

Two Case Reports of Situs Inversus Totalis

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Abstract

Case A, a 27-year-old Iranian male and case B, a 22-year-old Afghani male, both presented to the Emergency Department of Shohadaye Hafe-Tir Hospital with multiple traumas due to car accidents. Neither was aware of his unusual anatomy. One case unaware two just right heart. Both patients were referred to the Imaging Department for CT scans and some radiographic studies. We discovered they were a situs inversus totalis cases with right heart in side and left in the side liver. in CT scan of the thorax and abdomen for assessment of traumatic injuries. Thoracic and abdominal organs and the viscera were inversed complementary. It is important to inform medical personnel of the diagnosis of situs inversus totalis in order to decrease errors and prevent complications that arise from patient assessment and care, particularly in cases of appendicitis and abdominal organ lacerations.

Key words: Situs Inversus Totalis, Congenital Abnormalities, Computed Tomography, X-Ray

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Introduction

Situs inversus is a rare congenital anomaly characterized by the transposition of the abdominal organs, viscera and vasculature. When associated with dextrocardia it is known as situs inversus totalis. This condition is generally an autosomal recessive genetic condition. It may or

may not be associated with dextrocardia, also known as situs inversus totalis [1,2]. Generally, this rare genetic anomaly is discovered incidentally, often when a radiographic assessment of a patient is undertaken, particularly to investigate blunt abdominal trauma or infection.

The condition affects all major structures within the thorax and abdomen. The organs are simply transposed through the sagittal plane. The heart is located on the right side of the thorax; the stomach and spleen are located on the right side of the abdomen and the liver and gall bladder on the left side. The left lung has three lobes and the right lung two lobes; blood vessels, nerves, lymphatics and the intestines are also transposed (Figure 1). If the heart is located on the right side of the thorax, it is known as situs inversus with dextrocardia or situs inversus totalis. When the heart remains in its normal position on the left side of the thorax, there is a much rarer condition (1 in 22000 of the general population) known as situs inversus with levocardia or situs inversus incompletus. Situs inversus with levocardia or dextrocardia without situs inversus present with much higher rates of congenital defects than situs inversus with dextrocardia.

This report discusses two cases seen among 10000 patients who have referred to the Shohadaye Hafte-Tir Hospital Imaging Department for abdominal CT scans over a four year period.

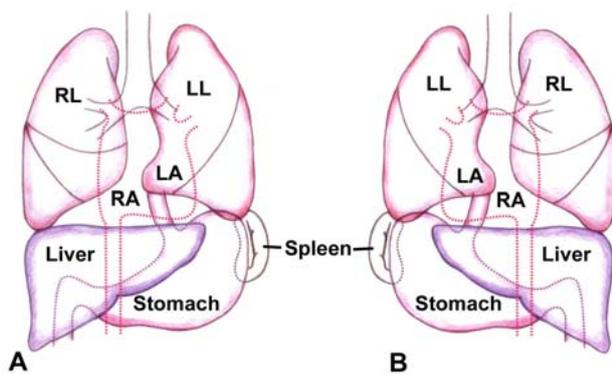


Figure 1. Schematic drawings illustrate the standard anatomy of situs solitus (A) and the mirror image of situs inversus (B). The right lung (RL), left lung (LL), right atrium (RA), and left atrium (LA) are shown [3].

We report a case of situs in versus discovered in the Medical Imaging Center following imaging

studies performed in relation to multiple traumas. This case was particularly interesting because of the scarcity of this association and the diagnostic difficulties and deferent approach in Jejunostomy.

Genetics and situs in versus

Situs in versus is an autosomal recessive genetic condition, although it can be X-linked or found in identical "mirror" twins. In the absence of congenital heart defects, individuals with situs inversus are phenotypically unimpaired and can lead normal healthy lives without any complications related to their medical condition (Figure 2).

Situs inversus occurs more commonly with dextrocardia [4]. A 3%-5% incidence of congenital heart disease is observed in situs inversus with dextrocardia, usually with transposition of the great vessels. Of these patients, 80% have a right-sided aortic arch. Situs inversus with levocardia is rare [5] and it is almost always associated with congenital heart disease [6-11].

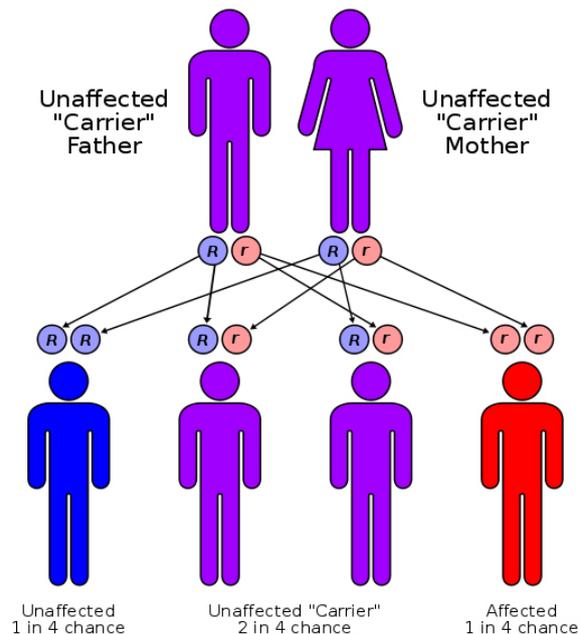


Figure 2. Situs in versus has an autosomal recessive pattern of inheritance [12].

Many people with situs inversus totalis are unaware of their unusual anatomy until they seek medical attention for an unrelated condition. The reversal of the organs may then lead to some confusion, as many signs and symptoms will be on the 'wrong' side. For example, if an individual with situs inversus develops appendicitis, they will present to the physician with lower left abdominal pain, since that is where their appendix lies. Thus, in the event of a medical problem, the knowledge that the individual has situs inversus can expedite diagnosis. People with situs inversus may inform their physicians before an examination, so the physician can redirect their search for heart sounds and other signs. But in people who present to an Emergency Department in a coma or those with a low GCS the condition is different, thus wearing a medical identification tag can help to inform health care providers in the event the person is unable to communicate.

Situs inversus also complicates organ transplantations as donor organs will almost certainly come from situs solitus (normal) donors. The geometric problems arise from placing an organ into a cavity shaped in the mirror image. because heart and liver are chiral [12] For example, a person with situs inversus who requires a heart transplant needs all the vessels to the transplant donor heart reattached to their existing ones. However, the orientation of these vessels in a person with situs inversus is reversed, necessitating steps so that the blood vessels join properly.

Case report

All study procedures were approved by the Medical Ethics Committee Shahid Behesht University of Medical Sciences, Tehran Iran. Additionally we got verbal informed consent from both of two cases.

Case A

A 27-year-old Iranian male presented to the Emergency Department of Shohadaye Hafe-Tir Hospital with multiple trauma due to a car accident. After admission and examination for assessment of traumatic injuries he was referred to the Medical Imaging Center for brain, thoracic and abdominal CT scans and chest and lower extremity imaging studies (Figure 3).

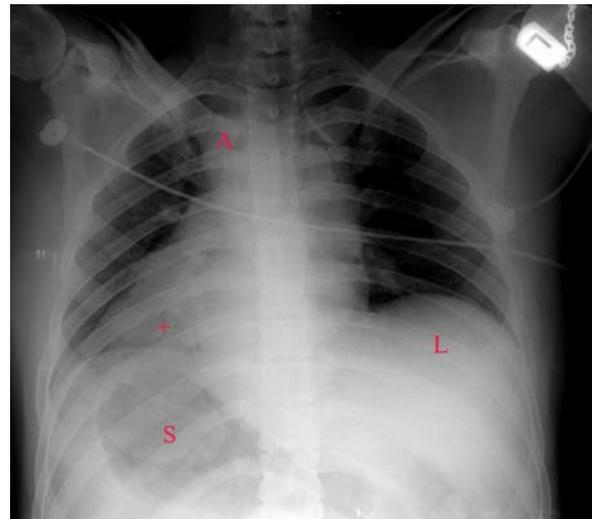


Figure 3: AP chest radiograph in a 27-year-old Iranian male with situs inversus and dextrocardia. This image shows that the cardiac apex (+) points to the right. Right-sided aortic arch (A). The stomach bubble (S) is visible in the right upper quadrant. (DDR ,MMD Italy)

The brain CT scan was normal, however there were FX in the right femur and FX and DX at base of the metatarsal and in sonography The hemoperitoneum was reported. We discovered a situs inversus totalis on the left side of his liver The heart was located on the right side of the thorax, the stomach and spleen on the right side of the abdomen and the liver and gall bladder on the left side. The patient's left lung had three lobes, whereas the right lung had two lobes. Bloodvessels (Aortic arc, carotid artery, SVC and IVC), nerves, the alimentary tract and intestines were also transposed. The thoracic,

abdominal organs and viscera were completely reversed. Free fluid was observed in his peritoneal cavity. The patient underwent surgery to repair a lacerated mesentery. He had a pulse rate of 88 beats per minute, a blood pressure of 140/90 mm Hg and his blood group was type B.

Case B

A 22-year-old Afghani male presented to the Emergency Department of Shohadaye Hafte-Tir Hospital with multiple trauma due to a car accident. After admission and examination for assessment of the presence of traumatic injuries, he was sent to the Medical Imaging Center for brain, thoracic and abdominal CT scans as well as chest and cervical spine imaging studies (Figures 4,5,6 and 7). We discovered a situs inversus totalis with the left side liver, The heart was located on the right side of the thorax, the stomach and spleen on the right side of the abdomen, and the liver and gall bladder on the left side of the patient's body. The patient's left lung had three lobes and the right lung had two lobes. The blood vessels (Aortic arc, carotid artery, SVC and IVC), nerves, the alimentary tract and intestines were also transposed. The thoracic, abdominal organs and viscera were completely inverted. According to the CT scans, the patient had a subarachnoid hematoma (SAH), intraventricular hematoma (IVH), a few contusions and edema in his brain. There was free fluid in his peritoneal cavity. The patient was transferred to the ICU. His pupils were asymmetric and unreactive to light. The right pupil size was 4 mm and the left pupil was 2 mm. He had a pulse rate of 96 beats per min, blood pressure of 100/60 mm Hg and his blood group was type O⁺. His physical examination was remarkable for tenderness and fracture of his right clavicle, and tenderness and fracture on his right superior and inferior pubic ramuses. In the ICU, he was in a vegetative condition and after seven days he

underwent a pre-operative Examination, after which a median laparotomy was performed, followed by a tracheostomy and Jejunostomy with foley catheter. The patient was returned to the ICU.

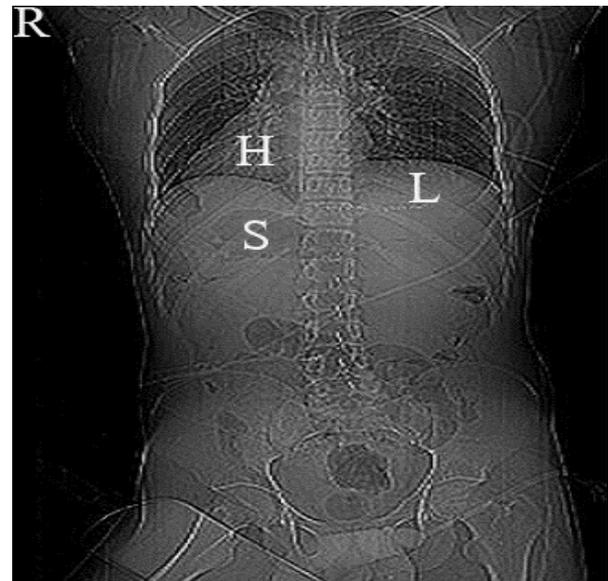


Figure 4. AP tomogram of the chest, abdomen and pelvis with left-sided liver (L) and right-sided heart (H) and stomach (S). (MDCT aqilion 16 Toshiba)

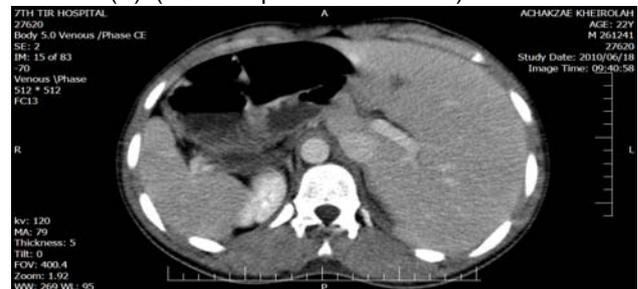


Figure 5. Axial CT scan slice of situs inversus with left-sided liver (L) and right stomach (S). (MDCT aqilion 16 Toshiba) from two cases

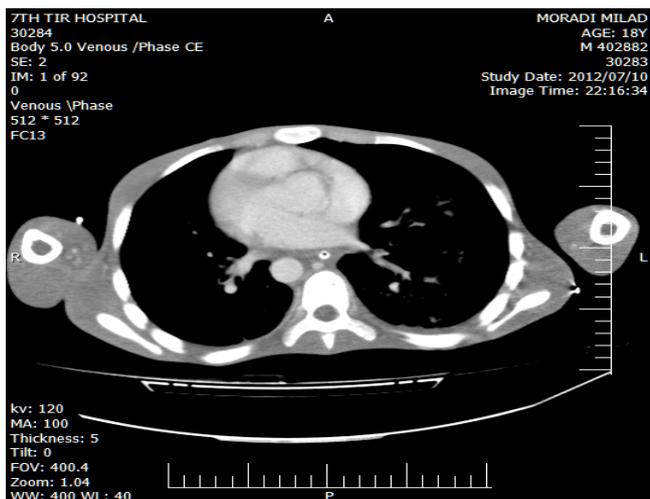


Figure 6. Axial CT scan slice of situs inversus with right-sided aorta and heart (H). (MDCT aquilion 16 Toshiba)

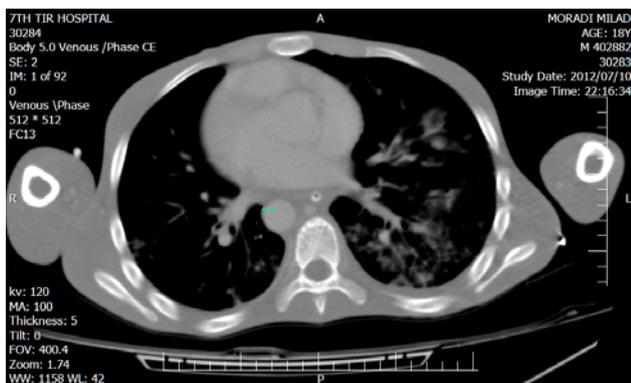


Figure 7. Axial CT scan slice of situs inversus with left-sided aorta. (MDCT aquilion 16 Toshiba)

Discussion

Situs inversus is a positional anomaly that rotates the abdominal internal viscera. It is known as situs inversus totalis when associated with a transposition of the thoracic organs. Situs inversus is a rare congenital anomaly with an incidence in the population of only 0.001 to 0.01 [1, 2] with a male-to-female ratio of 3:2 [13]. Its transmission mode is autosomal recessive, but its precise genetic mechanism has yet to be identified [1, 13]. Situs inversus results from a rotation in the opposite direction of the viscera and organ during the organogenesis of the embryo [2, 13]. Patients with situs inversus who present to the Emergency Department may have diagnostic problems in physical examination because of their unusual

anatomy, low GCS and unusual localizations of their pains and symptoms. In patients with situs inversus who present with multiple traumas, medical imaging can assist clinicians and surgeons to arrive at a correct and reliable diagnosis. Abdominal and chest X-rays, sonography and CT scan also facilitate a reliable and early diagnosis if the patient is unaware of his unusual anatomy and has a low GCS [1, 13, 14]. Medical imaging can also guide the appropriate therapeutic choice, biopsy, surgical indications and approaches, and type and location of the incision in Jujenostomy or other surgical procedures such as an appendectomy, chest tube insertion and organ transplantation. Laparoscopy is useful in these situations as it favors a minimally invasive surgical approach in diagnostics and treatment [15]. In patients with situs inversus totalis who present with an acute abdomen that suspended of having appendicitis that involves left iliac localization, the diagnosis is more complex and difficult. Until 2008, fewer than 10 cases of appendicitis associated with situs inversus were reported in the literature [13]. Half of these reported pain in their right iliac fossa despite the presence of situs inversus [1]. Informing physicians and surgeons of the diagnosis of situs inversus can decrease medical errors and prevent some of complications that may arise in assessment and care.

The occurrence of situs inversus is very rare. Very few cases have been reported in the literature. This condition poses a diagnostic problem that can be solved by medical imaging, particularly by the performance of a body CT scan and other imaging modalities such as sonography and laparoscopy. These procedures allow for early management of the disease and guidance for the best approaches.

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