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The challenges in successful anaesthetic management in a case of situs inversus totalis

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Abstract
Situs inversus totalis is an uncommon congenital abnormality in which the orientation of all asymmetric organs of the body are mirror image of normal morphology. In such patients not only is the diagnosis of abdominal pathology difficult due to transposition of abdomino thoracic viscera, they also pose several anaesthetic challenges during the surgical procedure. Here we successfully managed a case posted for whipples surgery who was previously diagnosed with situs inversus totalis.

Keywords: Situs inversus, dextrocardia, whipples surgery

Introduction
Situs inversus is a rare congenital anomaly with autosomal recessive pattern of inheritance in which all the major visceral organs are mirrored from their normal positions [1, 2]. The predicted incidence is 1:5,000 to 1:20, 000 among the general population [3]. At the embryological development, a 270-degree normal anti- clockwise rotation is substituted by an abnormal 270 degree clockwise of the developing thoraco-abdominal organs resulting in mirror image positioning of the abdominal and thoracic viscera [4]. Such patients are asymptomatic and have a normal life expectancy. In this case report, we aim to explain the anaesthesia management in a patient with situs inversus totalis was posted for an open whipples surgery.

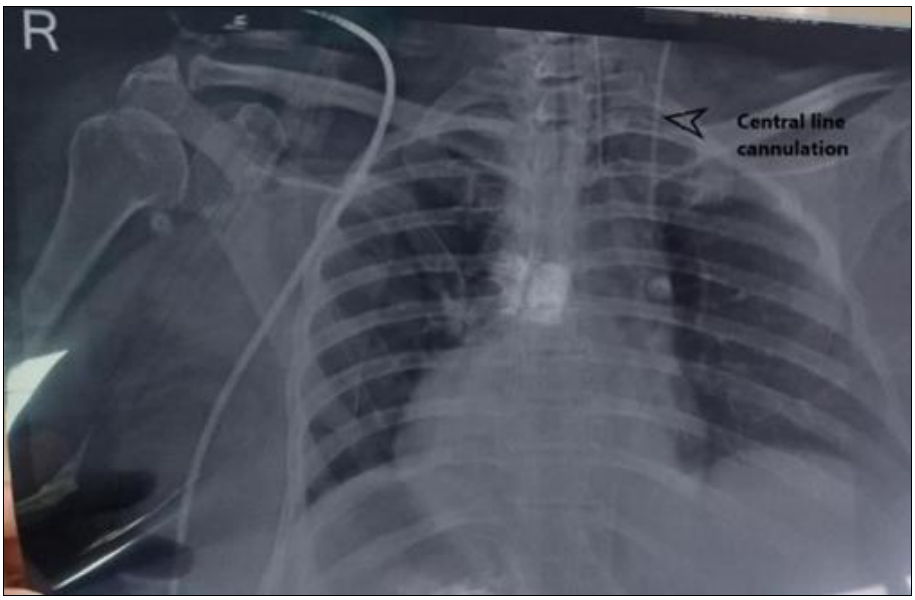

Fig 1: Chest radiograph showing cannulation of left IJV

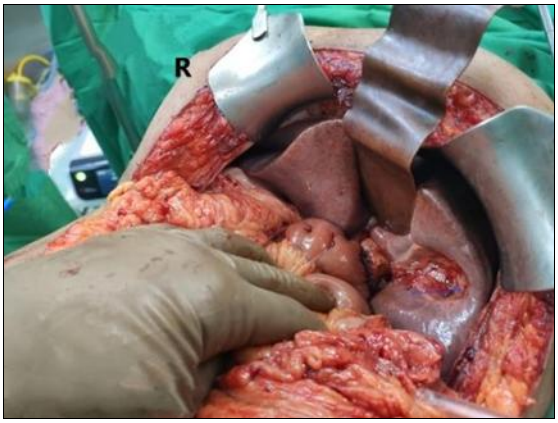

Fig 2: Transposition of thoraco abdominal organs

Case report

A 52-year-old female presented to our surgical outpatient department with history of jaundice since 3 weeks. Patient also complained of significant weight loss (10 kgs in 2 months) and generalised abdominal pain. Patient's diagnosis was confirmed with an upper GI scopy with biopsy.The Patient was advised whipples surgery (pancreatico duodenectomy) and subsequently evaluated with pre operative blood and radiological investigations.

It was only after the radiological examination that the diagnosis of situs inversus totalis was confirmed. Her chest x ray depicted dextrocardia. Computerised tomography (CT) revealed transposition of all major abdominal organs. Echocardiography was carried out which showed normal cardiac functions with ejection fraction of 60%.

Pre anaesthetic evaluation revealed the patient to be hypertensive with ongoing treatment of T.Amlodipine 2.5mg OD. On physical examination she had an adequate mouth opening with Mallampatti class I, pulse rate of 78/min, blood pressure of 140/78 mmHg and normal laboratory investigations; however, liver function tests were deranged. Examination of the cardiovascular system revealed location of the apex beat in the right 5th intercostal space 1.5 cms medial to the mid clavicular line. On auscultation heart sounds were heard on the right side of the chest. At present, the patient had no other systemic complaint Decision to operate the patient was taken and she was posted for open whipples surgery. General anaesthesia with epidural analgesia was decided as the mode of anaesthesia.

She was advised to continue with the antihypertensive drug on the morning of surgery. In the operation theatre, a wide bore intravenous (IV) access (18G) was secured. She was preloaded with ringer lactate solution. Non-invasive arterial blood pressure (NIBP), peripheral oxygen saturation (SpO2) and electrocardiography (ECG) monitoring was performed. ECG electrodes were placed paying heed to dextrocardia in the patient. After vital parameters were recorded patient was given sitting position for placement of epidural catheter for post operative analgesia. Epidural catheter was placed in the T10-T11 intervertebral space using 18 G tuohy's needle. After confirmation of the epidural catheter placement, she was made to lie in supine position for induction of anaesthesia.

Patient was pre medicated with Inj. Midazolam 0.05mg/kg and Inj. Fentanyl 2mcg/kg. Induction of anaesthesia was achieved with Inj. propofol 2mg/kg, and Inj. Vecuronium 0.1mg/kg. After adequate relaxation, patient was intubated with 7.5 mm endotracheal tube. Maintenance of anaesthesia was carried out with Inj. vecuronium and low concentration of isoflurane in oxygen and air mixture. Post induction an 18G triple lumen central line was inserted in the left IJV (figure 1) for monitoring of central venous pressure(CVP) and administration of fluids and blood components. The surgical procedure was smooth and uneventful, and lasted for 8 hours. Intra operative fluids included 6 units of crystalloids (4 units RL, 1 unit DNS and 1 unit of 0.9%NS) and 2 units of Packed cell blood volume (PCV). At the end of surgery, patient was shifted to Intensive care unit for post operative ventilation considering the prolonged duration of surgery. Thereafter, extubation was performed only after establishing the return of all protective airway reflexes and establishment of regular breathing pattern and normal haemodynamic vitals on the second postoperative day. Patient had an ICU stay period of 5 days after which she was shifted to the surgical ward. Epidural infusion of 0.0625% Bupivacaine was continued for post operative pain relief till the 3rd post operative day. The recovery period and hospital stay were uneventful and the patient was discharged in a satisfactory condition on the 15th post-operative day and was called for follow-up after 2 weeks.

Discussion

Sius inversus is a rare condition where the etiology is not well understood. There is no difference in distribution among gender or race. Situs inversus totalis usually remains undiagnosed, unless it is diagnosed incidentally while investigation of some other ailments as happened in our case. There is transposition of major thoracic organs and all the visceral organs of the abdomen to the side opposite to the normal position in the body. This poses a great challenge to the operating surgeon who is planning a pancreatico duodenectomy with anastomosis where in all the organs are in opposite position to normal (Figure 2).

Pancreatic cancers are among the rare cancers we see in our surgical outpatient department. The most common presenting symptom is jaundice followed by weight loss both of which were present in our patient. After confirmation of diagnosis further evaluation including CT scans, chest and abdominal radiographs incidentally confirmed the presence of situs inversus totalis in our patient. The diagnostic parameters of radiographs include the presence of dextrocardia, stomach bubble under the right dome of diaphragm, liver shadow on the left side.

The challenging aspects for anaesthesiologists in such patients are:

1. The electrocardiogram leads should be placed in reverse orientation for accurate interpretation [5].
2. The association of situs inversus with other syndromes like Kartagener's syndrome [5] is invariably associated with mucociliary dysfunction and can have considerable implications during induction of anaesthesia and intubation. Primary ciliary dyskinesia is present in 25% of the patients with situs inversus totalis with an increased probability of developing respiratory complications. [5] The role of bronchodilators, chest physiotherapy, postural drainage, antibiotics and incentive spirometry is mandatory in optimizing the pulmonary status before any surgical procedure [5]
3. A case of prolonged paralysis after administration of succinylcholine has been reported earlier in a patient with situs inversus totalis [6]. However, we did not use

succinylcholine since there was no anticipation of difficult airway.
4. Spinal deformities like split cord, spina bifida, meningo myelocele, scoliosis have been described in the literature, and one has to evaluate the patient carefully if any surgery is planned under neuraxial anaesthesia [7].
5. The syndrome is associated with numerous cardiac anomalies such as atrial septal defects, ventricular septal defects, transposition of great vessels, absent coronary sinus, and pulmonary valve stenosis either singly or in combination. [8] No such anomaly was detected in our patient.
6. In case of cardiac arrhythmias and cardiac arrest, great care has to be taken while applying direct current with defibrillator pads on the right side [5]. A successful resuscitation of such patients requires an in-depth knowledge and prolific skills on the part of the attending anaesthesiologist.
7. Mainstream intubation can occur on the left side and should be kept in mind while intubating the trachea [9].
8. In case of inversion of great vessels, preference should be given to the left internal jugular vein for cannulation to avoid thoracic duct and ensure direct access to the right atrium [9] which was performed in our patient.

From the above-mentioned implications in a case of situs inversus totalis, general anaesthesia can be administered safely provided that there is no associated cardiac, neurologic or any other associated syndromes.

Conclusion
Patients with situs inversus are asymptomatic and have a normal life expectancy. The diagnosis of situs inversus pre operatively can minimize the various dilemmas and challenges associated with anaesthetic management. With necessary precautions and meticulous planning such patients can be managed successfully intra operatively and post operatively in intensive care units irrespective of the duration and nature of the surgery.

Conflict of Interest
Not available.

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