

www.jmscr.igmpublication.org

Impact Factor 3.79

ISSN (e)-2347-176x

**crossref** DOI: <http://dx.doi.org/10.18535/jmscr/v3i9.65>



**Journal Of Medical Science And Clinical Research**

An Official Publication Of IGM Publication

## Situs Inversus Totalis- A Case Report and Review

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### Abstract

*Dextrocardia with situs inversus or situs inversus totalis is a condition that is characterized by abnormal positioning of the heart and other internal organs i.e. there will be reversal of positions of major thoracic and abdominal organs. In people affected by dextrocardia, the tip of the heart points towards the right side of the chest instead of the left side. Some affected people have no obvious signs or symptoms. However, a small percentage of people also have congenital heart defects. Here we are reporting a case of situs inversus totalis in a new born which was detected in the antenatal period itself.*

**Keywords –** *Dextrocardia, Situs Inversus, congenital heart diseases*

### Introduction

Situs inversus viscerum can be either total or partial. Total situs inversus, also termed as mirror image dextrocardia, is characterized by a heart on the right side of the midline while the liver and the gall bladder are on the left side. Patients are usually asymptomatic and have a normal lifespan. The exact etiology is unknown but an autosomal recessive mode of inheritance has been speculated. However, situs inversus abdominus, characterized by 'mirror image' of the normal bowel, is caused by a clockwise rotation of the viscera during early embryonic life. Very few cases of situs inversus totalis have been described in the literature <sup>[1]</sup>.

### Case report

A 24 year old Mrs X, who was Gravida 4 Para 2 Living 1 Abortion 1 with previous two vaginal deliveries, was admitted to labour ward in active labour at 38 weeks of gestation. Last child birth was four years back, it is a male child now active and healthy. In the present pregnancy the anomaly scan done at 24<sup>th</sup> week of gestation showed fetal situs inversus totalis. She delivered a male baby weighing 2.9 kgs vaginally which cried immediately after birth but the heart sounds were prominently heard on the right side. Since the anomaly scan had a mention of situs inversus totalis the baby was further evaluated by a chest X ray and a 2D echo, which showed dextrocardia with situs inversus with a small PDA of 2mm, left

to right shunt with mild tricuspid regurgitation, moderate PAH with good biventricular function.



X ray of the chest and abdomen showing the apex of the heart towards right side and liver on the left side

Postnatal period was uneventful and she was discharged on 4<sup>th</sup> postnatal day with both mother and baby in good general condition

### Discussion

The term “situs” means position, site, or location. Situs inversus totalis is a congenital condition in which the major visceral organs are reversed from their normal position. The normal arrangement is known as Situs Solitus in which the heart is on the left side, 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 [2]. The other rare case is known as Situs ambiguus or heterotaxy, where in situs cannot be determined. 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. Matthew Baillie described the complete mirror-image reversal of the thoracic and abdominal organs in situs inversus.

Situs inversus is present in 0.01% of the population. Situs inversus is generally an autosomal recessive genetic condition, sometimes it can be X-linked and also found in identical twins. The Situs inversus with Dextrocardia or Situs inversus totalis has been estimated to occur once in about 6-8,000 live births. Situs inversus

with levocardia or situs inversus incompletes is another rare condition (1 in 22,000 of general population) in which the heart is found on the normal left side of the thorax. Recent studies suggest that left-right asymmetry defects are due to genetic abnormalities in lefty genes, nodal genes, and ZIC 3, ACVR2B and Pitx2 genes and mutation of genes present on chromosome 12 [3]. Most people with situs inversus have no medical symptoms or complications resulting from the condition except difficulty in diagnosing appendicitis, auscultating heart sounds and palpation of liver etc. during routine clinical examination. Around 3-5% of people with situs inversus have any type of functional heart defect, this is higher than the rate of heart defects in the general population, which is less than 1%. Common congenital cardiac defects reported include transposition of the great arteries and ventricular septal defects. It is estimated that about 25% of people with situs inversus have an underlying condition called primary ciliary dyskinesia (PCD) [4].

### Conclusion

The individuals with situs inversus are phenotypically unimpaired, and can lead normal healthy lives, without any complications related to their medical condition. Many people with situs inversus totalis are unaware of their unusual anatomy until they seek medical attention for an unrelated condition.

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