

CASE REPORTS

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# Complicated acute appendicitis in a child with left atrial isomerism: a case report



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## Abstract

**Background:** Heterotaxy syndrome is a rare clinical entity that is characterized by abnormal visceral organ arrangement in the chest and abdomen. Left atrial isomerism is a subcategory of heterotaxy syndrome characterized by the presence of multiple spleens with or without cardiac anomalies. Patients may remain asymptomatic their whole lives until they are diagnosed incidentally. Given that patients with left atrial isomerism might demonstrate atypical presentations of acute intrabdominal pathologies, it is important to keep a high index of suspicion when encountering such cases.

**Case presentation:** In this report, we present a case of a 9-year-old boy with a known history of left atrial isomerism who presented with left lower quadrant pain and positive McBurney, psoas, and obturator signs on the left side. A computed tomography scan of the abdomen confirmed the diagnosis of perforated acute appendicitis on the left, which prompted an emergent laparoscopic appendectomy.

**Conclusion:** Our case highlights the importance of keeping a high index of suspicion for a heterotaxy syndrome that is complicated by acute appendicitis in pediatric patients presenting with vague abdominal pain. Planning the location of trocar placement in patients with situs anomalies is of paramount importance to avoid technical difficulties in laparoscopic procedures.

**Keywords:** Left atrial isomerism, Polysplenia, Heterotaxy, Appendicitis

## Background

Heterotaxy syndrome is a rare condition in which the internal organs are abnormally arranged in the chest and abdomen. Unlike “situs inversus” in which the organs are completely flipped from right to left, heterotaxy syndrome is known as “situs ambiguous” in which only some of the organs are flipped. Individuals with this condition present with different birth defects and the severity of presentation depends on the specific organs involved; out of which cardiac and liver abnormalities have the most severe health outcomes [1]. This congenital syndrome is usually diagnosed early in childhood due to manifestation of severe cardiac anomalies. A subcategory of heterotaxy syndrome is the left atrial isomerism,

a condition characterized by an abnormal arrangement of thoracoabdominal organs and associated with the presence of multiple spleens and most patients die by age five due to the cardiac defects [2]. Around 5 to 10% of patients have normal hearts or mild cardiac defects, demonstrate no symptoms, and remain undiagnosed until imaging is obtained at a later stage in life for an unrelated reason [3]. We are reporting a case of a 9-year-old boy who presented to the Emergency Department (ED) with left lower quadrant abdominal pain and was found to have a perforated acute appendicitis in the setting of a prior diagnosis of left atrial isomerism.

## Case presentation

### History and workup

A 9-year-old boy weighing 45 kg with an asymptomatic left atrial isomerism, diagnosed in utero, and presented to the pediatric ED with a 2-day history of left lower

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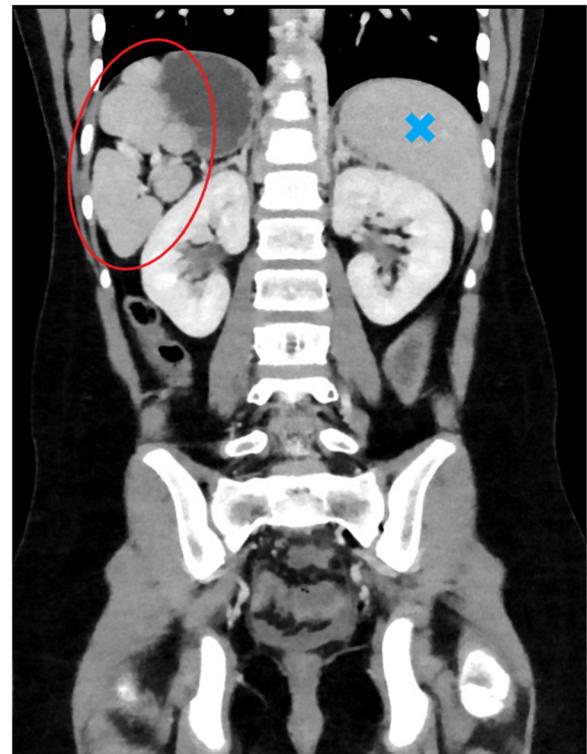
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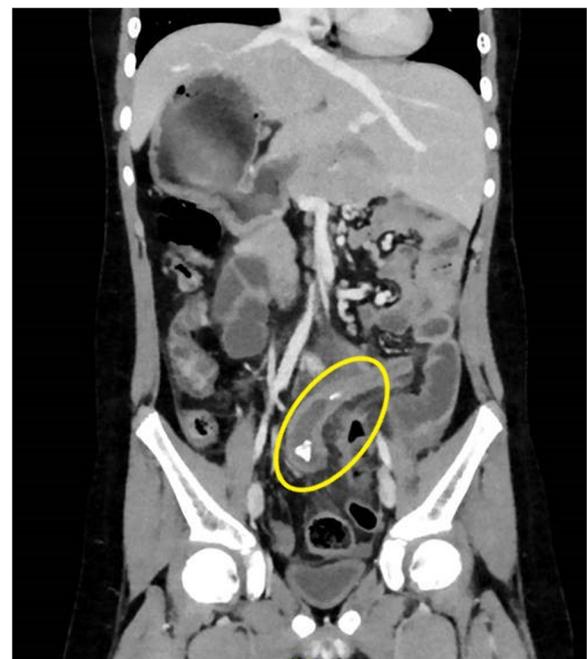
quadrant (LLQ) crampy abdominal pain not improving with antispasmodics (scopolamine butyl bromide). Pain was 8/10, radiating to his middle aspect of the abdomen, associated with nausea and four episodes of non-bloody, non-bilious vomiting as well as low-grade fever reaching 38°C, mild dysuria, and one episode of non-bloody, non-mucoid watery diarrhea the night prior to presentation. Upon presentation, the patient was not tolerating oral intake and was having difficulties ambulating on his left leg because of the abdominal pain. He denied any testicular pain or hematuria. Past medical history was negative for constipation; patient has had no previous surgeries. No history of symptoms suggestive of cardiac anomalies. No history of trauma or exposure to any toxic substance. Review of systems was otherwise negative. On arrival, his vital signs were stable with a blood pressure of 130/78 mmHg, oral temperature of 37.4°C, heart rate of 135, respiratory rate of 26, and oxygen saturation of 99%. On physical exam, the patient was pale and ill looking. Rebound tenderness was elicited in the LLQ and the suprapubic area. McBurney, psoas, and obturator signs were all positive on the left side. Costovertebral angle tenderness was negative on both sides. His initial complete blood count (CBC) showed significant leukocytosis with a left shift. Urine analysis was significant for pyuria with 8–10 WBC/HPF, 3+ ketones, and 1+ glucose. CT scan of the abdomen and pelvis with IV contrast was performed and confirmed the case of left atrial isomerism with intestinal malrotation and polysplenia located in the right upper abdominal quadrant (Fig. 1). The appendix base was located in the mid-lower abdomen with the tip extending to the LLQ. An obstructing appendicolith, with a perforation, was noted in the base of the appendix (Fig. 2). There were pockets of extraluminal air with features of peritonitis (Fig. 3).

### Management

Patient was given piperacillin/tazobactam 90 mg/kg for a broad-spectrum antibacterial coverage. Laparoscopic appendectomy was performed under general anesthesia using three-port technique; 12-mm umbilical trocar, and two 5-mm trocars in the right lower quadrant and the suprapubic region. Laparoscopic exploration confirmed the presence of the liver in the mid upper abdomen with extension to the left side and multiple small spleens in the right upper abdomen consistent with the patient's heterotaxy syndrome. The acutely inflamed appendix was identified in a retrocecal position in the mid lower abdomen; the appendix was perforated in its middle part with multiple patches of wet gangrene involving its middle and proximal portions up to its base. We found thick purulent peritoneal fluid in the lower abdomen bilaterally and in the pelvis, we also identified thick adhesions between the appendix and the surrounding bowels



**Fig. 1** CT scan of the abdomen with IV contrast demonstrating the polysplenia and left isomerism. Red circle: Polysplenia, located in the right upper abdominal quadrant. Blue cross: liver, located in the left upper abdominal quadrant



**Fig. 2** CT cut showing an obstructing appendicolith in the base of the perforated appendix. (Yellow circle)



**Fig. 3** CT cut illustrating the degree of inflammation around the appendicolith with extraluminal pockets of air. (Green circle)

that were diffusely dilated, and the peritoneum looked significantly inflamed and hyperaemic.

The appendix was freed using a combination of electrocautery and blunt dissection, it was then transected using endo-GIA; a small portion of the cecum was included with the specimen to ensure the excision of all gangrene and necrosis that was extending to the base of the appendix. A sample of the peritoneal fluid was sent for culture, and a Jackson-Pratt drain was inserted and kept in place for 5 days. Intravenous piperacillin/tazobactam was continued on the ward. Patient voided freely on day 0 postoperatively and passed flatus and stools on day 2. He was started on sips of water and ice chips on day 0 given the transient post-op ileus, and diet was advanced progressively on the subsequent days, which he tolerated very well. Intraoperative cultures grew *Escherichia coli extended-spectrum beta-lactamase* and *Enterococcus faecium*, so the patient was switched to meropenem and ampicillin for a total of 8 days.

## Discussion

Heterotaxy syndrome which illustrates an abnormal positioning of the visceral organs of the chest and abdomen is a particularly rare congenital entity with a prevalence of 1 in 10000 live births [1]. Although the exact etiology of this syndrome is still not well understood, it is believed to be due to altered embryogenesis caused by several internal and external influences, which may collectively contribute to the asymmetric anatomic patterning. Genetic factors play a role as it was found that the mode of inheritance follows an X-linked and autosomal recessive pattern, although some sporadic cases were reported in the literature [4, 5]. Patients with heterotaxy syndrome can be asplenic or polysplenic. Asplenic patients (right atrial isomerism) have a higher incidence of congenital heart disease and die at a very young age thus the earlier presentation and detection [6, 7].

In our case, the patient had a typical pattern of polysplenia heterotaxy syndrome with an additional vascular pattern associated with left atrial isomerism and an interrupted inferior vena cava with azygous continuation at the level of the right renal vein putting him also under the category of abernethy syndrome [8–10]. Although patients with heterotaxy syndrome who are polysplenic have a significant mortality rate close to 75% at 5 years of age, there is a lower incidence of cardiac defects compared to the asplenic counterpart [2]. However, some patients with left atrial isomerism might remain asymptomatic until their diagnosis is found incidentally by imaging done at a later stage in life [11].

Even though situs anomalies do not have a direct correlation with symptoms, the randomness of organ positioning can cause some confusion in the clinician's judgment especially in the setting of an abdominal infection or inflammation as the abdominal exam might be atypical given the location of the organs. That said, the clinical judgement of the physician complemented with the appropriate imaging modality is key for a prompt diagnosis in an unknown case of heterotaxy syndrome. In our case, the patient was already known to us to have the syndrome, so a probable left-sided complicated appendicitis was high on our differential. In fact, our patient was found to have sterile pyuria which is also a typical finding associated with acute appendicitis. Furthermore, some similar cases of acute uncomplicated appendicitis in the setting of left atrial isomerism were described in the literature. One must wonder whether such a syndrome predispose to developing appendicitis or that we tend to have a reporting bias by reporting more cases of appendicitis in such a syndrome [12, 13].

Laparoscopy remains the gold standard in situations where the diagnosis is uncertain. Not only can the location of the appendix be visualized, but the other organs can be inspected as well. There are no standard port positions in patients with situs anomalies and the surgeon has to modify the port placement adhering to the basic principles of ergonomics in laparoscopy. Most of the available literature describes the techniques of trocar placement in patients with situs anomalies in laparoscopic cholecystectomy [14] and bariatric procedures [15] but no available recommendations for laparoscopic appendectomy. In our case, we used a 12-mm supraumbilical port for the laparoscope, a 5-mm right lower quadrant port, and a 5-mm suprapubic port to pass the working instruments; this is the mirror image of our standard trocars positioning for appendectomy. In retrospect, we realized that while our choice of trocar placement was helpful to explore the left lower abdominal quadrant, it did not allow for an optimal exploration of the entire abdomen. We could have placed the working trocars in the right and left abdominal quadrants; this

may have provided a better triangulation and allowed for a wider angle for abdominal exploration including the main pathology that was more-or-less located in the mid-pelvis. That said, it was difficult to initially identify the exact location of the appendix because of the associated extensive inflammation.

Prophylactic removal of the appendix, in children with situs anomalies, eliminates any possibility of future misdiagnoses. It also excludes the risks of complications that come with delayed diagnosis, such as an appendicular rupture, which can cause infertility in young female patients. Song et. al concluded that patients with a history of situs inversus should undergo an appendectomy at the time of laparoscopy performed for other reasons [16].

## Conclusion

Diagnosing an acute appendicitis can be challenging to clinicians who encounter patients with atypical abdominal pain and with findings suggestive of incidental asymptomatic left atrial isomerism diagnosed on imaging. Thus, it is necessary to keep a high clinical suspicion in order to make prompt diagnosis for an efficient and appropriate plan of care. The surgeon must carefully plan the site of trocar placement, on a case-by-case basis, to create proper and suitable ergonomic arrangement and avoid technical difficulties.

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## Authors' contributions

FT was involved in the conception and design of the manuscript. FT, KWA, KA, AZ, and YA were involved in the literature search and manuscript writing. YA was responsible for the manuscript revision and listing of citations. The authors read and approved the final manuscript.

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## Availability of data and materials

Data is available upon request.

## Declarations

### Ethics approval and consent to participate

Not applicable. All personal information about the described case has been deidentified.

### Consent for publication

The decision to write the case report was taken months following the surgery, and hence verbal consent was obtained originally from the patient's parents over the phone. However, we currently have secured written consent from the parents.

### Competing interests

The authors declare that they have no competing interests.

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