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Case Report

A rare case of isolated dextrocardia

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ABSTRACT

Dextrocardia is a clinical condition in which heart is located on the right side of the chest cavity. Dextroversion is a result of early interruption of normal embryological development and therefore, rarely occurs without other cardiac and noncardiac anomalies. Isolated dextrocardia is a benign condition. It can often be associated with other congenital anomalies which can lead to failure to thrive, breathing difficulties, recurrent infections in new borns and neonates. It presents in situs solitus and situs inversus. Among them Dextrocardia with situs solitus is very rare. We are here presenting a case of dextrocardia with situs solitus.

1. Introduction

In an embryo, the heart is first organ to develop, which develops from an embryonic heart tube-formed by fusion of the endocardial tubes. The cranial aspect to the arterial trunk is attached by the heart tube, and the caudal aspect is connected to the venous channels. Then after formation of the heart tube, the important step in the development of the heart is looping. The position of the ventricle in relation to the atria is determined by looping. The heart tube may loop to the left (L-loop), or loop to the right (D-loop). With L-loop the morphological right ventricle is positioned to the left of the left ventricle, whereas, with a D-loop, it lies to the right of the left ventricle.[1–3] If a D-bulboventricular loop fails to migrate into left hemithorax, with the heart in the right hemithorax, it can result in dextrocardia.

The very first step in ultrasonic evaluation of fetal heart is fetal situs assessment. Fetus situs establishes the accurate determination of the ventricular and atrial situs.

There are three types of situs, which may exist are: 1) Situs solitus, 2) Situs inversus, 3) Situs ambiguous.

Table 1: Clinical findings related to different types of situs

Situs	Findings	
	Right side	**Left side**
Solitus	Right atrium	Left atrium
	Major hepatic lobe	Stomach
	Inferior vena cava	Descending aorta
	Trilobed lung	Bilobed lung
Inversus	Left atrium	Right atrium
	Stomach	Major hepatic lobe
	Descending aorta	Inferior vena cava
	Bilobed lung	Trilobed lung
Ambiguous	Variable	Variable

The fetal cardiac axis can be readily determined by obtaining a transverse view of fetal chest at the level of four-chamber plane of the heart. A straight line drawn from the spine to the anterior chest wall divides the chest into two equal halves. The angle that the interventricular septum line makes with this line is known as the cardiac axis. Normally the heart is deviated about 45± 20º (2 SD)towards the left

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side of the fetus.[4]

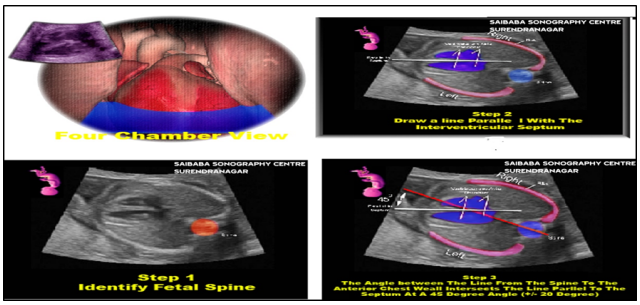

Fig. 1: Steps to determine fetal cardiac axis

Dextrocardia is used to describe a condition when an anomaly of embryological development is present. It is a rare congenital disorder in which the heart resides on the right side of the thoracic cavity. In most cases, it is diagnosed incidentally. Some developmental anomalies are often associated with this. Dextrocardia can occur by itself or it can be accompanied by a reversal in the position of other organs which is termed as situs inversus totalis.[5] Dextrocardia is a condition in which orientation of the heart changes with its base to the apex axis being directed to right, in contrast to normal heart orientation where apex is directed towards the left side. This orientation change differentiates it from cardiac dextroposition in which the heart is displaced to the right side as a result of some extra cardiac causes, such as a diaphragmatic hernia, right pneumonectomy, or right lung hypoplasia.[6] The intracardiac anomaly is also reversed in typical dextrocardia is termed as situs inversus. When there is association of dextrocardia with a normal position of other thoracoabdominal structures, it is termed as situs solitus.[7]

Dextrocardia is a very rare condition and incidence rates of dextrocardia have been revealed by studies to be around 1 in 12000 pregnancies.[8] Isolated dextrocardia with primary ciliary dyskinesia was seen in 0.6% of patients.[9] For dextrocardia there has been no ethnic or gender-related predilection described. Primary dextrocardia is most common with situs solitus (45.4%), situs ambiguous(36.3%) and then situs inversus totalis (18.1%).Structural cardiac malformation were found in 100%, 80% & 25% of fetus with situs ambiguous, situs solitus and inversus respectively, suggest wide spectrum of complex cardiac malformation are associated with fetal dextrocardia.[10] The exact etiology of dextrocardia is unknown; therefore it is during embryonic development thought to be secondary to the abnormal position of the organs. Kartagener syndrome is an inherited disorder of ciliary motility which is caused by an autosomal recessive mutation and is associated with chronic sinusitis, bronchiectasis and dextrocardia.[11]

2. Case Report

Here we present a case of Isolated Dextrocardia which is identified at 24-25 weeks of gestation. A 26 year old patient Gravida 2 Para 1 Live 1 (Female child 4 year of age at time of sonography), came for Anomaly scan at our centre. Her married life was 6 year, non-consanguineous marriage. Her personal and family history was unremarkable. Her last menstrual period (LMP) date was 29^{th} March 2018. During ultrasonography, in situs evaluation, we observed apex of heart was on right side and stomach, spleen were on left side. Detailed ultrasonography and fetal echocardiography was done which showed no positive finding other than Dextrocardia. Fetal biometry was compatible with gestational age. On physical examination of patient; her blood pressure, pulse, SpO2 and respiratory rate was normal. Female child delivered vaginally at 39 weeks of gestation. Primarily chest X-ray was advised to our baby which was suggested Dextrocardia. Rest investigation refused by patient because of a healthy baby. Baby is right now 3 years old and healthy. Here are pictures of ultrasonography at 24-25 weeks of gestation age, chest X-ray after delivery and baby's recent photograph.

2.1. Isolated dextrocardia

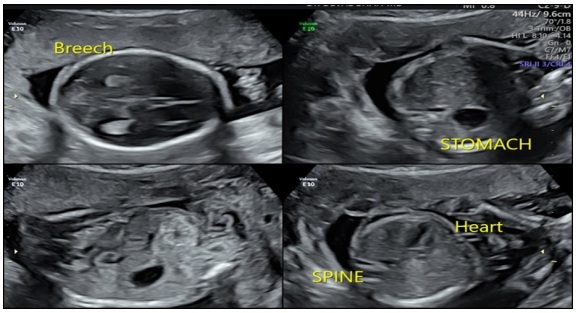

Fig. 2:

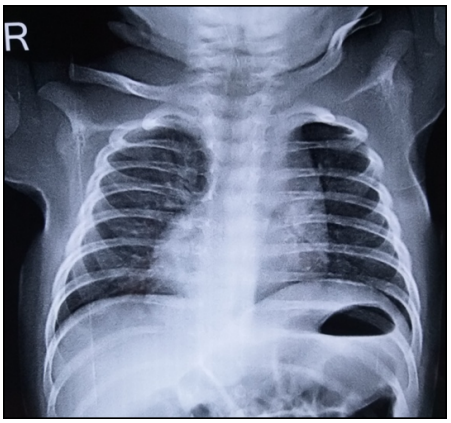

Fig. 3:

Fig. 4:

3. Discussion

There are three types of situses. In situs solitus, major part of liver, IVC & SVC, right atrium & its appendage, tri-lobed right lung with eparterial bronchus on right side and stomach, spleen, left atrium and its appendage, pulmonary vein, bi-lobed left lung with hyparterial bronchus on right side. Situs inversus totalis, mirror image arrangement of above explained structure to that of situs solitus. In Situs ambiguous, any arrangement of visceral &/or thoracic organs other than situs solitus or situs inversus.

In third week post conception, the embryo consists of three basic germ layers: ectoderm, mesoderm, and endoderm. The mesoderm differentiates into four compartments which are as follows: axial, paraxial, intermediate and lateral. The lateral mesoderm is involved in formation of circulatory system and viscera. In this lateral splanchnic mesoderm, clusters of angiogenic cardiac precursor cells develop form cardiogenic plate bilaterally.[12] Bilateral cardiogenic plate fuses to form midline primitive heart tube.[13] If heart tube bends to left instead of right there is transposition, in which the heart and its vessel are reversed left to right forms dextrocardia.[14]

Dextrocardia is characterized by the location of the heart on right side of the chest cavity clinically. Though isolated dextrocardia is benign, it can also often be accompanied with other congenital anomalies, which may lead to several problems.

Isolated dextrocardia is asymptomatic, whereas dextrocardia with situs inversus and kartagener syndrome can have symptoms depending on accompanying abnormalities as follows:[15–17]

• Cyanosis	• Repeated sinus or lung infections
• Dyspnoea	• Hydrocephalus
• Failure to thrive	• Arrhythmias, especially atrioventricular (AV) blocks
• Fatigue	• Intestinal obstruction
• Jaundice	
• Pallor	
• Decreased exercise tolerance	

The physical examination of a dextrocardia patient might reveal clubbing, cyanosis, a prominent apical impulse on the right side of the chest and easily audible heart sounds on the right side of the chest.

It is often diagnosed incidentally on the routine radiological examination which shows an abnormal location of the heart. The electrocardiogram can also reveal the presence of dextrocardia which is evidenced by right axis deviation, inversion of all complexes in lead I, upright p wave in AVL and an absent R wave progression in anterior leads.[18] For further evaluation of dextrocardia and associated anomalies echocardiogram, computed tomography (CT) scan of the chest and magnetic resonance imaging (MRI) of chest might help.[19,20] For diagnosis of kartagener syndrome nasal brush biopsy and nasal nitric oxide measurements screening tests are used.[21]

Most of the patients with isolated dextrocardia are asymptomatic and may lead to a normal life. Management of dextrocardia depends on the presence of associated congenital anomalies. In patients with dextrocardia having other associated cardiac malformations such as defects of walls of the heart, malposition of the blood vessels, tetralogy of fallot, septal defects, and severe valvular abnormalities; surgical reconstruction might need to be considered.[22,23]

Differential diagnosis of dextrocardia are cardiac dextroposition in which heart is displaced to right side as a result of extra cardiac causes such as diaphragmatic hernia, right pneumectomy or right lung hypoplasia; kartagener syndrome in which dextrocardia situs inversus is accompanied by primary ciliary dyskinesia; dextroversion in which heart is abnormally positioned to right and rotated; transposition of great vessels (TGA) in which major vessels of heart connect in reverse with a reversal of the heart chambers; heterotaxy; endocardial cushion defect.

The prognosis of dextrocardia patients depends on the presence or absence of any other accompanying congenital defects and the type of congenital anomalies. As mentioned earlier, patients with isolated dextrocardia will have a normal life span without any major complications.

4. Conclusion

If Dextrocardia (isolated dextrocardia) presents without any cardiac or extra cardiac anomaly, prognosis is good,

irrespective of situs solitus or situs inversus totalis, although incidence are very rare.

5. Source of Funding

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6. Conflict of Interest

None

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