A Case of Mirror Image Dextrocardia with Ostium Secundum Atrial Septal Defect

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ABSTRACT
Dextrocardia when associated with situs inversus usually has no structural defects. A case of dextrocardia with situs inversus and atrial septal defect (ASD) in a 14-year-old boy born of consanguineous marriage is reported because of its rarity.

Keywords: Dextrocardia, Situs inversus, Atrial septal defect.

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INTRODUCTION
The normal position of heart is situs solitus and levocardia. On the other hand, dextrocardia indicates the heart is mainly on the right and its apex points to the right. Dextrocardia when associated with situs inversus usually has no structural defects. A case of dextrocardia with situs inversus and atrial septal defect (ASD) in a 14-year-old boy born of consanguineous marriage is reported. The interest of this case lies in the rare association of congenital heart disease (CHD) in dextrocardia with situs inversus.

CASE REPORT
A 14-year-old male patient born of second degree consanguineous marriage presented with complaints of progressive breathlessness on exertion since past 4 months, associated with intermittent episodes of palpitations and swelling of feet. There was no orthopnea or paroxysmal nocturnal dyspnoea. There was no history of recurrent respiratory tract infection.

The patient is first child born by full term normal delivery. The mother’s antenatal period was uneventful. There was no history of fever or rash or exposure to drugs or radiation during pregnancy. The patient achieved normal developmental milestones and is presently doing well in school. The second child is 12-year-old male born of full term normal delivery; he is apparently normal.

On examination, the patient is moderately built and nourished. Pulse: 80 beats per minute, regular, normal volume, normal character, with no radio-radial or radio-femoral delay. Blood pressure: 110/70 mm Hg. Ankle edema: present; no pallor, icterus, cyanosis or clubbing.

Systemic examination—shape of chest wall is normal, the apical impulse visible in 4th intercostal space in right midclavicular line. No visible parasternal pulsations, dilated veins, scars or sinuses. On palpation apical impulse felt in 4th right intercostal space in mid clavicular line, normal in character. No palpable P2. On percussion, his cardiac borders corresponded to 2nd intercostal space on left parasternal, 2nd intercostal space on right parasternal and lower corresponded to right 4th intercostal space in midclavicular line. Liver dullness is in left 6th intercostal space in midclavicular line and tympanitic note is heard in right 7th intercostal space. On auscultation, S1 and S2 normal heard in right 4th intercostal space in midclavicular line; P2 heard loud in right 2nd intercostal space with ejection systolic murmur and wide fixed split A2 and P2. Per abdomen examination revealed liver in left hypochondrium, palpable 2 cm below left costal margin in midclavicular line. Rest of the systemic examination—unremarkable.

INVESTIGATIONS

Chest X-ray
Dextrocardia with situs inversus. The heart is in the right hemi-thorax with apex to the left.

Gastric air bubble is visualized under the right hemidiaphragm which is slightly at the lower level than the left hemi-diaphragm and liver shadow on left side (Fig. 1).

Electrocardiography
Complete right bundle branch block (RBBB) pattern with QRS complexes tallest in lead V1 and diminishing progressively toward lead V6, lead I and a VL showing cavity complexes, i.e. P wave and T wave inverted and QRS having dominant downward deflection.
Ultrasonography of Abdomen
Liver and gallbladder are in left hypochondrium, spleen is in right hypochondrium.

CT Abdomen
Liver is on left side and spleen is on right side. Aorta is located on right side, inferior vena cava (IVC) on left side.
There is accessory spleen on right side. Pancreas head is located on left side and body and tail on right side. Stomach is on right side, well distended. Kidneys are normal, ureters normal. Small bowel and large bowel are normal.

HRCT Chest
Trachea and bronchi appear normal. Mediastinum: dextrocardia noted with aorta on right and superior vena cava on left side. Mirror image of tracheobronchial anatomy with long left principle bronchus. No evidence of bronchiectasis. Chest wall is normal.

2D Echo and Color Doppler
Situs inversus with dextrocardia. Ostium secundum atrial septal defect measuring 1 cm with left to right shunt. Right atrium and right ventricle dilated. Normal pulmonary venous drainage (Figs 2A and B).
Normal systemic venous drainage. Interventricular septum is intact. Atrioventricular and ventriculoarterial concordance (aorta arises from the left ventricle and pulmonary artery arises from the right ventricle); AV valves are normal structurally and functionally. No regional wall motion abnormality. Normal left ventricular ejection fraction (LVEF) 60%. No hepatic congestion. No pulmonary arterial hypertension.

Karyotyping: His genetic karyotyping was normal.

DISCUSSION
Dextrocardia is a common cardiac malposition anomaly. This term refers to the condition in which the heart is in the right hemithorax and the left sided chambers are on the right, the left ventricular apex being palpable on the right side. Dextrocardia may be associated with situs inversus in which other organs, such as liver and stomach are also abnormally placed, i.e. liver is on the left and stomach is on the right. Such dextrocardias are termed mirror-image dextrocardias.

Incidence of congenital cardiac anomalies in dextrocardia with situs inversus is low as compared to congenital cardiac anomalies in isolated dextrocardia. Total situs inversus or mirror image reversal of visceral asymmetry itself is an uncommon condition. On review of literature, the incidence approximately appears to be 1:10,000 adults and although it appears to be genetically determined, the exact mode of inheritance is not clear. 1

Situs inversus with dextrocardia is a type of cardiac malposition most likely to exist with structurally normal heart.
Associated cardiac anomalies described in patients of dextrocardia with situs inversus are: ventricular septal defect (VSD), ASD, complete AV canal defect, pulmonary atresia (PA), tetralogy of fallot (TOF) and double outlet right ventricle (DORV).

Kulkarni and Inamdar from Government Medical College, Nanded, reported a case of large perimembranous VSD associated with dextrocardia and situs inversus.\textsuperscript{4}

In a population-based study of cardiac malformations and outcomes associated with dextrocardia, cardiac malformations were found in 96\% cases of situs solitus, 23\% cases of situs inversus, and 100\% cases of situs ambiguous or isomerism.\textsuperscript{2}

In dextrocardia with situs solitus the abdominal organs are not reversed (stomach on left and liver on right). Such dextrocardias are usually associated with multiple cardiac anomalies. The common congenital cardiac anomalies associated with isolated dextrocardia are atrial situs solitus (93\%), discordant AV connection (44\%), and discordant ventriculo-atrial (VA) connection (30\%).\textsuperscript{3}

Dextrocardia with situs inversus, though mostly having structurally normal heart poses a special problem in angina pectoris and acute myocardial infarction (AMI), where chest pain is felt on the right side of the chest and radiates to right arm. Cardiac interventions like transcatheter closure of PDA, ASD, VSD and procedures like balloon mitral valvotomy (BMV) also pose orientation problems for the operator.

The present case is a mirror-image dextrocardia with ostium secundum atrial septal defect, which makes it a very rare condition.

Cardiac catheterization study and ASD device closure is awaited in this case.

**CONCLUSION**

The presence of atrial septal defect in dextrocardia with situs inversus is a rare clinical condition. Can be easily diagnosed by clinical examination and basic investigations? The treatment is similar to atrial septal defect with situs solitus. The development of later complications needs to be evaluated. The incidence with consanguineous marriage needs to be investigated and the parents have to be advised accordingly.

**REFERENCES**