



Case Report of a Patient with Situs Inversus Totalis with Ciliary Dysfunction for Laparoscopic Cholecystectomy: Considerations for Safe Peri-Operative Anaesthetic Management

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Abstract

Patients with rare genetic disorders are associated with diverse congenital anomalies and present special challenges to the anaesthesiologist. We report the successful management of a 20 year old female with situs inversus totalis and suspected kartagener syndrome posted for laparoscopic cholecystectomy surgery.

Cases of situs inversus with or without ciliary dysfunctions are known to have several structural and functional abnormalities especially those as cardiac, spine and airway malformations, presenting a challenge to an inadequately equipped and mis-informed anaesthesiologist. This calls for an individualisation of the peri-operative concerns of such patients based on associated anomalies along with appropriate monitoring.

In this report we highlight the evaluations done and precautions taken during the management of such a patient who underwent successful management at our centre. It is recommended that such patients with multiple congenital anomalies be given a thorough and cautious assessment for a safe intra-operative and postoperative course.

Keywords: *Peri-operative management. Kartagener syndrome. Situs inversus totalis*

Key Messages: *Cases of situs inversus totalis with ciliary dysfunction reveal a very unique set of problems due to multiple functional and structural abnormalities that might coexist with it. A review of nomenclatures of the abnormalities of left-right asymmetry with specific organ system malformations reported in these patients is presented here and their implications for anaesthetic care are discussed.*

Introduction

Situs inversus is a disorder distinguished by mirror image symmetrical orientation of the abdomino-thoracic viscera relative to the midline. Transmitted by autosomal recessive genes, if present with a right sided heart or dextrocardia, it is known as situs inversus totalis.^{1,2} Situs inversus is one of the constituents of kartagener syndrome or immotile ciliary syndrome, a rare, ciliopathic, genetic disorder.³ The surgical management in such cases is highlighted in literature but information on

peri-operative anaesthetic difficulties is limited. We put forward the anaesthetic considerations and precautions taken during the care of such a patient in our report.

Case History

A 20-year-old woman was presented to the surgical OPD with chief complaints of acute epigastric pain, occasional vomiting, and dyspepsia. She also gave a history of recurrent rhinorrhoea since childhood. Per-abdominal examination revealed

tenderness in left hypochondrium. Initially, she was diagnosed to be a case of acute peptic ulcer disease but an abdominal ultrasound pointed to situs inversus with acute cholecystitis. Computerized Tomography studies confirmed the diagnosis. On pre anaesthetic evaluation, her pulse rate was 80/minute and blood pressure 120/80 mmHg. On examination of cardiovascular system, apex beat was located at fifth intercostal space in the right side, 1.5 cm medial to the mid clavicular line, and on auscultation heart sounds were heard on the same side of the chest. Respiratory system examination revealed normal air-filled lungs, vesicular sounds heard over most of the lung fields and broncho-vesicular sounds between the first and second interspaces on the anterior chest. The rest of the examination was within normal limits. Plain radiograph of the chest revealed clear lung fields with dextrocardia and right sided fundal gas shadow [Figure 1]. The patient was asked to undergo further cardiac evaluation with 12 lead ECG and 2-D echocardiography. ECG showed significant right axis deviation with negative p wave in lead 1 and aVL [Figure 2a, 2b]. ECG with reverse lead placement showing no abnormality. 2D-Echocardiography pointed to dextrocardia and normal cardiac anatomy.

In the operating room, routine monitors (pulse oximeter, temperature monitoring probe, five lead electrocardiogram and non invasive blood pressure) were attached. Left-sided ECG leads were reversed and attached on the right side and vice versa. Induction of anaesthesia was done with intravenous fentanyl (2micrograms/kg) and propofol (2mg/kg), and lungs were ventilated with oxygen with Fio₂ 0.8. After achieving neuromuscular blockade with atracurium (0.5 mg/kg), trachea was intubated using 7.5mm cuffed endotracheal tube. Capnography monitoring was also initiated. Anaesthesia was maintained with sevoflurane in oxygen and nitrous oxide (30:70) (MAC 1) using close circuit along with boluses of atracurium (0.03mg/kg) as required. The patient was kept warm using warm fluids and hydration maintained using normal saline.

The surgeon found the liver on left side with right lobe on the left and an inflamed gall bladder on the same side.

In the middle of surgery, there was one episode of sudden desaturation with the SPO₂ reading below 70%. On auscultation, breath sound was slightly decreased on the right lung fields. Endotracheal tube suctioning was done. The tube was fixed at a higher marking and repeat suction given. We suspected either endobronchial displacement of the tube or a mucus plug to be obstructing the endotracheal tube. Post this event ventilation became better. Surgery lasted 1.5 hours, without any significant blood loss. Reversal of neuromuscular blockade was performed with neostigmine/glycopyrrolate (0.05/0.02 mg/kg). Extubation was uneventful and smooth. The patient was transferred to the post-anaesthesia care unit. Postoperative analgesia was maintained with injection paracetamol one gram intravenously. The postoperative course was uneventful.

Discussion

Marco Severino first identified dextrocardia in 1643. More than a hundred years later, Matthew Baillie recognised the total mirror-image reversal of the abdominal and thoracic organs in situs inversus.

Situs inversus is an extremely rare disorder present in 0.01% of the population. Our literature search revealed very little information on the pre-operative evaluation and anaesthesia consideration for this subset of patients. We present here a brief review of the anomalies of right-left asymmetry that might help anaesthesiologists in better evaluation of these patients before proceeding with the case.

The normal left-right anatomical orientation of viscera is called situs solitus, and mirror image reversal of all asymmetrical structures is called situs inversus. The morphologic right atrium is to the left, and the left atrium is on the right in this condition. The normal pulmonary anatomy is reversed as well so that the right lung has two lobes and the left lung has three lobes.

The intermediate phenotypes between these two extremes is called situs ambiguus or heterotaxia.⁴ Situs inversus can be stratified further into situs inversus with levocardia or dextrocardia.⁵ The terms levocardia and dextrocardia point to the direction of the cardiac apex at birth. In dextrocardia, the base-to-apex axis is towards the right, and in levocardia, the axis is reversed.

Situs inversus occurs more frequently with dextrocardia.⁶ An incidence of 3-5% congenital heart disease is noted in situs inversus with dextrocardia, commonly with transposition of the great vessels. Among these patients, 80% have a right-sided aortic arch.

20% patients have Kartagener syndrome or primary ciliary dyskinesia, a mix of situs inversus, bronchiectasis, and infertility attributed to atypical motility of the ciliary lining of respiratory tract (upper and lower sinuses, middle ear, Eustachian tube) and fallopian tubes in females or in the flagella of sperm cells in males.⁷ It is a combination of dextrocardia with breathing difficulty due to chronic dilatation of the bronchi, recurrent respiratory tract infections (bronchiectasis) and sinusitis. It was a probability in our case because of a positive history of recurrent rhinorrhea with the presence of situs inversus totalis. Symptomatology although indicated the presence of ciliary dysfunction, yet we could not get definitive evaluation done by ciliary motility studies due to lack of such facilities at our centre. This calls for a need of optimisation of respiratory status before surgery and care for prevention of inspissation of secretions although it still happened in our patient. Taking into account the possibility of secretions obstructing the endotracheal tube one should have a low threshold for suctioning of endotracheal tube, and keep a vigilant eye on the changes in lung compliance and peak inspiratory pressures.

Dwarkanath et al have assessed the association between spinal cord anomalies and situs inversus.⁸ Scoliosis, spina bifida, split cord, meningomyelocele and tethered spinal cord have been noted in some cases. A meticulous neurological examination with diligent palpation of the

spine is crucial to detect such problems. Our patient did not show any symptoms related to the above listed disorders.

Cases of airway/craniofacial anomalies have been reported in literature in both adult and pediatric age groups in patients with situs inversus; goldenhar syndrome, aglossia, hypoglossia, cranial diaphysial dysplasia all being noted.^{9,10,11,12} Most of these would be obvious on clinical examination and the anaesthesiologist can modify his plan accordingly.

Alimentary system is affected in more than 50% of the cases. This is mostly because of defects during the rotation and fixation of the gut. A left sided gallbladder and appendix or a right sided duodenal ulcer pain can lead to misdiagnosis and delay of peri-operative care the patient.¹³

Cases of prolonged paralysis and apnea following the use of succinylcholine has been previously described in situs inversus totalis, hence use of the drug was withheld in our patient.¹⁴

It has to be kept in mind that patients may come for surgery formerly undiagnosed or diagnosed with situs inversus. The first step of analysis should probably be to ascertain if the patient has situs ambiguus or situs inversus totalis. As there are huge variabilities in presence of malformations, so meticulous physical exam is essential. The haemodynamic or cardiac monitoring should be assessed individually based on the type and complexity of the cardiac defect and kind of surgical procedure to be performed. In the absence of any spinal deformities, regional anaesthesia can also be used successfully in these patients if the need arises.

Conclusion

We present a discussion of the disorders of left right malformations and review the organ systems that can be critically affected. To conclude, patients with syndromic anomalies with congenital defects need to be exhaustively and cautiously assessed. The data presented in this review attempts to help anaesthesiologists take care of patients with such rare diseases safely.

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