



Volume :2, Issue :4, 220-221
April 2015
www.allsubjectjournal.com
e-ISSN: 2349-4182
p-ISSN: 2349-5979
Impact Factor: 3.762

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Successful Pregnancy outcome in a primigravida with twin gestation with complete situs inversus: A rare case report

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Abstract

Situs inversus is a rare congenital positional anomaly characterized by right sided heart (Dextrocardia) and transposition of abdominal viscera. This case report discusses about a unbooked 23 year old primigravida with twin gestation came with labour pains was found to have dextrocardia was taken up for emergency caesarean section in view of non-progression of labour and fetal distress. Intrapartum was eventful . Post surgery the mother was investigated in view of dextrocardia and found to have situs inversus . Situs inversus totalis is not associated with any significant morbidity or mortality and have a normal life expectancy in a patient possessing it. However, its timely diagnosis is crucial and documentation of situs inversus in an individual is important to correctly interpret any future symptoms and avoid any inadvertent clinical or surgical complications.

Keywords: situs inversus totalis , dextrocardia , twin gestation

1. Introduction

Situs inversus is a rare congenital positional anomaly characterized by right sided heart (Dextrocardia) and transposition of abdominal viscera. Dextrocardia was first seen by Leonardo da Vinci in 1452-1519 and then recognized by Marco Aurelio Severine in 1643. Situs inversus, first described by Aristotle in animals and fabricus in humans¹. In early twentieth century, it was Mathew Baille first described situs inversus, a complete mirror-image reversal of the thoracic and abdominal organs². Situs inversus is present in 0.01% of the population. In latin, situs inversus is inverted position of internal organs. Very few cases of situs inversus totalis have been reported in the literature.

Case Report

A 23 year old Primigravida came with 37 weeks twin gestation, Unbooked with us, all trimesters were uneventful. She got admitted with labour pains 5 hrs prior to admission. O/E: Pulse - 82/min, regular and BP of 130/ 90 mm Hg, CVS: apex beat localized in the right 4th intercostal space, S1, S2 are heard in the right side, RS- NAD. There was no apparent spinal and neurological abnormalities. P/A: Uterus over distended, acting (2c/20/10"), 1st twin in cephalic presentation, FHS¹- 132/min, FHS²-140/min. P/V: cervix - 50% effaced, OS 2 cm dilated, 1st twin- PPVx @ -3 station, membranes absent, liquor clear. Reactive NST.

Investigations done

- Blood Sugar -83 mg%
- HIV - Non reactive
- HBsAg - Negative
- ECG revealed right axis deviation.
- USG Abdomen(before delivery) - Situs inversus, Twin A - Gestational age corresponding to 33 weeks 3 days ± 3 weeks, Twin B - Gestational age corresponding to 33 weeks 5 days ± 3 weeks.

A - BPP: 8/8 AFI: 8-9 cms EFW: 2.3 kgs ± 15%

B - BPP: 8/8 AFI: 8-9 cms EFW: 2.2 kgs ± 15%



USG Abdomen showing right sided spleen and left sided liver

- USG Abdomen and pelvis (after delivery): Situs inversus, liver on left side and a left sided gall bladder, spleen on right side with normal echostructure.
- 2D Echo - Dextrocardia with normal sized cardiac chambers and normal LV function.
- X-Ray- Chest PA view- Showed Dextrocardia

Patient was unaware of this condition until this point, with no significant family history.

Patient underwent emergency LSCS in view of non-progression of labour under spinal anaesthesia.

POF: 1st twin – an live term SGA female baby of Birth Weight -2.07 kgs with Apgar: 1[']- 7/10, 5[']- 9/10, 2nd twin- an live term SGA male baby of Birth Weight -1.69 kgs with Apgar: 1[']- 7/10, 5[']- 8/10 was delivered. Placenta – Dichorionic Diamniotic. There was complete situs inversus with the liver on the left side and spleen on the right side. The stomach was on the right and the first part of the duodenum to the left of the midline. The rest of the abdomen was explored showed features typical of situs inversus totalis, that is, the caecum and appendix in the left iliac fossa and the sigmoid colon on the right side. Intra and post-operative recovery was uneventful.

2. Discussion

Situs inversus totalis is a rare anomaly with an incidence varying from one in about 6-8,000 live births², the exact etiology is not known and is an autosomal recessive condition with equal distribution in both gender³. Situs inversus viscerum can be either total or partial. Situs inversus totalis

with Dextrocardia is associated with only 3-5% incidence of congenital heart disease and in about 80% of individuals have right-sided aortic arch¹. Levocardia with situs inversus will almost always associated with congenital heart disease.

Usually remains asymptomatic and undiagnosed, as typified by the present case, and is diagnosed incidentally while investigating for another ailment. In our case the diagnosis was made pre-operatively and she underwent emergency LSCS.

Certain congenital anomalies such as polysplenia (left isomerism) /asplenia (right isomerism) or Kartagener's syndrome (primary ciliary dyskinesia often leading to infection of the paranasal sinuses and lungs) are known to occur in some patients. However, our patient did not have any of these abnormalities².

Various modalities such as ECG, chest x-ray, MRI /CT scans with oral and intravenous contrast, ultrasound, and barium studies can be used to diagnose situs inversus, and possibility of situs ambiguous as well. In our case, we diagnosed situs inversus by abdominal ultrasonography and echocardiogram.

Our patient failed to follow up, as she was advised to review for further investigations like CT scan and other diagnostic modalities⁴.

Life expectancy in individual with situs inversus and Dextrocardia is same as that with the general population. In case of isolated Dextrocardia, prognosis depends on the structural and functional defects.

3. Conclusion

Situs inversus totalis is not associated with any significant morbidity or mortality and have a normal life expectancy in a patient possessing it. However, its timely diagnosis is crucial and documentation of situs inversus in an individual is important to correctly interpret any future symptoms and avoid any inadvertent clinical or surgical complications. An preceded diagnosis of this condition will form a baseline reference for future intervention if any.

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