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Case Report

Incidental Finding of Dextrocardia with Situs Inversus in A 4-Year-Old Child: A Case Report

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Abstract

Dextrocardia with situs inversus means a situation in which vital organs in the chest and abdomen are on the opposite side (most often right side) of the body from the normal location. The heart is positioned abnormally where the tip is positioned on the right side of the chest, and visceral organs like the liver and spleen appear reversed (situs inversus).

This is a four-year-old male child that presented for chest radiograph on account of cough and fever with a diagnosis of bronchopneumonia. Following the plain radiograph, the tip of the heart was noticed on the right side of the chest, the gastric and splenic shadows were also noted on the right hemi-abdomen and the hepatic shadow on the left hemi-abdomen. A complimentary abdominal ultrasonography further confirmed the location of the spleen and stomach on the right hemi-abdomen while the liver on the left. An echocardiogram further confirmed the change in normal positioning of the chambers of the heart.

We present a case of dextrocardia with situs inversus totalis incidentally diagnosed in a four-year-old male child because of its rare nature and to review the literature.

Keywords: situs inversus: dextrocardia: liver: spleen

Introduction

Dextrocardia with situs inversus is often present at birth, this condition has no sex predilection, since both males and females are equally affected. This must be differentiated from conditions like dextroversion in which the tip of the heart is located on the right side of the chest but the left ventricle is remains on the left with abnormality on the electrocardiogram, and dextroposition of the heart where the tip of the heart is on the right side of the chest with a normal electrocardiogram and caused by acquired diseases of the lungs, pleura or diaphragm^{1-3.}

Dextrocardia has an estimated incidence of about 1/10000 live births with an unknown definitive cause, but has been linked with some etiologies like autosomal recessive gene inheritance, maternal diabetes, conjoined twinning and cocaine use^{1,4-6}.

Situs inversus totalis is a rare clinical condition in which there is a complete mirror image reversal of the normal positioning of the internal organs^{7.}

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Dextrocardia with situs inversus totalis may be associated with some congenital anomalies, some of which are atrial situs solitus, discordant ventrculo-Atrial connection, polysplenia and Kartegener's syndrome (primary ciliary dyskinesia). Katergener's syndrome which is seen in about 25% of individuals with situs inversus comprises of situs inversus, chronic sinusitis and bronchiectasis^{1,8,9}. Dextrocardia with a normal abdominal situs is known to have a higher association with congenital heart disease¹⁰.

Individuals with dextrocardia and situs inversus are most often asymptomatic and most times lead a normal life, only occasionally do some present with chest findings like bronchiectasis often associated with Katagerner's syndrome^{10,11}.

Dextrocardia with situs inversus can be diagnosed by imaging, some of the imaging methods are plain radiography, ultrasonography, electrocardiography, echocardiography, barium studies, computed

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tomography (CT) and magnetic resonance imaging (MRI), with CT and MRI being particularly important in the diagnosis and assessment and demonstrating the mirror anatomy of the viscera in dextrocardia and situs inversus⁷⁻¹⁰.

Most individuals with dextrocardia do not display symptoms and subsequently require no treatment, except where anomalies like heart defects occur, treatment is required and directed at the cause following appreciation of associated pathogenesis and appropriate diagnosis¹².

Case Report:

This is a four-year-old male child that presented for chest radiograph on account of cough and fever with a diagnosis of bronchopneumonia. Past medical history was not significant in this case.

Physical examination revealed a stable child, without features to suggest anemia, dehydration, cyanosis and jaundice. The cardiac apex beat was palpated at the fifth right intercoastal space.

Following the plain radiograph, the apex of the heart and aortic arch were noted on the right side of the chest, the gastric fundal gas and splenic shadows were also noted on the right hemi-abdomen and the hepatic shadow on the left hemi-abdomen (Figures 1 and 2). A complimentary abdominal ultrasonography further confirmed the location of the spleen and stomach on the right hemi-abdomen while the liver on the left. The pancreatic head and inferior vena cava were demonstrated on the left hemi-abdomen, while the pancreatic tail and abdominal aorta were noted on the right hemi-abdomen. An echocardiogram further confirmed the change in normal positioning of the chambers of the heart.

A diagnosis of dextrocardia with situs inversus was made in this fouryear-old male child that presented with symptoms suspicious of bronchopneumonia, we decided to present the case due to its rare nature.

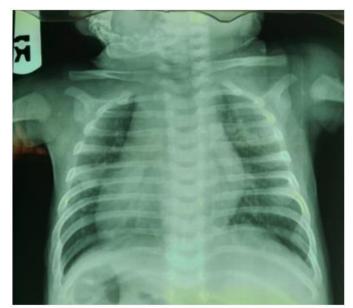


Figure 1: An anterior-posterior chest radiograph of a child demonstrating a displaced apex of the heart to the right in keeping with dextrocardia (left blue arrow).



Figure 2: An anterior-posterior chest radiograph of a child demonstrating a displaced apex of the heart to the right (right blue arrow), gastric fundal (right yellow arrow) and splenic (right red arrow) shadows on the right with the hepatic shadow (left blue arrow) on left in keeping with dextrocardia with situs inversus.

J. Thoracic Disease and Cardiothoracic Surgery **Discussion:**

Dextrocardia with situs inversus is often present at birth, this condition has no sex predilection, since both males and females are equally affected¹⁻³. The index case is a male child of four years with features of dextrocardia and situs inversus and most likely from birth, thereby conforming to these literatures.

Dextrocardia must be differentiated from conditions like dextroversion in which the tip of the heart is located on the right side of the chest but the left ventricle remains on the left with abnormality on the electrocardiogram, and dextroposition of the heart where the tip of the heart is on the right side of the chest with a normal electrocardiogram and caused by acquired diseases of the lungs, pleura or diaphragm¹⁻³. The index case had features suggestive of true dextrocardia clinically and following imaging ruling out differentials like dextroversion and dextroposition.

Dextrocardia has an estimated incidence of about 1/10000 live births with an unknown definitive cause, but has been linked with some etiologies like autosomal recessive gene inheritance, maternal diabetes, conjoined twinning and cocaine use^{1,4-6}. The index case has no known etiology of dextrocardia, no family history of dextrocardia among siblings, and no family history of diabetes mellitus, conjoined twinning and substance abuse, thereby conforming to these literatures.

Dextrocardia with situs inversus totalis may be associated with some congenital anomalies, some of which are atrial situs solitus, discordant ventrculo-Atrial connection, polysplenia and Kartegener's syndrome (primary ciliary dyskinesia). Katergener's syndrome which is seen in about 25% of individuals with situs inversus comprises of situs inversus, chronic sinusitis and bronchiectasis^{1,8,9}. This index case had no feature to suggest any associated congenital anomalies following clinical evaluation and available imaging currently, thereby not conforming to these literatures.

Dextrocardia with situs inversus can be diagnosed by imaging, some of the imaging methods are plain radiography, ultrasonography, electrocardiography, echocardiography, barium studies, computed tomography (CT) and magnetic resonance imaging (MRI), with CT and MRI being particularly important in the diagnosis and assessment and demonstrating the mirror anatomy of the viscera in dextrocardia and situs inversus⁷⁻¹⁰. The case under review was confirmed following chest radiography, ultrasonography and echocardiography, conforming to these literatures. The patient had no CT or MRI during this review partly because of non-availability of these imaging modalities in our environment.

Most individuals with dextrocardia do not display symptoms and subsequently require no treatment, except where anomalies like heart defects occur, treatment is required and directed at the cause following appreciation of associated pathogenesis and appropriate diagnosis¹². The case under review has no associated congenital anomalies and also

asymptomatic, and no treatment was administered, thereby conforming to this literature.

Conclusion:

Dextrocardia with situs inversus is relatively uncommon and most often asymptomatic, but when suspected, basic plain chest radiography with ultrasonography and additional echocardiography can confirm the diagnosis and rule-out presence of associated anomalies.

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