

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

Yogesh Galphade^{1*}, Smita Galphade², S S Dhapate³

¹Speciality Medical Officer} ²Assistant Professor, Department of Anatomy} TNMC and BYL Nair Ch. Hospital, Mumbai, Maharashtra, INDIA.

³Professor and HOD, Department of Anatomy, SRTR Government Medical College, Ambajogai, Beed, Maharashtra, INDIA.

Email: yash141985@gmail.com

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 situsinversus. **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.

*Address for Correspondence:

Dr. Yogesh Galphade, Speciality Medical Officer} ²Assistant Professor, Department of Anatomy} TNMC and BYL Nair Ch. Hospital, Mumbai, Maharashtra, INDIA.

Email: yash141985@gmail.com

Received Date: 30/10/2015 Revised Date: 22/11/2015 Accepted Date: 26/12/2015

Access this article online

Quick Response Code:



Website:

www.statperson.com

DOI: 10 February
2016

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

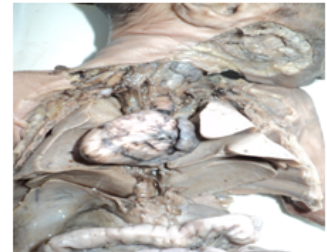


Figure 4

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.

REFERENCES

1. Korth, C., And Schmidt, J.: Die Klinik der Dextrokardien. Arch. f. Kreislaufforsch. 21: 188, 1954.
2. Tabry, I. F.; Calabrese, J.; Zammar, H.; AbouKasem, K.; Akeilan, H.; Gharbieh, N.; Zinati, H.; Noureddine, W.; elHout, A.; Tayah, M.; Khalidy, L. and Yaghi, M.; 2001. Case Report: Off-Pump Total Myocardial Revascularization for Dextrocardia and Situs Inversus. Heart Surg. Forum, 4(3):251-3. 3
3. Ofusori DA, Okwuonu CU, Ude RA, Adesanya OA; 2009. Dextrocardia and situs inversus totalis in a Nigerian cadaver: A case report of rare anomaly. Int J Morphol; 27(3): 837-840.
4. Nawaz, H.; Matta, M.; Hamchou, A. and Jacobez, A. H. A. Salem; 2005. Situs inversus abdominis in association with congenital duodenal obstruction: a report of two cases and review of the literature. Pediatr. Surg. Int., 21:589-92.
5. Russ PD, Weingardt JP. Cardiac malposition. In: Drose JA, ed. Fetal echocardiography. 1st ed. Philadelphia: WB Saunders, 1998:59-73.
6. Calcaterra G, Anderson RH, Lau KC, et al. Dextrocardia: value of segmental analysis in its categorisation. Br Heart J 1979; 42:497-507. [PMC free article] [PubMed]
7. Garg N, Agarwal BL, Modi N, et al. Dextrocardia: an analysis of cardiac structures in 125 patients. Int J Cardiol 2003; 88:143-55. [PubMed]
8. Huhta JC, Hagler DJ, Seward JB, et al. Two-dimensional echocardiographic assessment of dextrocardia: a segmental approach. Am J Cardiol 1982; 50:1351-60. [PubMed]
9. Squarcia U, Ritter DG, Kincaid OW. Dextrocardia: angiographic study and classification. Am J Cardiol 1974; 33:896-903.
10. Stanger P, Rudolph AM, Edwards JE. Cardiac malposition: an overview based on study of 65 necropsy specimens. Circulation 1977; 56:159-72. [PubMed]
11. Uchenna DI, Jesuorobo DE, Anyalechi JI; 2012. Dextrocardia with Situs Inversus Totalis in an Adult Nigerian: a case report, department of Internal Medicine, University of Port Harcourt Teaching Hospital, Port Harcourt, Nigeria.
12. Ivemark, B. J.: Implications of agenesis of spleen in pathogenesis of cono-truncus anomalies in childhood. Acta Paediat. 41: suppl. 104, 1955, p. 590.
13. Putschar, W. G., And Manion, W. C.: CDngenital absence of spleen and associated anomalies. Am. J. Clin. Path. 26: 429, 1956.
14. Foxj, P., And Crawford, O. W.: Duodenal obstruction, situs inversus and non-rotation of the colon. Surgery 27: 896, 1950.
15. Walmsley R, Hishitani T, Sandor GG, et al. Diagnosis and outcome of dextrocardia diagnosed in the fetus. Am J Cardiol 2004; 94:141-3. [PubMed]
16. Calcaterra G, Anderson RH, Lau KC, et al. Dextrocardia: value of segmental analysis in its categorisation. Br Heart J 1979; 42:497-507. [PMC free article] [PubMed]
17. Stanger P, Rudolph AM, Edwards JE. Cardiac malposition: an overview based on study of 65 necropsy specimens. Circulation 1977; 56:159-72. [PubMed]
18. A Bernasconi, A Azancot, J M Simpson, A Jones, and G K Sharland, Fetal dextrocardia: diagnosis and outcome in two tertiary centres. Heart. 2005 Dec; 91(12): 1590-1594. doi: 10.1136/hrt.2004.048330

Source of Support: None Declared
Conflict of Interest: None Declared