

Situs Inversus- Not Just a Mirror Image

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INTRODUCTION

Situs inversus totalis, a rare condition presents with multitude of problems due to multiple structural and functional abnormalities that coexist with it like cardiac, spine and airway malformations. We are reporting a patient who had situs inversus totalis and was operated for upper limb fracture. The present case report emphasises on the challenges during anaesthetic management of a patient with situs inversus.

Situs inversus (also called situs transversus or oppositus) is a congenital condition in which the major thoracic and abdominal visceral organs are reversed or mirrored from their normal positions.¹ It is a rare disease with a predicted incidence of one in 10,000 among the general population.² We are reporting a case of situs inversus totalis who was operated for upper limb fracture for which we gave a nerve block, an anaesthesia modality which has not been published in a situs inversus patient earlier. These patients pose a challenge to anaesthesiologists because of the associated conditions like Kartagener's syndrome, cardiac anomalies, spleen malformations and other such clinical entities, all of which have specific anaesthetic implications, and not just for the anatomical mirroring.

PRESENTATION OF CASE

A 28-year-old lady presented to our orthopaedic out-patient department (OPD) with chief complaints of pain over the right elbow after history of trauma 10 days ago. The radiograph of the right elbow showed a capitulum fracture. She also had complaints of intermittent productive cough for the last two months for which chest radiograph was done, which showed clear lung fields, although the heart and gastric shadow were both on the right side (dextrocardia) as shown in Figure 1. These findings led us to suspect situs inversus, which was further confirmed on ultrasonography of the abdomen, which showed liver and gall bladder in the left hypochondrium and spleen in the right hypochondrium. Her electrocardiogram (ECG) showed p wave inversion in Leads I and aVL and T wave inversion in lateral leads, the mirror image of any normal ECG as shown in Figure 2, suggestive of dextrocardia. Echocardiography was carried out and revealed normal cardiac parameters and anatomy with an ejection fraction of 60% along with confirming the finding of dextrocardia. She was born as a second of twin children and her brother did not have any features suggestive of situs inversus.

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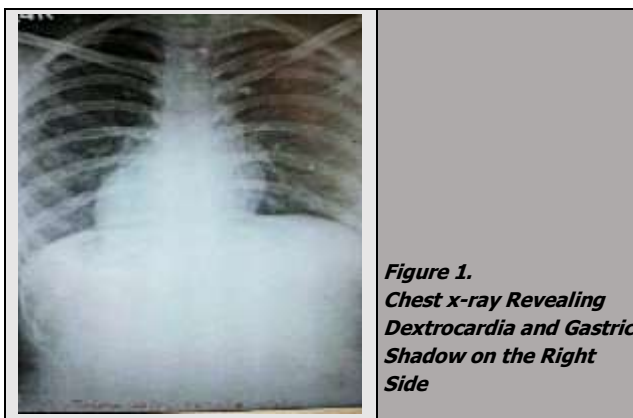


Figure 1.
Chest x-ray Revealing
Dextrocardia and Gastric
Shadow on the Right
Side

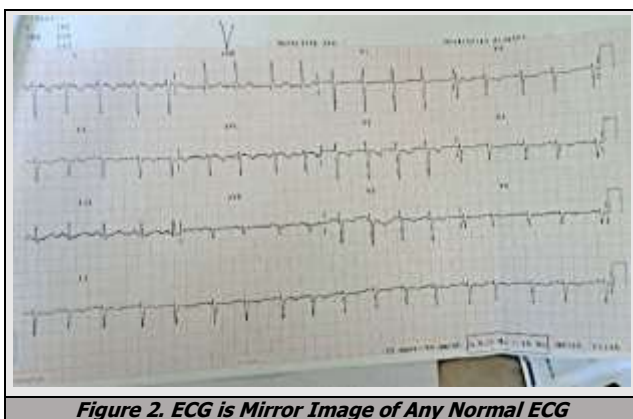


Figure 2. ECG is Mirror Image of Any Normal ECG

Pre-anaesthetic evaluation revealed a normal airway examination, a pulse rate of 92/min, blood pressure of 106/70 mmHg and normal profile of laboratory investigations. On auscultation, heart sounds were heard more on the right side, apex beat was palpated on the right side of the chest and there were no added lung sounds. She was put on steam inhalation twice daily, deep breathing exercises, incentive spirometry and good respiratory hygiene practices till the morning of surgery. A decision to operate the patient was taken and she was posted for open reduction and internal fixation with Herbert screw fixation of the right elbow. In the operation theatre, 18G intra venous cannula was secured over the left dorsum of hand and all routine monitors (ECG, Non-invasive blood pressure, SpO₂) were attached as per the ASA guidelines³ and baseline parameters were noted.

ECG electrodes were placed in reverse position of what we do in normal patients. She was given ultrasound guided nerve stimulator assisted right sided brachial plexus supraclavicular block with 15 ml of 0.25% bupivacaine and minimal sedation (injection midazolam 1 mg IV). Once adequate motor and sensory blockade of the right arm was achieved, surgery was done. The surgery was uneventful with minimal blood loss and patient was shifted to post anaesthesia care unit. The post-operative period was uneventful, and the patient was discharged in a satisfactory condition on the fourth postoperative day.

CLINICAL DIAGNOSIS

Fracture right radius in a patient with situs inversus.

DISCUSSION OF MANAGEMENT

Situs inversus totalis (SIT) is a rare condition, the etiologic factors for which are still not completely understood. During the embryological development, a normal 270 degree anti-clockwise of the developing thoraco-abdominal organs occurs. If there is a 270 degree clockwise rotation, it results in mirror image positioning of the thoraco-abdominal organs.⁴ There is no established gender or racial difference in its incidence, but genetic predisposition and familial occurrence point towards multiple inheritance patterns.^{5,6} There are some studies done which show no correlation to the occurrence of SIT in twins.⁷

SIT may either occur independently or in association with other syndromes such as Kartagener's syndrome, mucociliary dysfunctions, cardiac anomalies, spleen malformations, airway anomalies, spine deformities etc. The challenge for the anaesthesiologist increases if it occurs with other syndromes like airway difficulties and pulmonary infections that can affect conduct of anaesthesia. Kartagener's syndrome, which manifests as a triad of dextrocardia, chronic sinusitis and bronchiectasis, is frequently associated with SIT.⁸ Situs inversus in Kartagener's syndrome is invariably has mucociliary dysfunction and is basically a immotile ciliary syndrome - a rare, ciliopathic, genetic disorder.⁹ It is present in 25% of the patients with SIT with an increased probability of developing respiratory complications. Impaired mucociliary clearance leads to recurrent lung infections, bronchiectasis, chronic sinusitis, and otitis media. Ciliary dysfunction also causes reduced fertility in women and sterility in men from reduced motility of spermatozoa.¹⁰

Therefore, moist and filtered mixture of gases should be administered during mechanical ventilation in ICUs and during general anaesthesia. The role of bronchodilators, chest physiotherapy, postural drainage, antibiotics and incentive spirometry cannot be underestimated and is mandatory in optimizing the pulmonary status before any surgical procedure in these patients. All these conditions require adequate treatment and optimisation perioperatively. Where possible, local or regional anaesthesia should be preferred to general anaesthesia to avoid the aforementioned complications.¹¹ The syndrome of situs inversus is also associated with several cardiac anomalies like atrial septal defects, ventricular septal defects, transposition of great vessels, absent coronary sinus, double-outlet right ventricle, total pulmonary anomalous venous defect and pulmonary valve stenosis either singly or in combination, all of which have specific anaesthesia concerns.¹⁰ Hence a routine cardiac screening echocardiography is suggested for all patients of situs inversus in the pre anaesthetic evaluation and specific optimisation should be done after specialty consult when present.

Many spinal deformities like meningomyelocele, scoliosis, split cord, spina bifida, etc. have also been described in these set of patients, hence as perioperative physicians, we should evaluate the patients suspected of

having situs inversus carefully in case central neuraxial anaesthesia is planned and do a thorough neurological examination as well as palpation of spine.¹²

Although the airway in our patient was without any abnormality or difficulty, patients with situs inversus have been known to have associated difficult airways and abnormalities like congenital aglossia,¹³ congenital hypoglossia,¹⁴ craniodiaphyseal dysplasia¹⁵ and Goldenhar syndrome.¹⁶

The ECG electrodes have to be applied in the opposite direction as the changed surface electric polarity may give a false picture of perioperative ischaemia. Left internal jugular vein cannulation should be avoided to prevent thoracic duct injury and ensure direct access to the right atrium in case of inversion of great vessels. The anatomy of the bronchi should be considered selecting a double-lumen tube for surgeries which require lung isolation.

One case of prolonged paralysis after administration of succinylcholine has been reported in a patient with situs inversus totalis hence should be avoided.¹⁷ In case of cardiac arrhythmias and cardiac arrest, care should be taken while applying direct current with defibrillator pads on the right side. A successful resuscitation of such patients requires a thorough knowledge and skills on the part of the attending anaesthesiologist and intensivists. For the surgeons, the reversed anatomy should be kept in mind, especially in laparoscopic surgeries where port placement and handedness can be concerns.¹⁸

Various case reports of general and central neuraxial anaesthesia being performed on situs inversus patients have been published, but in our literature search, we could not find any published data on nerve blocks given in a patient with situs inversus hence this is a pilot article describing the same.

FINAL DIAGNOSIS

The precise diagnosis of situs inversus and a thorough preoperative evaluation can minimize, to a large extent, the difficulties and the various potential challenges associated with its anaesthetic management. Keeping in mind all these considerations and implications, it becomes easier and safer to successfully manage the patients of situs inversus totalis in the operation theatre and intensive care units.

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