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CASE REPORT

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Situs inversus totalis in a newborn: A case report

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Abstract: *Introduction:* Situs inversus with dextrocardia is the complete inversion of position of the thoracic and abdominal viscera. It may be isolated or associated with malformations, especially cardiac and/or alimentary. It may be discovered in infancy because of associated anomalies but often remains asymptomatic and discovered incidentally in adult life.
Case presentation: We present a 19-day-old female newborn found to have dextrocardia with situs inversus totalis. A chest X-ray showed her heart in the right hemithorax with the cardiac apex pointing towards the right. The findings at echocardiography confirmed the location of her heart in the right hemithorax with Transposition of great arteries (TGA) and ventricular septal defect (VSD). An abdominal sonogram showed her liver on her left side whereas her stomach and spleen were located on the right side.
Conclusion: There is need for newborn babies to have a thorough physical examination after delivery and before discharge to enable early diagnosis of congenital anomalies for appropriate referral.

Keywords: Situs inversus, Dextrocardia, Transposition of Great Arteries, Newborn

Résumé: *Introduction*: Le situs inversus avec dextrocardie est l'inversion complète de la position des viscères thoraciques et abdominaux. Il peut être isolé ou associé à des malformations, notamment cardiaques et/ou digestives. Il peut être découvert dans la petite enfance en raison d'anomalies associées, mais reste souvent asymptomatique et découvert fortuitement à l'âge adulte.
Observation: Nous présentons le cas d'un nouveau-né de sexe féminin, âgé de 19 jours, qui a présenté une dextrocardie avec un situsinversus total.Une radiographie du thorax a montré son cœur dans l'hémithorax droit, l'apex cardiaque étant orienté vers la droite. Les résultats de l'échocardiographie ont confirmé la localisation du cœur dans l'hémithorax droit, avec une transposition des gros vaisseaux (TGV) et une communication interventriculaire (CIV). L'échographie abdominale a montré que le foie se trouvait sur le côté gauche, tandis que l'estomac et la rate se trouvaient sur le côté droit.
Conclusion: Les nouveau-nés doivent subir un examen physique minutieux après l'accouchement et avant leur sortie de l'hôpital afin de permettre un diagnostic précoce des anomalies congénitales et une orientation appropriée.

Mots-clés: Situs inversus, dextrocardie, transposition des gros vaisseaux, nouveau-né.

Introduction

As a rare congenital disorder, situs inversus totalis, also known as dextrocardia with situs inversus, occurs when the heart's anatomical position is completely reversed to the right side, with all of the visceral organs also inversely rotated.[1,2]

The direct etiology of this condition is not known. However, this autosomal recessive condition has been linked to several other conditions such as conjoined twinning, cocaine usage, and maternal diabetes.[2,3] Situs inversus totalis is reported to occur in 1 in 8000 to 1 in 25,000 patients,[4] with no racial predilection. Individuals with this disorder may also present with primary ciliary

dyskinesia, congenital heart defects, and splenic abnormalities.[3] We describe a case of situs inversus totalis in a newborn, the first case to be reported in Freetown, Sierra Leone.

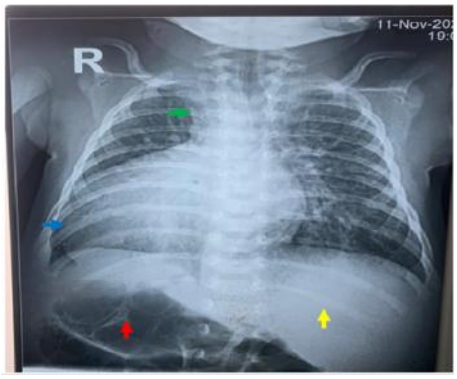
Case Presentation

A 19-day-old female newborn of non-consanguineous parents; who was delivered via spontaneous vaginal delivery at 38 weeks of gestation. She had a good extrauterine transition and did not require in-hospital care after birth. Her mother had an uneventful antenatal period, as she reported normal vital signs and investigation results, no history of febrile illness with rashes during early pregnancy, no exposure to ionizing radiation, no history suggestive of gestational diabetes, and she received only prescribed medications.
Baby was however admitted on the fifteenth day of life at a peripheral health centre, where she received care for community acquired pneumonia following acute onset of fever, cough with fast breathing. She was referred to our facility on account of subnormal oxygen saturation in spite of notable clinical improvement.
On examination, she was cyanosed, not febrile and not pale. Cardiovascular system examination revealed normal volume pulses, regular with a capillary refill time of <3seconds. Her oxygen saturation in both pre- and post-ductal limbs ranged between 79-84% on intra-nasal oxygen. Her apex beat was at the 4th right intercostal space mid clavicular line. There was cardiac dullness on the right side of her chest and her heart sounds were also louder on the right side with a grade 3 pansystolic murmur heard. She had tachycardia with a heart rate of 168/minute. Respiratory examination showed mild dyspnoea with normal respiratory rates and breath sounds. Her abdominal examination demonstrated a palpable liver 2cm below the left costal margin that was firm, non-tender with smooth surface, and tympanic note was present over the right hypochondrium.

A chest X-ray (Fig 1) showed her heart in the right hemithorax with the base to apex axis pointing towards the right. She had cardiomegaly with a cardiothoracic ratio of 0.66. Her lung fields were clear. Her thoracic cage was normal. Gastric air bubble was seen under the right hemidiaphragm. Echocardiography demonstrated dextrocardia with situs inversus. The interatrial septum was intact. The interventricular septum was defective (measuring 3.3mm) with her Aorta arising from the Right Ventricle, Pulmonary Artery from the Left Ventricle (ventriculo-arterial discordance), and both vessels were in parallel orientation. There was no ventricular hypertrophy. Retrograde blood flow from the descending aorta in to the main pulmonary artery was reported and her Aortic arch was right sided (Fig 1). Abdominal ultrasound visualized the liver in the left; spleen and stomach in the right.

A diagnosis of Situs inversus totalis with transposition of great arteries (TGA) and ventricular septal defect (VSD) was made. She received suspension spironolactone, furosemide and captopril, and progressively maintained normal heart rates and respiratory rates. Oxygen was weaned off with her oxygen saturation at 85% in room air. She was discharged on the 10thday of admission to the Cardiology clinic for outpatient follow-up. She remained stable on follow up visits, maintained on suspension spironolactone, furosemide and captopril as anti-failure medications.

Fig 1: Chest radiograph demonstrating the cardiac apex on the right side (blue arrow), left-sided ascending aorta and right-sided aortic knuckle (green arrow) with a gastric bubble on the right (red arrow) and left-sided hepatic shadow (yellow arrow).



Discussion

Dextrocardia with complete situs inversus is rare, usually discovered incidentally in otherwise normal subjects.[5] It may be discovered in infancy because of associated anomalies but often remains asymptomatic and discovered by chance in adult life.[6]Many people with this condition are unaware of their unusual anatomy until they seek medical attention for an unrelated condition.[7]

The arrangements of the position of the abdominal viscera in dextrocardia may be normal (situs solitus), reversed (situs inversus), and indeterminate (situs ambiguous or isomerism) in 32 to 35%, 35 to 39%and 26 to 28% of cases respectively.[8] Dextrocardia with a normal abdominal situs has a high incidence of associated congenital cardiac anomalies including among others, transposition of the great vessels and atrial septal defects (ASDs)[9] and ventricular septal defects (VSDs)[10] in 90 to 95% of cases. However, dextrocardia with situs inversus is associated with a lower incidence of congenital heart-disease (0 to 10%) as was the case in our patient. Presentation varies depending on associated malformation.[4,11,12] Situs inversus may be associated with other congenital anomalies such as duodenal atresia, asplenism, multiple spleens, ectopic kidney, horseshoe kidney and various pulmonary and vascular abnormalities.[13]

In this patient, situs inversus was associated with dextrocardia. Cardiac anomalies identified on echocardiography were VSD and transposition of the great arteries (TGA). This case is reported because of the situs inver

sus, dextrocardia with early symptomatic presentation due to complex pattern of cardiac malformation.

Conclusion

Dextrocardia with situs inversus is a rare congenital malformation that must be fully evaluated when noticed because in rare instances it may result in fatal outcome. There is need for newborn babies to have a thorough physical examination after delivery and before discharge to enable early diagnosis of congenital anomalies for appropriate referral.

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