CASE REPORT

Dextrocardia with Situs Solitus in a Neonate – an Overview

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ABSTRACT

Dextrocardia, a rare congenital heart condition, can occur in about 1 out of every 12,000 pregnancies. Dextrocardia with situs solitus refers to when the heart is on the right side of the thorax while other viscera are found in their normal positions. The condition can go unnoticed in cases of limited prenatal care and newborn evaluation, leading to patients never receiving pertinent cardiac evaluations and condition progression monitoring throughout their lives. This is the first case reported of isolated dextrocardia with situs solitus in a neonate without any additional cardiovascular abnormalities. This case report highlights the importance of prenatal and postnatal evaluation to ensure the identification of neonates with dextrocardia and improve their quality of life and outcomes.

KEYWORDS: dextrocardia; situs solitus; prenatal diagnosis; pediatric echocardiography; isolated dextrocardia

INTRODUCTION

The normal anatomic position of the heart is on the thorax's left side, with the cardiac apex typically pointing to the left. Variations on the normal position of the heart and other viscera are fairly uncommon. The term ''dextrocardia'' specifically describes the rare congenital condition whereby the heart and apex are instead positioned on the thorax's right side.

Dextrocardia has an estimated incidence of around 1 in 12,000 pregnancies [1] and may be associated with other cardiac anomalies. No predilection for race, ethnicity, or gender have been described for dextrocardia.

The term ''situs'' is used to describe the positioning of thoracoabdominal organs in the body, including the heart itself. Dextrocardia can occur with a normal position of abdominal visceral organs (situs solitus [2]), with reversal in the position of abdominal visceral organs (situs inversus), or with abnormal distribution of major abdominal visceral organs (situs ambiguous), with varying reported incidence rates, ranging from around 22-33%, 37-39%, and 30-39% of cases in past studies, respectively [1,3].

Dextrocardia should be distinguished from dextroposition, where the heart is positioned in the right hemithorax but the cardiac apex points towards the left [2]. Cardiac dextroposition can result from extracardiac malformations such as a left-diaphragmatic hernia or right lung hypoplasia [2].

We present a case of a neonate with dextrocardia and situs solitus, as well as an overview of the clinical presentation and diagnosis for this condition.

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CASE PRESENTATION

We present a case of a baby girl born at 39 weeks of gestational age by spontaneous vaginal delivery, first child of a non-consanguineous Hispanic couple, weighing 3020 g. Apgar scores were 8 and 9 at 1 and 5 min, respectively. The pregnancy was uneventful and there was no relevant family history, specifically no history of neonatal cyanosis or congenital heart disease. On clinical examination, vital signs were stable, and no respiratory distress or cyanosis was discovered. However, mild swelling in the supero-occipital area on the head was identified, most likely caused by trauma during the delivery. Other physical findings were normal, with a regular rate and rhythm, without murmurs, but heart tones were louder and clearer in the right chest instead of the left. Prenatal records indicated a highly suspicious finding for dextrocardia in a fetal maternal ultrasound and follow up was recommended after delivery. For this reason, a chest X-Ray, electrocardiogram, 2D echocardiogram, and doppler echocardiogram were ordered by recommendations of the pediatric cardiology division as soon as the patient was delivered. Also, the patient was screened for situs inversus with an abdominal ultrasound and for primary ciliary dyskinesia with genetic testing by recommendation of the pediatric pulmonary division three weeks later.

At birth, the patient passed neonatal pulse oximetry screening for congenital heart disease, with saturations detected between 96–98%. A chest X-ray confirmed the heart was normal in size, clear bronchovascular markings and costophrenic angles and absence of consolidations or effusions. The chest X-ray confirmed the presence of a left-sided stomach gas bubble, with a right-sided heart (Figure 1). Upon evaluation with the echocardiogram, the patient was

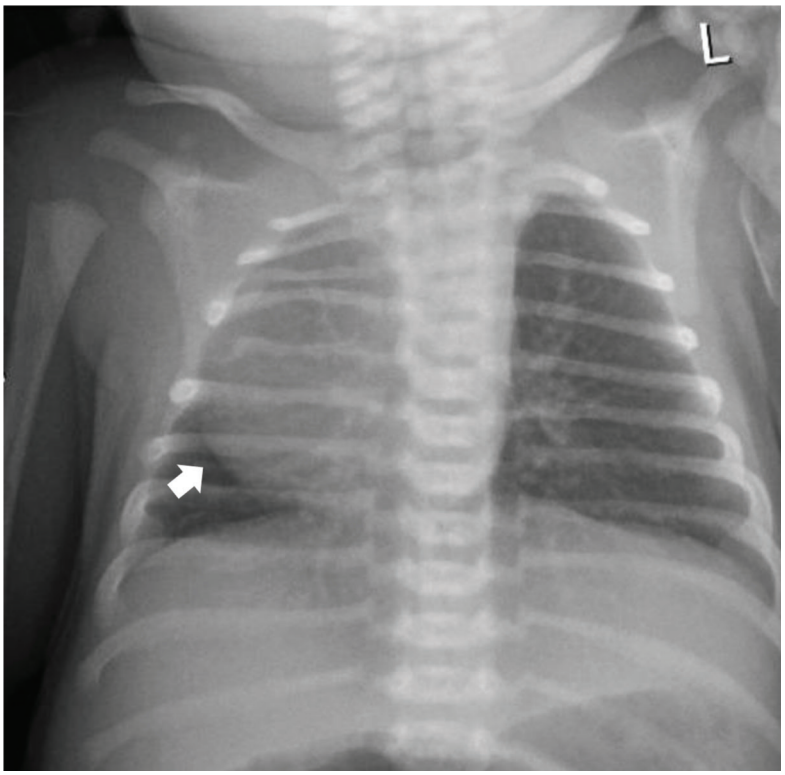

Fig. 1. Chest X-ray film which confirms the presence of a left-sided stomach gas bubble and a right-sided heart.

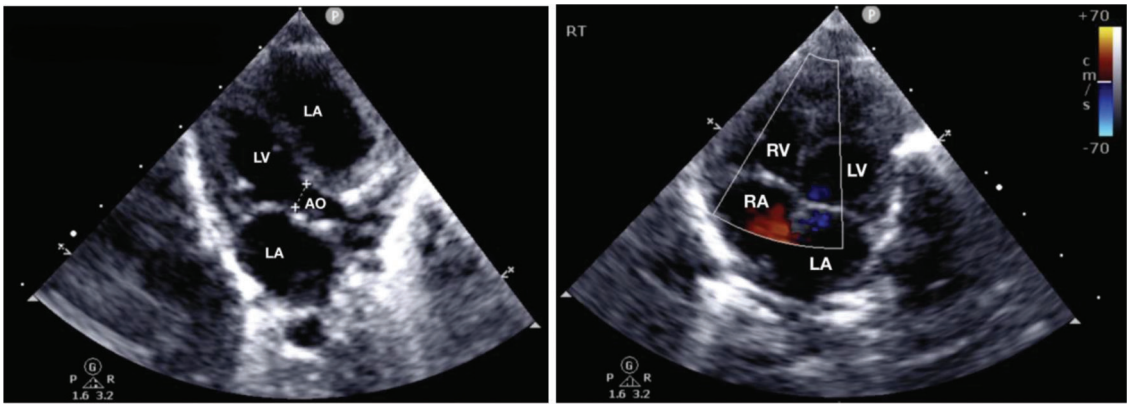

Fig. 2. Transthoracic echocardiogram in dextrocardia with situs solitus without other cardiovascular abnormalities.

found to have a dextroversion of the heart with concordance of atrioventricular connections, and normal size of the heart and its chambers (Figure 2). This means that the morphological right atrium was to the right and posteriorly positioned, while the morphological left atrium was to the left and anteriorly positioned. The aortic orifice was situated posteriorly but lied somewhat to the left of the pulmonary trunk. The pulmonary trunk was positioned towards the left, almost horizontally, and then shifted to the left posteriorly adjacent to the ascending aorta to reach its bifurcation. Doppler echocardiogram revealed left and right ventricles with preserved function, a left ventricle ejection fraction greater than 55%, and a patent ductus arteriosus and foramen ovale. These last findings were physiologically expected as the ductus arteriosus and foramen ovale close after approximately 12-24 hours and 6-12 months, respectively. The electrocardiogram findings had characteristic changes previously described in the literature for dextrocardia with situs solitus, such as negative P waves and QRS complexes in lead 1 and reverse R wave progression across the precordium (Figure 3).

Transverse and longitudinal images of the abdomen by ultrasound confirmed the presence of a right-sided liver and a left-sided stomach, without any morphological abnormalities or location discrepancies of the other organs in the abdomen. Lastly, the genetic diagnostic test 'Invitae Respiratory Distress Panel' was insignificant for the presence of mutations related to primary ciliary dyskinesia and other genes associated with conditions that cause respiratory distress in newborns. However, a variant of uncertain

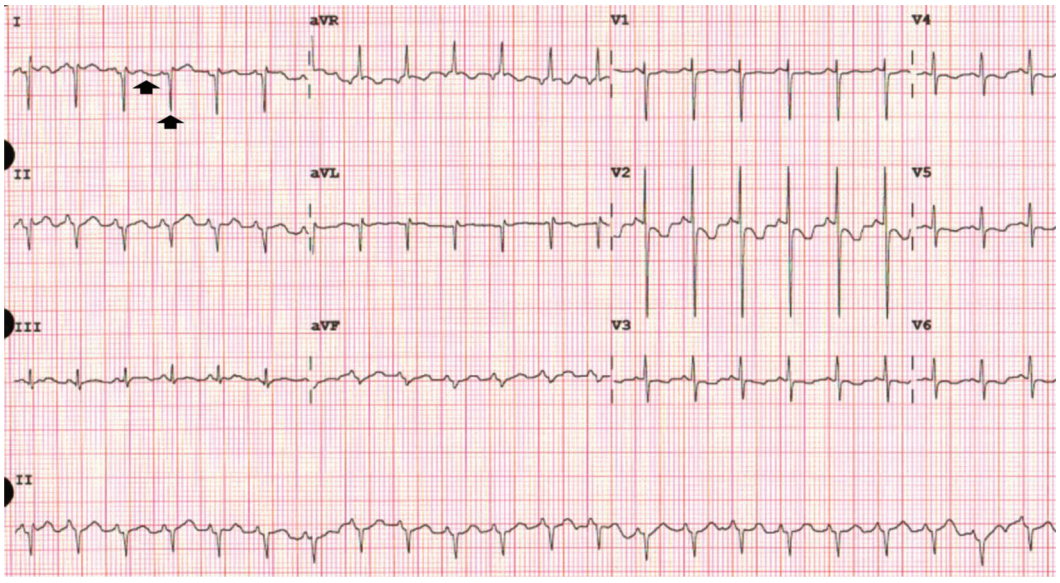

Fig. 3. Electrocardiogram in dextrocardia with situs solitus.

significance, c.352_360del (p.Ser118_Gly120del) was identified in the CHD7 gene, which is associated with autosomal dominant CHARGE syndrome and Kallman syndrome.

Three days after delivery, the patient was discharged home and recommended to continue follow up with the pediatric cardiologist of their preference. For the past three months, Pediatric Cardiology consults have found the patient stable with a normal development and function of the heart.

DISCUSSION

The exact cause of dextrocardia is unknown, but the condition occurs when there are abnormalities in the formation of the heart during embryonic development. The autosomal recessive genetic ciliary disorder Kartagener's syndrome has been associated with bronchiectasis, sinusitis, and dextrocardia with situs inversus [4].

The fetal heart develops from an embryonic cardiac tube formed from the endocardial tube fusion. The embryonic cardiac tube is initially straight but transforms into a helically wound loop in a process termed cardiac looping [5]. Cardiac looping determines the relative positions of the ventricles in relation to the atria.

The cardiac tube may loop to the right (forming a D-loop) or left (forming a L-loop), with the D-loop resulting in the morphologic right ventricle being on the right of the morphologic left ventricle and the L-loop resulting in the morphologic right ventricle being on the left of the left ventricle, respectively. The D-loop is the normal (solitus) cardiac loop while the L-loop is a mirror-image (inversus) loop [6].

Dextrocardia can occur when a D-bulboventricular loop fails to move into the left hemithorax or when a L-bulboventricular loop completes apical migration while localized in the right hemithorax. In the case of dextrocardia with situs solitus, also termed isolated dextrocardia, the D-loop ventricles and normally positioned great arteries result specifically from the D-loop failure of migration [6].

In around 90% of cases, dextrocardia with situs solitus presents with concomitant congenital cardiac malformations, including anomalous pulmonary venous return, tetralogy of Fallot, septal defects, pulmonic stenosis, coarctation of the aorta, and corrected transposition of the great arteries; other common abnormalities include atrioventricular discordance, single ventricle, and atrial or ventricular septal defect [2,7].

Cases of isolated dextrocardia without associated congenital cardiac malformations are rare; such patients are typically asymptomatic, and the condition can go without being discovered in these patients until when they present as adults with symptoms of acquired heart disease [8]. We provide the first case report on diagnosed dextrocardia with situs solitus without additional cardiovascular abnormalities in a neonate in at least the past 20 years. While there exist other case reports in this time frame on isolated dextrocardia, these patients have either been found to possess various other congenital cardiac anomalies or have been diagnosed much later in life [8,9].

The incidence of coronary heart disease and the life expectancy of patients with isolated dextrocardia and no other congenital cardiac abnormalities are the same as in the general population. Our report's findings serve to exemplify this fact, given that our patient passed the neonatal pulse oximetry screening for congenital heart disease, detecting saturations of 96%–98%, despite atypical physical exam findings and later diagnostic studies confirming the patient's condition. In this way, this case serves as a unique learning opportunity, providing insight on an infrequently observed phenomenon. Moreover, this case highlights the importance of providing thorough prenatal and postnatal care to prevent congenital anomalies from going unidentified and minimize possible future cardiac complications.

CONCLUSION

Isolated dextrocardia without associated congenital cardiovascular abnormalities is rare. Our experience in this case demonstrates that successful prenatal and postnatal evaluation optimizes the identification of neonates with dextrocardia and improves the delivery of better outcomes.

Conflict of interests

The authors declare that there is no conflict of interests.

Informed Consent

Written informed consent for the case to be published was obtained from the patient's legal guardian for publication of this case report, including the accompanying images.

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