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(CASE REPORT)

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Echocardiographic detection of mesocardia, situs solitus associated with multiple congenital heart defects in a neonate: Rare case report

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Abstract

Mesocardia is a condition in which the heart is longitudinally oriented along its long axis in the midline. Cardiac position refers to the anatomical position of the left ventricular apex in the thorax. The heart has three positions: dextrocardia or right-sided heart, levocardia or left-sided heart, mesocardia or midline heart. Mesocardia is extremely rare. It represents only 0.2% of cardiac anomalies. Mesocardia is usually associated with cardiac structural abnormalities, heterotaxy syndrome, chromosomal disorders and genetic syndromes. Occasionally, it is an isolated finding.

We are presenting an interesting case report of severely symptomatic neonate afflicted with mesocardia, situs solitus, moderate sized atrial septal defect (ASD) and large ventricular septal defect (VSD).

Keywords: Mesocardia; Cardiac malposition; Inlet VSD; Cardiac position

1. Introduction

Mesocardia means "heart in the middle of the thorax". In mesocardia, the heart is centrally located in the chest (Figure 1, 2) [1].

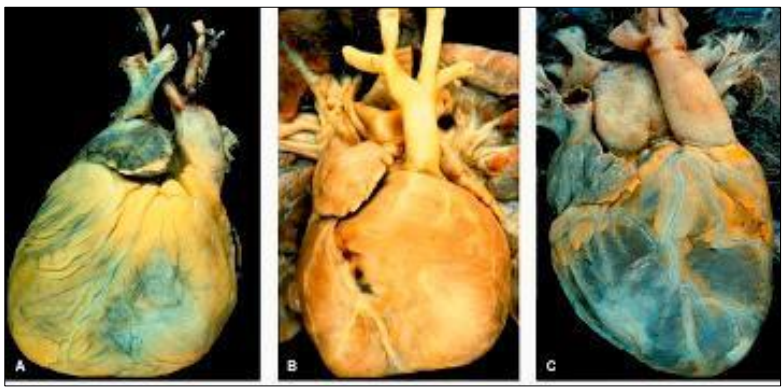

Figure 1 Pathological specimens. (A) Dextrocardia, (B) Mesocardia and (C) Levocardia

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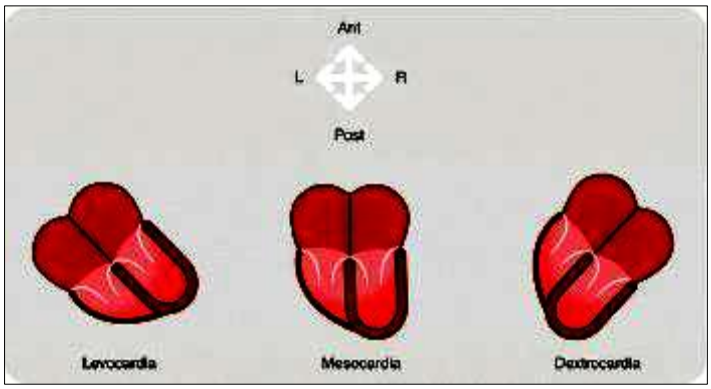

Figure 2 Diagramatic depiction of three possible cardiac situs positions: Levocardia, mesocardia, dextrocardia

Mesocardia in an uncommon cardiac abnormality, in which the heart is positioned in the center of the thorax and its longitudinal axis lies in the thoracic mid-sagittal plane.

Mesocardia is usually associated with other structural cardiac abnormalities, heterotaxy, chromosome disorders, and genetic syndromes, but occasionally it is found alone [1, 2]. Most of the reported prenatal cases are related to heart defects or extracardiac anomalies. Displacement of the heart to the thoracic midline can occur secondary to an intrathoracic mass, pulmonary abnormalities, or diaphragmatic hernia [1].

Prevalence: extremely rare: 0.2:10.000 deliveries; 0, 2% of congenital heart disease. Strong male preponderance; male/female ratio being 11:5 [3, 4].

Here, we are narrating a rare case of a 45 day male infant suffering from mesocardia, situs solitus, moderate sized ASD and large inlet VSD.

2. Imaging modalities for detection of mesocardia (Figures 3-6)

- Xray chest (PA)
- Cardiac CT
- Cardiac MR
- Transthoracic echocardiography
- Fetal Echocardiography

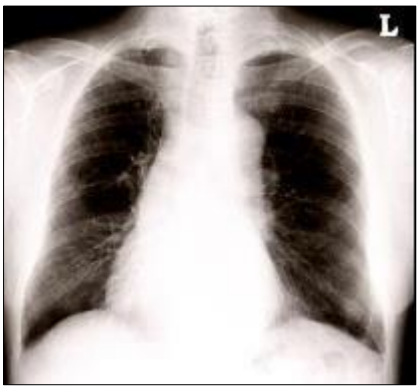

Figure 3 X-ray chest PA view identifies mesocardia

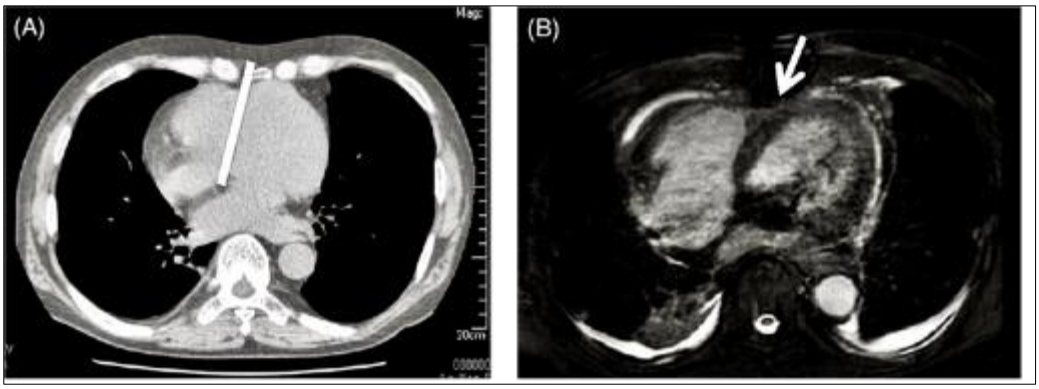

Figure 4 Computed tomography (CT) (A) and magnetic resonance imaging (MRI) (B) revealed counterclockwise rotation of the heart. Differences from the normal cardiac position comprise the right ventricle located directly over the right atrium, the interventricular septum located in an upright position, and the apex of the heart located in the mid-thorax. These findings indicated the presence of mesocardia. The white line and white arrow show the interventricular septum

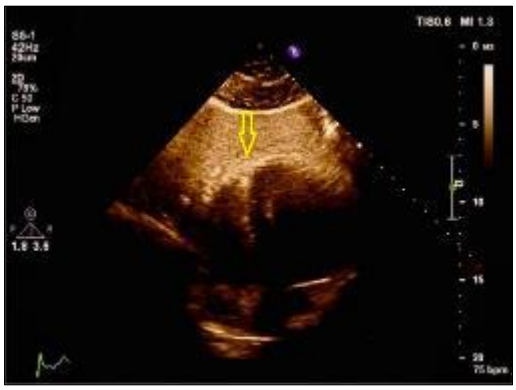

Figure 5 Transthoracic echocardiography in the subcostal view shows mesocardia. The arrow shows the cardiac apex, which is in the midline and is suggestive of mesocardia.

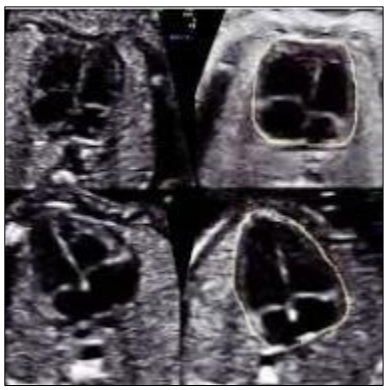

Figure 6 Fetal Echocardiography images of cardiac situs positions. Comparison of heart morphology. Transverse view in 2 fetuses with mesocardia (A and B) and 2 fetuses with levocardia (C and D). In the fetuses with mesocardia (A and B), the heart is positioned at the center of the thorax and the interventricular septum is aligned with the thoracic midline. In both cases, the hearts have a squared-off shape that contrasts with the triangular shape of the hearts in levocardia (C and D)

3. Case Report

A forty five day old male infant was referred to us for comprehensive cardiac evaluation and transthoracic echocardiography.

He was a full term normal delivery born out of non-consanguineous marriage. There was no history of maternal risk factors of CHD (obesity, diabetes, febrile illness, smoking, alcohol intake, teratogenic drug use, or radiation exposure). On clinical examination, the child was very "sick-looking" and was having severe respiratory distress, intercostal retractions and tachypnea (Figure 7). He was thin built, highly irritable and persistently crying. His weight was 4.2 kg, respiratory rate was 38/min, pulse rate was 151/mm, blood pressure was 100/70 mmHg and SPO2 was 95% at room air. There was a conspicuous pectus excavatum deformity without any other musculoskeletal anomalies. All the peripheral pulses were normally palpable without any radio femoral delay. The parents denied any bluish discolouration of the lips and mucous membranes or dusky discolouration of the skin. Likewise, clubbing was absent.

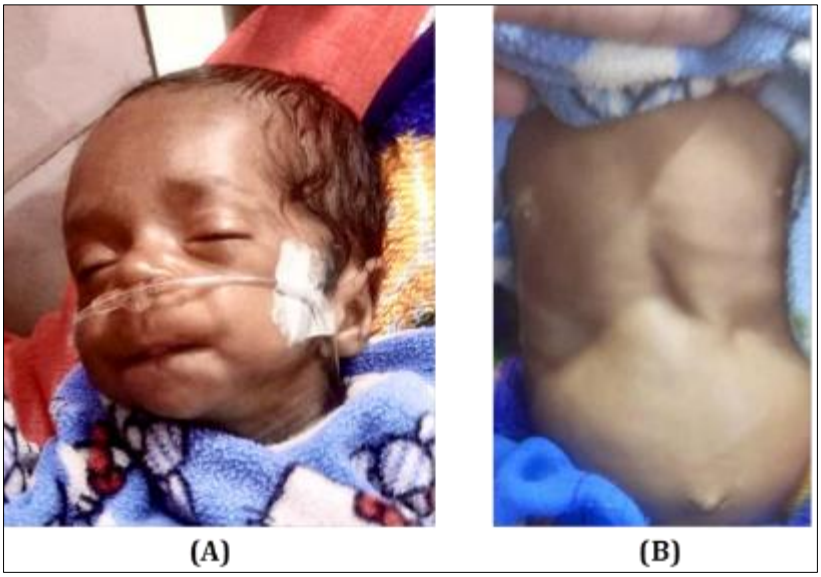

Figure 7 (A) "Sick looking" child. (B) marked respiratory distress, intercostal refraction and pectus excavatum chest deformities

On cardiovascular examination there was presence of Grade 2/6 short ejection systolic murmur heard in the pulmonary area. First and second heart sound were normal and there was no clicks or gallop sound heard. Rest of the systemic examination was unremarkable.

Xray chest AP view was consistent with mesocardia, a midline heart position. Nonetheless there was presence of cardiomegaly with evidence of increased pulmonary blood flow (Figure 8).

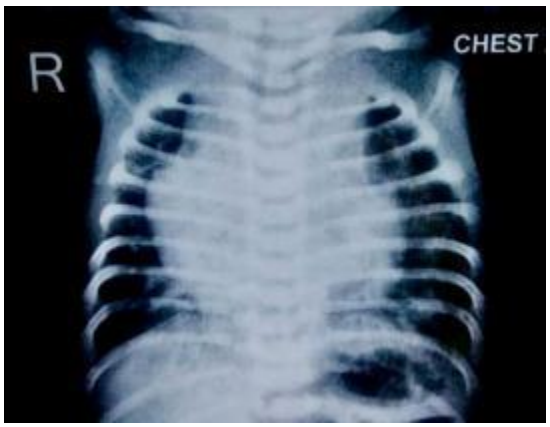

Figure 8 Xray chest A-P view. There is mesocardia, cardiomegaly, and increased pulmonary blood flow

Resting ECG revealed inverted P and QRS in lead II; positive P and QRS waves in AVR (Figure 9A) and reverse progression of QRS from leads V_1-V_6 (Figure 9B).

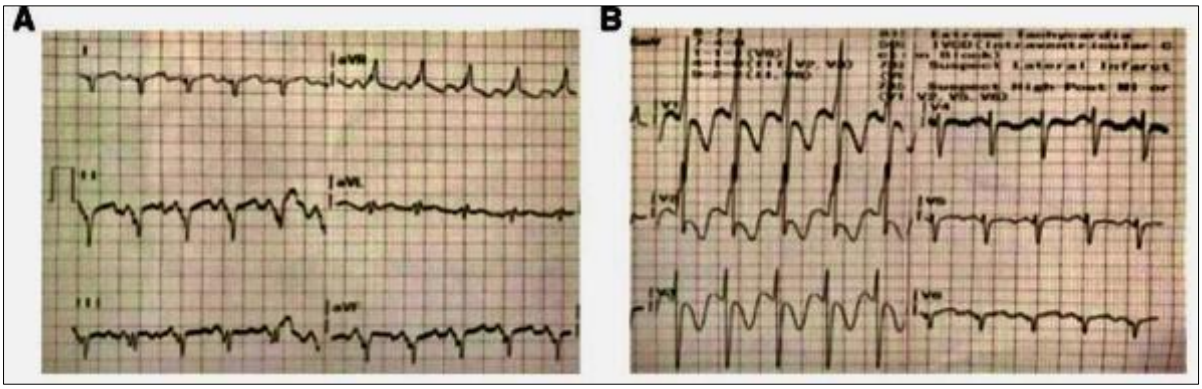

Figure 9 (A) Inverted P and QRS complex in lead II; positive P and QRS waves in AVR. (B) Reverse progression of QRS from leads V_1-V_6

4. Transthoracic Echocardiography

All echocardiography evaluations were performed by the author, using My Lab X7 4D XStrain echocardiography machine, Esaote, Italy. The images were acquired using a pediatric probe equipped with harmonic variable frequency electronic single crystal array transducer while the subject was lying in supine, left lateral decubitus and also frequently in right lateral decubitus positions.

Conventional M-mode, two-dimensional and pulse wave doppler (PWD) echocardiography and continuous wave doppler (CWD) echocardiography was performed in the classical subcostal, parasternal long axis (LX), parasternal short axis (SX), 4-Chamber (4CH), 5-Chamber (5CH) and suprasternal views. Contemporary sequential segmental approach for echocardiographic analysis of our index patient was accomplished and the characteristics are outlined (Figures 10-12):

4.1. M-mode Echocardiography

The features of M-mode echocardiography are mentioned:

Table 1 Features of M-mode echocardiography

LV	
IVS d	4.1 mm
LVID d	19.1 mm
LVPW d	3.4 mm
IVS s	5.2 mm
LVID s	12.2 mm
LVPW s	6.4 mm
EF	69 %
%LVFS	36 %
LVEDV	11.4 ml
LVESV	3.5 ml
SV	7.8 ml
LV Mass	10 g

4.2. 2-Dimensional Color Echocardiography

- Mesocardia
  - Situs solitus
  - Concordant D-bulboventricular loop
  - AV concordance
  - VA concordance
  - NRGA
  - Left aortic arch, confluent pulmonary arteries.
  - Normal systemic venous drainage.
  - Normal pulmonary venous drainage.

- Atrial septal defect (moderate)
  - Size : 3.4 mm.
  - Ostium secundum type.
  - Peak/mean gradient across ASD = 5.3/2.8 mmHg.
  - Lt to Rt shunt.
- Ventricular septal defect (large)
  - Size : 6.3 mm.
  - Inlet type.
  - Peak gradient across VSD = 20 mmHg (peak velocity 2.24 m/sec)
  - Lt to Rt shunt.
- Qp/Qs ratio = 3.5:1.
- Normal biventricular dilation with normal systolic function.
  - Normal LVEF = 69 %
  - No regional wall motion abnormality seen.
- Moderate PAH (estimated RVSP/PAP =45 mmHg).
- No evidence of PDA, COA, AS, PS.

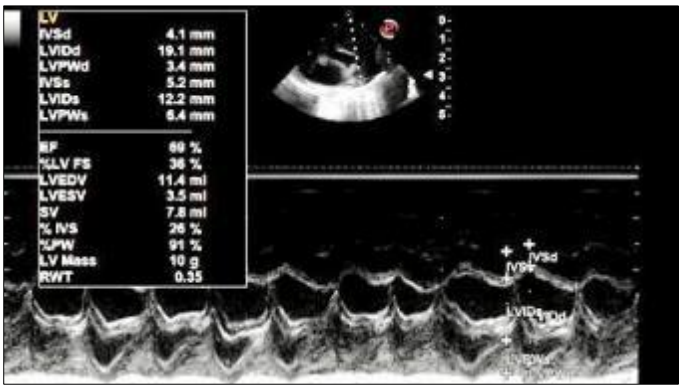

Figure 10 M-mode echocardiography. There was left ventricular dilation and normal LV systolic functions

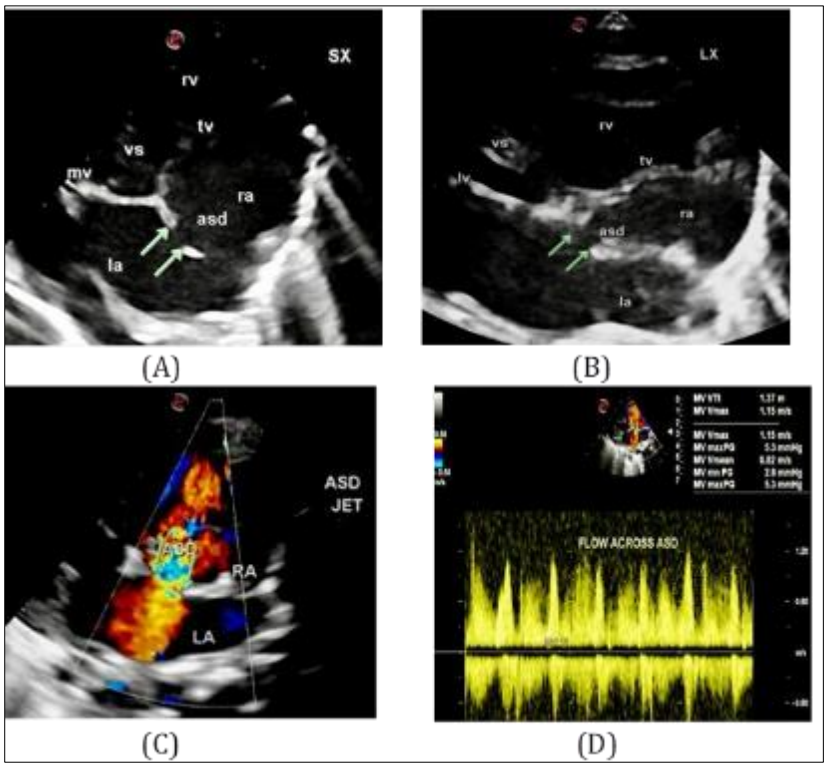

Figure 11 2-Dimensional Transthoracic Echocardiography: (A) SX View shows a moderate size ostium secundum ASD. (B) LX view reveals the same ASD. (C) Color Echocardiography shows a turbulent mosaic pattern jet across the ASD. (D) Pulse wave doppler (PWD) across ASD depicts a peak and mean gradient of 5.3/2.8 mmHg

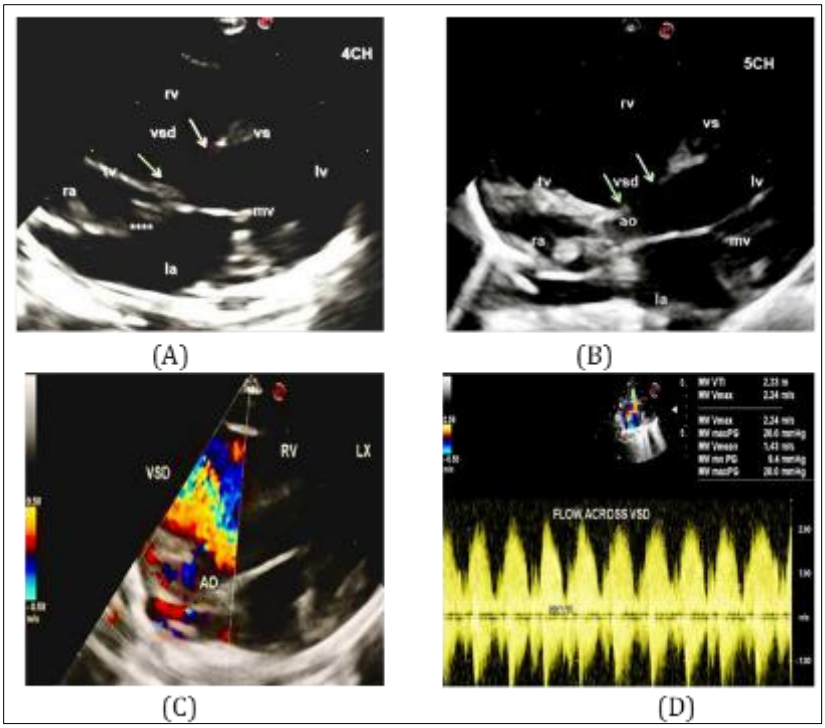

Figure 12 (A) 4CH View and (B) 5CH View demonstrates a large (6.3 mm) inlet VSD. (C) On Color flow mapping, a turbulent mosaic pattern jet is recognized across the VSD with a left to right shunt. (D) On CWD analysis a peak gradient across the VSD was 20 mmHg

In view of severely symptomatic infant with presence of moderate ASD alongwith large PDA, Qp/Qs ratio of 3.5:1 and moderate pulmonary hypertension; the parents were advised to consult a tertiary care pediatric cardiovascular institute for suitable corrective surgical procedure to cure the infant of the current illness.

5. Discussion

Mesocardia is that condition in which the longitudinal axis of the heart lies in the mid-sagittal plane and the heart has no apex [5]. It is a very rare congenital cardiac anomaly wherein the cardiac apex points to midline. It is seen in 0.2 of 10,000 deliveries and 0.2% of congenital heart disease, and has a strong male preponderance with the male to female ratio being 11:5 [3, 4]. Adult cardiac surgeries with mesocardia have been extremely rare [6].

5.1. Cardiac Axis

The cardiac axis describes the orientation of the apex of the heart within the thorax (Figures 13) [7].

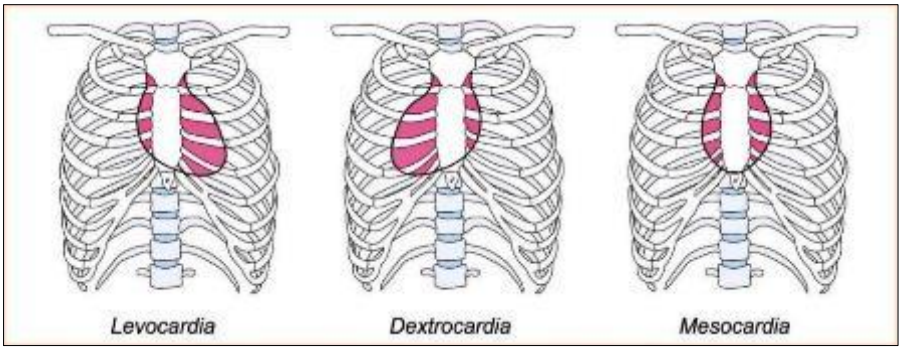

Figure 13 Cardiac position within the thorax. In levocardia, the heart is predominantly in the left hemithorax. In dextrocardia, the heart is predominantly in the right hemithorax. In mesocardia, the heart is midline and the apex typically points anteriorly and inferiorly

5.2. Terms used to describe cardiac axis orientation [7]:

Levocardia: The cardiac apex points to the left (normal). Greater than 57 degrees of leftward axis deviation may be considered abnormal.

Mesocardia: The cardiac apex points to the midline.

Dextrocardia: The cardiac apex points to the right.

Cardiac Position

Cardiac position describes the location of the heart within the thorax.

Terms used to describe cardiac position:

Levoposition: The heart is in the left thorax.

Dextroposition: The heart is in the right thorax.

Mesoposition: The heart is in the middle thorax

Mesoversion Type I: heart vertical and central with both mixed mesocardia with ventricular inversion and mixed mesocardia with atrial inversion.

Mesoversion Type II: inversion of both right and left chambers.

Etiology: Cardiac location is affected by many factors including underlying cardiac malformation, abnormalities of mediastinal and thoracic structures (eg., Bronchogenic cysts, lung mass, tumors, esophageal atresia, diaphragmatic

hernia, kyphoscoliosis, abnormalities of the diaphragm, etc.), Otherwise mesocardia reflects that embryonic malrotation before the formed heart points to the right or left and acquires an apex, situs viscerum inversus [3, 8].

Pathogenesis: Bulboventricular loop A portion of the embryonic cardiac tube that grows so that th tube assumes an S-shape. The ventricles evolve from the loop. The type of bulboventricular loop describes the relationship of the ventricles to eachother. In a D-loop (dextro loop) the morphological right ventricle is located to the right of the morphological left ventricle. In L-loop (levo loop) the morphological right ventricle is located to the left of the morphological left ventricle [3, 8].

Comparison of the morphology of hearts in mesocardia with those in levocardia shows a more rounded left ventricle than normal, without the typical inverted cone shape. The tip of the heart does not have the usual apical form, resulting in a morphology different from that of a heart in levocardia [2].

In 1971, Lev et al [5] described the anatomic characteristics of midline hearts. These authors performed autopsy study of 13 cases and reported that the abnormal morphology has an embryologic origin. In normal conditions, after the heart has completely formed and reached a size of 20 to 25mm, there is no true apex and it has a shape similar to that of a heart in mesocardia. In the following phase, the heart points toward the left or right and attains an apical shape. Lev et al. reported that the apex may be hidden or "absent" in midline hearts. In the area where the apex should be, there is a wide stretch of ventrally oriented muscle fibers. It is this distribution that confers the tear shape seen on chest radiology studies.

Allan et al [9]. reported that the cardiac apex points in an anterior direction at 9 weeks and rotates toward the fetal left at 11 weeks. McBrien et al [10]. evaluated the cardiac axis in 188 fetuses between 8 and 15 weeks of gestation and also concluded that the fetal cardiac axis has a midline position at 8 weeks' gestation and undergoes levorotation at the end of the first trimester. The normal, levorotated position is attained at approximately 12 weeks.

The looping that occurs in the fetal cardiac axis at the end of the first trimester is one of the last phases of cardiac embryogenesis. During normal looping, a complex series of changes takes place in the position of the heart [11]. At the beginning of this process, the ventricles have a craniocaudal relationship. In the final phase, the heart rotates in a counterclockwise direction around the basal-apical axis, with the right ventricle moving ventrally. This final phase and the relatively late changes occurring in the shape of the left ventricle can explain the change in the cardiac axis seen in early fetal echocardiography [10]. Thus, midline hearts that do not complete this final embryological phase show both a change in the cardiac axis and an abnormal apical morphology.

Persistence of the LSVC tends to be more common in hearts in mesocardia than those in levocardia associated with congenital heart disease [5], Of the 13 cases described by Lev et al [5], (46%) were associated with this vascular anomaly. Development of the innominate vein and LSVC involution occur at the end of cardiac morphogenesis, at the same time as looping of the cardiac axis.

In most cases, mesocardia is associated with major cardiac or extracardiac defects; hence, meticulous study of the fetal anatomy should be performed [3, 7, 8, 12-14].

5.3. Major cardiac or extracardiac anomalies associated with mesocardia

Myriads of cardiac and extracardiac defect associated with mesocardia [3, 7, 8, 15-18] are detailed:

5.3.1. Thoracic defect

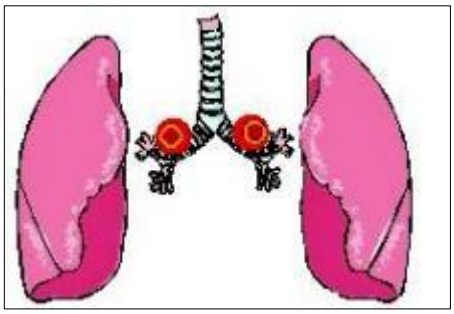

Figure 14 Thoracic defects

- Situs inversus
- Bilobed lung, bilateral
- Polylobulation of both upper lobes of lung
- Abnormal lobation of lungs
- Incomplete lung transverse fissure
- Kyphoscoliosis
- Spondylocostal dysplasia
- Lung mass
- Bronchogenic cyst
- Thymic mass

5.3.2. Abdominal situs

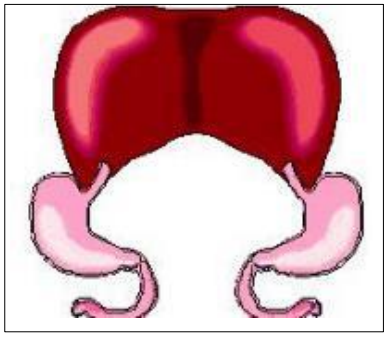

Figure 15 Abdominal situs

- Solitus
- Inversus
- Ambiguous

5.3.3. Spleen

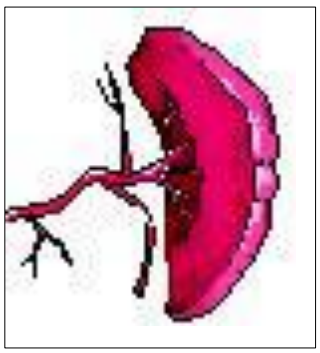

Figure 16 Spleen

- Asplenia
- Polysplenia
- Accessory spleen

5.3.4. Heart

Figure 17 Heart

- Coronary artery distribution
- Dominant left
- Ostia exchange vessels
- Ostia rotated
- Ventricular septal defect
- Atrial septal defect
- Tricuspid stenosis
- Pulmonary stenosis
- Complete transposition
- Partial inverted transposition
- Partial transposition with common A-V orifice
- Common A-V
- Persistent ostium primum complex
- Single ventricle
- Ebstein's anomaly
- Bicuspid pulmonic valve
- Bicuspid aortic valve
- Right subclavian from descending aorta
- Absent coronary sinus
- Complete A-V block
- Cleft aortic leaflet of mitral

5.3.5. Chromosomal anomaly

- Trisomy 18
- Trisomy 13

5.3.6. Other

- Esophageal atresia
- Diaphagmatic hernia
- Hydrops Fetalis
- Renal agenesis

Prognosis: Good for isolated form, without associated anomalies [7].

Recurrence risk: Unknown. Depend upon the underlying etiology in the secondary form [7].

6. Conclusion

Mesocardia is one of the rarer outcomes of abnormal morphogenesis of the heart. It is often associated with other congenital cardiac and extracardiac defects of vasculature as well as organ isomerism. While extremely rare and with

questionable prognostic significance, its association with other cardiac and vascular anomalies warrants a workup for the identification of such associated conditions.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest statement to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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