

CASE REPORT

SITUS INVERSUS TOTALIS WITH SINGLE VENTRICLE

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ABSTRACT: Situs Inversus Totalis is a rare congenital anomaly characterized by transposition of thoracic and abdominal viscera. Its anaesthetic plans & implications have not been thoroughly discussed. It poses a magnitude of problems to the anaesthesiologist in terms of airway management, endotracheal intubation, lung separation during thoracic surgeries, providing ventilation, placement of ECG leads and electrodes during defibrillation. So we report the problems encountered in handling the case of 4 days old baby diagnosed as Situs Inversus Totalis with cardiac abnormalities scheduled for duodenal atresia corrective surgery. The surgery was abandoned due to single ventricle which is not compatible with life.

KEYWORDS: Situs Inversus Totalis, Duodenal atresia, single ventricle, TGA, PDA.

INTRODUCTION: Situs Inversus Totalis is rare congenital anomaly characterized by transposition of thoracic and abdominal viscera, including dextrocardia. Incidence is about 1 in 10,000 of normal population¹ but its association with duodenal atresia is rare and cardiac anomaly is extremely rare. A review of English medical literature reveals only 25 cases of this association had been so far reported. There is a 270 degree clockwise rotation instead of normal 270 degree anticlockwise rotation during embryogenesis. The association of SIT with cardiac, splenic anomalies and syndromes such as Kartagners pose a challenge to anesthesiologist. A rare condition with such complexity has been described only by few journals and its anaesthetic plans & implications have not been thoroughly discussed.

CASE REPORT: A 4 days old full term male baby was born via naturalis weighing 2.1 kg, baby did not cry immediately after birth, it cried after initial resuscitation of 30 secs. Baby was not fed in view of duodenal atresia which was detected in third trimester. Mother who had 2nd degree consanguineous marriage had regular ANC visits and adequately immunized, gave history of PROM for 2 days.

Examination revealed:

Baby was afebrile,

HR-138bpm, RR-44 cpm, Capillary refilling time<3 secs

SPO₂-86% with 5litres O₂ on head box

Head to toe examination –normal.

Spine-normal

CVS: Absence of heart sounds on left side, S1S2 heard on right side. Systolic murmur present.

RS: Normal vesicular breath sounds heard on all lung fields.

PA: Spleen on right side and liver on left side'

CNS: Normal.

INVESTIGATIONS: Hb-15.7%, Hct-43.8, TLC-11000 cells per cubic mm, Platelet count-1.7 lakhs per cubic mm.

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Coagulation profile: Normal.

Total bilirubin: 10.6 mg/dl.

Renal function test and Serum electrolytes – within normal limits.

CHEST RADIOGRAPHY: Revealed dextrocardia with Situs Inversus.

Infantogram showed double-bubble sign with gastric shadow in the right.

ECG displayed inverted P wave in lead I and aVL and positive in avR which suggests Situs Inversus.

USG: Liver in left hypochondrium, Spleen in right hypochondrium.

Superior mesenteric artery and superior mesenteric vein-reversed in position.

Reversal of Aorta and Inferior venacava position.

The above features are highly suggestive of Situs Inversus Totalis with dextrocardia.

ECHO

Single ventricle

IAS shows primum ASD

AV normal, PV atretic

LVEF 50%

TGV

PDA

L to R shunt

Due to single ventricle and complex cardiac anomalies surgery was abandoned. Although case was abandoned it deserves to be discussed on grounds of rarity of its incidence and grave anesthetic complications associated with its surgical management.

DISCUSSION: Situs Inversus Totalis is associated with lower rate of congenital heart disease than incomplete Situs Inversus. Heart is the most common affected intrathoracic organ in insitus anomaly, and in most instances cardiac symptoms are usually the first ones that lead to the detection of this anomaly. The associated cardiac anomalies are TGA, VSD, ASD, absent coronary sinus, Single ventricle and pulmonary atresia. There is no sex or racial difference, but genetic predisposition and familial occurrence favours multiple inheritances.

USG is useful in bedside screening. CT scan is best for thoracic and abdominal organ anomaly detection.

Anaesthetic implications:

Preoperative evaluation:

- A) Absent heart sounds on left side during auscultation should arise a high degree of suspicion
- B) Association with other co-morbid diseases like Kartageners and cardiac anomalies to be looked for
- C) Investigations like ECG, CXR, USG, INFANTOGRAM and ECHO will reveal same findings as shown above.

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Preparation:

- A) Optimization of pulmonary status increase of primary mucociliary dyskinesia (like kartageners) with postural drainage, physiotherapy, antibiotics, bronchodilators and incentive spirometry.
- B) In neuraxial blockade associated spina bifida, scoliosis, and meningomyelocele should be looked for.
- C) In thoracic surgery, insertion of double lumen tube can be difficult and main stem endobronchial intubation can occur on left side.
- D) Central line insertion should be done on left side with USG guidance,
- E) For monitoring, ECG electrodes should be applied in opposite direction.
- F) During resuscitation defibrillation pads applied in opposite direction.
- G) For LSCS, uterine tilt to be given on the right side.
- H) Due to deficiency of pseudocholine esterase, rarely prolonged paralysis may be seen with use of succinyl choline.
- I) Premedication that depresses ventilation or ciliary activity should be avoided especially in patients with Kartageners.
- J) Intracardiac catheterization can be hazardous as it may provoke arrhythmias.

CONCLUSION:

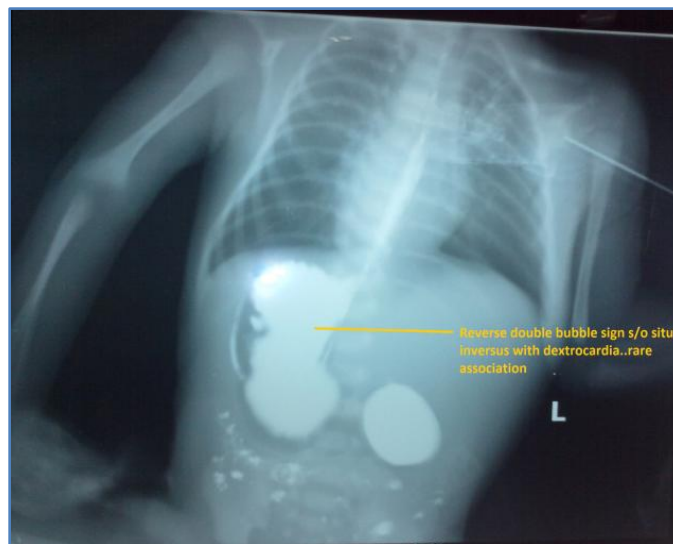
- Patient with Situs Inversus Totalis is asymptomatic and gave a normal life expectancy but its association with complex cardiac anomalies imposes a great challenge to anaesthesiologist.
- Detection and documentation of Situs Inversus Totalis is important to prevent inadvertent complications and confusions.
- So a thorough preoperative evaluation can minimize the difficulties and varies potential challenges associated with anaesthetic management.

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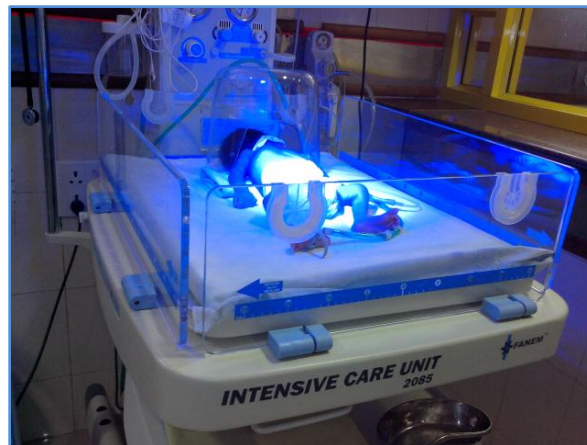


Chest x ray showing dextrocardia, reverse double bubble sign

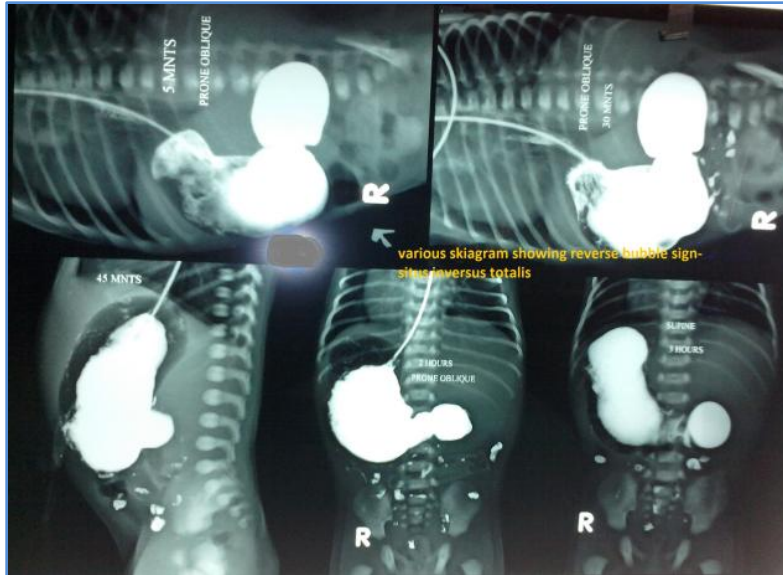
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Chest X ray showing dextrocardia



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Infantogram showing duodenal atresia in various views



Infantogram

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Saturation monitoring

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