

## Case Report

# Dextrocardia with *Situs Inversus Totalis* - A Rare Anatomical Variation: A Case Report

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## ABSTRACT

A 35-year-old female dead body was brought to the mortuary for post-mortem examination as a burn case. During examination, incidentally, we found that there was an anatomical variation termed as *situs inversus totalis*. The term *situs inversus* is a short form of the Latin phrase '*situs inversus viscerum*', meaning inverted position of the internal organs. The *situs inversus totalis* is a rare syndrome, with an estimated prevalence of 1 in 10,000 births, characterised by the inverted position of the thoracic and abdominal organs with respect to the sagittal plane. *Situs inversus totalis*, a not so uncommon congenital positional anomaly, can be a diagnostic problem at times. The condition affects all major structures within the thorax and the abdomen. Surgeons and radiologists need to be aware of this anomaly during preoperative and surgical management. Routine premedical examination helps the patient to be aware of his/her condition, thereby preventing wrong diagnosis and possibly death due to delay in surgical management; it can also be used as an identification parameter.

**Keywords:** *Situs inversus totalis*, Anatomical, Sagittal plane, Syndrome, Abdominal organ

## INTRODUCTION

*Situs inversus* is a short form of the Latin phrase '*situs inversus viscerum*' meaning 'inverted position of the internal organs', as first described by Marco Severino in 1643.<sup>[1]</sup> The incidence is seen in about 1 in 10,000 live people. Normal human development results in an asymmetrical arrangement of the organs within the chest and the abdomen. Typically, the heart lies on the left side of the body (*levocardia*) and the liver and the spleen lie on the right. Mirror image, i.e. transposition of the abdominal and thoracic viscera, is termed as *situs inversus totalis*. It exhibits anomalies, such as rotation, fixation during the development and a perfect mirror image of opposite side. Such relationship between the organs does not lead to functional problems in most cases<sup>[1]</sup>. It is also called *situs transversus or oppositus*. It is a congenital condition in which the major visceral organs are reversed from their normal position. The normal

arrangement is known as *situs solitus*. The other rare case is known as *situs ambiguous or heterotaxy*, wherein the *situs* cannot be determined. Dextrocardia was first observed by Leonardo da Vinci in 1452-1519, and then recognised by Marco Aurelio Severino in 1643 and described more than a century later by Matthew Baillie. Matthew Baillie described the complete mirror-image reversal of the thoracic and abdominal organs as *situs inversus*. *Situs inversus* is present in 0.01% of the population<sup>[2]</sup>. It is generally an autosomal recessive genetic condition, and sometimes can be X-linked and also found in identical twins<sup>[3,4]</sup>. 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*<sup>[5]</sup> is another rare condition (1 in 22,000 general population) in which the heart is found on the normal left side of the thorax<sup>[2]</sup>. Many people with *situs inversus* are unaware of their unusual anatomy until

they seek medical attention for unrelated conditions<sup>[6]</sup>. One case report found that the stomach and the spleen were located on the right side, while the liver was on the left side<sup>[7]</sup>. In a related report of chronic gallbladder inflammation, *situs inversus* with the liver and the gallbladder on the left side was observed. The gallbladder located in the epigastric area towards the left side and the spleen on the right was discovered in a 76-year-old man<sup>[8]</sup>. A case of dextrogastric, multiple jejunal atresia and inverse rotation of the intestine with levocardia in a neonate was reported<sup>[9]</sup>.

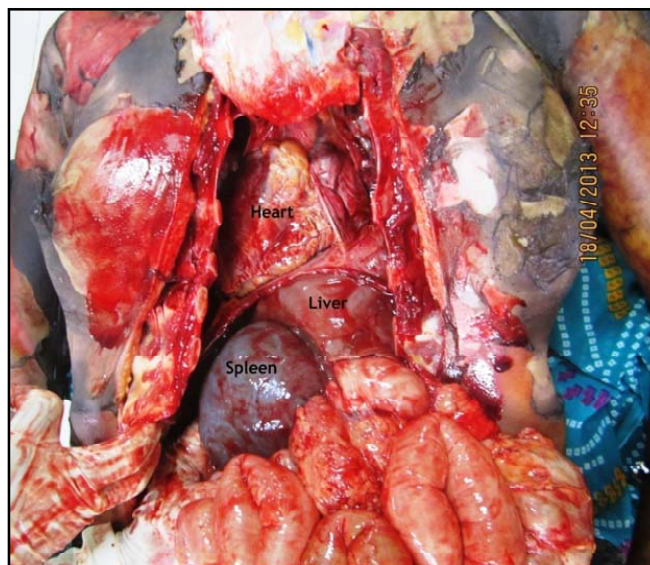
### CASE REPORT

A 35-year-old female dead body was brought to the mortuary for post-mortem examination with an alleged history of death due to burn injuries. She was burnt at home, accidentally, by the bursting of the stove. She was initially admitted at the District Hospital, Badwani, where they noted 96% superficial-to-deep burns present all over the body. Then, she was referred to MY Hospital and MGM Medical College, Indore, for further treatment at the burns ward. When she was brought here, she was declared dead and the body was sent to mortuary for post-mortem examination by the police with a requisition form. During post-mortem examination, after opening the thoracic and abdominal cavity, we found that the viscera were not present at their normal locations. Variations were found in the positions of the viscera and they showed dextrocardia with transposition of great vessels, with the base of the heart situated at the right side of the chest cavity. The right lung had two lobes and the left lung contained three lobes. The spleen was situated on the right side of the abdominal cavity and the liver was situated on the left side of the abdominal cavity. Greater curvature of the stomach was on the right side and no organ enlargement was observed. Kerosene oil-like smell was present on the scalp hairs, the uterus was enlarged and gravid and the vascular dimensions were 48x22x20 cm<sup>3</sup>. After opening the uterus, we found a well-developed male foetus, weighing 2.8 kg, with the umbilical cord found *in situ*; the foetal measurements were: foetal length 44 cm, head circumference 31 cm, chest circumference 24 cm, abdominal circumference 20 cm, umbilical cord length 46 cm, placenta weight 450 gm, mid-arm circumference

07 cm, mid-thigh circumference 11 cm of foetus present in the uterus.

### DISCUSSION

Dextrocardia with *situs inversus* is a rare anomaly occurring in about 1 in 10,000 people<sup>[10]</sup>. Its frequency is between 1 in 8000 and 1 in 20,000<sup>[11]</sup>. It may be total (*situs inversus totalis*) or incomplete in less than 10% of the cases. Many people with *situs inversus totalis* are unaware of their unusual anatomy until they seek medical attention for an unrelated condition. In our case report, we observed a female dead body with dextrocardia and *situs inversus totalis* (Figure 1). It is pertinent to note that people with this anomaly have higher chances of suffering from other problems of the heart, especially if other organs are affected too. Some individuals may find that they suffer no symptoms at all until they are in the later stages of life, whilst others may suffer from some serious renal, respiratory, gastrointestinal and cardiovascular diseases<sup>[12]</sup>. For instance, Danbauchi and Alhassan<sup>[13]</sup> reported two cases of dextrocardia with *situs inversus*. The first patient was a 35-year-old man presented for the first time with respiratory symptoms, but no cardiac symptoms, and the second patient (14-year-old) presented with cardiac symptoms<sup>[13]</sup>. In



**Figure 1: Anterior view of the chest and abdominal cavity showing the heart and the spleen on right side and the liver on left side of the sagittal plane**

addition, Treiger *et al.*<sup>[11]</sup> reported a clear-cell carcinoma in the kidneys of a patient with *situs inversus*, even though the two kidneys were normal on macroscopic examination. Nawaz *et al.*,<sup>[10]</sup> in their report, described a case of two newborns with *situs inversus* in association with congenital duodenal obstruction. In our study, the left lung had three lobes and the right lung had two lobes, but in a case reported in Nigeria, Ofusori *et al.*<sup>[12]</sup> observed that the right lung was uni-lobed and the larger of the two lungs, while the left lung had three lobes. Our case report finding is almost similar with that reported by Sharma *et al.*<sup>[1]</sup> The apex of the heart is located on the right side of the thorax, the stomach and spleen on the right hypochondriac region in the abdomen and the large lobe of the liver and the gallbladder on the left side. The left lung is tri-lobbed and the right lung is bi-lobbed<sup>[1]</sup>.

## CONCLUSION

Because dextrocardia and *situs inversus* are asymptomatic and do not cause any long-term problems, it would be very dangerous if the conditions are not diagnosed prior to surgery. Often it is diagnosed during a medical examination or during a routine visit to the hospital, when the cardiac function is examined. The surgeons and the radiologists must be sensitised to be aware of this anomaly during the presurgical and surgical management, and also a routine medical examination must be encouraged, which could give a signal to the existence of *situs inversus* condition if present. This will help the patient when afflicted with certain clinical conditions, such as appendicitis, where the referred pain will be on the left side rather than the right, thus leading to wrong diagnosis and possibly death due to delay in surgical management. It can be an important parameter for the identification of unidentified bodies, if previously diagnosed with this type of anomaly.

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