

CHAPTER 1...INTRODUCTION

Marfan's syndrome which is the results of mutation in the gene that encodes fibrillin-1.

The right and left ventricle do not function in series in fetus as they do in extra uterine circulation.

About 60% of the umbilical venous return that is oxygenated in the placenta bypasses the liver through the ductus venosus and enters IVC and right atrium.

The Electrocardiogram developed in 1903 by William Einthoven.

The x-ray was developed in 1895 by ROENTGEN.

CHAPTER 2.....NORMAL OR INNOCENT MURMURS

Murmurs that occur in the absence of either morphologic or physiologic abnormalities of heart or circulation are called innocent murmurs.

BOX 2-1 NORMAL MURMURS
A. Systolic
1. The vibratory systolic murmur of Still
2. The pulmonary artery systolic murmur
3. The branch pulmonary artery systolic murmur
4. The supraclavicular systolic murmur
5. The systolic mammary souffle
6. The aortic sclerotic systolic murmur
7. The cardiorespiratory systolic murmur
B. Continuous
1. The venous hum
2. The continuous mammary souffle
3. The cephalic continuous murmur

All normal systolic murmur are midsystolic except mammary souffle.

PULMONARY ARTERY SYSTOLIC MURMUR

Midsystolic...

Maximal intensity in second left intercostal space.

Commonly heard in pregnant women with anemia.

BRANCH PULMONARY ARTERY SYSTOLIC MURMUR

Found in neonates.

SUPRACLAVICULAR SYSTOLIC MURMUR

Maximal above clavicle.

SYSTOLIC MAMMARY SOUFFLE

Systolic murmur

The murmur is heard over breast late in pregnancy.

AORTIC SYSTOLIC MURMUR IN OLDER ADULTS

Aortic sclerosis systolic murmur of older age.

GALLAVARDIN DISSOCIATION

Musical high frequency murmur within left ventricular cavity and the harsh impure right basal murmur within aortic root.

NORMAL BICUSPID AORTIC VALVE

Midsystolic murmur that is most prominent in the second right intercostal space in children.

CARDIORESPIRATORY MURMUR

Benign nature.

VENOUS HUM

Normal continuous murmur.

Maximal intensity in SUPRACLAVICULAR fossa.

CONTINUOUS MAMMARY SOUFFLE

Occur in late pregnancy or early postpartum.

CHAPTER... 3..CARDIAC MALPOSITION

Dextrocardia in Situs inversus was one of the first recognised congenital malformations of heart.

Left sided heart.. Levocardia

Right sided heart... Dextrocardia

Midline heart.. Mesocardia

Situs... site or position

Solitus... usual or normal

Situs solitus.. Normal position

Inversus.. Reverse or opposite

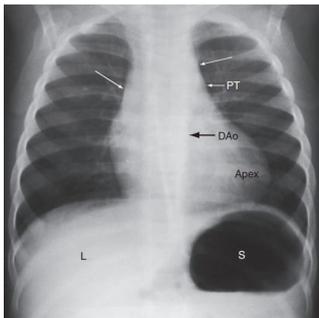
Situs inversus... opposite or reverse of normal

Ambigus.. Uncertain

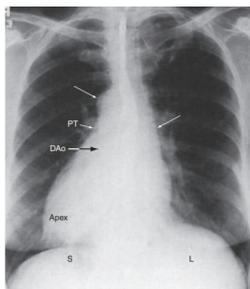
Situs Ambigus.. Uncertain position

Ectopia Cordis... extrathoracic location of heart

Heterotaxy.. Different arrangements



NORMAL HEART AND VISCERA IN SITUS SOLITUS...



DEXTROCARDIA WITH SITUS INVERSUS....

Asplenia.... Congenital absence of spleen.

Polysplenia.. Multiple spleen

Accessory spleen.. Splenules

Concordant... to agree

Discordant.. Not agreeing

Transposition of great arteries... each great artery is connected to a morphologically Discordant ventricle. The aorta arises from morphologic right ventricle and pulmonary trunk arises from morphologic left ventricle.

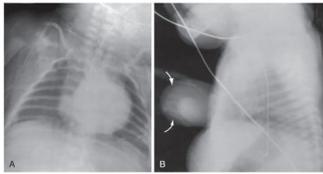
Atrioventricular Discordance..

ATRIOVENTRICULAR Discordance applies when a morphologic right atrium connects to a morphologic left ventricle via morphologic mitral valve and when morphologic left atrium connect to a morphologic right ventricle via morphologic tricuspid valve.

Ventriculoarterial Discordance..

When a morphologic right ventricle give rise to aorta and morphologic left ventricle give rise to pulmonary trunk.

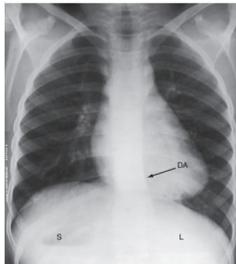
Congenitally Corrected Transposition of great arteries.. Double Discordance.



X-RAY of 2 day old male with ectopia cordis.

SITUS SOLITUS

Atrial Situs and abdominal Situs are usually Concordant.



Chest x-ray showing Situs inversus with levocardia.

SITUS INVERSUS WITH DEXTROCARDIA

The heart is right sided.

SITUS SOLITUS WITH DEXTROCARDIA

The lungs and abdominal viscera are Situs solitus but heart is right thoracic.

SITUS INVERSUS WITH LEVOCARDIA

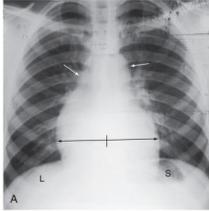
Situs inversus of thoracic and abdominal viscera in the presence of left thoracic heart.



X-RAY showing Situs inversus with levocardia.

MESOCARDIA

Midline cardiac position in presence of thoracic and abdominal Situs solitus.



SITUS SOLITUS WITH MESOCARDIA.

HISTORY

KARTAGENER TRIAD...

Sinusitis+Bronchiectasis+Situs inversus.

SITUS inversus with levocardia invariably occurs with coexisting congenital heart disease.

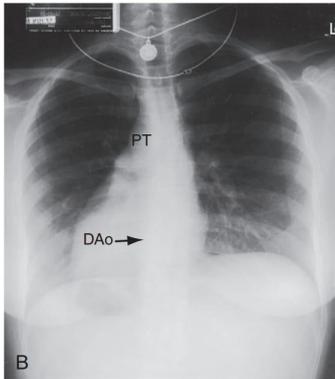
POLAND SYNDROME

Absence of Pectoralis major muscle.

GOLDENHAR'S SYNDROME

Occuloauricular Vertebral dysplasia, hemi facial microsomia.

A right anterior chest bulge with asymmetry arouse suspicion of dextrocardia.



SITUS INVERSUS WITH DEXTROCARDIA.

RIGHT ISOMERISM

BOX 3-2 RIGHT ISOMERISM

Congenital Cardiac Malformations

- Common atrium
- Common atrioventricular valve
- Morphologic or functional single ventricle
- Pulmonary stenosis or atresia
- Total anomalous pulmonary venous connection
- Absent coronary sinus

BOX 3-1 RIGHT ISOMERISM

Bilateral superior vena cavae
 Bilateral sinoatrial nodes
 Paired atrioventricular nodes
 Bilateral morphologic right atria (appendages)
 Bilateral morphologic right bronchi
 Bilateral morphologic right lungs
 Dextrocardia or levocardia
 Asplenia
 Transverse liver
 Right-sided or left-sided stomach

LEFT ISOMERISM**BOX 3-3 LEFT ISOMERISM**

Bilateral superior vena cavae
 Bilateral morphologic left atria (appendages)
 Absent or atretic sinoatrial node
 Bilateral morphologic left bronchi
 Bilateral morphologic left lungs
 Transverse liver
 Polysplenia
 Stomach usually right-sided

BOX 3-3 LEFT ISOMERISM

Bilateral superior vena cavae
 Bilateral morphologic left atria (appendages)
 Absent or atretic sinoatrial node
 Bilateral morphologic left bronchi
 Bilateral morphologic left lungs
 Transverse liver
 Polysplenia
 Stomach usually right-sided

Visceral Heterotaxy with left ISOMERISM tend to occur in females and right ISOMERISM in Males.

CHAPTER.. 4...ISOLATED CONGENITAL COMPLETE HEART BLOCK

Complete heart block... atrial impulses are not conducted to ventricles.

An association between Maternal lupus erythematosus and congenital complete heart block was reported in 1966.

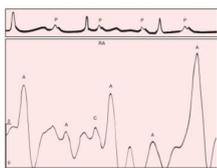
CHB is passively transferred autoimmune disease that affects offspring of mother with Ro/SSA auto antibodies.

Dilation of the ascending aorta is present in large proportion of pediatric patients with isolated congenital heart block.

A pulse rate slow for age leads to diagnosis of congenital CHB.

The upstroke is brisk and pulse pressure relatively wide.

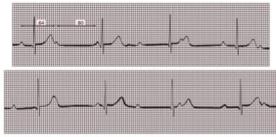
Cannon waves are found in CHB.



CANNON WAVES IN CONGENITAL CHB.

Fourth heart sound are found. They are not heard in healthy young people.

The P wave of atrial activity and the QRS of ventricular activity occur independently.



ECG SHOWING CHB.

CHAPTER 5...CONGENITAL ABNORMALITIES OF PERICARDIUM

The pericardium consists of two layers. The layer attached to the surface of heart is the visceral pericardium.

The layer that is not attached to the surface of heart is parietal pericardium. It is separated from the visceral pericardium by 20 to 50 ml of serous ultrafiltrate of plasma.

CANTRELL'S SYNDROME..

Ectopia cordis

Epigastric omphalocele

Cleft of distal sternum

Defects of anterior diaphragm

Defect of diaphragmatic pericardium

HISTORY

In partial or complete absence of pericardium, the most common symptoms is chest pain that appears suddenly in previously asymptomatic adults.

The pain is stabbing, left sided, brief, unrelated to exertion but aggravated by position especially left lateral decubitus, awakens patients from sleep and is relieved in upright position.

The pain associated with partial absence of left pericardium is attributed to herniation of left atrial appendage through pericardial defect.

The pain associated with complete absence of pericardium is believed to originates from torsion of thoracic inlet.

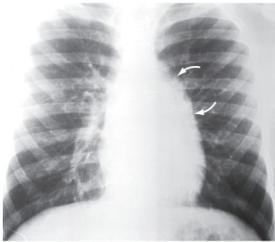
Additional symptoms include Dyspnea, Palpitations, Dizziness, syncope and most commonly sudden death.

PHYSICAL APPEARANCE

Abnormal facies
GH deficiency
VATER defects

ARTERIAL PULSE AND JVP

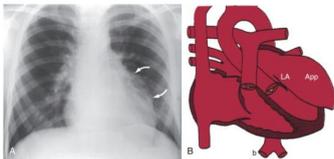
Normal.



CONGENITAL PARTIAL ABSENCE OF PERICARDIUM

With congenital partial absence of left pericardium, the position and movement of the heart are normal.

With complete absence, a striking mobility of heart shifts to the left.

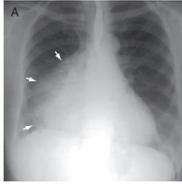


- A. .. Xray from 10 year old girl shows a relatively long left paracardiac convexity caused by congenital Intra pericardial aneurysm of left atrial appendage..**
- B. Schematic illustration of the congenital Intra pericardial aneurysm of left atrial appendage**

X-RAY

Congenital partial absence of the left pericardium is accompanied by herniation of the left atrial appendage, represented in the x-ray by convexity immediately below pulmonary trunk or if herniation is larger by extension of convexity to the second and third left interspace.

A congenital pericardial cyst typically present as a smooth homogeneous radio density in the right cardiophrenic angle, touching anterior chest wall and anterior portion of right hemidiaphragm.



X-RAY from a 23 year old women with an usually large congenital pericardial cyst (arrow) that occupies entire right cardiac border.

A congenital complete absence of pericardium is characterised by dramatic mobility of heart that results in striking leftward and posterior displacement.

ECHOCARDIOGRAPHY

The echocardiogram may strongly suggest the diagnosis of congenital absence of pericardium and can be used to identify Coexistent cardiac defects.

SYMPTOMS

LEFT sided chest pain

Dyspnea

Palpitation

Dizziness

Syncope

Sudden death

DEFINITIVE DIAGNOSIS

Thoracic MRI

CT SCAN

CHAPTER 6....CONGENITALLY CORRECTED TRANSPOSITION OF THE GREAT ARTERIES

Karl Von Rokitansky applied the term corrected to a undescribed form of transposition of great arteries.

The aorta is positioned left and anterior.

The right sided ventricle is finely trabeculated as usually seen in left sided ventricle.

The left Atrioventricular valve and left sided ventricle resembled the usual right Atrioventricular valve and right ventricle.

From the right sided ventricle arises somewhat right and posteriorly positioned pulmonary artery.

The atria are normal.

Transposition of the great arteries are characterised.. by chambers that are joined Concordant at the Atrioventricular junction but discordantly at the ventriculo great arterial junction.

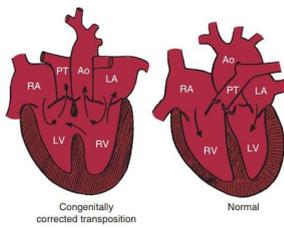
The pulmonary artery arises from a morphologic left ventricle and the aorta arises from morphologic right ventricle.

The circulation are in parallel rather than in series.

Congenitally Corrected Transposition of the great arteries is characterised by chambers that are joined discordantly at the Atrioventricular junction and ventricles that are joined discordantly at the ventriculo Great arterial junction.

Atrioventricular alignment and Ventriculoarterial alignment are both Discordant.

The double Discordance physiologically corrects the Discordance intrinsic to each other.



Also called as L-transposition.

TO.. BE CONTINUED.....