

# A patient with Situs Inversus without ciliary dysfunction presenting for urgent Cesarean Section: considerations for a safe anesthetic

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## Abstract

Introduction: Patients with situs inversus without ciliary dysfunction have multiple structural and functional abnormalities especially in terms of cardiac, spine and airway malformations, posing a challenge to a safe anesthetic.

Case: A 40 year old female was admitted with spontaneous onset of labor with a breech presentation for urgent cesarean section. Her medical history included dextrocardia, which prompted a thorough physical examination for other possible structural anomalies, especially pertaining to her cardiovascular system, airway and the spine. The case was found to be total situs inversus without any evidence of ciliary dysfunction. An epidural anesthetic was planned, with careful dosing of and monitoring for hemodynamic changes. The case proceeded uneventfully and the epidural catheter was also used for postoperative pain control.

Conclusion: We describe here the evaluation of and the precautions taken during the care such a patient who underwent a successful cesarean section delivery and post-operative pain management with an epidural anesthetic.

## INTRODUCTION

Situs inversus is a congenital visceral malrotation anomaly that results from disturbances in establishment of left-right asymmetry. It can exist as part of the Kartagener's syndrome or Immotile cilia syndrome, and also independent of them. Anesthetic management of patients with Kartagener's syndrome has been described [1]. Patients with situs inversus without ciliary dysfunction present a very different set of problems due multiple structural and functional abnormalities that coexist with it [2]. We describe here such a case that presented for an urgent cesarean section at our hospital. We present the evaluation of and precautions taken during the care of this patient. We could not find any reports on peripartum management of these patients during our literature search. A review of nomenclature of the anomalies of left-right asymmetry, specific organ system malformations described in these patients is presented and their implications for anesthetic care are discussed.

## CASE REPORT

A 40 year old female was admitted in the Labor and

Delivery suite with spontaneous rupture of membranes and onset of labor. She had a breech presentation and the obstetrician planned to deliver the baby with cesarean section. She had one prior uneventful vaginal delivery two years ago. Her medical history included dextrocardia, good functional capacity with no deterioration in cardiorespiratory function during the pregnancy. Her history of dextrocardia prompted a thorough physical examination for other possible structural anomalies, especially pertaining to her cardiovascular system, airway and the spine. She was 5' 2", 145 lbs at that time and the airway exam revealed no apparent abnormalities with Mallampatti Class 1 and good range of motion of the neck. There were no apparent spinal deformities on examination and palpation. A neurological examination of lower extremities was normal. Auscultation of the heart revealed a Grade 2/6 systolic murmur at the left parasternal border. Vesicular breath sounds without crackles or wheeze were heard on chest auscultation. An abdominal examination for checking left sided liver was not conclusive due to her pregnancy and labor. A presumptive diagnosis of situs inversus totalis was made. She had minimal pedal

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edema which had been with her during most of her pregnancy. Her labs showed a hematocrit of 42.0 and white cell count of 6.8. Liver function tests from a month ago were all within normal range. According to the patient she had a 2D-Echocardiogram of the heart during the first pregnancy two years ago, which had shown dextrocardia, but neither details nor the report was available for review.

An epidural anesthetic was planned for the cesarean section to allow a gentle induction of neuraxial block as compared to a spinal anesthetic. An 18 ga intravenous line was placed. In the operating room, instead of the usual right side up, her left side was put up for vena caval decompression.

Electrocardiogram leads were placed on exactly opposite spots to what is done for levocardia. With the patient in sitting position, an epidural catheter was placed uneventfully at L3-L4 level. Test dose of 1.5% lidocaine with epinephrine 1:200,000 (3 ml) was negative. A total of 15 ml of 2% Lidocaine with epinephrine 1:200,000 was administered, in 5ml increments, to achieve a T-6 level block to pin-prick stimulus. Patient's heart rate, blood pressure and oxygen saturation remained stable during the induction of anesthetic. Surgery was uneventful and healthy male baby was born, with Apgar scores 9 and 9 at 1 and 5 minutes respectively. She lost about 1000ml of blood during the surgery and received 2800ml of Ringers Lactate intravenously. The epidural catheter was left in place for post operative patient controlled epidural analgesia and discontinued uneventfully 2 days later.

### **DISCUSSION**

This patient presented with abnormal finding of dextrocardia for an urgent surgical procedure. The finding of dextrocardia and lack of previous medical evaluation lead to a search for other possible abnormalities. The fact that she had an uneventful vaginal delivery two years ago and no cardiorespiratory deterioration during this pregnancy indicated that may not have serious cardiovascular anomalies. But as these could not be ruled out totally, the decision was taken to use an epidural technique instead of a spinal anesthetic. The patient did not reveal any abnormalities of spine or airway.

Our literature search did not reveal any information on anesthesia for this subset of patients. We present here a brief review of the anomalies of right-left asymmetry for anesthesiologists to be able to evaluate these patients before an anesthetic. The normal left-right anatomical arrangement is called situs solitus, and mirror image reversal of all

asymmetrical structures is called total or complete situs inversus. The intermediate phenotypes between these two extremes have been called situs ambiguus or heterotaxia [3]. Situs ambiguus with symmetrical morphology can be a bilateral left sidedness, called as left isomerism or polysplenia syndrome, or a bilateral right sidedness, called as right isomerism or asplenia syndrome. Majority of patients with these syndromes will present in infancy or childhood with these associated anomalies; however, some do survive to adulthood remaining undiagnosed [2,4].

A list of structural abnormalities in these patients is given in Table 1. The most important abnormalities involve the cardiovascular system. The prevalence of congenital heart disease (CHD) in normal levocardiac population is less than 1% while that in situs inversus totalis goes up to 3-5%. We think our patient belonged to this group. A normal situs with dextrocardia and polysplenia syndromes have CHD in the range 90-95%. Patients with asplenia syndrome almost universally have CHD which leads to their diagnoses in infancy, with rare reports of discovery in adulthood [5]. The anomalies are usually 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 [2]. In most cases of asplenia syndrome the aorta and the vena cava are situated in a "piggy back" fashion on same side of the spine [6]. The absence of the hepatic segment of IVC with azygos or hemiazygos continuation occurs in 50-85% of patients with polysplenia [7].

Neuraxial techniques have an important place in obstetric anesthesia. Dwarkanath et al have reviewed the association between situs inversus and spinal cord malformations [8]. Scoliosis, split cord, spina bifida, meningomyelocele and tethered spinal have been described. Though most of these are symptomatic in childhood their presentation in asymptomatic adults has been found rarely. A diligent neurological examination and palpation of the spine is essential to detect these problems. Our patient did not reveal any symptoms related to the above listed disorders.

**Figure 1**

Table 1: Structural anomalies in Situs Inversus

| System         | Anomalies  |
|----------------|--|
| Cardiovascular | Congenital heart disease including atrial septal defect (ASD), ventricular septal defect (VSD), transposition of great arteries, total anomalous pulmonary venous connection, absent coronary sinus, pulmonary valve stenosis etc. (incidence is 5% situs inversus with dextrocardia, but >90% in most other defects of laterality)<br>Interruption of inferior vena cava (IVC), IVC & aorta on same side, anomalies of azygos-hemiazygos vein |
| Spine          | Scoliosis<br>Meningomyelocele<br>Spina bifida, split cord<br>Tethered cord   |
| Airway         | Aglossia, hypoglossia<br>Goldenhar syndrome<br>Cranial diaphysial dysplasia  |
| Alimentary     | Left side appendix, right side gall bladder<br>Atresia, defects of rotation, fixation<br>Liver structural defects  |

Cases of airway/craniofacial anomalies have been described in literature in both pediatric and adult age groups in patients with situs inversus. Goldenhar syndrome [9], aglossia [10], hypoglossia [11], cranial diaphysial dysplasia [12] have been reported. It seems that most of these would be evident on clinical exam and the anesthesiologist can tailor his plan accordingly. We did not come across any report of unanticipated difficult airway due to a structural abnormality in these patients.

Alimentary system is involved in more than half of the cases. This is mostly due to defects during the rotation and fixation of the gut, and rarely atresias. A left sided appendix and gallbladder or a right sided duodenal ulcer pain can lead to misdiagnosis and possibly delay, prolong or complicate perioperative care the patient [13]. Asplenic patients have hematological findings consistent with the absence of spleen (Howell-jolly bodies, Dohle bodies, Heinz bodies and target cells) [13]. They can also be predisposed to infections due to this. A case of prolonged paralysis after use of succinylcholine has been reported in a patient with situs inversus totalis [14].

Patients may come for surgery previously diagnosed or undiagnosed with situs inversus. The first step in evaluation should probably be to determine if the patient has situs

inversus totalis or situs ambiguus. As there is a huge variability in presence of malformations, a thorough physical exam is essential. Table 2 summarizes the common findings for diagnosis. If time permits, an echocardiogram of the heart may help in confirming absence of major defects. A cardiology consult may help to determine if further testing is needed. The cardiac or hemodynamic monitoring needs to be individualised based on the severity and complexity of the cardiac defect and type of surgical procedure. In the absence of any spinal deformities, regional anesthesia can be used successfully in these patients, as was in our case.

**Figure 2**

Table 2: Common clues to anomalies of right-left asymmetry

| Examination          | Sign  |
|----------------------|---|
| Physical examination | Right sided cardiac apical impulse                          |
|                      | Auscultation of cardiac sounds on right side                |
|                      | Left sided liver  |
| Electrocardiogram    | Lead I has negative QRS with inverted P waves               |
|                      | AVL and AVR are reversed                                    |
|                      | V1-6 'mirror image' of normal R wave progression            |
|                      | EKG changes of associated cardiac anomalies (e.g. VSD, ASD) |
| X-ray                | Right sided cardiac shadow                                  |
|                      | Gastric sided 'gastric bubble'                              |

**CONCLUSION**

We describe a case of situs inversus totalis, who successfully underwent a Cesarean section with epidural anesthesia. We present a review of the disorders of left right malformations and discuss the organ systems that can be severely affected. The information presented would help anesthesiologists take care of such patients with a rare disease safely.

Competing interest: The author(s) declare that they have no competing interests

Authors' contributions: JS and YF were involved in direct care of the patient, searching literature and writing manuscript. IM was involved in searching literature, writing/preparing and reviewing manuscript.

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