

Dextrocardia with left lung lobe variation in foetus: A rare case report

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Abstract

Introduction: Dextroversion has been recognized under several different names in the past: "isolated" dextrocardia, incomplete rotation of the heart, dextrotorsion, etc. However, the comprehensive studies by Korth and Schmid have made it clear that it is a distinctive syndrome with broader clinical and embryologic implications than these names imply, justifying the special term dextroversion. In general, dextroversion consists of a rotation of the ventricular part of the heart to the right, as in turning the page of a book, with the atria remaining in normal position. Dextrocardia is an abnormal congenital positioning of the heart. Instead of the heart forming in the fetus on the left side, it flips over and forms on the right side. **Case Report:** In this case report Dextrocardia was observed in one of the fetal cadavers, the stomach and spleen were seen in their normal position on the left and the liver on the right. The heart is somewhat flattened and flipped towards the right with its cardiac apex facing the right. This also known as SitusSolitus or Isolated Dextrocardia. Left lung was also found to be defective having two fissures (horizontal and oblique) and three lobes (superior, middle and inferior) representing partial situs inversus. **Conclusion:** Situs anomalies are rare conditions which present with diagnostic and management challenges. Surgeons and radiologists should be aware of this anomaly during preoperative and surgical management of their patients. It is very dangerous if this condition is not diagnosed prior to surgery. Doctors should encourage routine medical examination of their patients to identify this anomaly and prevent wrong diagnosis.

Keywords: Dextrocardia, SitusSolitus.

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INTRODUCTION

Dextroversion has been recognized under several different names in the past: "isolated" dextrocardia, incomplete rotation of the heart, dextrotorsion, etc. However, the comprehensive studies by Korth and Schmidt¹ have made it clear that it is a distinctive syndrome with broader clinical and embryologic implications than these names imply, justifying the

special term dextroversion. In general, dextroversion consists of a rotation of the ventricular part of the heart to the right, as in turning the page of a book, with the atria remaining in normal position. Dextrocardia is an abnormal congenital positioning of the heart. Instead of the heart forming in the fetus on the left side, it flips over and forms on the right side². Dextrocardia is frequently diagnosed in a routine prenatal sonogram, although not every radiologist will catch it, particularly if there are no cardiac structural abnormalities. There are several types of dextrocardia, also called looping defects. 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 also.³, as seen in a case study by Nawaz *et al* in their case report which they discovered that the stomach and spleen were located on the right side, while the liver was on the left side, the gallbladder was located in the epigastric area toward the left side⁴. Foetal dextrocardia is a condition in which the major axis of the heart (from the base to the apex along the

interventricular septum) points to the right. Dextrocardia should be distinguished from dextroposition, in which the heart is shifted into the right chest as a consequence of pathological states involving the diaphragm, lung, pleura, or other adjoining tissues.⁵ The term dextrocardia describes only the position of the cardiac axis and conveys no information regarding chamber organisation and structural anatomy of the heart.⁶⁻¹⁰ In the postnatal period a broad spectrum of cardiac malformations is observed associated with dextrocardia, and the incidence varies according to the atrial situs. Complex cardiac heart malformations are found more often with situs solitus and situs ambiguus than with situs inversus.^{7,8,9,10} Diagnostic modalities like a chest radiograph and an electrocardiogram are sufficient to make a diagnosis of dextrocardia while more recent imaging modalities like echocardiography and magnetic resonance imaging puts the diagnosis beyond doubt¹¹.

CASE REPORT



Figure 1



Figure 2



Figure 3

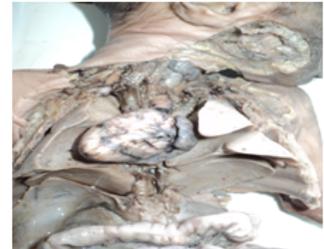


Figure 4

During a supervision of dissection of formalin-fixed foetal cadavers as part of the medical project in the Department of Anatomy, dextrocardia was observed in one of the fetal cadavers. Our observation became interesting as it provided an avenue for anatomists to better understand and appreciate anatomy of the thorax well in such a very rare anomaly. In our case report, the stomach and spleen were seen in their normal position on the left and the liver on the right. The heart is somewhat flattened and flipped towards the right with its cardiac apex facing the right. This type of dextrocardia is known as Dextrocardia with Situs Solitus or Isolated Dextrocardia where there is transposition of the heart without accompanying alteration of the abdominal viscera. Left lung was also found to be defective having two fissures (horizontal and oblique) and three lobes (superior, middle and inferior). Also the presence of three lobes in left lung appears to be an instance of the group of malformations representing partial situs inversus.

DISCUSSION

Though the exact cause is unknown, dextrocardia has been linked with a number of factors including autosomal recessive gene with incomplete penetrance, maternal diabetes, cocaine use and conjoined twinning. The arrangements of the position of the abdominal viscera in dextrocardia may be normal (Situs Solitus), reversed (Situs Inversus), and indeterminate (Situs Ambiguus or Isomerism). In our case report, we observed situs solitus dextrocardia in a foetal cadaver. Also the presence of three lobes in left lung appears to be an instance of the group of malformations representing partial situs inversus. Various studies in literature state that Dextrocardia with situs solitus results from failure of final leftward rotation of ventricles into right chest during embryogenesis. Calvin *et al*¹² in his work discovered dextrocardia with situs solitus in a 3-year-old boy presented with a chest infection. Dextrocardia with a Situs Solitus has been highly associated with congenital cardiac anomalies like transposition of the great vessels, atrial and ventricular septal defects. Often it is diagnosed during a medical examination or during a routine visit to

hospital when cardiac function is examined or even during surgery. Surgery in such cases is rendered difficult by the unfamiliarity of the surgeon with such an orientation of the heart. A curious anomaly often associated with dextroversion, and one which further emphasizes the difference between dextroversion and mirror-image dextrocardia with situs inversus, is congenital absence of the spleen, a subject that has recently been reviewed by Ivemark¹² and Putscher and Manion.¹³ Ten of the 69 autopsied cases of dextroversion (including case 2 of the present report) had asplenia, while among hundreds of cases of situs inversus totalis reported in the literature, only 1 had asplenia and in this case the diagnosis was made during surgical exploration and not at autopsy.¹⁴ Situs solitus was the most common type in A. Bernasconi *et al*'s study (47%), which is in contrast to the study of Walmsley *et al*,¹³ in which situs solitus was least frequent (22%).¹³ All the fetuses with dextrocardia and situs solitus in the study of Walmsley *et al*¹³ had a cardiac malformation compared with 66% in our study. In postnatal series, the incidence of a normal heart varied between 0–9%²⁻⁷ The fetuses

with a normal heart in our population were mostly referred because of associated extracardiac malformations and not because there was concern about the fetal heart. This may explain the discrepancies between prenatal and postnatal studies. With Situs inversus had structurally normal hearts Walmsley *et al.*¹⁵ In those with a cardiac malformation a wide spectrum of congenital heart disease was seen, most of which were complex.¹⁶⁻¹⁷ However, the best survival was observed in this group. Situs ambiguus as Walmsley *et al.*¹⁵ it is higher incidence in fetal series may be explained by the possibility that many of the fetuses may not survive, either because of interruption of pregnancy or because of death in utero or during the early neonatal period.

CONCLUSION

Situs anomalies are rare conditions which present with diagnostic and management challenges. Surgeons and radiologists should be aware of this anomaly during preoperative and surgical management of their patients. It is very dangerous if this condition is not diagnosed prior to surgery. Doctors should encourage routine medical examination of their patients to identify this anomaly and prevent wrong diagnosis.

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