

CASE REPORT

DEXTROCARDIA WITH SITUS INVERSUS TOTALIS IN NEONATE: CASE REPORT

Nagraj Jawali¹, Prakash H. K.², Naseema Banu³, Teeka Rao⁴

HOW TO CITE THIS ARTICLE:

Nagraj Jawali, Prakash H. K., Naseema Banu, Teeka Rao. "Dextrocardia with Situs Inversus Totalis in Neonate- Case Report". Journal of Evidence based Medicine and Healthcare; Volume 2, Issue 13, March 30, 2015; Page: 2114-2119.

ABSTRACT: Dextrocardia with situs inversus are rare congenital anomalies which can be asymptomatic and compatible with normal life. They are characterized by mirror images of all intra-thoracic and intra-abdominal viscera. Our aim is to report an incidental finding of dextrocardia with situs inversus in a neonate with sepsis and also to report other associated anomalies with it. Most cases of situs inversus have been reported either in adults or in cadavers during dissection. Very few in number have been detected in neonates. Here is a one such case report. A day -old term male neonate presented with features of septic shock. Physical examination revealed cardiac apex on the 4th right intercostal space, along the mid-clavicular line. Chest radiograph and abdominal ultrasound confirmed the diagnosis of dextrocardia with situs inversus along with renal agenesis and ectopia.

KEYWORDS: Dextrocardia, Situs inversus, Neonate.

INTRODUCTION: Dextrocardia (Also called looping defect) is an abnormal congenital positioning of heart on the right side. Situs inversus totalis (Also called situs transversus), is a congenital condition in which major visceral organs are reversed or mirrored from normal positions. Many people with situs inversus are unaware of their unusual congenital anomaly until they seek medical attention for unrelated conditions. Individuals with dextrocardia and situs inversus totalis may have associated congenital heart malformations, primary ciliary dyskinesia or other malformations. Very few in number have been detected in neonates. Here is a one such case report.

CASE SUMMARY: A day old term male neonate weighing 2350gms, was brought to our hospital with complaints of respiratory distress and cyanosis. On examination, baby was hypothermic, tachypneic, with signs of shock (Prolonged capillary refilling time, feeble peripheral pulses, and tachycardia and oxygen saturation 65 -68% in room air). However, apical impulse was at 4th right intercoastal space, with heart sounds better heard on right side, liver being palpable on left side. He was a 3rd child born to 2nd degree consanguineous married couple with other two siblings being normal. Present Pregnancy was UN booked case and not medically supervised. Baby was delivered at home with no relevant birth details. On Evaluation, Sepsis work up showed leucocytosis, positive CRP and no growth on blood culture. Chest x-ray showed normal heart size with apex located to the right in keeping with dextrocardia. Hepatic shadow was noted on the left hypochondrium and possible fundus shadow on the right hypochondrium [Fig. 1]. Abdominal scan demonstrated liver in left hypochondriac region with normal in size and echo pattern. Head of pancreas situated in left paravertebral region with altered relationship of superior mesenteric

CASE REPORT

artery and vein. Both kidneys could not be noted in their normal situ and only one reniform structure was noted in left lower lumbar region suggesting left renal ectopia and right renal agenesis. 2D echo revealed dextrocardia, with VSD, pulmonic stenosis and aortic overriding. Diagnosis of Early onset neonatal sepsis in shock was made with background dextrocardia and situs inversus. Supportive treatment was initiated with IV fluids, Antibiotics as per our Nicu protocol. Shock resolved with IV fluids and inotropes. However oxygen saturation and cyanosis did not improve owing to cyanotic congenital heart disease. Baby was referred to cardiac centre for further evaluation and definitive treatment.

DISCUSSION: The term "situs" means position, site, or location.^[1, 2, 3,] "Situs solitus" - This brief word summarizes lot of expectations, namely that the stomach and spleen lie to the left, the right lobe of the liver is larger than the left, and the appendix is right-sided. This normal asymmetry of the abdominal organs is accompanied by differences in lobulation between the right and left lung, with a trilobed right and a bilobed left lung. This arrangement is termed by different authors' normal situs or situs solitus. ^[1, 2, 3,]

Situs inversus totalis is a condition in which the organs of the chest and abdomen are arranged in perfect mirror image of normal. In situs inversus, the morphologic right atrium is on the left, and the morphologic left atrium is on the right. The normal pulmonary anatomy is also reversed so that the left lung has 3 lobes and the right lung has 2 lobes. In addition, the liver and gallbladder are located on the left, whereas the spleen and stomach are located on the right. The remaining internal structures are also a mirror image of the normal. [Fig. 2] This anomaly has autosomal recessive mode of inheritance. The incidence of situs inversus for the neonates with hereditary tendency is 1/5000-1/10000 with distribution being same for both genders (1:1). There is no difference between races.^[4] Clinically, the condition can be divided into complete or partial types. In complete visceral transposition, organs in chest and abdomen are all mirror imaged but such case is pretty rare. In partial visceral transposition, only certain viscera are reversed and are usually associated with other complex abnormalities. The complete visceral transposition accounts for 86% and the partial accounts for 14%.^[5]

In human and in vertebrates anatomic asymmetry is established during embryogenesis. The left-right axis is determined at the beginning of the embryonic development with the formation of the dorso-ventral and cephalo-caudal axes. The cardiac tube curve to the right is the first sign of asymmetry. The left-right gradient has been established at cellular level. The left-right relation of the asymmetric viscera is conserved; and when there is complete inversion of the lateralization of the organs (mirror image), it is known as Situs inversus totalis.^[4,5] Recent studies suggest that left-right asymmetry defects to be due to genetic abnormalities in lefty genes, nodal genes, and ZIC 3, ACVR2B and Pitx genes and mutation of these genes are present on chromosome 12.^[5,6] So far, no definite theories can explain the cause of this phenomenon. It is generally believed that visceral transposition occurs in the third to eighth week during embryonic development. In the initial or early stage of embryo, the abnormal rotation of cilia or parental chromosome aberration or gene mutation can lead to the happening of this disease. There are mainly four theories to explain the formation of visceral transposition. 1) All organs transposition theory believes that the position disordering of progenitors of viscera during embryo period causes visceral transposition.

CASE REPORT

2) Uneven heat theory believes local high temperature of embryo leads to the inversion of organs. 3) Twins theory is based on observing the frequency of visceral transposition among twins. 4) Embryo rotation highlights the rotation of embryo during development leads to the anomaly. [5, 6]

The individuals with situs inversus are phenotypically unimpaired and can lead normal healthy life, without any complications related to their medical condition. Many people with situs inversus are unaware of their unusual anatomy until they seek medical attention for an unrelated condition.^[7] Situs inversus may be associated with other anomalies or malformations. A 3-5% incidence of congenital heart disease is observed in situs inversus with dextrocardia. Of these patients, 80% have a right-sided aortic arch. Situs inversus with levocardia is rare, and it is almost always associated with congenital heart disease.^[8] Kartagener syndrome is typified by bronchiectasis, sinusitis, and situs inversus and this syndrome affects 20% of patients with situs inversus; however, only 50% of patients with Kartagener syndrome have situs inversus. [9]

Situs abnormalities may be recognized first by using radiography or ultrasonography. [10, 11] However, computed tomography (CT) scanning is the preferred examination for the definitive diagnosis of situs inversus with dextrocardia. CT scanning provides good anatomic detail for confirming visceral organ position, cardiac apical position, and great vessel branching. Magnetic resonance imaging (MRI) is usually reserved for difficult cases or for patients with associated cardiac anomalies. Attention to the left and right sides of the neonate as well as left and right labeling of images need to be done by the radiologists to prevent technical errors that occur during diagnosis of this anomaly. These Technical errors are the most common cause for both false positive and false negative diagnosis of situs inversus. [11, 12]

Long-term prognosis depends on associated congenital defects and the presence of cardiovascular compromise. Most patients with dextrocardia with situs inversus do not have any significant cardiac defects, and their life expectancy is normal. Those who have ciliary defects require frequent treatment for chronic pulmonary infections and complications. Since prognosis is variable depending on associated malformations, knowledge of the entire spectrum is important for prenatal and postnatal management and counseling. Fetal MRI, due to high soft tissue contrast, provides excellent organ anatomy and therefore improves the classification of the heterotaxy. [14, 15]

Special Concerns of Situs Inversus:

1. Clinicians should have high index of suspicion since this anomaly is asymptomatic
2. All newborn babies should receive thorough physical examination by paediatricians immediately after delivery and any abnormality need to be evaluated.
3. The recognition of situs inversus is important for preventing surgical mishaps that result from the failure to recognize reversed anatomy or an atypical history. For example, in a patient with situs inversus, appendicitis causes left lower quadrant pain.
4. Situs inversus also complicates organ transplantation operation as donor organs will almost certainly come from situs solitus (Normal), as heart and liver have geometric problems while placing the organs into cavity, shaped in mirror image.

CASE REPORT

Figure 1: [X-ray of neonate, AP view showing cardiac apex pointing towards right, liver on left side and stomach shadow on right]



Fig. 1

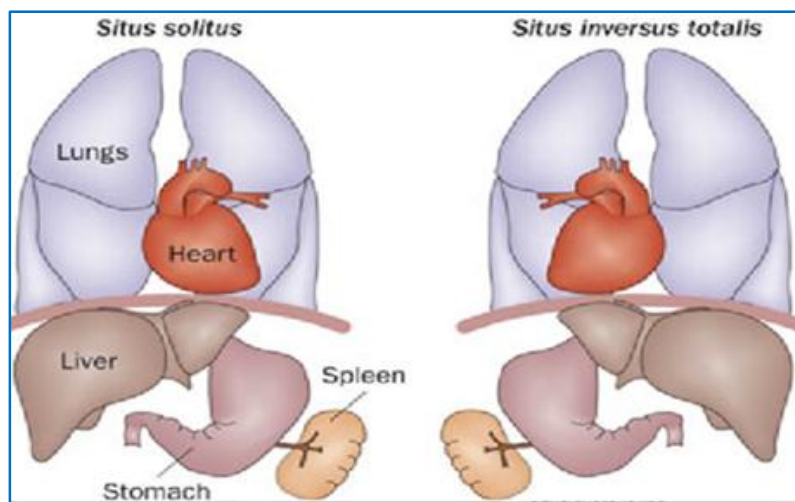


Fig. 2

REFERENCES:

1. Fraser RS, Muller NL, Colman NC, Pare PD. Fraser and Pare's Diagnosis of Diseases of the Chest. Vol 3. 4th ed. Philadelphia, Pa: WB Saunders Co; 1999:2281-3.
2. Gutgesell HP. Cardiac malposition and heterotaxy. In: Garson AG Jr, Fisher DJ, Neish SR, eds. Science and Practice of Pediatric Cardiology. Vol 2. 2nd ed. Baltimore, Md: Williams & Wilkins; 1998:1539-61.
3. Hagler DJ, O'Leary PW. Cardiac malpositions and abnormalities of atrial and visceral situs. In: Emmanouilides GC, Riemenschneider TA, Allen HD, Gutgesell HP, eds. Moss and Adams' Heart Disease in Infants, Children, and Adolescents: Including the Fetus and Young Adult. Vol 2. 5th ed. Baltimore, Md: Williams & Wilkins; 1995:1307-3.

CASE REPORT

4. M. João marta, I. Menezes falcão, j. A. Saavedra, Luciano ravara Serviço de Medicina I, Hospital de Santa Maria, Lisboa Rev Port Cardiol 2003; 22 (1):91-104.
5. Brett Casey Two rights make a wrong: human left–right Malformations. Human Molecular Genetics, 1998, Vol. 7, No. 10 Review 1565–1571.
6. Jiang-Kun Jia, Huan-Zhou Xue, Quan Shen. One case of transposition of viscera and hepatic and extrahepatic bile duct stones. Life Sci J 2013; 10(3):1522-1523] (ISSN: 1097-8135). <http://www.lifesciencesite.com>. 228.
7. Tabry IF, Calabrese J, Zammar Het al. Case report: off-pump total myocardial revascularization for dextrocardia and situs inversus. Heart Surg Forum 2001; 4: 2513.
8. Gindes L, Hegesh J, Barkai G, and Jacobson JM, Achiron R. Isolated levocardia: prenatal diagnosis, clinical importance, and literature review. J Ultrasound Med. Mar 2007; 26(3):361-5. [Medline].
9. Ortega HA, Vega Nde A, Santos BQ, Maia GT. [Primary ciliary dyskinesia: considerations regarding six cases of Kartagener syndrome.] [Portuguese, English]. J Bras Pneumol. Oct 2007; 33(5):602-8. [Medline].[Full Text]
10. Tonkin IL, Tonkin AK. Visceroatrial situs abnormalities: sonographic and computed tomographic appearance. AJR Am J Roentgenol. Mar 1982; 138(3):509-15. [Medline].
11. Partridge J. The radiological evaluation of atrial situs. Clin Radiol. Jan 1979; 30(1):95-103. [Medline].
12. Situs Inversus Totalis CPT Paul J. Shogan, MC USA * †; Col Les Folio, USAF MC SFS (Ret.) ‡ MILITARY MEDICINE, 176, 7:840, 2011.
13. Prenatal Imaging of Situs Anomalies: Utility of Fetal MRI Preeyacha Pacharn, MD; Leann Linam, MD; Maria Calvo Garcia, MD; Eva Rubio, MD; Beth Kline-Fath, MD.
14. Fung TY, Chan DL, Leung TN, Leung TY, Lau TK. Dextrocardia in pregnancy: 20 years' experience. J Reprod Med. Jul 2006; 51(7):573-7. [Medline].
15. Lee SE, Kim HY, Jung SE, et al. Situs anomalies and gastrointestinal abnormalities. J Pediatr Surg. Jul 2006; 41(7):1237-42. [Medline].
16. Douglas YL, Jongbloed MR, den Hartog WC, Bartelings MM, Bogers AJ, Ebels T, et al. Pulmonary vein and atrial wall pathology in human total anomalous pulmonary venous connection. Int J Cardiol. Dec 29 2008; [Medline].
17. Van Mierop LH, Eisen S, Schiebler GL. The radiographic appearance of the tracheobronchial tree as an indicator of visceral situs. Am J Cardiol. Oct 1970; 26(4):432-5. [Medline].

CASE REPORT

AUTHORS:

1. Nagraj Jawali
2. Prakash H. K.
3. Naseema Banu
4. Teeka Rao

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor, Department of Paediatrics, RIMS.
2. Senior Resident, Department of Paediatrics, RIMS.
3. Assistant Professor, Department of Paediatrics, RIMS.

4. Junior Resident, Department of Paediatrics, RIMS.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. Prakash H. K.
S/o. H. K. Chandra Shekar,
Commission Agent, Plot No.7, Shop No.53,
Rajendra Gunj, Raichur-58410
E-mail: pakkudoc@gmail.com

Date of Submission: 17/03/2015.
Date of Peer Review: 18/03/2015.
Date of Acceptance: 26/03/2015.
Date of Publishing: 30/03/2015.