

Situs Inversus- Report Of Twins

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Abstract: Dextrocardia with situs inversus (situs inversus totalis) is rare congenital anomaly which can be asymptomatic and compatible with normal life. Incidence is 1 in 10000. But in cases of situs inversus with levocardia association of congenital heart disease is high. We are presenting a pair of twins, born out of nonconsanguineous marriage, from a primigravida mother, with an uneventful antenatal period, at 34 weeks 5 days of gestation. Both the twins did not require active resuscitation. On routine examination it was found that, both the twins had situs inversus and chest x ray, echocardiography and abdominal ultrasound was done, which confirmed the same. Our 1st twin was a case of situs inversus totalis (cardiac apex on right side) without any congenital heart disease and 2nd twin was a case of situs inversus with levocardia (cardiac apex on left side) with complex congenital cyanotic heart disease. To conclude newborn babies should have a thorough physical examination after delivery before discharge to enable early diagnosis of congenital anomalies for appropriate referral. Though situs inversus in single neonate is well described, but we have not come across any case report in which both twins have situs inversus with or without dextrocardia, so we think the case is worth reporting.

Keywords: neonate, twins, dextrocardia, situs inversus.

I. Introduction

Situs inversus is a congenital positional anomaly, characterized by transposition of abdominal viscera, and when associated with a right sided heart (Dextrocardia) is termed as situs inversus totalis [1]. The incidence is about 1:10,000 live people. Many people with situs inversus are unaware of their unusual congenital anomaly, until they seek medical attention for any unrelated condition. Individuals with isolated dextrocardia and situs inversus totalis may have associated congenital heart malformations [2], primary ciliary dyskinesia or splenic malformations [3]. We are presenting two newborns (twins) one have situs inversus with dextrocardia (situs inversus totalis) and other have situs inversus with levocardia, which is very rare.

II. Case

Pair of male twins were delivered through caesarean section (indication- twin pregnancy with rupture of membrane) at 34 weeks and 5 day from a primigravida mother. Both the babies did not require active resuscitation, only routine care was given. Antenatal, natal and immediate postnatal periods were normal. Anthropometry of the babies were as follows, 1st twin- weight 2018 grams, length 43cm, head circumference 32 cm and that of the 2nd twin- weight 1937 grams, length 41cm, head circumference 31.5cm. On examination of the 1st twin revealed normal vital parameters and on auscultation, heart sounds were more heard on the right side of the chest and palpation of the abdomen showed liver on the left side. While examining the 2nd twin we found that he had cyanosis and oxygen saturation is between 82-87% and no significant improvement on supplemental oxygen, heart sounds were on the left side and a pansystolic murmur is heard on the left lower sternal border, liver is palpated below the left costal margin. With these findings diagnosis of situs inversus was thought of in both the cases and investigations done for confirmation. X ray of chest and upper abdomen of the 1st twin (Fig 1) showed dextrocardia with situs inversus of the abdominal organs (fundic gas on right side and liver on left side of abdomen) and in case of 2nd twin heart position was normal but there was transposition of abdominal organs (Fig 2). Echocardiography was normal in the 1st baby except for dextrocardia but in case of 2nd baby echocardiography showed levocardia and congenital heart disease in the form of AV canal defect, ostium primum ASD and subaortic VSD with normal biventricular function. Abdominal ultrasound confirmed transposition of abdominal viscera. Both babies were on breast feeding and kept on follow up, especially the second twin for any sign of congestive heart failure.

III. Discussion

In the intrauterine life, early in the normal development of an embryo, the tube-like structure that becomes the heart forms a loop towards the left, identifying the left/right axis along which the other organs should be positioned. Although the mechanism that causes the heart loop to go left is not fully understood. However, it is thought that many factors may be involved in causing situs inversus. Most often situs inversus is an isolated and accidental event occurring in an individual for the first time in the family but rarely, it can occur

in families. Situs inversus is generally an autosomal recessive genetic condition, although it can be x-linked or found in identical twins [4]. Our cases were identical twins of same sex. Situs inversus totalis is a condition in which the organs of the chest and abdomen are arranged in a perfect mirror image reversal of the normal positioning. 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. Dextrocardia with situs inversus has been known to be associated with a lower incidence of congenital heart disease (0-10%) unlike its association with situs solitus (the normal levoposition of the heart) with up to 90% of cases [5, 6]. Although only 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 [7, 8, 9].

It is estimated that about 25% of people with situs inversus have an underlying condition called primary ciliary dyskinesia (PCD) [10].

Our 1st twin was a case of situs inversus totalis without any congenital heart disease and 2nd twin was a case of situs inversus with levocardia with complex congenital cyanotic heart disease.

IV. Summary

Situs inversus totalis should be detected by a thorough physical examination. To confirm the suspected diagnosis of situs inversus, imaging studies should be done depending on the patient such as MRI, CT, or ultrasound may be ordered, and a referral may be made to a cardiologist. Imaging studies will also rule out the possibility of random arrangement of the organs, or heterotaxy, which has a much higher risk for serious medical complications. Documenting situs inversus in an individual is important in order to correctly interpret any future symptoms and avoid any inadvertent clinical or surgical mishap. Our case is one of the rarest reported combinations of situs inversus totalis and situs inversus with levocardia in identical twins, which we thought is worth reporting.

References

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Figure legends and headings:

Figure 1- (x ray of chest and upper abdomen of twin 1) cardiac apex towards right side, Liver shadow below left diaphragm and fundic gas on right side



Figure 2-(x ray of chest and upper abdomen of twin 2) cardiac apex towards left side,
Liver shadow below left diaphragm and fundic gas on right side

