### Situs Inversus with Levocardia (Fig. 1.5)

- All the thoracic and abdominal viscera are placed: Mirror image of normal position except the heart.
- Heart is placed on the left side of the thoracic cavity.
- Left cupola of the diaphragm is lower than right.

*Cardiac chambers*: LA and LV are on right of RA and RV.

#### *Mesocardia (Heart Midline)*

- Heart is in the middle of thoracic cavity.
- All the abdominal and thoracic visceral structures are in normal position (situs solitus).

#### *Congenital anomalies associated with cardiac malposition*

*Situs inversus with dextrocardia*: Not associated with cardiac abnormalities.

*Isolated dextrocardia*: Can be associated with septal defects or pulmonary or aortic outflow obstruction.

*Levocardia with situs inversus*: Can be associated with complicated congenital heart disease.

#### *Cardiac examination and symptoms of heart disease in different cardiac malpositions*

*Situs inversus with mirror image dextrocardia*: Chest pain of IHD is felt on the anterior part of the chest radiates to ulnar border of right upper limb.

#### *Cardiac examination*

Situs inversus with mirror image dextrocardia:

- Cardiac apex—right side.
- Cardiac dullness—percussed on the right side of the chest.
- Liver dullness—on the left side of the abdomen.
- Fundal resonance of stomach—on the right side.
- Heard sounds—1st and 2nd heart sounds are heard better on the right side.
- 2nd heart sound—splitting is better on the right 2nd intercostal space.

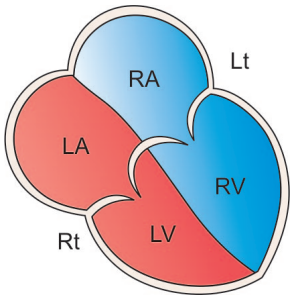


**Fig. 1.5:** Situs inversus with levocardia

#### *ECG and X-ray Chest in Situs Inversus with Dextrocardia*

##### *X-ray chest*

- Fundal gas—on the right side
- Liver shadow left of the abdomen.
- Cardiac shadow—right side of the chest

##### *ECG*

- QRS—negative in lead I and aVL
- P wave negative lead I and aVL
- Prominent R wave in V1.

### Dextroposition of the Heart

Position of the heart is in the right side of the chest due to noncardiac causes like right lung fibrosis or eventration of the diaphragm on the left side.

#### *Dextroversion of the Heart*

- Due to severe right-sided rotation of the heart.
- Heart is in the right side of the chest. LV lies anteriorly. Relation of the cardiac chambers is normal. LV lies to the left of RV.

## CHEST PAIN

#### *Four life-threatening disorders which cause acute chest pain or discomfort*

- Acute myocardial infarction
- Aortic dissection
- Pulmonary embolism
- Pneumothorax.

#### *Other causes of acute chest pain—cardiac causes*

- Stable angina

- Bilateral mass palpable in the abdomen: Polycystic kidney.
- Auscultate for renal artery bruit.
- Sudden raise of blood pressure—on massaging the abdomen—possible pheochromocytoma.
- For evidence of complications of hypertension.

## Secondary Hypertension

### Causes

#### *Renal parenchymal disease*

- Acute glomerulonephritis
- Chronic glomerulonephritis
- Chronic pyelonephritis
- Polycystic disease of kidney
- Renal tumors especially renin producing

#### *Renovascular*

- Renal artery stenosis
- Fibromuscular dysplasia
- Atherosclerosis.

### Endocrine Disease

- Acromegaly
- Hypothyroidism—diastolic hypertension
- Thyrotoxicosis—systolic hypertension
- Hyperparathyroidisam
- Primary aldosteronism
- Cushing's syndrome
- Pheochromocytoma
- Adrenal enzyme deficiency.

*Vascular causes*: Coarctation of aorta, vasculitis syndrome.

#### *Drug induced*

- Intake of corticosteroids, large dose of estrogen.
- NSAIDs.
- Nasal decongestants
- Cyclosporin

#### *Pregnancy-induced hypertension*

*Neurological*: Increased intracranial pressure, porphyria, Guillian-Barré syndrome.

### Investigation of a Patient of Secondary Hypertension

- Routine investigations—already discussed under hypertension
- Urine—RBCs, casts, protein: Parenchymal renal disease.
- Serum electrolytes—potassium low:
  - Conn's syndrome
  - Cushing's syndrome
  - Adrenal enzyme defect.
- 24 hours urinary VMA and catecholamines—pheochromocytoma.
- 24 hours urinary cortisol and serum cortisol levels.
- Serum calcium, thyroid function tests.
- Renal artery Doppler/angiogram for renal artery stenosis.
- Echocardiogram/ECG for evidence of coarctation of aorta/hypertensive heart disease.
- Sleep study—to rule out obstructive sleep apnea syndrome.
- MRI brain—for pituitary pathology and intracranial lesions.
- Ultrasound abdomen/CT scan for kidney size, cysts, adrenal pathology.

### Pseudohypertension and Oslerís Sign

Pseudohypertension occurs due to thickening of arterial walls and occurs in conditions like—Monckeberg's sclerosis of arteries. Manual recording of blood pressure is overestimated even though intra-arterial recording of blood pressure is normal.

### Oslerís Sign

Normally when the blood pressure cuff is inflated above systolic pressure both the radial pulse and the artery are not palpable. In patients with positive Osler's sign radial pulse is absent but vessel is still palpable when the BP cuff is inflated above the systolic pressure. This occurs in a patient of pseudohypertension.

- Right ventricular hypertrophy occurs in patients with chronic cor pulmonale.
- Right ventricular dilatation occurs in patients with acute cor pulmonale, e.g. acute massive pulmonary embolism.

### Causes

*Lung parenchymal disease*

- Chronic obstructive pulmonary disease.
- Bronchiectasis.
- Cystic fibrosis.
- Interstitial lung disease.
- Pneumoconiosis.

*Pulmonary vascular disease*

- Acute pulmonary embolism (acute cor pulmonale)
- Chronic pulmonary embolism
- Veno-occlusive disease of the lungs.
- Chronic pulmonary vascular disease.

*Conditions causing chronic hypoxia*

- Chronic obstructive pulmonary disease.
- Obesity hypoventilation syndrome.
- Neuromuscular dysfunction.
- High altitudes.
- Diseases of the chest wall.

*Clinical manifestations*

- Symptoms of primary disorder causing cor pulmonale.
- Symptoms of dyspnea, orthopnea.
- Symptoms of abdominal pain and anasarca (due to cardiac failure).

*Clinical signs*

*Signs of pulmonary hypertension*

- Prominent 'a' wave in the JVP
- Left parasternal heave—due to RVH
- $P_2$ palpable
- $P_2$ loud (narrow splitting of 2nd heart sound—except in a patient of acute pulmonary embolism)
- ESM pulmonary area
- Early diastolic murmur of PR (Graham Steell murmur)
- TR murmur.

*Signs of congestive cardiac failure—in late stages*

- JVP increased
- Enlarged tender liver
- Pedal edema
- Central cyanosis occurs either due to chronic lung disease or due to opening of patent foramen ovale
- *ECG*: P pulmonale, RVH, right axis deviation.
- *Chest X-ray*
  - Enlarged main pulmonary artery
  - RVH
  - Evidence of chronic lung disease

## CLINICAL ASPECTS OF RHEUMATIC FEVER

*Organism*: Any strain of streptococcus can cause rheumatic fever.

*Common strains responsible for rheumatic fever*:

Serotypes: 1, 3, 5, 6, 14, etc.

Rheumatic fever is more likely to be associated with HLA class II alleles DR7 and DR4.

*Immune response*: Cross-reacting antibodies attacking valvular endothelium allowing the entry of primed CD4 cells leading onto subsequent T cell-mediated inflammation.

### Pathogenesis

- Autoantibodies against cardiac endothelium.
- There is cross-reaction between streptococcus M protein and cardiac myosin.
- There is also cross-reaction between autoantibodies and neuronal cell surface gangliosides and dopamine receptors.
- There is a genetic susceptibility with more incidence with HLA class II antigens.

### Clinical Features

- 50% may not give classical history of rheumatic fever except for pharyngitis
- Usual latent period between streptococcal infection and acute rheumatic fever is 3 weeks.

*MR due to papillary muscle dysfunction*: Ischemia and acute MI causing posterior mitral leaflet involvement (posterior papillary muscles are more vulnerable to ischemia as it has got single blood supply).

*MR due to abnormality of mitral annulus*: Causes dilatation of mitral annulus causing MR (any cause of LV enlargement).

*MR due to prolapse of the leaflet*: MVP with MR.

## Pathophysiology of MR

### Chronic MR

- There will be steady increase in the volume of left ventricle with decreased function of left ventricle.
- In chronic MR—cardiac output is reduced with slow development of LV dysfunction and increase in the left atrial pressure with increase in the pulmonary venous pressure.
- Massive LA enlargement predisposes to atrial fibrillation.
- Chronic MR predisposes to endocarditis.

### Acute MR

*Causes*

- Acute MI/acute infective endocarditis.
- Rapid increase in the left atrial pressure and pulmonary venous pressure leads to rapid left heart failure.

## Clinical Features of MR

### Symptoms

*Acute MR*: Presents with acute pulmonary edema.

*Chronic MR*

- Mild to moderate MR—asymptomatic for a long period of time
- Chronic rheumatic MR—may be asymptomatic up to 15–20 years after the attack of rheumatic fever.
- Chronic severe MR—causes decrease in the cardiac output causing fatigue.
- There will be symptoms of dyspnea, PND and orthopnea due to pulmonary venous congestion.
- Irregular palpitation occurs due to AF.
- Symptoms like hemoptysis, CVA and embolism are less common compared to mitral stenosis.
- Long standing MR can result in pulmonary hypertension and right heart failure.
- Infective endocarditis may be a presenting feature of chronic MR.

### Signs

- Radial pulse—high volume pulse
- Cardiac apex—out and down and hyperdynamic
- Massive enlarged LA–pulsation in the left parastenral area.
- Systolic thrill at the apex.

*Heart sounds*

- 1st heart sound—rheumatic MR—soft
- MR with normal 1st heart sound—MVP and papillary muscle dysfunction.
- 2nd heart sound in severe MR—widely split—A2 occurs early—early aortic closure—due to less aortic flow.
- Left-sided 3rd heart sound—S3 in severe MR due to rapid filling of LV—rules out significant associated mitral stenosis.

*Murmur*

- Rheumatic—pan systolic—due to constant pressure gradient between LA and LV.
- MR with late systolic murmur—MVP and papillary muscle dysfunction
- MR in HOCM—papillary muscle is displaced anteriorly and there will be systolic anterior motion of anterior mitral leaflet.
- Cooing/seagull murmur of MR—chordae tendinae rupture.
- Musical quality of murmur—due to flail leaflet.
- MDM at mitral valve in MR—due to increase flow across mitral valve during diastole (functional narrowing of mitral valve) in severe MR.

### Important clinical points

- Mitral annular calcification occurs in elderly hypertensives and diabetes mellitus and can cause MR.