ISSN: 0975-833X

Available online at http://www.journalcra.com

International Journal of Current Research
Vol. 8, Issue, 02, pp.26934-26937, February, 2016

INTERNATIONAL JOURNAL OF CURRENT RESEARCH

CASE STUDY

DEXTROCARDIA AND SITUS INVERSUS TOTALIS IN A NEWBORN- A RARE CASE REPORT

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ARTICLE INFO

Article History:
Received 14th November, 2015
Received in revised form
25th December, 2015
Accepted 09th January, 2016
Published online 27th February, 2016

Key words:

SitusInversus Totalis, Dextrocardia, Congenital.

ABSTRACT

Situs Inversus Totalis is a rare condition with not much known sex discrimination. The cause for the same is still unknown although some studies describes certain maternal medical conditions like diabetes mellitus as a reason. The term Situs Inversus is a short form of the later meaning inverted position of internal organs. Such individuals are generally asymptomatic and have a normal life expectancy and it may go unnoticed for a long period of lifetime. Other congenital abnormalities can also be seen with this condition like cardiac defects, pulmonary infections. Being a rarecondition, the literature for it is not that extensive. In our report we describe a case of Dextrocardia with SitusInversus Totalis in a newborn with respiratory distress.

Citation: **Rajiv Jasrotia, Gauri Chauhan, Gurdeep S. Dhanjal and Ritambhara Lohan, 2016.** "Dextrocardia and situs inversus totalis in a newborn- A rare case report", *International Journal of Current Research,* 8, (02), 26934-26937.

INTRODUCTION

'Situs' means the position of heart i.e. the cardiac atria and viscera . When there is occurrence of mirror image it is termed as 'Situs Inversus' i.e. mirror image of as that supposed to be on the normal position. Dextrocardia is a term used exclusively for defining the positioning of heart i.e. the tip of heart points to the right side. A very few cases of situsinversustotalis has been described in literature i.e. Dextrocardia with Situs Inversus. We report such a case further in a newborn.

Case presentation

An out born 3.012 kg term male infant born by normal vaginal delivery to primigravidamother, was admitted to our N.I.C.U. with the complaints of fast breathing and refusal of feeds for 2 days. According to parents, the child was born by Normal Vaginal Delivery in a civil hospital and after sometime noted to have fast breathing and was taken to some local practitioner from where he was referred to a higher center. On presentation his vitals were temperature of 36.8 C, RR of 84/min, HR of 136/min, SpO2- 88% and B.P. of 72/46 (56) mmHg in his right upper arm in supine position. Cardiovascular system examination showed visible apex beat in right fifth intercostal space in mid-clavicular line. There was cardiac dullness on his right side. Heart sounds were louder on the right side of the chest. Abdominal examination showed no palpable organomegaly but on percussion liver dullness was on left side. After initial stabilization with oxygen, Chest X-ray was advised which showed his heart in the right hemithorax with base to apex pointing towards the right; the trachea and lung fields were normal. For evaluation of heart echocardiography was planned which demonstrated dextrocardia. IVC and aorta on right side with situsinversus with patent foramen ovale with no significant shunt. Further, abdominal ultrasound revealed liver and gallbladder located in left hypochondrium, spleen is located in the right hypochondrium and apex of heart is directed on the right side and normal kidneys. Other investigations: CBC suggestive of normal values, CRP was positive and blood culture suggestive of actinobacter and candida growth after incubation. Antibiotic coverage was broadened and extended to 14 days. Symptomatically baby improved by day 12th but his unexplained oxygen dependence continued. Chest physiotherapy was done and multidisciplinary follow up arranged.

DISCUSSION

Dextrocardia was first seen by Leonardo da Vinci in 1452-1519 and then recognized by Marco Aurelio Severine in 1643 and described more than a century later by Matthew Ballie.

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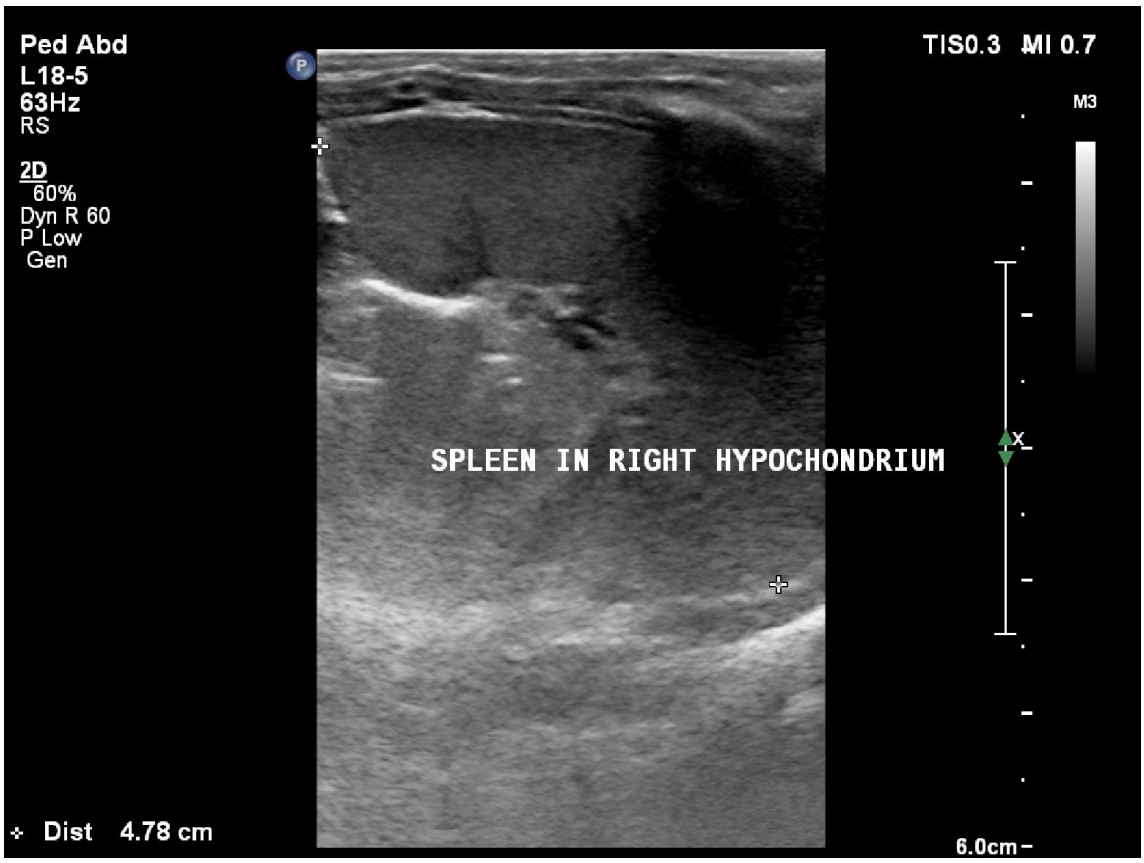

Fig. 1. USG finding showing spleen in the right hypochondrium

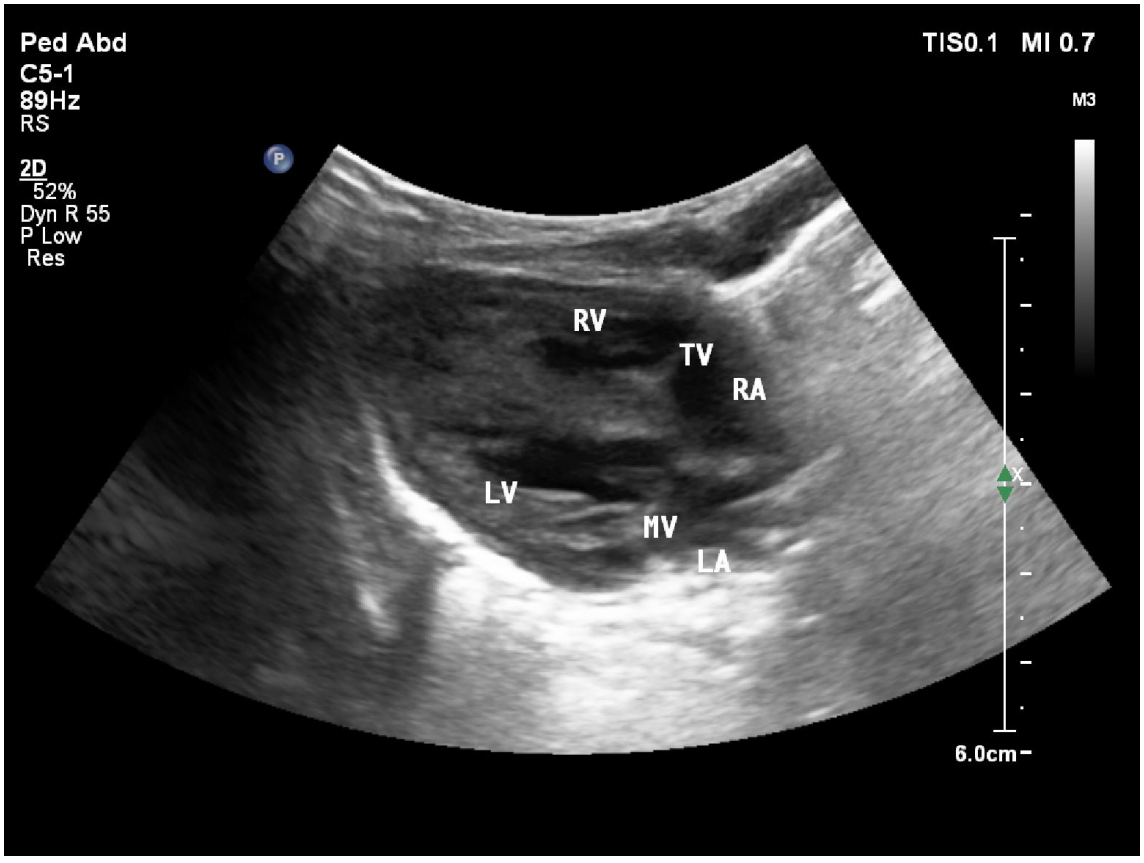

Fig. 2. ECHO showing normal valves and chambers

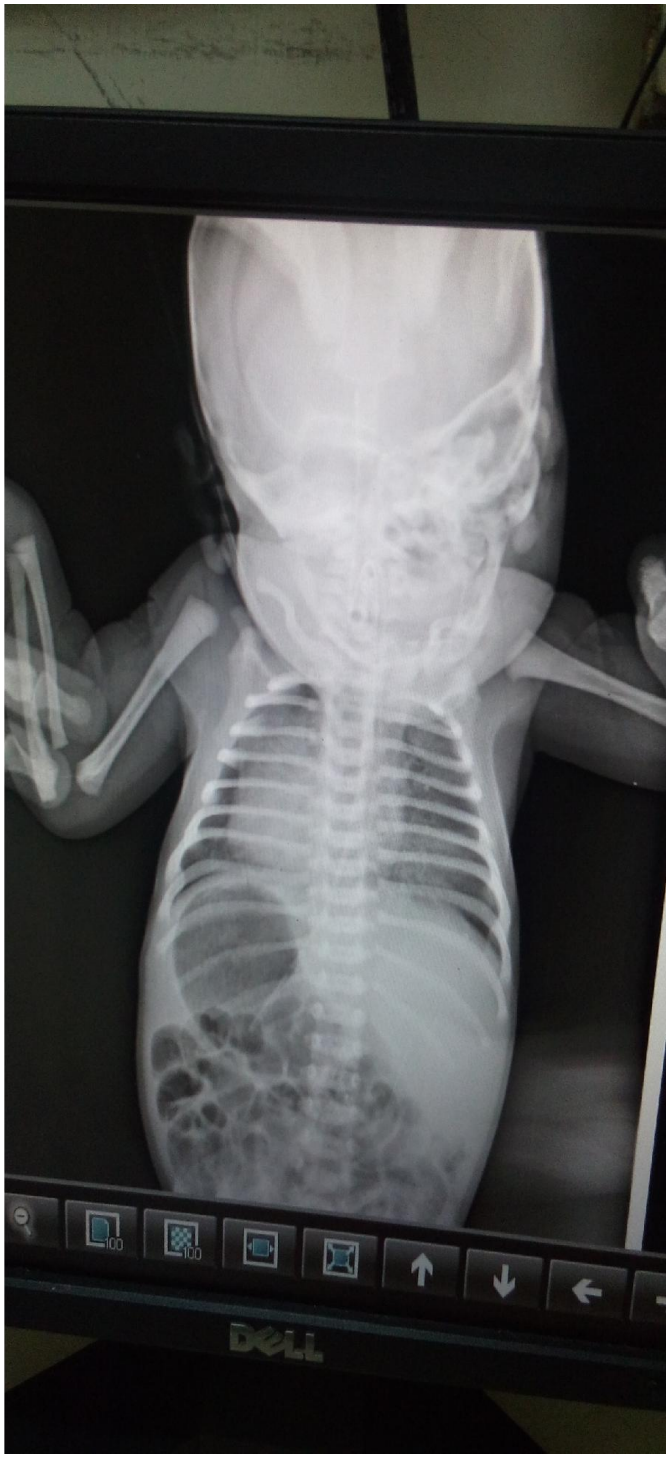

Fig. 3. X-ray showing the position of heart and fundus shadow on the right side

Also called a looping defect, it is an abnormal congenital positioning of heart on the right side (Tabry *et al*., 2001). Situs tells the position of cardiac atria and the viscera. 'Solitus' is the 'normal' position and 'Inversus' is the 'reverse' position. SitusInversus Totalis is SitusInversus with Dextrocardia i.e. mirror image of normal anatomy. It is a rare condition with a prevelance of 1:10,000 in some population (Oppido *et al*., 2004). It is generally an autosomal recessive genetic condition, although it can be X – Linked or found in identical twins (Radhika *et al*., 2011). In human, 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 (Ryan *et al*., 1998). The condition is often associated with other heart defects like ASD and VSD; as in our case there is patent foramen ovale, although the incidence of this is low (0-10%) (Kulkarni and Inamdar, ?). SitusInversus Totalis is found associated with primary ciliary dyskinesia termed as Kartagener Syndrome (Ortega *et al*., 2007). The exact cause is unknown, dextrocardia has been linked with several factors including recessive gene with incomplete penetrance, maternal diabetes, cocaine use and conjoined

twinning (Agirbashi *et al*., 2000). Although in our case, the cause could not be well known or found. The management of such patients is required only in presence of congenital heart defects or other associated syndromicfeatures, otherwise this condition goes unnoticed with normal life expectancy (Garba *et al*., 2014). Coronary angiography was first reported in dextrocardia in 1974 (Ilia *et al*., 1988). The most important modification in performing coronary angiography in such patients are opposite – direction catheter rotations and mirror image angiographic angles i.e. anticlockwise rotation needed in the ascending aorta for right coronary artery and reversing the required right anterior oblique angles, keeping cranial / caudal tilts the same (Zhang *et al*., 2008).

Conclusion

The condition is rare and with limited literature. Our case is one such reported in a term newborn with unknown causes and presentation with respiratory difficulty which was symptomatically relieved and follow up advised. Parents should be counseled and made aware of this near normal & benign condition and further role of intervention described in any condition of abnormality or association found.

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