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# Isolated Levocardia with Situs Inversus in a Child: A Rare and Frequently Missed Diagnosis
# Isolated Levocardia with Situs Inversus – A Rare & Frequently Undiagnosed

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## ABSTRACT

A rare condition in which the heart is located within the left hemithorax with its base-apex axis pointing caudally to the left and with partial or complete inversion of the abdominal viscera. This condition has an estimated incidence of 1 in 22,000 in the general population. This condition is comorbid with various heart defects and is associated with poor prognosis, with only 5-13% of the population surviving more than 5 years.[1]. Despite high mortality and being associated with congenital forms of severe heart defects, this condition is frequently undiagnosed and is often an accidental finding. In this case report, we describe a 13-year-old girl diagnosed as having complete situs inversus with levocardia, with symptoms only involving the gastrointestinal tract.

**Keywords:** Phylogenetic Diversity, Ethnobotany, Medicinal Plants, Species Richness, Conservation Biology

## LEARNING OBJECTIVE

The objective of this case report is to describe a rare presentation of isolated situs inversus with levocardia in a 13-year-old girl exhibiting only gastrointestinal symptoms. It aims to highlight the importance of accurate diagnosis in atypical presentations and the challenges faced in resource-limited settings. Additionally, this report seeks to contribute to the limited literature on situs inversus with levocardia and raise awareness about its clinical and diagnostic intricacies.

Isolated levocardia with situs inversus (ILSI) is

**CASE PRESENTATION**

A 13-year-old girl was brought to the OPD (outpatient department) with a history of epigastric pain that started four years ago. The pain was sudden in onset, moderate in intensity, non-radiating, and non-colicky. It was associated with intermittent diarrhea and was aggravated by taking meals. For the last two months, the pain increased in severity and radiated to the left flank. For the last two weeks, the patient's pain was associated with four to five episodes of projectile vomiting daily. The vomitus consisted of food particles and didn't contain bile or blood. Physical exam of the chest and abdomen didn't reveal any abnormalities.

The patient has visited various physicians and hospitals over the last four years and was diagnosed and treated for gastroenteritis. The most recent hospital visit was two months ago, with the complaint of a productive cough. Sputum cultures were sent for Mycobacterium tuberculosis and were negative, and the patient was treated with antibiotics. The patient was delivered via C-section at full term. The patient vomited one hour after birth. The rest of the neonatal and developmental history was unremarkable. The patient's parents and two siblings were alive and had no known medical illnesses.

The patient first underwent abdominopelvic ultrasonography four months ago, which indicated the presence of liver in the left hypochondrium with a span of 12.0 cm and no gross abnormalities **(Figure 1)**. The spleen was seen as 9.4 cm in size, present in the right hypochondrium. The gallbladder, pancreas, kidneys, bladder, ovaries, and uterus were unremarkable. Minimal free fluid was seen in the pouch of Douglas (POD). A repeat ultrasound on the day of patient presentation confirmed the previous findings.

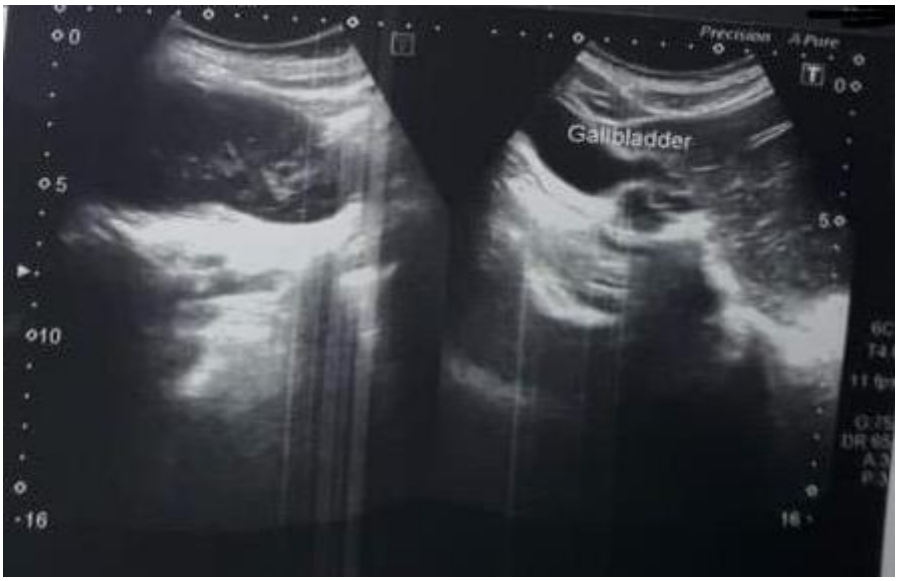


**FIGURE 1. ABDOMINAL ULTRASOUND SHOWING LIVER IN THE LEFT HYPOCHONDRIUM AND SPLEEN ON THE RIGHT, CONFIRMING SITUS INVERSUS**

A CXR confirmed the findings of the ultrasound, showing levocardia (cardiac apex pointing towards the left side), liver on the left side, and a right-sided stomach and spleen **(Figure 2)**. The patient also had situs inversus with an ascending colon on the left side and a right-sided descending and sigmoid colon

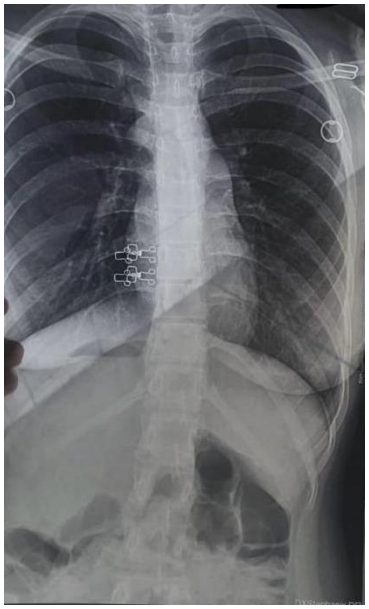

**FIGURE 2. CHEST X-RAY SHOWING LEVOCARDIA WITH RIGHT-SIDED STOMACH AND SPLEEN, AND LIVER ON THE LEFT.**

The patient didn't have any structural cardiac defects and had normal electrocardiography and echocardiography. Therefore, the patient was offered symptomatic treatment for her gastrointestinal symptoms and offered watchful waiting.

## DISCUSSION

The term 'situs' refers to the positioning of cardiac atria and visceral abdominal organs relative to the midline. The normal positioning, i.e., left-sided heart and proper visceral positioning (left-sided stomach), is referred to as situs solitus. Conversely, situs inversus is characterized by mirror imaging of the cardiac atria and abdominal organs with the midline as the plane. The reported incidence of situs inversus is 1:10,000 and is almost always associated with dextrocardia (right-sided heart).[2] According to the Genetic and Rare Diseases Information Center (GARD), situs inversus is a genetic disease, and there are several known variants of the gene(s) that may cause it. These include ANKS3, NME7, NODAL, CCDC11, WDR16, MMP21, PKD1L1, and DNAH9, and these may be inherited in an autosomal dominant or autosomal recessive manner.[3]

As mentioned above, however, situs inversus with levocardia is an extremely rare presentation of maldevelopment, which is usually associated with high mortality. According to the literature, we could identify only one case of situs inversus with levocardia in Pakistan, which likely makes this case the second 'reported' case presenting this disease.[4] The extreme rarity of this condition in Pakistan may be attributed to the rarity of the disease, lack of diagnosis/misdiagnosis, or under-reporting of the cases.

Countries like Pakistan, with inadequate healthcare resources and a lack of public awareness, harbor a plausible cause for the late diagnosis and presumable underreporting of cases like this. This is evident as this patient was diagnosed with gastroenteritis multiple times over the last couple of years due to persistent abdominal symptoms. Due to a lack of resources, the patient couldn't undergo extensive testing early on, which, if done, might have prevented the deterioration of her symptoms.

## TREATMENT

In most cases, Situs Inversus (SI) may present with no symptoms or clinical picture and, therefore, does not require treatment. However, SI associated with other heart defects or visceral abnormalities should be treated for the symptoms that appear.[5] Attempting to surgically reverse

the organ positions in such cases is generally discouraged due to the high risks involved and the unpredictable impact on a patient's overall well-being.[6]

However, when it comes to isolated situs inversus with levocardia, a typical presentation often includes significant cardiovascular defects. Consequently, a patient's prognosis and survival largely hinge on the severity of these heart-related issues. In our patient's unique case, her heart exhibits no concurrent structural abnormalities. As a result, the management plan for this patient will primarily focus on treating any specific medical conditions that may arise, along with regular monitoring for potential cardiac concerns using electrocardiography and echocardiography.

## CONCLUSION

Situs inversus with levocardia may present with symptoms depicting another pathology, and proper diagnostic modalities should be utilized to identify and treat the underlying cause. Symptomatic treatment is usually opted for in patients with no structural cardiac abnormalities.

**CONSENT:** Patient's assent and parental consent were obtained from the patient and her parents.

## CONFLICT OF INTEREST

The authors declare that they have no conflicts of interest relevant to this case report.

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