

Misdiagnosis of Dextrocardia and Situs Inversus in a two Year Old Female

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Abstract

Dextrocardia with situs inversus is a congenital anomaly involving transposition of the heart to the right side with its apex pointing towards the right side associated with left handed mal rotation of visceral organs. We report a case of 2 year old female, weighing 11 kgs, who presented herself at the Out-Patient Department of Pediatrics Unit II, Abbasi Shaheed Hospital. She complained of flu since 2 days, along with cough and difficulty in breathing. The importance of detailed physical and systematic examination in asymptomatic newborn children is highlighted to assure early diagnosis. (ASH & KMDC 20(1):73;2015).

Introduction

Recognized first by Marco Severino in 1643, Dextrocardia is the abnormal hereditary form of the positioning of the heart on the right side whereas Situs inversus was first described by Aristotle in animals and Fabricius in humans. It has a variable occurrence rate of 4000 to 20000 live births¹. Dextrocardia with Situs inversus is the mirror-image reflection of the organs and cavities in the thorax. It most commonly occurs with a visually normal heart structure, where many people are oblivious of their condition² and are usually diagnosed via chest x-ray or regular physical examination performed due to various reasons. The occurrence of coronary heart disease in such individuals is independent of the

described condition³ though individuals may be susceptible to inborn heart malformation⁴, primary ciliary dyskinesia or splenic malformation. The exact physiological cause of the disease is undiscovered but an autosomal recessive inheritance is proposed⁵. We hereby discuss and describe the diagnosis of Dextrocardia with situs inversus in a 2 year old female.

Case report

A 2 year old female, weighing 11 kg, presented herself at the Out-Patient Department of Paediatrics Unit II, Abbasi Shaheed Hospital. She complained of flu for 2 days, along with cough and difficulty in breathing. The child was malnourished and her dietary intake was poor. According to the patient's attendant, patient was in usual state of health 2 days ago, when she developed mild cough, which was non-productive in nature. It increased gradually, while occurring in bouts, accompanied with flu and difficulty in breathing due to which her oral intake was disturbed.

The patient also exhibited a history of chest infections, inconsistent for 1½ years and was admit-

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ted to Abbasi Shaheed Hospital due to similar illness 1½ year back. The patient had no history of blood transfusion. Spontaneous Vaginal Delivery was held at a private hospital. No history of delayed cry, cyanosis or Neonatal Intensive Care unit (NICU) admission was reported. Child was vaccinated and BCG scar was positive. The patient was breastfed up to 1½ year of age, weaned at 6 months and administered cow milk with a bottle after 1½ years. She is reported to have successfully achieved her milestones eventhough the mother could not exactly recall the age of achievement. The patient's father and mother are both alive. Her father was diagnosed with Tuberculosis two years ago. The patient has two siblings, both are alive and healthy. The family confers no history of asthma and their socio-economic status is satisfactory.

The patient's general physical examination manifested the following facts. She turned up as an ill looking child, afebrile, well oriented to time, person and place. She lay in bed displaying signs of respiratory distress, mild tachypnea with positive subcostal and supraclavicular recession, and was severely anaemic. The anthropometric parameters (according to CDC charts) included the patient's length as 79cms (<5th percentile) and weight as 11 kg (<25th percentile). Her heart rate was 140 beats/min, respiratory rate was 50 breaths/min, and temperature was 101°F. The peripheral capillary oxygen saturation was 97% without O. The differential diagnosis conferred was Pneumonia with Dextrocardia and Tuberculosis. Further Investigation tests performed confirmed haemoglobin level of 9.5 g/dl, total leukocyte count of 14.6, and platelets 256,000. Tests performed were Chest X-Ray (CXR), Erythrocyte Sedimentation Rate (ESR) and abdominal Ultrasound.

On chest examination, heart sounds were heard on the right side, resulting in an initial impression of Dextrocardia. Chest X-ray and echocardiography demonstrated dextro position of the heart, showing agenesis of right lung. The left lung has an area of consolidation and a large pleural effusion.

The considered provisional diagnosis was Pneumonia with Dextrocardia with Situs inversus. The plan of treatment prescribed was Injection ceftriaxone, ventolin, nebulization with atrovent and syrup calpol.

Discussion

The location of the heart governs the long axis of the apex (base-to-apex axis). The normal position of the heart is levocardia and situs solitus with its apex pointing towards the left side. There are rare congenital cardiac anomalies reported in which the location of the heart is oriented exactly to the opposite side, leading to mirror image orientation. This condition is called Dextrocardia. When the organs along with Dextrocardia are oriented on the opposite side, this situation is called Situs inversus⁶.

The ratio of congenital cardiac anomalies of Dextrocardia with Situs inversus is 1:10,000, which is low as compared to isolated congenital dextrocardia⁷. The usual abnormality of Situs inversus with Dextrocardia is atrial situs solitus (93%), discordant AV connection (44%) and disconcordant ventriculo-atrial (VA) connection (30%)⁸.

The orientation of organs, due to Situs inversus is mirror image of the normal position, the liver and inferior vena cava are on the left, whereas the stomach and the descending aorta are on the right. Due to Dextrocardia the right atrium and right ventricle are on the left anteriorly and the left atrium and left ventricle are on the right posteriorly and the heart axis points to the right anterior thorax.

Patients suffering from Dextrocardia and Situs inversus usually remain asymptomatic and may be diagnosed in later life when they undergo examination and test for some unrelated disease⁹, as in our patient. Dextrocardia may be associated with following signs and symptoms, bluish skin, difficulty in breathing, fatigue, jaundice, repeated sinus or lung infections.

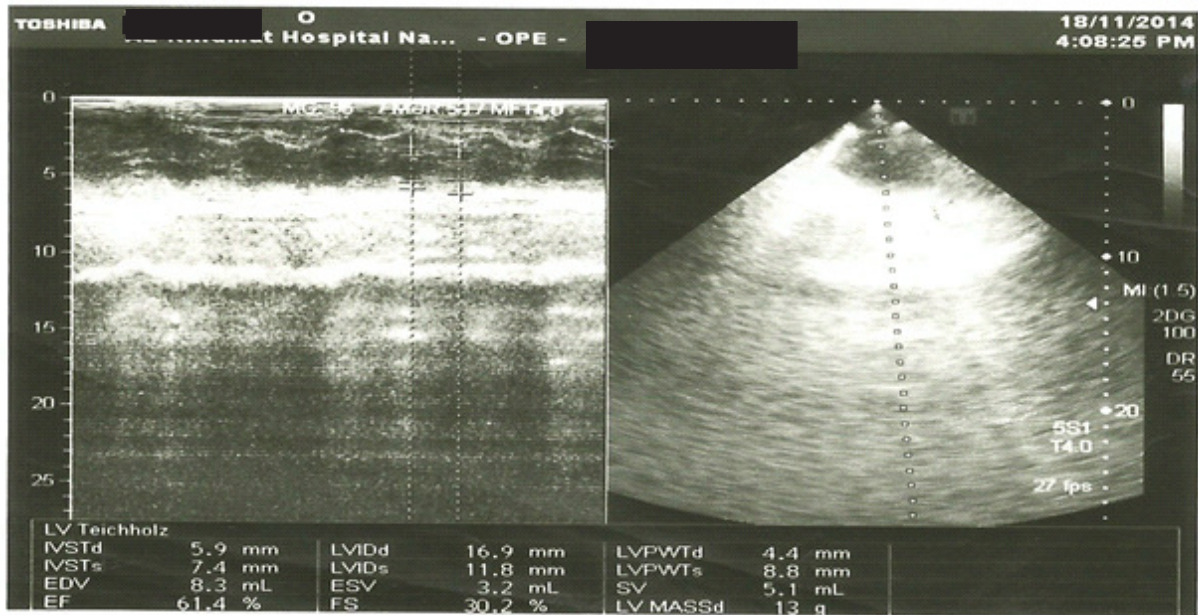
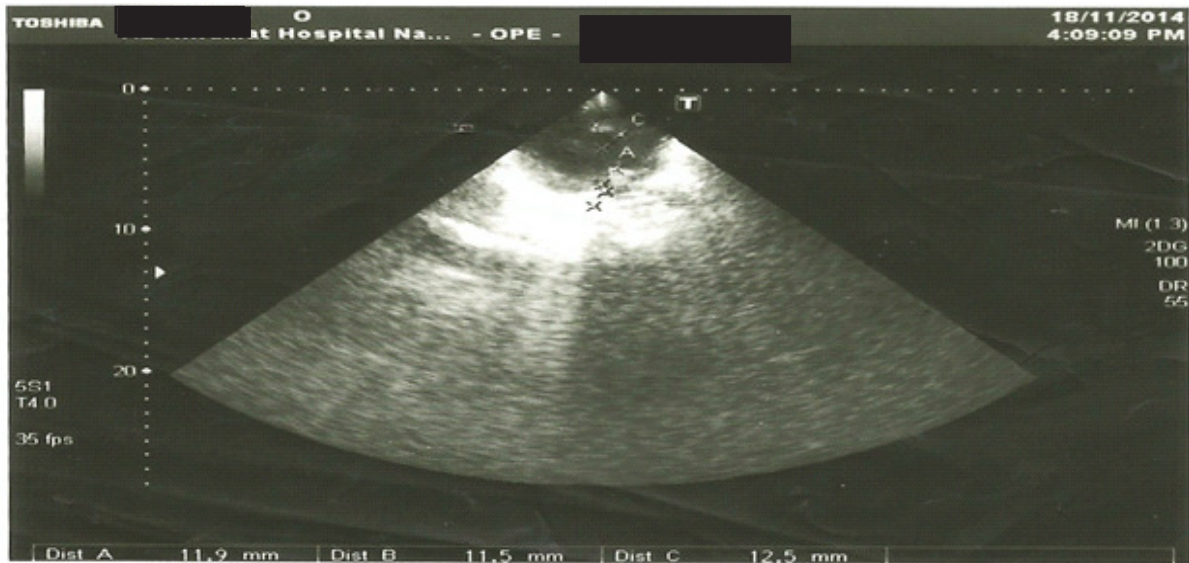


FIG. Showing Echocardiography with Dextro position of heart.

Transposition of atrial chambers determines the type of cardiac Situs inversus, which are of 3 types: 1) Situs solitus: it is the normal position of the atria that is right atrium on the right side and left atrium on the left side. Dextrocardia associated with situs solitus is called dextroversion, which is the second most common type of dextrocardia characterized by D-loop ventricles, normal great arteries and the apex of heart pointing towards the right side whereas the organs remain in their normal position. 2) Situs inversus: which is the mirror image of situs solitus with right atrium on the left side and left atrium on the right side. This is the most common form of Dextrocardia associated with situs inversus, also called situs inversus totalis characterized by L-loop ventricle with inverted great vessels. The apex points towards the right side with the organs being the mirror image of the normal position¹⁰. 3) Situs ambiguous or heterotaxy which is characterized by difficulty to determine the situs of atria¹¹. In this condition many of the organs are not in their morphologic places. Heterotaxy may be associated with midline liver, asplenia or multiple splenia, abnormal gall bladder system and malrotated intestine. Situs ambiguous is divided into 2 subtypes: Right isomerism or asplenia syndrome, and left isomerism or polysplenia syndrome.

In right isomerism or asplenia syndrome, there is bilateral right sidedness. Patients have bilateral right atria, asplenia, midline liver and 3 lobes in each lung with inferior vena cava and descending aorta on the same side. In contrast to right isomerism, left isomerism is characterized by bilateral left sidedness, polysplenia, bilateral left atria with 2 lobes in each lung and interruption commonly present due to azygous or hemiazygous vein in inferior vena cava. In situs ambiguous most of the organs are not in their usual place, it therefore requires thorough examination of the body⁶.

Situs abnormalities along with Dextrocardia can be diagnosed by using radiograph or ultrasonography.

The location of visceral organs, cardiac position along with position of great vessels can be confirmed by Computed Tomography (CT) scan which is also a conclusive diagnosis of Situs-inversus with Dextrocardia. Magnetic Resonance Imaging (MRI) is preferred for complicated cardiac anomalies.

Conclusion

A case on Dextrocardia with Situs inversus was reported and the importance of detailed physical and systematic examination in asymptomatic newborn children was highlighted to assure early diagnosis of this asymptomatic anomaly.

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