

# Situs Inversus Totalis with Left-Sided Appendicitis: A Case Report

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## Abstract

**Introduction:** Left-sided acute appendicitis (LSAA) develops in association with two types of congenital anomalies: situs inversus totalis (SIT) and midgut malrotation (MM). A Left sided appendicitis is an ambiguous and difficult diagnosis to make. **Aim:** To present a proven case of left-sided acute appendicitis (LSAA) associated with situs inversus totalis (SIT). **Case Report:** A case of Left appendicitis was evaluated in a 28-year-old Asian male, who presented to our hospital in Feb. 2016, with lower abdominal pain more on left side and suspected diverticulitis or acute appendicitis with unusual appendix location. The patient doesn't recall any history of abdominal surgery or about situs inversus totalis, abdominal and pelvic ultrasound was done, left iliac fossa appendicitis was diagnosed, Erect chest X-ray including upper abdomen revealed dextrocardia and stomach air on right side (situs inversus totalis), the patient underwent diagnostic Laproscop and Endoscopic resection of the appendix, with no incidents, and then discharged without complications, follow visits went unremarkable. **Conclusion:** The diagnosis of left lower quadrant pain is based on well-established clinical symptoms, physical examination and physician's experience.

## Keywords

Left Lower Quadrant Pain, Left-Sided Appendicitis (LSAA), Midgut Malrotation (MM), Situs Inversus Totalis (SIT)

## 1. Introduction

Acute appendicitis is a common condition requiring emergency surgery. The diagnosis is based on clinical symptoms, basic radiologic findings and surgeon experience [1] [2]. Approximately one third of patients with acute appendicitis have pain localized outside of the right lower quadrant because of the various

positions of the appendix vermiformis, *i.e.* retrocecal, pelvic, subcecal, preileal and postileal, while subhepatic, meso-cealic, mid-inguinal and left-sided are seen more rarely [1] [2].

Appendicitis causing pain in the left lower quadrant is extremely rare and can occur with congenital abnormalities that include true left-sided appendix or as an atypical presentation of right-sided, but long appendix, which projects into the left lower quadrant [2]. Left-sided acute appendicitis (LSAA) develops in association with two types of congenital anomalies: situs inversus totalis (SIT) and midgut malrotation (MM) [1]-[65]. I am presenting an unusual case of SIT with acute appendicitis presenting as left lower abdominal pain.

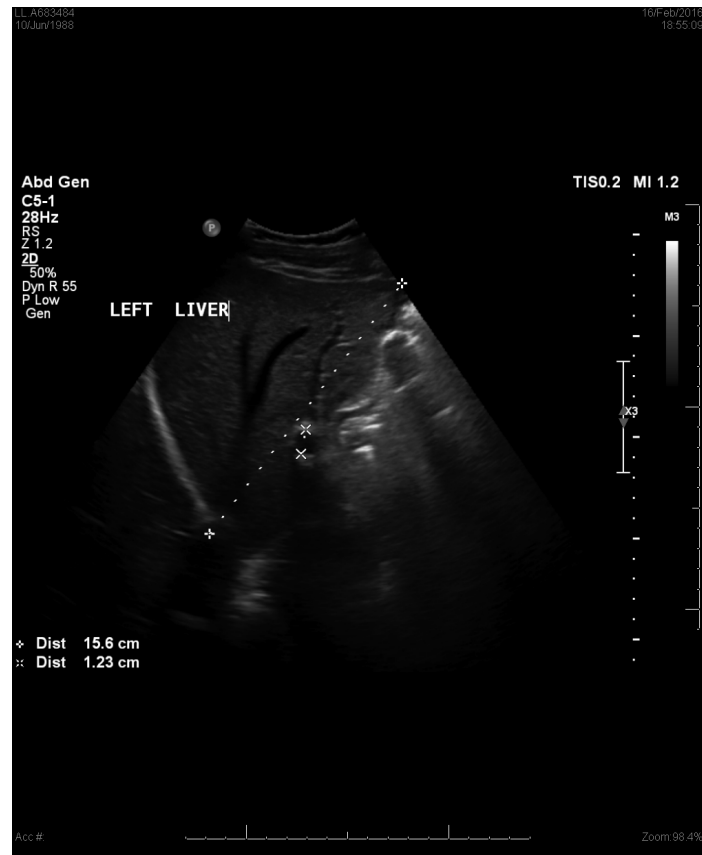
## 2. Case Report

A 28-year-old male presented to the emergency unit on Feb., 16, 2016 (private famous hospital in Abu Dhabi, UAE) with severe abdominal pain, which started the previous night. The patient stated that the pain has begun first in the epigastric area and later expanded through the left lower quadrant. Patient history revealed no previous illness or surgery. The patient recalled no information about having intestinal malrotation or situs inversus. On physical examination, rebound tenderness was observed in the lower quadrant more on left side. Laboratory tests, showed high CRP titer, and high leukocyte count, normal urinalysis, normal liver and kidney function tests, normal ECG. Based on the patient's clinical status, the ER doctor send him for abdominal ultrasonography for suspected diverticulitis or acute appendicitis.

Abdominal ultrasonography (USG) done first, We usually examine the upper abdominal organs first then the lower, the liver was located in left hypochondrium and spleen on right side suggesting Situs inversus which later confirmed by taken chest X-ray. The appendix was not visualized in right iliac fossa but a dilated non-compressible blind intestinal loop consistent with acute appendicitis was located in the left lower quadrant (**Figure 1(a)** & **Figure 1(b)**). Chest X-ray was taken 4 hours later after admission, revealed dextrocardia (**Figure 2**) and the patient was immediately taken to surgical suite and laparoscopic appendectomy was performed, the surgeon confirmed the diagnosis, the operative details was videotaped and kept in hospital records. The patient was going well, and discharged on the third postoperative day with no complications. Follow-up visits for him was unremarkable and went smoothly. The patient was informed of his condition for future medical consultation or surgical intervention.

## 3. Review of Literature about Left-Sided Appendicitis

PubMed and Google Scholar databases was reviewed (all articles from 1893 to July 2010) and 64 reports concerning 95 cases of LSAA meeting the above-mentioned criteria were included in this review [1]-[64]. The patients were aged from 8 to 82 years (mean:  $29.1 \pm 15.9$  years). Fifty-seven were male and 38 were female. Sixty-six patients had SIT, 23 had MM, 3 had cecal malrotation, in two the anomaly was previously unnoted and in one case, the end of the

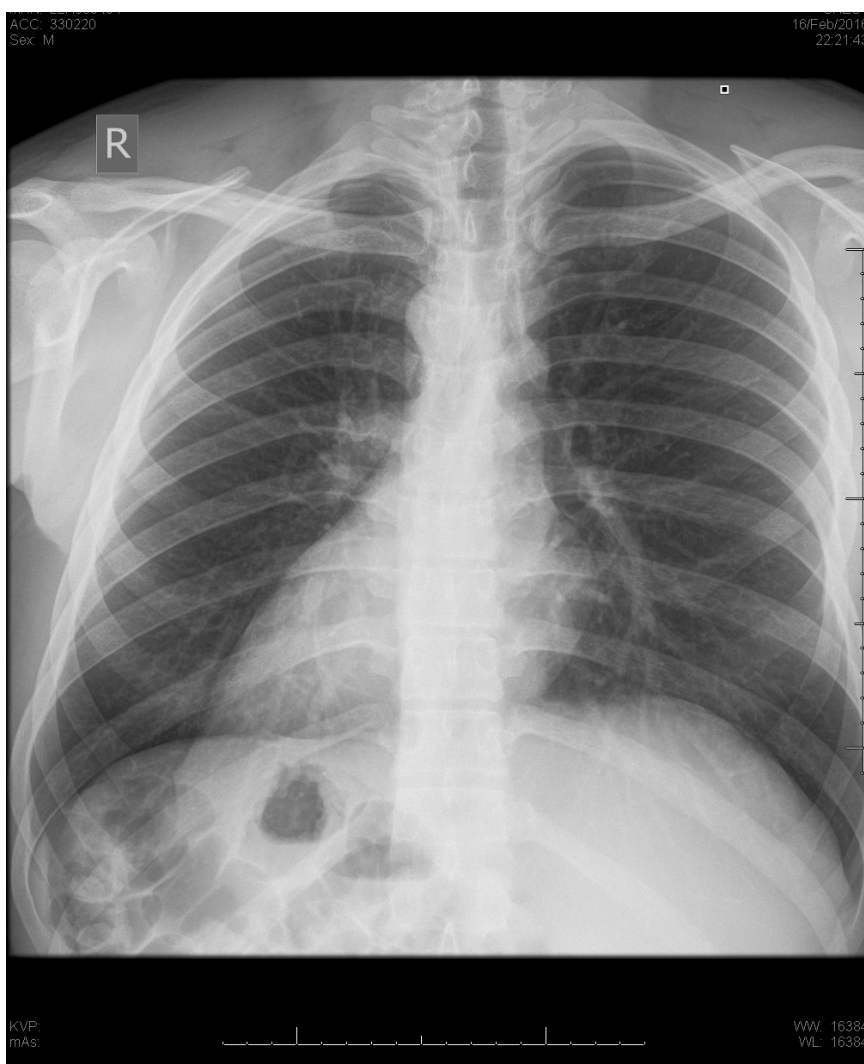


(a)



(b)

**Figure 1.** (a) and (b) Ultrasonography of abdomen and pelvis, showed situs inversus totalis including reversed position of liver and spleen. The right iliac fossa was examined, the appendix was not visualized but the left iliac fossa showed signs of acute appendicitis, in the form of distended fluid-filled appendix at 12.2 mm in diameter and seen non-compressible with thick wall, no periappendiceal fluid, and no appendicolith seen, the same patient name, the same ID and the same hospital were displayed on the 3 original images provided (all patient identity information will be removed in final version).



**Figure 2.** Chest X-ray was done 4 hours later for confirmation of dextrocardia.

appendix running along the anterior side of sacrum was found in the left side. According to localization of the symptoms, 59 patients presented with left and 14 with right lower quadrant pain, 7 with bilateral lower quadrant pain, 7 with left upper quadrant pain, 6 with peri-umbilical, and two presented with pelvic pain. With regard to the diagnosis, 49 patients were diagnosed with appendicitis during the pre-operative period, in 19 patients, the diagnosis was established intraoperatively and in 5 postoperatively; 14 patients were previously known to have SIT and/or MM. No information was available in eight patients. Of 95 patients included in this literature review, 13.6% (13 cases) of patients underwent Laparoscopic appendectomy [66].

#### 4. Discussion

It is important to differentiate LSAA associated with malrotation from that associated with situs inversus totalis, in which every organ, including duodenum, duodenojejunal junction, small and large bowel, cecum, and appendix, is located

in a mirror position to situs solitus [1]. In addition to these features, a left-sided liver and right-sided spleen and stomach serve as clues to the correct diagnosis of situs inversus totalis. Chest X-ray is important to obtain at this point for ruling out situs inversus totalis which may be confused with intestinal malrotation. More than two-thirds of the left-sided appendicitis is due to situs inversus totalis rather than intestinal malrotation [66].

MM is the term used to describe a spectrum of congenital positional anomalies of the intestine caused by nonrotation or incomplete rotation of the primitive loop around the axis of the superior mesenteric artery (SMA) during fetal life. Although about 80% of cases are diagnosed in patients younger than 1 month, malrotation has also been reported in adults [8]. The incidence of MM cited in the literature varies from 0.03% to 0.5% in live births [1] [3] [9] [10]. The situs inversus may be complete (SIT), when both thoracic and abdominal organs are transposed, or partial, when only one of those cavities is affected [1]. The incidence of SIT reported in the literature varies from 0.001% to 0.01% in the general population [12] [13] [14], whereas the incidence of acute appendicitis associated with SIT is reported to be between 0.016% and 0.024% [5] [13] [14].

In the literature, LSAA occurs between the age of 8 and 63 years and is 1.5-fold more frequent in men than in women [3] [13].

The differential diagnosis of left lower quadrant abdominal pain includes: diverticulitis, renal colic, ruptured ovarian cyst, Meckel's diverticulitis, epididymitis, incarcerated or strangulated hernia, bowel obstruction, regional enteritis, psoas abscess, and right- and left-sided appendicitis (LSAA) [1] [4].

LSAA is a diagnostic dilemma, because the appendix is located in an abnormal position. The differential diagnosis of LSAA may not be promptly established in the emergency setting and is often delayed due to lack of uniformity in the clinical signs [11] [58]. It is assumed that even though the viscera are transposed, the nervous system may not show the corresponding transposition, which may result in confusing symptoms and signs. In about 18.4% - 31% of patients with SIT and MM, the pain caused by LSAA has been reported in the right lower quadrant [1] [5] [11] [12] [13]. In this literature review, it was observed that 14.7% of patients had pain localized in the right lower quadrant, which indicates the importance of accurate preoperative diagnosis in order to avoid incorrect incision.

The diagnosis of acute appendicitis in patients with SIT or MM can be based on physical examination, electrocardiogram, chest X-ray, barium studies, USG, CT scan and diagnostic laparoscopy [1] [2].

Plain radiographs are usually not helpful for establishing the diagnosis of appendicitis. However, the detection of dextrocardia on chest X-ray and right-sided gastric bubble on abdominal plane X-ray is of considerable value in establishing the diagnosis of SIT, as demonstrated in our case the chest X-ray was conclusive. Barium enema with gastrografin can reveal MM or SIT, when there are difficulties in making the diagnosis of acute left lower quadrant pain [11]. Over the last

two decades, there has been an increasing use of imaging modalities, such as USG and CT, in the diagnosis of acute appendicitis, we didn't use enhanced CT scanning of the abdomen, because our case was straight-forward and because of insurance limitations for private hospital. USG is widely used in cases of appendicitis, however, it has significant limitations: it is operator-dependent, and examination of the lower quadrant can be compromised in patients with large body habitus or by overlying bowel gas. The value of CT in the diagnosis of acute appendicitis has been well-documented, with a reported accuracy of 90% - 98% [2] [13]. USG and CT may also be helpful in the detection of SIT and MM. Of the patients included in this literature review, CT has been used in the diagnosis of 28 patients and USG in 22 patients since 2000 [1] [6] [8] [17]-[24] [26] [27]. USG in our case was accurate in diagnosing situs inversus and in depicting non-complicated acute appendicitis because was not fat and bowel distension.

After establishing the diagnosis of SIT or MM, the surgical options are the same as for normal patients [1]. According to the reviewed literature, it was observed that many open and a few laparoscopic procedures have been performed [1] [6] [8] [15]. Laparoscopic appendectomy was first carried out in 1998 by Contini *et al.* [58] in a 34-year-old male patient with SIT. Since then, laparoscopic appendectomy has been performed in a total of 20 cases (12 with MM and 8 with SIT), of which two have undergone cholecystectomy at the same surgical session [3] [6] [7] [10] [12] [13] [15] [21] [24] [41] [58] [65]. Laparoscopy may be very useful both in establishing the differential diagnosis and in performing the definitive surgery [1].

As in patients with normally localized appendix, appendectomy specimens in LSAA should be sent for pathological evaluation. In the literature, only two of 95 patients (59 male, 76 female), who underwent appendectomy due to LSAA, were pathologically diagnosed with malignancy. Ascendent hemicolectomy was performed in both patients after pathological evaluation, which revealed mucinous adenocarcinoma and mucinous cystadenocarcinoma [19] [26]. In our case, the diagnosis was acute left sided appendicitis with no complications, no fluid collection, or abscess formation and successful endoscopic appendectomy was done, the caecum was located in left lower abdomen, the SIT was confirmed the surgery was videotaped and saved in hospital records, the pathological specimen showed signs of acute inflammation in the appendix. Ultrasonography before discharging the patient was unremarkable. The patient was doing well in his follow up visits.

## 5. Conclusion

LSAA should be considered in the differential diagnosis of young patients presenting with pain localized in the left lower quadrant. Chest X-ray, abdominal USG and CT provide very useful information. Endoscopic appendectomy in straight-forward cases and Diagnostic laparoscopy is the gold standard in cases with complicated differential diagnosis.

## Patient Consent for Publication

The patient consented for images or clinical information relating to his case to be reported in a medical publication.

## Conflicts of Interest

The author declares no competing interests in relation to their work.

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### **Abbreviation**

LSAA: Left Sided Acute Appendicitis  
SIT: Situs Inversus Totalis  
MM: Midgut Malrotation  
CT: Computed Tomography  
HSG: Ultrasonography  
UAE: United Arab Emirates