



## Perforated Appendicitis with Generalized Peritonitis in a Patient with Situs Inversus Totalis: A Case Report and Literature Review

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### Article History :

Received date : 2025/07/14  
Revised date : 2025/08/09  
Accepted date : 2025/09/21  
Published date : 2025/10/14



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E-ISSN :

ISSN 3048-1368



P-ISSN

ISSN 3048-1376



### ABSTRACT

**INTRODUCTION:** Situs inversus totalis (SIT) is a rare congenital condition characterized by the mirror-image transposition of thoracic and abdominal organs. Its co-occurrence with acute appendicitis, one of the most common surgical emergencies, presents a significant diagnostic challenge due to the atypical, left-sided clinical presentation. This can lead to diagnostic delays and an increased risk of complications.

**CASE ILLUSTRATION:** A 40-year-old male presented to the emergency department with a three-day history of generalized abdominal pain that originated in the left lower quadrant (LLQ). Physical examination revealed tachycardia and signs of generalized peritonitis. Laboratory investigations showed marked leukocytosis. A chest radiograph incidentally revealed dextrocardia, raising suspicion for situs inversus. A subsequent abdominal ultrasound confirmed the transposition of visceral organs and identified findings suggestive of acute appendicitis in the LLQ. An emergency exploratory laparotomy was performed,

which confirmed a perforated, gangrenous appendix located in the left iliac fossa, with approximately 200cc of purulent fluid in the peritoneal cavity. An appendectomy was performed, and the patient experienced an uneventful post-operative recovery.

**DISCUSSION:** The patient's initial presentation with LLQ pain is a classic "mirror image" of typical appendicitis, highlighting the critical importance of considering anatomical variations in the differential diagnosis of abdominal pain. The diagnostic pathway, initiated by the serendipitous finding of dextrocardia, underscores the pivotal role of basic and advanced imaging in unmasking the underlying condition and facilitating timely surgical intervention, thereby preventing further morbidity.

**CONCLUSION:** Although exceedingly rare, left-sided appendicitis in patients with situs inversus must be included in the differential diagnosis for patients presenting with LLQ pain. A high index of clinical suspicion, coupled with prompt and appropriate imaging, is paramount to avoiding diagnostic delays and reducing the risk of severe complications such as perforation and generalized peritonitis.

**KEYWORDS:** Situs Inversus, Left-Sided Appendicitis, Perforated Appendicitis, Dextrocardia, Diagnostic Challenge, Atypical Presentation.

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## INTRODUCTION

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### Background

Acute appendicitis is one of the most frequent causes of acute abdominal pain globally, necessitating emergency surgical intervention. The classic clinical presentation is well-established: it typically begins with vague, periumbilical pain that subsequently migrates to the right lower quadrant (RLQ), localizing at McBurney's point. This is often accompanied by anorexia, nausea, vomiting, low-grade fever, and localized tenderness on physical examination (Ahmed et al., 2007).

This archetypal presentation, however, is contingent upon normal anatomical positioning, known as *situs solitus*. Situs Inversus (SI) is a rare congenital anomaly characterized by the transposition of thoracic and/or abdominal organs to the opposite side of the body. When this transposition involves all viscera, including dextrocardia (heart on the right side), the condition is known as Situs Inversus Totalis (SIT). The estimated prevalence of SIT is approximately 1 in 8,000 to 1 in 25,000 live births, arising from a failure of the primitive gut to rotate correctly during embryological development (Akbulut et al., 2010; Eitler et al., 2015).

The convergence of these two conditions—acute appendicitis in a patient with SIT—creates a significant clinical conundrum. The anatomical transposition results in a "mirror-image" clinical picture, with the appendix located in the left iliac fossa. Consequently, the inflammatory process manifests as left lower quadrant (LLQ) pain, a presentation that can mislead even the most experienced clinicians and is a well-documented diagnostic pitfall (Nelson and Pesola, 2001).

### Study Objectives

The primary objectives of this case report are:

- To present a detailed account of a rare case of perforated gangrenous appendicitis with generalized peritonitis in a 40-year-old male with previously undiagnosed situs inversus totalis.

- To analyze the diagnostic pathway, emphasizing the clinical reasoning and the sequential role of different imaging modalities that led to the correct pre-operative diagnosis.
- To review the relevant literature and discuss the surgical management strategies and challenges associated with this unique anatomical configuration.

### **Study Benefits**

This report aims to serve several key purposes:

- To increase awareness among clinicians, particularly emergency physicians, radiologists, and general surgeons, about this rare but critical diagnosis.
- To provide a practical educational resource that illustrates the potential for misdiagnosis when faced with atypical abdominal pain, thereby aiming to reduce diagnostic delays and improve patient outcomes.
- To contribute to the sparse body of medical literature on this subject, especially from the Indonesian and broader Southeast Asian context, where such case reports are limited.

### **Hypothesis**

The atypical, left-sided presentation of acute appendicitis in patients with situs inversus significantly increases the likelihood of misdiagnosis or delayed diagnosis. This delay, in turn, leads to a higher incidence of advanced complications, such as perforation and generalized peritonitis, compared to appendicitis in patients with normal situs. The patient in this report presented after a three-day history of pain and was found to have both perforation and peritonitis. This delay is a direct consequence of the diagnostic confusion caused by the LLQ pain. The standard clinical algorithms for abdominal pain are geographically based; LLQ pain triggers a differential diagnosis that does not typically include appendicitis. This case demonstrates how this anatomical anomaly can cause a "system failure" in standard diagnostic reasoning, directly contributing to increased patient morbidity (Molla et al., 2024).

### **Research Gap**

While cases of left-sided appendicitis in patients with SIT have been documented in global literature, there is a notable paucity of published reports from Indonesia. This gap limits the understanding of the clinical presentation, diagnostic challenges, and management outcomes within this specific regional and demographic context. Documenting this case helps to fill this void and provides valuable data for local and international medical communities.

### **Novelty**

This report offers a comprehensive, step-by-step illustration of the diagnostic journey, from a confusing initial presentation to a definitive diagnosis and successful surgical management. The novelty lies in its detailed documentation of a classic diagnostic pitfall and the serendipitous yet crucial role of a routine chest X-ray in redirecting the entire clinical investigation. It serves as a powerful reminder of the enduring importance of foundational diagnostic principles and the need for clinicians to maintain a high index of suspicion for anatomical anomalies in an era dominated by advanced imaging.

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## **CASE ILLUSTRATION**

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### **Patient Presentation and History**

A 40-year-old male, identified as U.S., presented to the emergency department with a chief complaint of severe, diffuse abdominal pain that had been ongoing for three days. The patient reported that the pain initially began in the left lower quadrant (LLQ) before progressively worsening and spreading to involve the entire abdomen, with the most severe pain localized to the lower abdomen. The pain was described as constant and was rated as 6-7 on the Visual Analog Scale (VAS). Associated symptoms included nausea, vomiting, and the passage of mucoid stool. He reported normal urination. There was no significant past medical history, and he denied any previous abdominal surgeries.

### **Clinical Findings on Examination**

On general examination, the patient appeared to be in moderate distress but was fully conscious and alert (Glasgow Coma Scale 15/15). His vital signs were notable for tachycardia: blood pressure was 140/80 mmHg, heart rate was 97 beats per minute, respiratory rate was 20 breaths per minute, body temperature was 36.9°C, and oxygen saturation was 96% on room air. Head and neck examination was unremarkable. Cardiovascular examination revealed normal S1 and S2 heart sounds with a regular rhythm. The pulmonary examination was clear to auscultation bilaterally.

The abdominal examination was highly significant. The abdomen was supple but revealed generalized tenderness to palpation, which was most pronounced in the left lower quadrant. There were clear signs of peritoneal irritation, evidenced by positive guarding and muscular defense (defans muscular). Bowel sounds were present but hypoactive. The extremities were warm with a capillary refill time of less than two seconds.

## **Diagnostic Assessment**

### **Laboratory Analysis**

Initial laboratory investigations were performed upon admission. The complete blood count was significant for marked leukocytosis with a neutrophilic predominance, strongly indicating a severe underlying infectious or inflammatory process. The hematocrit and erythrocyte count were at the lower limit of normal. Other hematological and biochemical parameters were within normal limits. The detailed laboratory findings are presented in Table 1.

**Table 1: Laboratory Findings on Admission**

Parameter	Patient's Result	Reference Range	Interpretation
Haemoglobin	14.3	13.5 – 17.5 g/dL	Normal
Hematokrit	39.5	42 – 52 %	Low
Eritrosit	4.63	4.7 – 6.1 $10^6/??L$	Low
<b>Leukosit</b>	<b>18.6</b>	<b>4.8 – 10.8 <math>10^3/??L</math></b>	<b>High (Leukocytosis)</b>
Trombosit	348	150 – 450 $10^3/??L$	Normal
MCV	85.4	80 – 94 fL	Normal
MCH	30.9	27 – 31 pg	Normal
MCHC	36.1	33 – 37 %	Normal
RDW-SD	50	37 – 54 fL	Normal
<b>PDW</b>	<b>16.2</b>	<b>9 – 14 fL</b>	<b>High</b>

Parameter	Patient's Result	Reference Range	Interpretation
MPV	8.9	8 – 12 fL	Normal

### Radiological Evaluation

As part of the standard workup for an acute abdomen, a chest radiograph was performed. This routine examination yielded a pivotal and unexpected finding: the cardiac apex was pointed towards the right side of the chest (dextrocardia), the aortic arch was on the right, and the gastric bubble was located under the right hemidiaphragm. This constellation of findings was the first objective evidence strongly suggesting a diagnosis of situs inversus totalis. This discovery immediately reframed the entire clinical picture, transforming the "atypical" LLQ pain into the expected location for a "mirror-image" pathology like appendicitis (Oh and Kim, 2012).

Following this critical finding, an emergency abdominal ultrasound was performed to evaluate the visceral anatomy and identify the source of the peritonitis. The sonographic examination confirmed the anatomical transposition: the liver and gallbladder were located in the left upper quadrant, while the spleen was situated in the right upper quadrant. More importantly, in the left lower abdomen, the sonographer identified a non-compressible, non-peristaltic, blind-ended tubular structure with a diameter of 8.4 mm, consistent with an inflamed appendix. The final radiological impression was "Situs inversus" and "Suspect appendicitis with appendix located in the left lower abdomen." The radiologist recommended a contrast-enhanced CT scan for further delineation; however, given the clear clinical signs of peritonitis and the compelling evidence from the initial imaging, this was deferred in favor of immediate surgical intervention.



**Figure 1. Chest X-ray result**

### **Pre-operative Diagnosis and Surgical Plan**

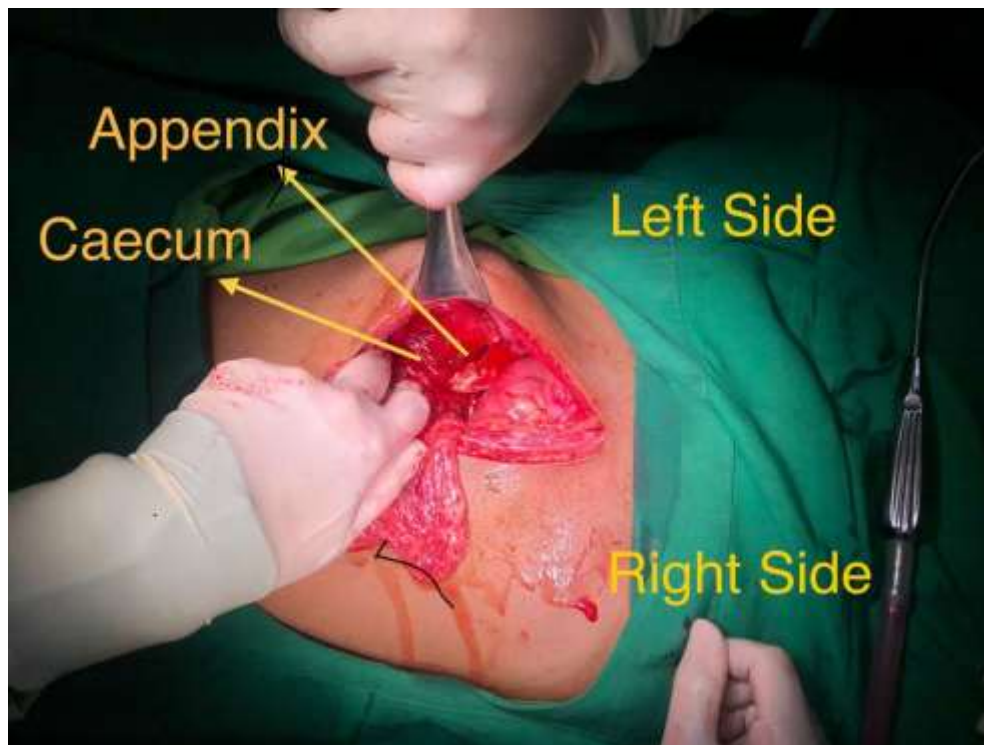
Based on the synthesis of the clinical presentation (LLQ pain with peritonitis), laboratory findings (marked leukocytosis), and definitive imaging results (dextrocardia on chest X-ray and a left-sided inflamed appendix on ultrasound), a confident pre-operative diagnosis was established: **Generalized peritonitis secondary to perforated appendicitis in a patient with situs inversus totalis.** The decision was made to proceed with an emergency exploratory laparotomy.

### **Surgical Intervention and Intraoperative Findings**

The patient was taken to the operating theatre for an emergency exploratory laparotomy. A midline incision was made. Upon entering the peritoneal cavity, approximately 200cc of purulent,

foul-smelling fluid was encountered and immediately suctioned. The intraoperative exploration confirmed the situs inversus anatomy. The caecum and the terminal ileum were found in the left iliac fossa. The appendix was identified arising from the base of the left-sided caecum.

The appendix was grossly inflamed, edematous, and covered in fibrinous exudate, consistent with a gangrenous state. It measured approximately 8 cm in length and 1.5 cm in diameter. A distinct perforation was identified at its base, which was the source of the peritoneal contamination. A fecalith was also found within the appendiceal lumen. There were significant inflammatory adhesions involving the appendix, caecum, and adjacent small bowel loops. A standard appendectomy was performed. The peritoneal cavity was thoroughly irrigated with copious amounts of warm saline until the effluent was clear. A surgical drain was placed in the pelvic cavity, and the abdomen was closed in layers.



**Figure 3. Surgical Intervention**

## Post-operative Course and Follow-up

The patient had an uneventful post-operative recovery. He was managed with intravenous antibiotics, analgesia, and fluid resuscitation. An early feeding protocol was initiated, and he was mobilized on the first post-operative day. He demonstrated a rapid return of bowel function and was discharged home on the third post-operative day in good general condition with a clean and healing surgical wound.

Follow-up appointments were scheduled at one and two weeks post-discharge. At both visits, the patient was found to be in excellent health. His abdomen was soft and non-tender, he was mobilizing well without pain, and his bowel function had returned to normal. The surgical wound had healed completely. Crucially, the patient and his family were provided with comprehensive education about his condition of situs inversus totalis and its implications for any future medical care, and they were advised to inform healthcare providers of this condition at every medical encounter.

## Histopathology

The excised appendix was sent for histopathological examination. The final pathology report confirmed the intraoperative findings, describing features of "acute gangrenous appendicitis with perforation at the base," thereby providing definitive pathological confirmation of the diagnosis.

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## DISCUSSION

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### Embryological and Genetic Basis of Situs Inversus

Situs inversus totalis (SIT) is a rare congenital anomaly with an autosomal recessive inheritance pattern (Manyacka Ma Nyemb et al., 2021). It originates from a critical error during embryogenesis related to the establishment of left-right asymmetry. In normal development (*situs solitus*), the primitive gut undergoes a complex 270-degree counter-clockwise rotation around the superior mesenteric artery, which positions the caecum and appendix in the right lower quadrant. In

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SIT, this process is mirrored, with a 270-degree clockwise rotation resulting in the transposition of all abdominal and thoracic viscera (Benjelloun et al., 2024; Song et al., 2004). This complex process is regulated by a cascade of over 100 genes, and mutations in any of these can lead to laterality defects (Farkas et al., 2022). While often an isolated finding, SIT can be associated with other conditions, most notably Primary Ciliary Dyskinesia (PCD), a disorder of motile cilia. The combination of SIT, chronic sinusitis, and bronchiectasis is known as Kartagener's syndrome, which accounts for approximately 20-25% of all SIT cases (Goyal et al., 2012; Kennedy et al., 2007).

### **The Diagnostic Labyrinth: Atypical Symptoms and Misleading Scores**

The primary challenge in diagnosing appendicitis in a patient with unknown SIT is the atypical clinical presentation. The patient's LLQ pain is a classic "mirror-image" symptom, but for the unsuspecting clinician, it triggers a differential diagnosis that typically includes sigmoid diverticulitis, renal colic, ruptured ovarian cyst, or Meckel's diverticulitis, with appendicitis being a remote consideration (Akbulut et al., 2010). This diagnostic confusion is a major contributor to delays in treatment, which directly increases the risk of progression to perforation and peritonitis, as seen in this case (Benjelloun et al., 2024; Kamran et al., 2022).

Furthermore, standard clinical scoring systems, such as the Alvarado score, are rendered unreliable. The Alvarado score, developed by Dr. Alfredo Alvarado in 1986, heavily weights "right lower quadrant tenderness" with two points, making it a cornerstone of the diagnostic algorithm (Alvarado, 1986). In a patient with SIT, this key criterion is absent, leading to a falsely low score and potentially misleading the clinician away from the correct diagnosis (Lee et al., 2023). Compounding the confusion is the phenomenon of paradoxical pain localization. Due to the complex and sometimes non-transposed visceral afferent neural pathways, up to 31% of patients with left-sided appendicitis may still report pain in the right lower quadrant, creating a profound disconnect between symptoms and anatomy (Akbulut et al., 2010; Kamran et al., 2022).

## **The Decisive Role of Imaging in Unmasking the Truth**

Given the clinical ambiguity, medical imaging becomes the cornerstone of diagnosis. The diagnostic pathway in this case exemplifies a logical and efficient progression. The initial chest X-ray, a routine and low-cost investigation, was the pivotal turning point. The incidental discovery of dextrocardia immediately raised the suspicion of SIT and re-contextualized the entire clinical picture, making left-sided appendicitis the leading diagnosis (Oh and Kim, 2012; Lee et al., 2023). This highlights the immense value of basic radiological studies in the workup of an acute abdomen.

Following this clue, the abdominal ultrasound served as an excellent confirmatory test. It is non-invasive, readily available, and avoids ionizing radiation. Sonography successfully confirmed the visceral transposition and identified the key features of appendicitis: a non-compressible, blind-ended tubular structure in the LLQ with a diameter exceeding 6 mm (Ratani et al., 2002; Patlas et al., 2017). While a contrast-enhanced CT scan is considered the gold standard for diagnosing appendicitis—offering superior detail of mural thickening, peri-appendiceal fat stranding, and potential complications like abscesses—it was correctly deferred in this case (Lee et al., 2023; Patlas et al., 2017). The patient had clear signs of generalized peritonitis requiring urgent surgery, and the combination of the chest X-ray and ultrasound provided sufficient diagnostic certainty. Proceeding directly to surgery without the delay of a CT scan was the most appropriate clinical decision to ensure timely source control (Benjelloun et al., 2024).

## **Navigating Mirrored Anatomy: Surgical Management and Considerations**

The management of perforated appendicitis with peritonitis requires prompt surgical intervention for source control and peritoneal lavage. The choice between an open laparotomy and a laparoscopic approach is a key consideration. In this case, with established generalized peritonitis, an open exploratory laparotomy via a midline incision was a sound and safe choice. This approach provides excellent exposure, facilitates the removal of widespread purulent contamination, and allows for thorough irrigation of the entire peritoneal cavity (Molla et al., 2024).

Laparoscopic appendectomy is now widely adopted and offers benefits such as reduced post-operative pain, lower rates of wound infection, and shorter hospital stays (Kim et al., 2011). It can also serve as a valuable diagnostic tool when the preoperative diagnosis is uncertain (Di Buono et al., 2020). However, laparoscopy in a patient with SIT presents unique technical and ergonomic challenges. The mirror-image anatomy requires the surgeon to adapt to a reversed view, which can be disorienting (Hoang et al., 2023). For the majority of surgeons who are right-handed, operating on the left side of the patient is ergonomically awkward and requires significant mental adjustment for instrument handling and dissection (Farkas et al., 2022; Oh and Kim, 2012). Pre-operative awareness of SIT is therefore crucial, allowing the surgical team to plan port placement and mentally prepare for the reversed anatomical landscape to ensure a safe and efficient procedure.

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## CONCLUSION AND RECOMMENDATIONS

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### Conclusion

This case of perforated appendicitis with generalized peritonitis in a patient with situs inversus totalis underscores a rare but critical clinical scenario where a common disease presents in an uncommonly deceptive manner. The patient's atypical left lower quadrant pain led to a delayed presentation with advanced complications. The diagnosis was ultimately achieved through a high index of suspicion, which was crucially prompted by the incidental finding of dextrocardia on a routine chest radiograph and confirmed by abdominal ultrasound. This case reaffirms that successful outcomes in such rare presentations hinge on the clinician's ability to think beyond standard presentations, consider anatomical anomalies, and utilize imaging judiciously to overcome the diagnostic challenge posed by mirrored anatomy.

### Recommendations

Based on the experience from this case and a review of the literature, the following recommendations are proposed:

- Clinicians should maintain a broad differential diagnosis for abdominal pain and actively consider the possibility of anatomical variations like situs inversus, especially when the clinical picture is atypical or does not conform to classic textbook patterns.
- A chest X-ray should be considered a valuable, low-cost screening tool in patients presenting with severe abdominal pain of unclear etiology. The presence of dextrocardia can be the single most important clue to the correct diagnosis.
- Surgeons preparing to operate on a patient with a known diagnosis of situs inversus should mentally rehearse the "mirror-image" procedure to anticipate and mitigate the ergonomic and technical challenges posed by the reversed anatomical landscape, thereby ensuring a safe and efficient operation.

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