields). The end on vessel diameter is more than that of accompanying bronchus. Normally it is 1.2:1 and in plethora it is $\geq 1.5:1$. The size of the RDPA in nomally < 14 mm and if it increased (> 16 mm in male and >14 mm in female), it indicates a shunt lesion. In normal children diameter of RDPA is less than that of trachea. If the ratio of RDPA to trachea is more than 1 it indicates significant left to right shunt. Plethora occurs in left to right shunts, admixture lesions and transposition of the great arteries (d-TGA) without PS. To have plethora left to right shunts should be at least 1.5:1.



Figures 13A and B: X-ray showing oligemia: A. Fluoroscopic image in a 10 month infant with pulmonary stenosis, shows oligemia with right atrium (RA), right ventricle ~~(RV)~~ and main pulmonary artery ~~(MPA~~) dilatation; B. CXR image in a 4 months infant of tetralogy of Fallot ~~(TOF)~~ with severe oligemia, boot-shaped heart, empty pulmonary bay and right sided aortic arch

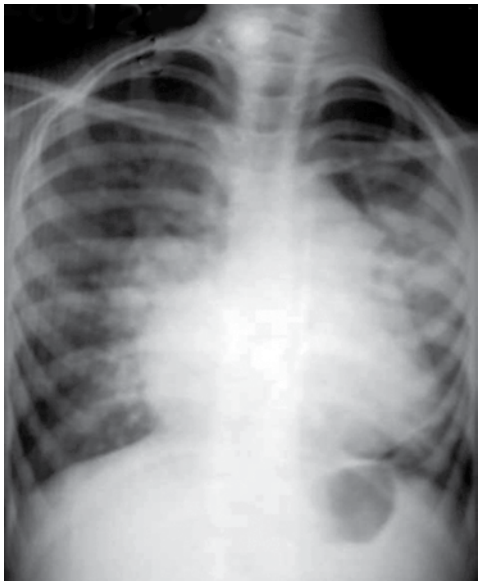


Figure 14: CXR image of a 12-year-old boy with large ventricular septal defect ~~(VSD)~~ and atrial septal defect ~~(ASD)~~ with pulmonary arterial hypertension ~~(PAH)~~, shows right atrium ~~(RA)~~, right ventricle ~~(RV)~~, main pulmonary artery ~~(MPA)~~ dilated with plethora

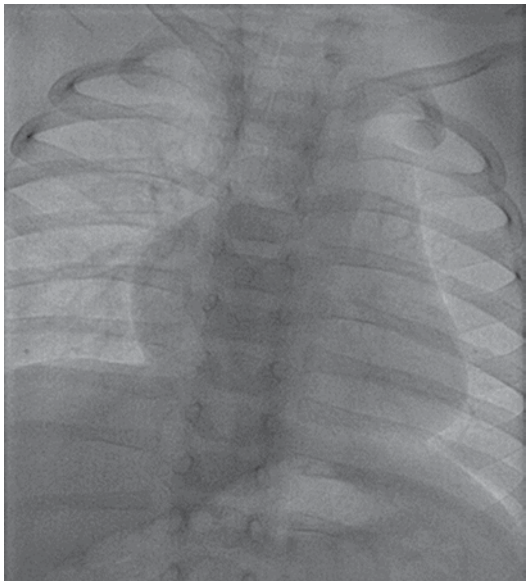
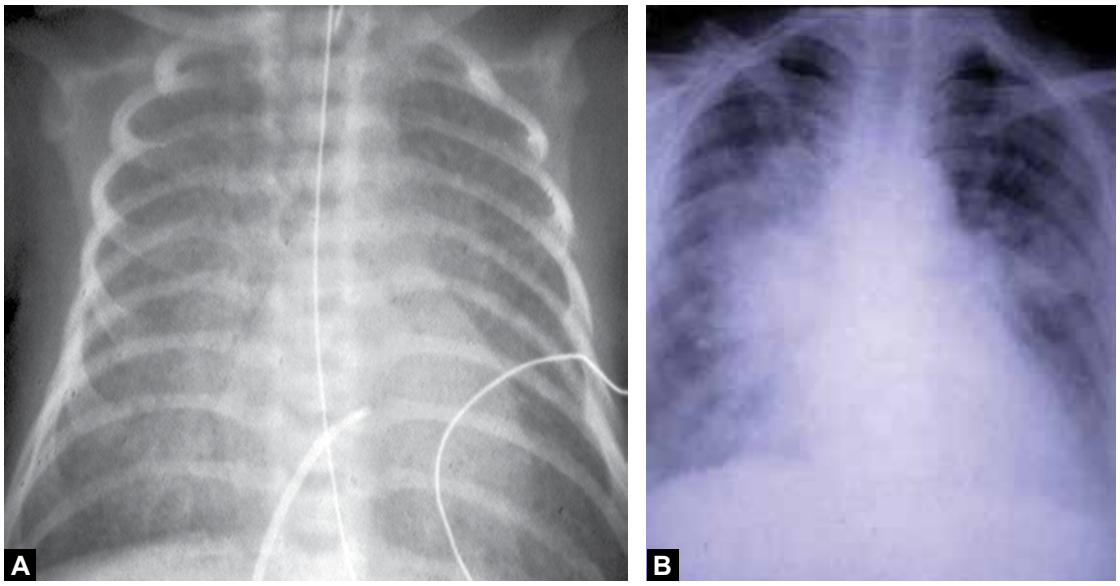


Figure 15: Fluroscopic image of 4 months infant tetralogy of Fallot ~~(TOF)~~ with absent left pulmonary artery ~~(LPA~~), shows oligemia in right lung and absent vascular markings in left lung

Occasionally there can be unilateral plethora as in BT shunt and in unilateral major aortopulmonary collateral artery (MAPCA). Asymmetry in lung vascularity can also occur after Glenn surgery and in pulmonary artery branch stenosis or absent right pulmonary artery (RPA) or LPA (Figure 15).

Pulmonary Venous Hypertension

Normally the upper lobe veins are less prominent than the lower lobe veins. In PVH or postcapillary hypertension, there is equalization of the vascularity. When there is severe obstruction to the pulmonary veins, the CXR shows ground glass appearance (Figure 16A). The ground glass appearance due to pulmonary venous hypertension is an important feature and should be distinguished from hyaline membrane disease. The normal pulmonary capillary wedge pressure (PCWP) is < 12 mm Hg. Larry Elliot has graded PVH into four stages. In stage I PVH, the PCWP is between 13 to 17 mm Hg. The pulmonary veins in the upper lobe are more prominent than that in the lower lobe. This is cephalization of the veins and



Figures 16A and B: A. CXR image of a 2-month-old infant with obstructed infradiaphragmatic total anomalous pulmonary venous connection; B. CXR of a 50-year-old lady with large atrial septal defect ~~(ASD)~~ with pulmonary hypertension shows bat-wing appearance

is also called as 'staghorn' or 'inverted mustache' appearance. In stage II PVH, as PCWP increases to between 18-25 mm Hg, interstitial edema occurs and causes perihilar haziness, peribronchial cuffing (increased thickness of the bronchial walls seen end-on, usually near the hilum), subpleural effusion and the appearance of Kerley B lines (short, thin, 1 to 2 cm long parallel lines seen at right angles to the pleura, laterally at the lung bases). These lines represent fluid in the interlobular septa. In stage III PVH, the PCWP is > 25 mm Hg. There is frank pulmonary edema reaching to the hilum and a typical 'bat-wing' appearance on CXR (Figure 16B). PVH and *pulmonary edema* occur in mitral valve disease, obstructed total anomalous pulmonary venous connection (TAPVC), hypoplastic left heart syndrome (HLHS), dilated cardiomyopathy (DCM). Stage IV PVH is seen in chronic pulmonary hypertension and there is hemosiderosis and ossification.

Pulmonary Artery Hypertension

The characteristic radiological features of pulmonary arterial hypertension (PAH) or precapillary hypertension are prominent central pulmonary arteries and with tortuosity, deviation, diminution in the size of arteries in the middle and lateral thirds of the lungs ("Pruned-tree appearance"). Pruning is defined as > 50% loss of vessel diameter at any degree branching and is suggestive of PAH. With onset of PAH there RV and RA enlargement. The heart size tends to normalize with the severe PAH, though in patients of ASD with PAH the right sided chambers remain enlarged.

Cardiac Silhouette

In the frontal projection, the right border of the cardiac silhouette consists of the following structures from top to bottom: superior vena cava (SVC), ascending aorta, right atrial appendage RA and IVC. The left border of the cardiac silhouette is formed from top to bottom by the aortic knuckle (aortic knob), pulmonary trunk, left atrial appendage and the LV. Any enlargement or hypoplasia of a particular component of the heart will alter the normal shape of the cardiac silhouette. An upturned apex without cardiac enlargement occurs in right ventricular hypertrophy.

The features of LV enlargement (lateral (L) and frontal (F) views) are:

- Mild-obliteration of retrocardiac (prevertebral) space (L)
- Moderate-cardiac shadow overlies vertebral column (L)
- Marked-cardiac shadow overshoots vertebral column (L)
- Left cardiac border elongated and more convex (Figure 17) (F)
- Left cardiac border dips below left dome of diaphragm (F)
- Apex rounded off (F).

The features of RV enlargement are:

- Obliteration of retrosternal space.
- Clockwise rotation of heart—apex moves posteriorly and RV comes to form the left cardiac border—rounded and elongated apex away from the left dome of diaphragm.

The features of RA enlargement are:

- Right cardiac border becomes more convex and elongated
- It forms greater than 50 percent of right cardiac border
- Dilation of SVC

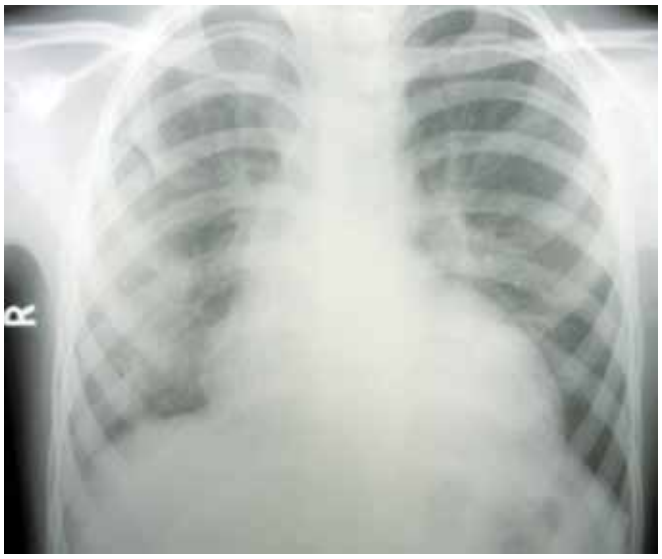


Figure 17: CXR image in a 12 year old patient with Shone's syndrome showing cardiomegaly with left ventricular enlargement with pulmonary venous hypertension

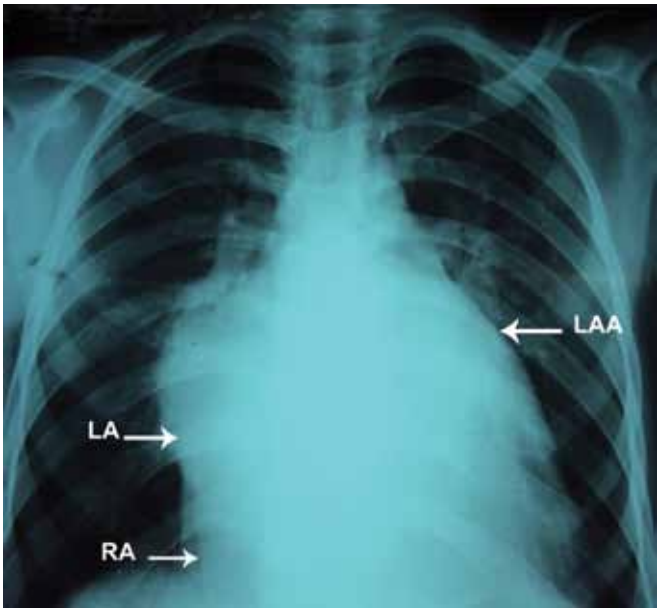


Figure 18: CXR image illustrates giant left atrium (LA), the border of LA is seen beyond the right atrium (RA) with a prominent left atrial appendage (LAA)

- Distance from midline to maximum convexity of right cardiac border greater than 5 cm in adults and > 4 cm in children
- RA border exceeds greater than 3 intercostal spaces.

Increased height of the RA border occupying more than half of the vertical distance between medial end of right clavicle and dome of right hemidiaphragm.

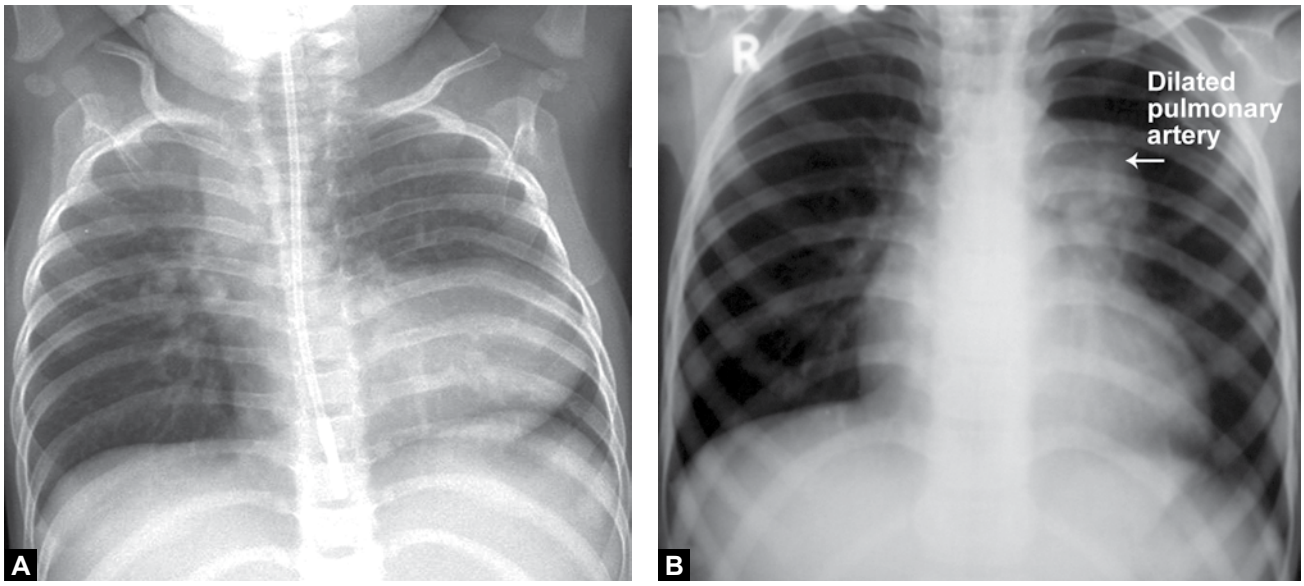
The features of LA enlargement are:

The LA is the uppermost and posterior most chamber in subcarinal angle. The first evidence of LA enlargement is the presence of a bulge, on the left heart border below the pulmonary artery, caused by the left atrial appendage, the 'third mogul sign'. The next evidence is a double density through the central heart shadow, followed by widening of the carinal angle with elevation or posterior displacement of the left main bronchus. There is also posterior esophageal displacement in the right anterior oblique view. The displacement ends well above the diaphragm. The LA shadow does not touch whereas the RA shadow touches the right dome of the diaphragm. The LA enlargement can be graded as follows.

- Grade 1—double cardiac density (contour)
- Grade 2—LA enlargement in flush with right cardiac border
- Grade 3—oveshoots RA and itself forms right cardiac border (Figure 18)

The characteristic cardiac contour in some anomalies are:

1. **Boot-shaped heart (couer en Sabot):** Tetralogy of Fallot (Figure 19A): This deformity is due to the uplifting of the cardiac apex because of RV hypertrophy and concavity of the MPA. But in cases of TOF with absent pulmonary valve instead of a empty pulmonary bay it is filled with aneurysmally dilated PA (Figure 19B) .
2. **Egg on side or egg on string:** Transposition of great vessels (TGA). The malposition of great vessles, in association with stress-induced thymic atrophy and hyperinflated lungs, results in the apparent narrowing of the superior mediastinum on radiographs, which is the the most consistent sign of TGA. The cardiovascular silhouette varies from normal in the first few days after birth to enlarged and globular, with the classic appearance described as an egg on a string (Figure 20).
3. **Figure-of-eight sign or snowman in snow storm:** It is seen in supracardiac TAPVC (Figure 21). The pulmonary veins converge behind the heart to form a common pulmonary vein that connects to the dilated vertical vein on the left, which joins the left innominate vein on the top. This drains into the SVC on the right to form the head of the snowman. The body of the snowman is formed by the enlarged RA along with cardiac shadow. Plethoric lung with ground glass appearance looks like snow storm.
4. **Scimitar sign:** Scimitar is a Turkish sword with the curved blade traditionally used by Persian and Turkish warriors. The shadow resembling the shape of scimitar is produced by an anomalous pulmonary vein that drains any or all of the lobes of the right lung (Figure 22). The so-called scimitar vein curves outward along the right cardiac border, usually from the middle of the lung to the cardiophrenic angle and usually empties into the IVC but also may drain into the portal vein, hepatic vein or right atrium.



Figures 19A and B: A. CXR image of Tetralogy of Fallot (TOF) with boot shaped heart with a empty pulmonary bay with reticular pattern due to collaterals; B. TOF with absent pulmonary valve with dilated pulmonary artery

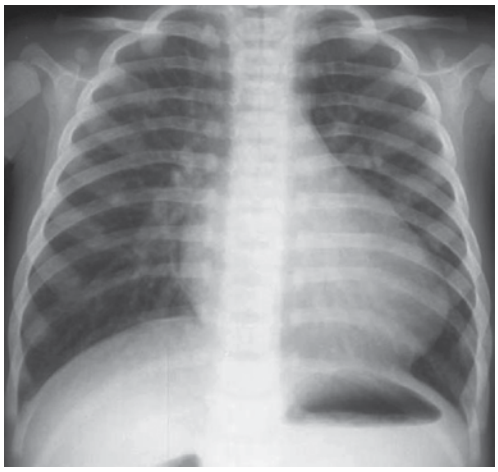


Figure 20: CXR image shows narrow pedicle with egg on string appearance in a 8-year-old with transposition of great arteries ~~(TGA)~~

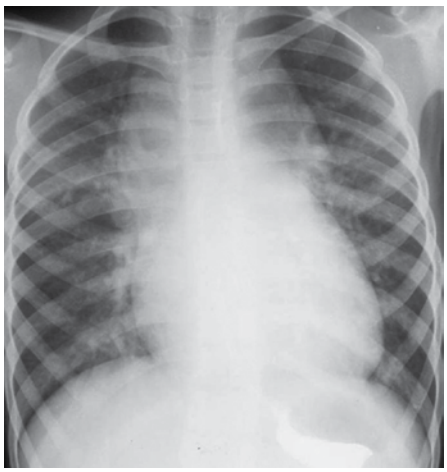


Figure 21: CXR image of posteroanterior view shows 'snowman in snow storm' appearance in total anomalous pulmonary venous connection

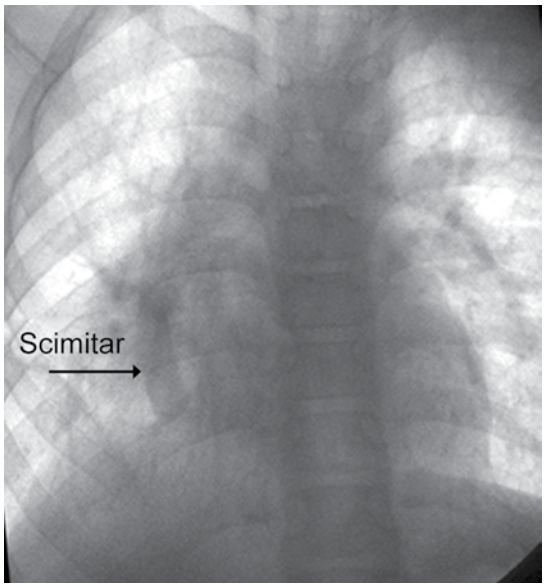
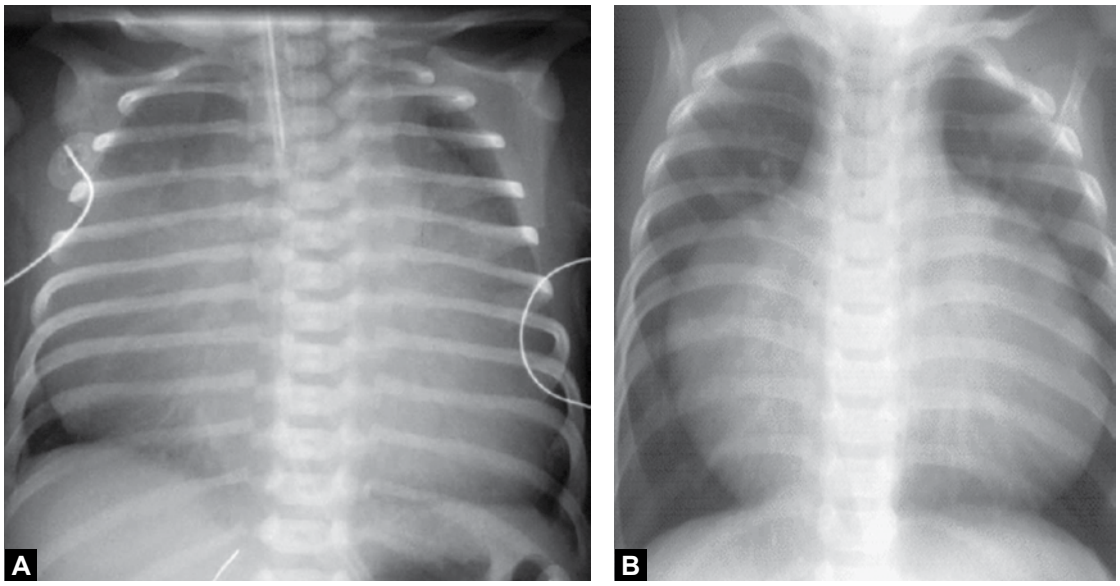
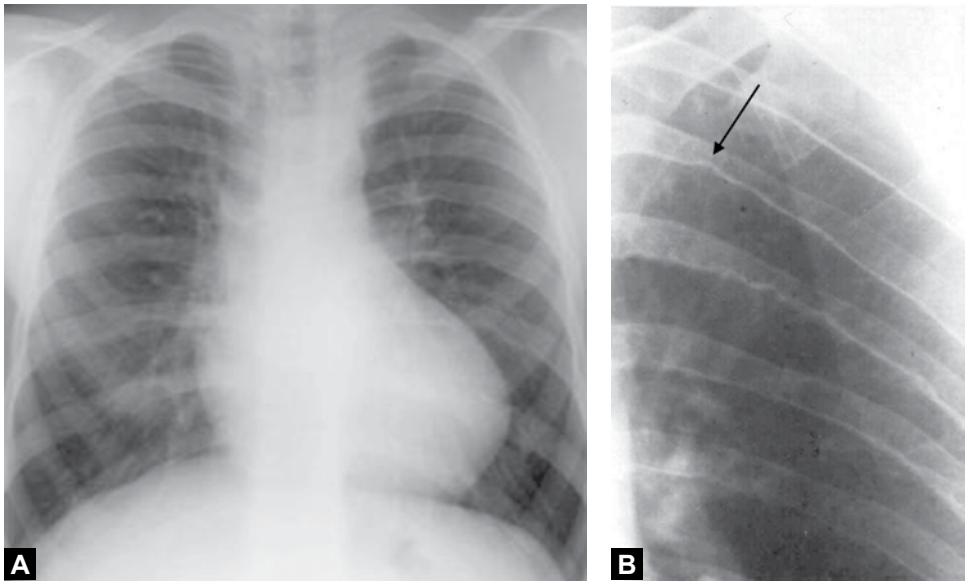


Figure 22: Fluoroscopic image of a 10-year-old girl with scimitar syndrome (arrow)

5. **Box-shaped heart:** The box-shaped or money bag shaped cardiac silhouette is seen in Ebstein anomaly. The RA is huge and fills the entire right hemithorax. The left atrium is normal in size, but the left cardiac contour has a shelved appearance because of the dilated right ventricular outflow tract. The aorta is small and the pulmonary trunk which normally appears as a discrete convex bulge is absent. This combination of features produces a cardiac silhouette that has been described as box shaped (Figures 23A and B).
6. **3 sign:** It is seen in COA. The number 3 is formed by the dilatation of the left subclavian artery and aorta proximal to



Figures 23A and B: CXR image shows A. Huge right atrium ~~(RA)~~ almost filling the entire right hemithorax in a case of severe Ebstein anomaly; B. Narrow pedicle with box shaped or money bag appearance in a case of Ebstein anomaly



Figures 24A and B: CXR image showing 3 sign and rib notching: A. Chest X-ray in coarctation of aorta (COA) shows concentric left ventricular hypertrophy with figure-of-3 sign; B. Rib notching is seen in the X-ray of 16-year-old boy of COA with collaterals

the site of coarctation, indentation of the site and dilatation of the aorta distal to the site (Figure 24A).

7. **Rib notching:** It is seen in the lower margin of the third to eighth ribs (Figure 24B) in aortic coarctation due to the enlarged, tortuous intercostal arteries supplying blood to the descending aorta. The notching is not usually seen in children younger than 5 years.

Apart from specific signs in some condition there are other conditions which have characteristic features on radiography, which are diagnostic, e.g. in cases with corrected transposition of great arteries (CTGV), the malposed ascending aorta produces a long convexity on the left upper mediastinal contour and cardiomegaly with increased pulmonary vascular markings secondary to a ventricular septal defect (Figure 25). The right pulmonary artery appears to have a high take off because of an absent aortic shadow and is also quite prominent indicating ventricular inversion. Also in tricuspid atresia, CXR shows a mildy enlarged LV with a gap between RA and the diaphragm,

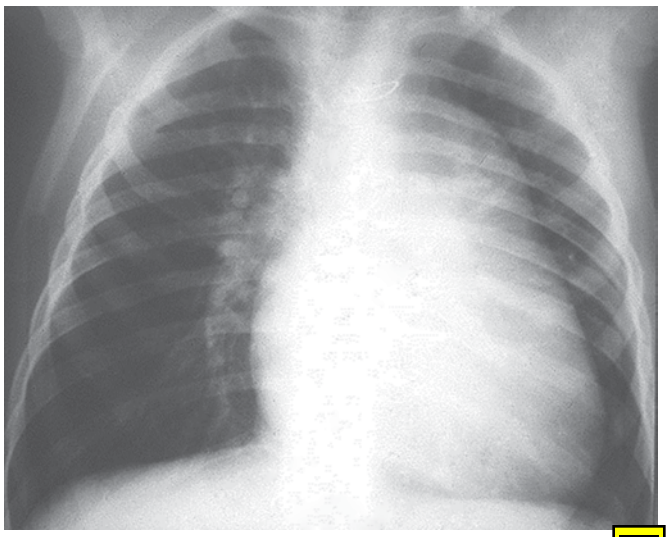


Figure 25: Corrected transposition of great arteries ~~(TGA)~~ ascending aorta producing a long convexity on the left upper mediastinal contour

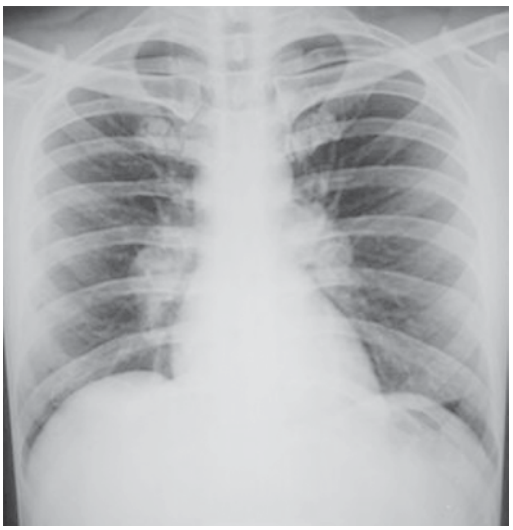


Figure 27: CXR image of 32-year-old patient with large patent ductus arteriosus ~~(PDA)~~ with severe pulmonary hypertension ~~(PH)~~ shows dilated right pulmonary artery ~~(RPA)~~ and left pulmonary artery ~~(LPA)~~ with peripheral pruning

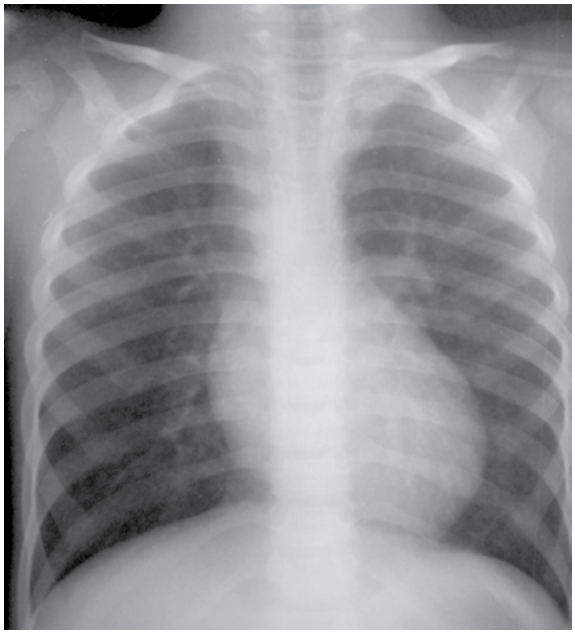


Figure 26: CXR image of film of tricuspid atresia shows normal sized heart wi[illegible]ldly enlarged left ventricular configuration with a gap betwee[illegible]nd the diaphragm, indicating hypoplastic ~~RV~~

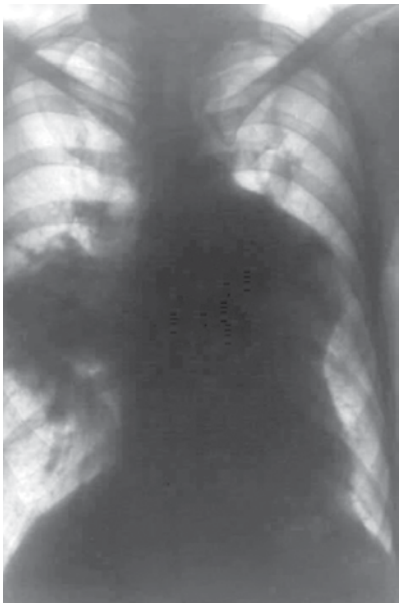


Figure 28: Fluroscopic image of 22-year-old patient with large[illegible] with severe[illegible] ~~with R to L~~ shunt sho[illegible]e the classic 'jug-handle appearance' due to aneurysmally dilated[illegible] ~~and RPA~~. Note the normal sized heart with peripheral pruning

indicating hypoplastic RV (Figure 26). The pulmonary artery can be enlarged as in shunt lesions with PAH (Figure 27). In Eisenmenger's, the classic 'jug-handle appearance' due to aneurysmally dilated MPA and RPA can be seen on CXR (Figure 28). There is a normal sized heart with peripheral pruning. The CXR can be diagnostic in idiopathic dilatation of pulmonary artery where there is aneurysmally dilated pulmonary artery with normal cardia and lung fields (Figure 29).

Lung Fields

Apart from the vascularity, the lung fields can give important clue for the diagnosis of pulmonary embolization of either thrombus or large vegetation from RV or pulmonary valve. The *Fleischner lines* are the linear shadows seen in the region of an embolus. The *knuckle sign*—Abrupt tapering or termination of a pulmonary vessel. The *Westermark sign* is the focal hyperlucency (Figure 30A). The *Hampton's Hump*

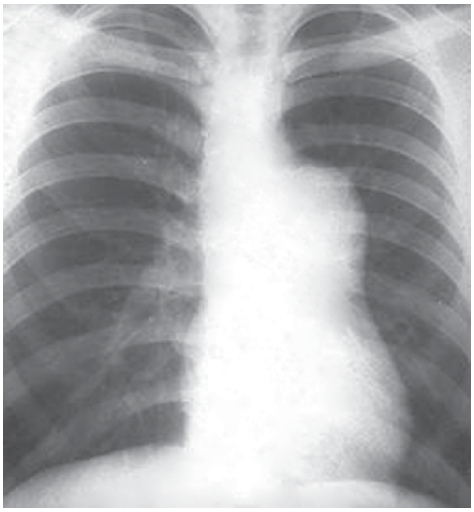


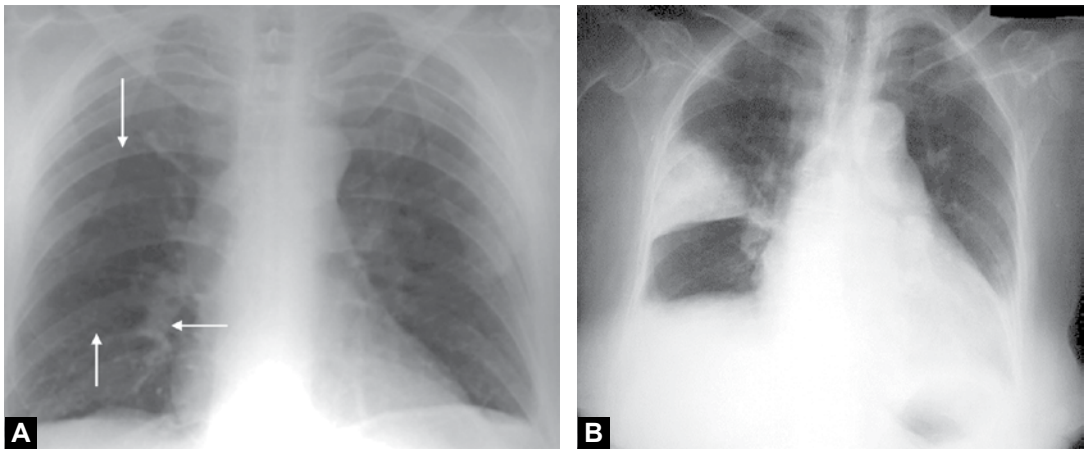
Figure 29: CXR image shows aneurysmally dilated pulmonary artery with normal cardia and lung field of 18-year-old girl with idiopathic dilatation of pulmonary artery

is the classical peripheral wedge-shaped area of opacification with apex towards the hilum (Figure 30B). The *melting ice cube sign* is the decrease in the size of opacity like melting ice cube on serial CXR/CT taken during the course of treatment.

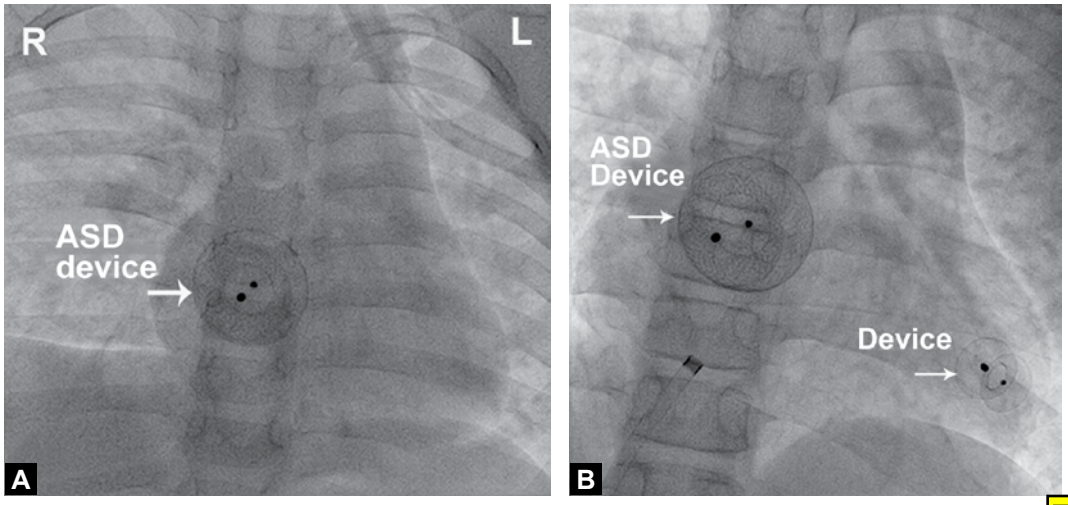
With the advent of non-surgical transcatheter interventions the radiographs are useful for documenting the position of the device (Figures 31A and B, Figure 32A) and also detect the embolization of the device (Figure 32B). The other devices like pacemakers and position of pacing leads can be detected on CXR.

Cardiophrenic Angles

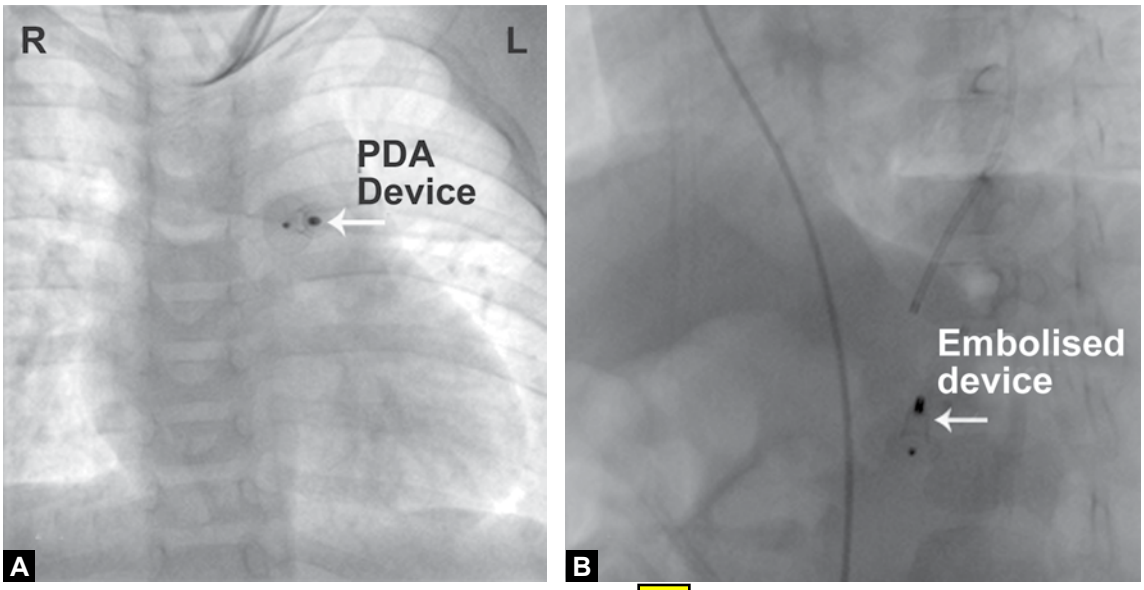
The cardiophrenic (CP) angle is obliterated in pericardial effusion (Figure 33). It is obtuse and helps to distinguish from cardiomegaly, in which the CP angle remains acute, however great the enlargement is. In patients of CHD with congestive heart failure along with the cardiomegaly the pleural effusion can be detected on CXR as the costophrenic angle is obliterated.



Figures 30A and B: A. CXR image of a patient with pulmonary embolism shows the abrupt tapering and termination of a pulmonary vessels (horizontal arrow) and hyperlucency (vertical arrows); B. CXR image of a patient with pulmonary infarction with peripheral wedge-shaped area of opacification with apex towards the hilum



Figures 31A and B: A. Fluoroscopic image showing atrial septal occluder *in situ*; B. Fluoroscopic image illustrating *in situ* device and a device, in an apical position, deployed simultaneously in 12 years old boy



Figures 32A and B: A. Fluoroscopic image illustrates the device *in situ*; B. Fluoroscopic image of the abdomen shows the PDA device embolized in aorta

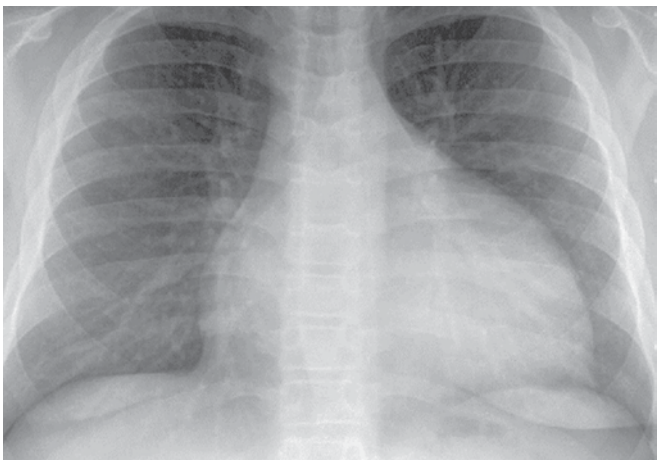


Figure 33: X-ray film shows the cardiomegaly with obtuse cardiophrenic angle, indicating the presence of pericardial effusion

CONCLUSION

Along with clinical evaluation CXR serves as a screening tool for the detection of congenital cardiovascular diseases. It is also a useful tool in short-term and long-term follow-up of post-surgical and post-procedure cases especially device closures. It is simple documentation tool for situs, cardiac silhouette, radio-opaque surgical and percutaneous interventional devices. All importantly CXR is a stimulating teaching tool and helps understanding the complexity of CHD. CXR is not any more a primary diagnostic tool in CHD but appreciating it as poor man's CT will not be overstating its utility in routine clinical practice.

Correct interpretation of pulmonary vasculature requires considerable experience and cannot simply be learned from a book.

—Kurt Amplatz

BIBLIOGRAPHY

1. Ahmed MZ, Gera R and Kailas L. Chest X-rays in pediatric cardiology.
2. Chokkalingam A, Chokkalingam V. Cardiac X-rays. Elsevier.
3. Sharada K and Gouthami V. Radiological Diagnosis of Congential Heart Diseases.
4. Sutton D. Text Book of Radiology and Imaging, 7th edition. Churchill Livingstone.