

## Anaesthetic implications in situs inversus totalis: A case report

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### Abstract

Situs inversus totalis (SIT) is a rare congenital condition which is characterised by the reversal of orientation of abdominal and thoracic organs where heart is on the right side of the thoracic cavity and liver on the left side, whereas stomach and spleen are on the right side in the abdomen. The reported prevalence of this anomaly is one in 5,000-20,000 live births. This case reports the anaesthetic management of situs inversus totalis in a 38-year-old male patient, with a history of poorly differentiated adenocarcinoma of the colon, who underwent laparoscopic intervention converted to open nodular excision with incisional hernia repair. The report analyses the anaesthetic implications and challenges associated with situs inversus totalis during surgery, including pre-operative evaluation, monitoring techniques and potential complications.

**Keywords:** Situs inversus totalis, Anaesthesia, Kartagener's syndrome, Dextrocardia.

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### Introduction

The anatomical variations associated with situs inversus totalis make the anaesthetic management challenging during surgery and resuscitation. Moreover, patients with situs inversus totalis may also have other associated disorders, for example, primary ciliary dyskinesia (Kartagener syndrome), cardiac abnormalities such as tetralogy of fallot, septal defects, transposition of great vessels and extra-cardiac abnormalities like gastrointestinal problems, liver and splenic dysfunctions, problems with the respiratory system, and midline defects which can pose challenges to the anaesthesiologists. Therefore, to ensure safe anaesthesia delivery and improve patient outcome, careful pre-operative evaluation and planning, and close intra-operative and post-operative monitoring are essential components of a safe healthcare delivery in patients with situs inversus totalis.

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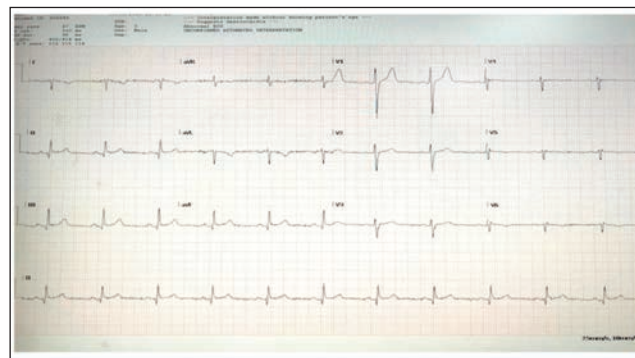
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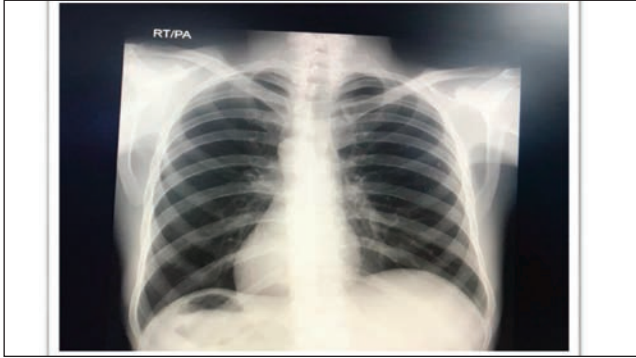
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### Case Report

A 38-year-old male who was a diagnosed case of colon cancer presented for his day care laparoscopic lymph node excision with incisional hernia repair surgery. Before undergoing surgery, the patient was seen in the pre-operative anaesthesia assessment clinic on March 6, 2023, at Shaukat Khanum Memorial Cancer Hospital, Peshawar, where a focussed history was taken and physical examination performed. He denied any other medical illness apart from his congenital condition of situs inversus totalis. There was no history of drug allergies and addiction. His exercise tolerance was good as he could climb one flight of stairs without any difficulty and there was no history of orthopnoea or ankle swelling. In addition, there was no history of repeated chest infections or sinusitis. His previous history of general anaesthesia was uneventful for right hemicolectomy. On airway assessment his mouth opening was more than three finger breadths, Mallampati Class II - full visualization of the faucial pillars and soft palate with no limitation of neck extension, and he was labelled as American Society of Anaesthesiologists Physical Status Classification system (ASA) 2.<sup>1</sup> His cardiovascular examination was consistent with dextrocardia and no murmurs were auscultated on his heart auscultation. His chest was clear on auscultation with no added sounds. His ECG with conventional lead position showed right-axis deviation, positive QRS complexes in aVR and abnormal R-wave progression in the chest leads, and his chest X-ray findings were consistent with dextrocardia and there was no evidence of bronchiectatic changes (Figure 1 and 2). His baseline investigations were normal and no abnormality was noticed in his liver function tests. The anaesthetic plan



**Figure-1:** Standard 12 lead ECG of the patient done pre-operatively showing Right axis deviation, positive QRS complexes (with upright P and T waves) in aVR, Lead I: inversion of all complexes and absent R-wave progression in the chest leads.



**Figure-2:** Chest X-ray of the patient showing apex of the heart towards the right side, Dextrocardia with normal lung fields without any suggestive findings of bronchiectasis. Below right hemi-diaphragm is the normal air shadow in gastric fundus, suggestive of dextrogastria; Situs inversus totalis.

included general anaesthesia with standard monitoring. An echocardiography was requested as it was unavailable at the time of his anaesthesia pre-operative assessment.

On the day of surgery, his echocardiography results were available which showed no cardiac defects and his ejection fraction was more than 60%. A written and informed consent was taken and NPO status confirmed. He was then moved to the operation theatre and "Sign-In" was performed before induction of anaesthesia as per the World Health Organization Surgical Safety Checklist.<sup>2</sup> Pre-induction vitals of the patient were: blood pressure=100/65 mmHg, Pulse=74/min, Temp=36.5 degree Celsius, Respiratory rate=18/min, SpO<sub>2</sub>= 100% on room air with normal sinus rhythm on electrocardiographic trace. ECG leads were applied in the reverse direction. Anaesthetic induction was performed using Fentanyl 100 mcg, Propofol 230 mg, Atracurium 30 mg and the patient was intubated with tracheal tube of size 7.5 mm ID, fixed at 20 cm at lips. On direct laryngoscopy, a full view of the glottis was obtained and he was labelled as Cormack-Lehane grade 1. Anaesthesia was maintained with oxygen, air, and Isoflurane. Intra-operatively, the patient was continuously monitored for blood pressure, temperature, pulse oximetry, end-tidal capnography and electrocardiography, and record was maintained every five minutes. Fibre optic bronchoscopy was performed for academic purposes which showed mirror images of the airways. The patient was positioned supine for surgery and the procedure took three hours with no complications. After reversal of the neuromuscular blockade, the patient was successfully extubated and then was observed in the post-anaesthesia care unit for one hour without any complications. Upon fulfilment of the Post-Anaesthesia (PACU) discharge criteria, as assessed by the Modified Aldrete Score, with a score of 9, he was relocated to the designated ward.<sup>3</sup> The rest of his post-operative course was uneventful and he was discharged home on post-operative day four.

## Discussion

Situs inversus totalis (SIT) is a rare congenital abnormality characterised by a mirror-image transposition of both the abdominal and thoracic organs. This uncommon congenital anomaly is characterised by a transposition of the abdominal and thoracic organs in a mirror image.<sup>1</sup> Its aetiology is still unclear and multiple underlying inheritance patterns are likely responsible as evident by its genetic predisposition and familial occurrence. An uneventful case of SIT has been presented here but this condition can pose challenges to the anaesthesiologists and emergency physicians during resuscitation and elective surgical procedures.

A chest X-ray or ultrasound imaging, which are usually the first choices for diagnostic imaging, are used to detect situs inversus totalis. Typical findings noted in SIT are dextrocardia, liver on left hypochondrium, and a right-sided spleen. Further details can be assessed by the use of advanced imaging techniques like computer tomography (CT) or magnetic resonance imaging (MRI). Prenatal MRI of the foetus can be used for research purposes.<sup>2</sup>

Approximately 50 percent of the patients with Kartagener syndrome have situs inversus totalis.<sup>3</sup> This syndrome is the triad of situs inversus, bronchiectasis, and sinusitis. Special attention should be paid to lung function during the peri-operative period, especially in major surgeries involving upper abdomen and thoracic cavity. These patients definitely benefit from physiotherapy and incentive spirometry. Thoracic epidural is a good choice for pain relief in surgery of the chest and upper abdomen, particularly in cases with a long history of chest infections.<sup>4</sup> A smaller sized nasal endotracheal tube should be preferred due to long history of sinusitis resulting in narrow nasal cavity. There are chances of left-sided endobronchial tube placement because the left bronchus is more in line with the trachea resulting in absent right-sided lung sounds. Single lung ventilation can be safely carried out using standard double lumen tubes and bronchial blockers. Single lumen tube can be used in children for one lung ventilation, another option is bronchial blocker. A flexible fibre-optic bronchoscope is always used to confirm the double lumen tube position at this institution and high risk patients are transferred to the intensive care unit for observation and prevention of complications.

Some of the examples of cardiac anomalies seen in situs inversus totalis include pulmonary valve stenosis, transposition of the major vessels, pulmonary venous stenosis, and a right ventricle with two outlets, atrial and ventricular septal defects. A detailed pre-operative history and physical examination should be followed by the

relevant investigations including echocardiography, and in some cases an input from a cardiologist is required.<sup>5</sup> In case of cardiac arrest and arrhythmias, a modified approach will be required when placing the defibrillator pads, which is self-explanatory.<sup>6</sup> Nitrous oxide should be avoided in patients with cardiac septal defects. Finally, because of the altered anatomy, trans-oesophageal echocardiography could be challenging.

In these patients, a left-sided central venous line placement using ultrasound guidance is preferable because of a straighter course. But in those with right superior vena cava and total pulmonary venous anomaly, right internal jugular vein is preferred. In situations where ultrasound is unavailable, right-sided subclavian central cannulation can be safely performed. Following right subclavian vein cannulation, a post-CVC chest X-ray should show a CVC moving towards the morphological right atrium through the right brachiocephalic vein, crossing the midline, and entering the left superior vena cava.<sup>7</sup> In other words, a correctly placed central venous line crosses the midline in patients with SIT when subclavian approach is adopted.

Patients should be carefully evaluated for spinal deformities during the pre-anaesthesia assessment if the surgery is planned under neuraxial anaesthesia. Rarely seen spinal deformities include scoliosis, split cord, spina bifida, and meningomyelocele.<sup>8,9</sup>

Succinylcholine is not an ideal muscle relaxant to use as it can lead to prolonged apnoea due to the presence of atypical cholinesterase enzyme in some cases.<sup>9</sup>

## Conclusion

In conclusion, situs inversus totalis presents a complex clinical scenario, necessitating meticulous planning and tailored approaches in anaesthesia and surgery. Understanding its unique anatomical and physiological characteristics is paramount for successful management, ensuring patient safety and optimal outcomes amidst the challenges posed by this rare congenital anomaly.

**Patient consent for publication:** Consent was obtained directly from the patient for publishing his case report.

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**Conflict of Interest:** None.

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### Author Contribution:

FM: Concept, correction and motivation.

FIQ: Introduction and discussion writing.

QTA: Reference management.