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Left Upper-Quadrant Appendicitis in a Patient with Congenital Intestinal Malrotation and Polysplenia

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Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
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Conflict of interest: None declared

Patient: Female, 13
Final Diagnosis: Left upper quadrant appendicitis
Symptoms: Left upper quadrant abdominal pain
Medication: —
Clinical Procedure: Laparoscopic ladd's procedure
Specialty: Surgery

Objective: Congenital defects/diseases

Background: Appendicitis is the most common cause of abdominal pain requiring emergent surgical intervention. Although typically presenting as right lower-quadrant pain, in rare cases it may present as left upper-quadrant pain secondary to abnormal position due to intestinal malrotation. Since atypical presentations may result in diagnostic and management delay, increasing morbidity and mortality, accurate and prompt diagnosis is important. Therefore, acute appendicitis should be considered in the differential diagnosis of left upper-quadrant abdominal pain. In this setting, medical imaging plays a key role in diagnosis. We report a case of a 13-year-old female with undiagnosed intestinal malrotation presenting with left-sided acute appendicitis.

Case Report: A 13-year-old Hispanic female presented at the emergency room with anorexia and left upper-quadrant abdominal pain with involuntary guarding. The laboratory work-up was remarkable for elevated white blood cell count and elevated erythrocyte sedimentation rate. A nasogastric tube was placed and abdominal x-rays performed to rule-out bowel obstruction, showing distended bowel loops throughout all abdominal quadrants, with sigmoid and proximal rectal gas, raising concern for ileus rather than an obstructive pattern. Lack of symptomatic improvement prompted an IV contrast-enhanced abdominopelvic CT, revealing intestinal malrotation and with an inflamed left upper-quadrant appendix. Surgical management proceeded with a laparoscopic Ladd's procedure.

Conclusions: Acute appendicitis may present with atypical symptoms due to unusual appendix locations, such as in malrotation. Most cases are asymptomatic until development of acute complications, requiring imaging for diagnosis. Clinicians and radiologists should have a high index of suspicion and knowledge of its clinical presentations to achieve early diagnosis and intervention.

MeSH Keywords: Appendectomy • Appendicitis • Heterotaxy Syndrome

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/908276

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Background

Appendicitis is the most common cause of abdominal pain requiring emergent surgical intervention. Since delay in diagnosis increases morbidity and mortality, its diagnosis should be emergent. Usually, it presents with vague periumbilical abdominal pain that subsequently radiates to the right lower quadrant. One should be aware that approximately 33% of patients with acute appendicitis present with pain in locations other than the right upper quadrant, for which left-sided abdominal pain should raise suspicion for such diagnosis. Left-sided acute appendicitis is most commonly seen with congenital anomalies such as midgut malrotation and situs inversus totalis [1]. Its rarity may lead to a wrong diagnosis. False-negative diagnosis increases adverse outcomes such as appendiceal rupture or abscess formation. Despite radiological imaging advances, diagnosis of acute appendicitis remains a challenge. This case demonstrates the importance of recognizing appendicitis as a cause of left upper-quadrant pain.

Case Report

We present a case of a 13-year-old Hispanic female with past medical history of epilepsy, autism, and bilateral hip dysplasia, who presented at the emergency room with anorexia and left upper-quadrant abdominal pain. The patient had no prior abdominal surgical history. Physical examination revealed a depressible abdomen with epigastric tenderness and left upper-quadrant pain upon palpation, as well as involuntary guarding. Laboratory work-up was remarkable for elevated white blood cell count (13.4 with neutrophilic predominance of 10.60) and elevated erythrocyte sedimentation rate of 82. A nasogastric tube was placed and initial imaging was performed with supine antero-posterior and lateral cross-table abdominal x-rays to rule-out bowel obstruction. These showed distended small and large bowel loops throughout all abdominal quadrants, with sigmoid and proximal rectal gas, raising concern for ileus rather than an obstructive pattern (Figure 1). In view of the lack of symptomatic improvement, an IV contrast-enhanced abdominopelvic CT was performed, showing a non-obstructive gas pattern with a left upper-quadrant fluid-filled dilated appendix, with hyper-enhancing walls, surrounding inflammatory fat stranding, and multiple pericecal mesenteric lymph nodes (Figure 2). Additionally, there was inversion of the superior mesenteric artery (SMA) and superior mesenteric vein (SMV) relationship, with the SMA identified in the right (Figure 3), the small bowel located at the right hemi-abdomen, and the ascending colon at the left abdominal quadrant with the cecal tip at the left upper quadrant. Findings were consistent with midgut malrotation and acute, non-complicated left upper-quadrant appendicitis with associated reactive lymphadenopathy. Incidentally, multiple spleniculi were seen at the left upper quadrant (Figure 4), as well as a partial dorsal pancreatic agenesis (Figure 5). Emergent surgical intervention was performed with laparoscopic Ladd's procedure.

Discussion

Acute appendicitis is a common cause of surgical emergencies, with an incidence rate of approximately 1 in 400 in the

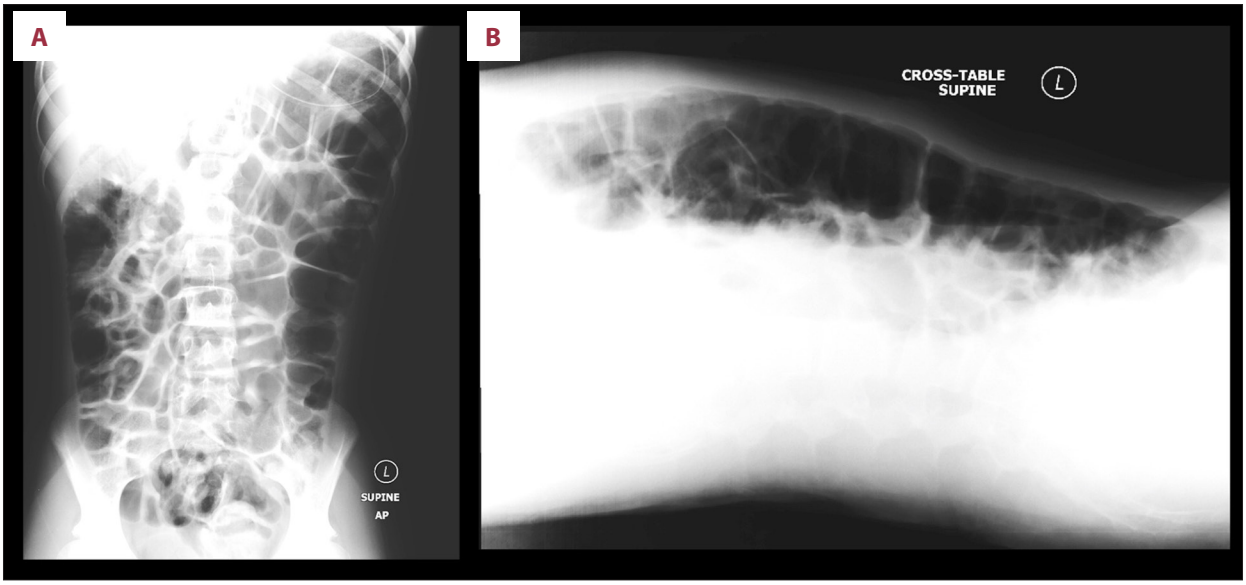

Figure 1. Supine antero-posterior (**A**) and lateral cross-table (**B**) abdominal x-rays show distended small and large bowel loops throughout all abdominal quadrants, with sigmoid and proximal rectal gas, raising concern for ileus rather than an obstructive pattern. A few nonspecific air fluid levels are also seen.

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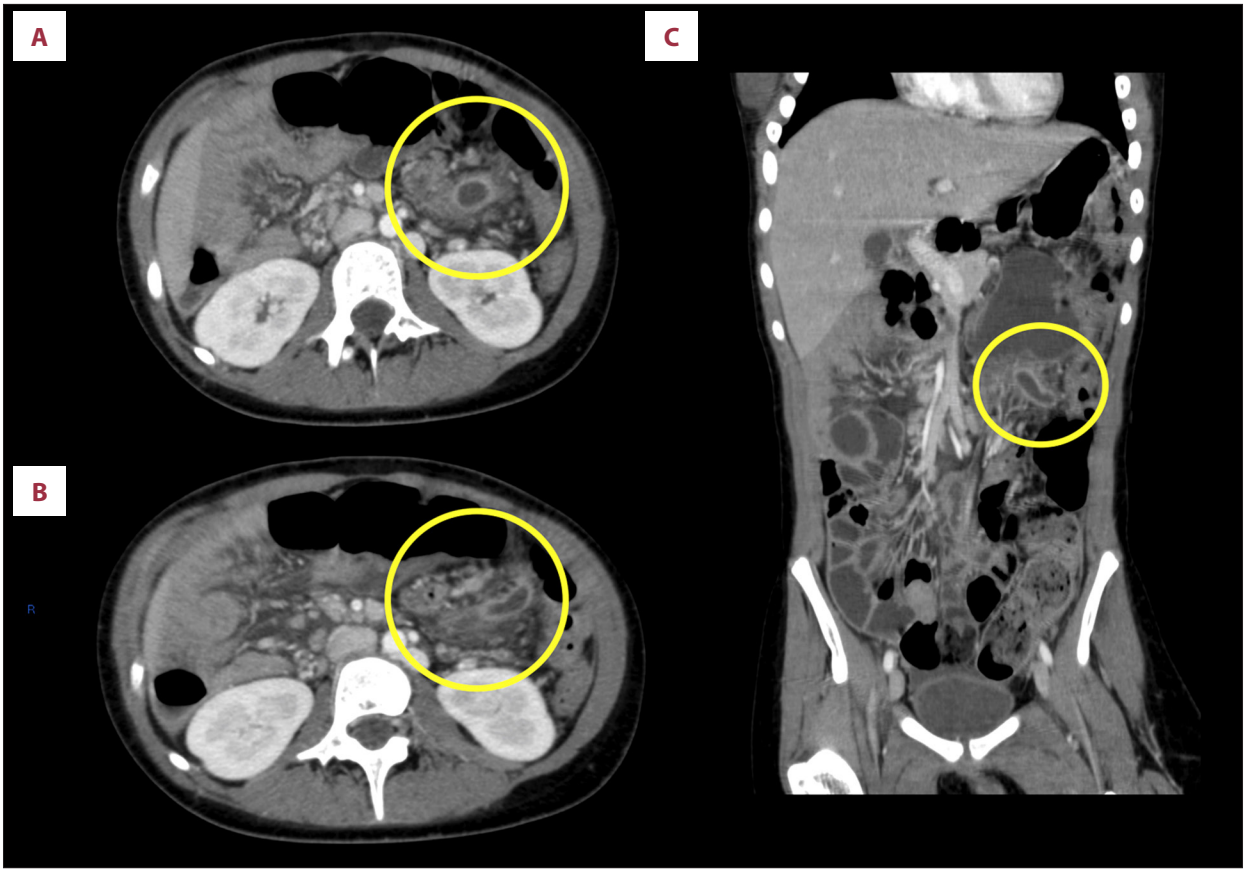

Figure 2. Axial (**A, B**) and coronal (**C**) Images of an IV contrast-enhanced abdominopelvic CT demonstrates a left upper-quadrant fluid-filled dilated appendix, with hyper-enhancing walls, surrounding inflammatory fat stranding, and multiple pericecal mesenteric lymph nodes.

United States. Usually, acute appendicitis is a straightforward clinical diagnosis, but sometimes may present with atypical symptoms, which may lead to a different diagnosis. Normally, the appendix is in the right iliac fossa, leading to right lower-abdominal pain when inflamed. During growth and development, the cecum undergoes changes in form and position, affecting its final anatomical location [2]. Since its position can vary considerably, appendiceal abdominal pain may manifest as an acute abdominal pain radiating to the left iliac fossa, sub-hepatic region, promontoric, pelvic, and splenic regions [2]. Congenital anomalies such as midgut malrotation also affect these atypical presentations, such as in our case of a 13-year-old female presenting with LUQ pain due to her congenital intestinal malrotation, which was diagnosed after performing CT imaging. Malrotation in adolescence and adulthood is usually not considered due to its nonspecific presentation, and the radiologist may be the first to encounter this important diagnosis as an incidental imaging finding or as the cause of acute abdominal symptoms [3].

Intestinal malrotation is a congenital anomalous rotation of the gut around the superior mesenteric artery axis during embryogenesis, resulting in the misplacement of the duodeno-jejunal junction to the right midline [1]. Even though its symptoms present early in life, some individuals (0.1–0.5%) progress to adulthood and remain asymptomatic [4]. However, it may also lead to complications such as midgut volvulus, or as in our case, to left-sided acute appendicitis. The best way to diagnose it is by CT imaging showing focal inflammation within the abdomen, and inversion in the SMA/SMV relationship with the SMA on the right and the SMV on the left, and large bowel predominantly on the left and small bowel predominantly on the right [4].

Additionally, as described in many studies, polysplenia (left-sided isomerism) may be seen in patients with intestinal malrotation or other intestinal anomalies, predominantly in female patients. Usually, patients die during their first years of life due to severe cardiac defects, but 5–10% will have normal heart or minor cardiac defects, reaching adulthood without complications [3,5]. Left isomerism tends to present later in life with abdominal complications, as is less associated with congenital heart disease. As seen in the literature, not all characteristics of this syndrome are present in all cases,

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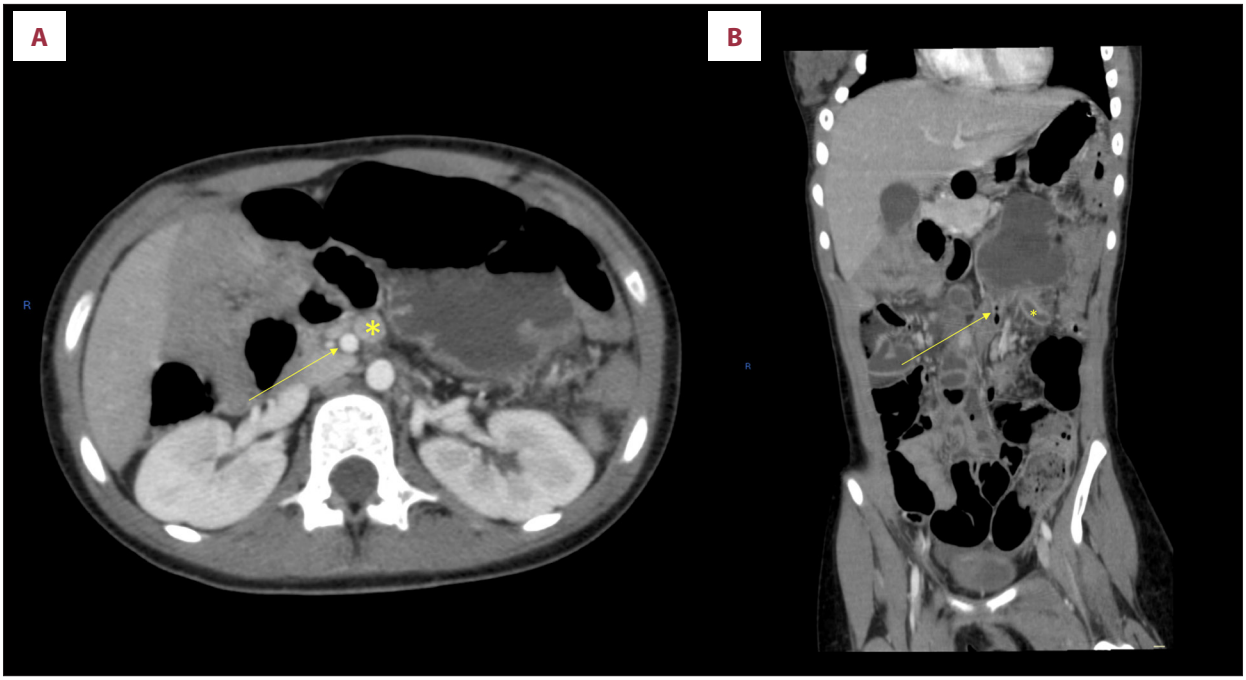

Figure 3. Axial (**A**) images of an IV contrast-enhanced abdominopelvic CT show inversion of the superior mesenteric artery (SMA arrow) and superior mesenteric vein (SMV*) relationship, with the SMA identified in the right. Coronal (**B**) images show a dilated cecum and ileocecal valve(arrow) with an inflamed appendix (*) at the left upper quadrant. The small bowel is located at the right hemi-abdomen.

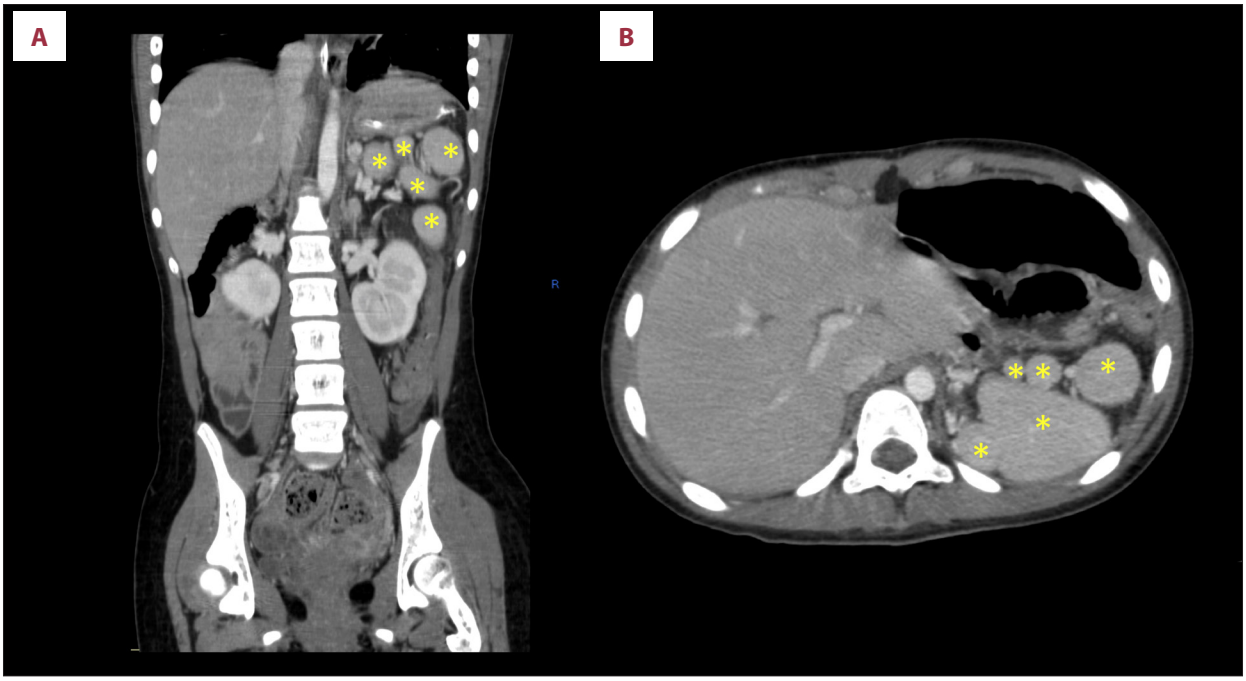

Figure 4. Coronal (**A**) and axial (**B**) images incidentally show multiple spleniculi (*) seen through the left upper quadrant.

which is why there is no single pathognomic abnormality to characterize this rare syndrome [6]. However, it may include multiple spleens, visceral heterotaxia, right-sided stomach, intestinal malrotation, short pancreas, preduodenal portal vein, and inferior vena cava anomalies [7]. The liver and gallbladder may be normal in shape and location or may be at midline. Unless they present for imaging, these patients will go undiagnosed [3]. Besides polysplenia and intestinal malrotation,

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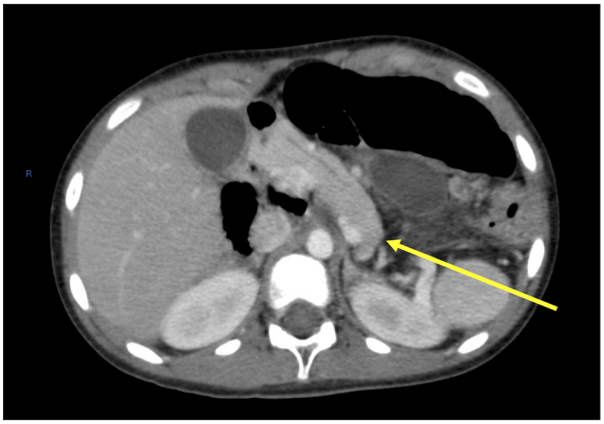

Figure 5. Short and truncated pancreas, as can be seen with partial dorsal pancreatic agenesis.

our patient had some of these CT findings, including a partial dorsal pancreatic agenesis "short pancreas", another common congenital defect that is associated with heterotaxy-linked polysplenia [8,9].

Heterotaxy syndrome is not clearly described in the literature. Burton et al. found variations in how heterotaxy-associated anomalies were described, diagnosed, and reported [10]. The definition, nomenclature, and classification of this syndrome and its presenting defects are not well established, but it has been described as an incomplete lateralization disorder that results in any arrangement of the viscera across the left-right axis that differs from its normal position. However, Burton et al. classified patients according to whether they had asplenia or polysplenia and at least 1 other known developmental association with heterotaxy and/or polysplenia syndrome. They found that only 55.6% of patients presented with total anomalous systemic venous return, and 48.9% had bilateral bilobed lungs [10]. Due to our inability to evaluate our patient's cardio-pulmonary anatomy, this syndrome cannot be ruled out. Thus, since our patient did not present with interrupted IVC anomalies, preduodenal portal vein, or other associated vascular anomalies, but presented with intestinal malrotation, multiple spleens, and partial agenesis of the dorsal pancreas, a milder presentation of polysplenia syndrome was suspected.

Despite controversies, surgical correction by Ladd's procedure continues to be the criterion standard for all operative candidates with malrotation. Laparoscopic Ladd's procedure seems to be as effective as the standard open Ladd's procedure [3,11]. It consists of complete division of all right-sided peritoneal bands, reduction of any noted volvulus, widening of the small bowel mesentery, and appendectomy with rearrangement of the small bowel on the right side of the peritoneal cavity and the colon on the left side. Long-term follow-up is recommended due to risks of new symptoms of midgut volvulus, or small bowel obstruction due to band adhesions [11]. Open versus laparoscopic approaches have been described but there is no consensus on which approach is best. However, the laparoscopic approach has been shown to be associated with shorter postoperative length of stay [12,13].

However, several other pathological conditions such as splenic abscess, peptic ulcer, colitis, and nephrolithiasis may present with LUQ pain. Thus, a high suspicion of this presentation is imperative to perform appropriate imaging and correctly diagnose acute appendicitis in such rare cases.

To the best of our knowledge, the most extensive review of left upper-quadrant appendicitis was performed in 2010, reporting 95 cases, in which there was an incidence of 24% associated with malrotation and 7.3% presented with left upper-quadrant pain. In our study, ages ranged from 8 to 82 years old (mean, 26.5±16.4 years), leading to the conclusion that malrotation is not exclusively diagnosed during infancy [1].

Conclusions

Acute appendicitis may present with atypical symptoms due to its numerous variants in anatomical position. Intestinal malrotation is a rare but important cause of left-sided acute appendicitis. Other congenital and associated vascular anomalies are rarely encountered and incidentally detected when patients are being evaluated for other reasons. Delay in diagnosis may be detrimental to the prognosis of these patients. Imaging is key to accurately diagnosing this condition, with helical computed tomography (CT scan) being the preferred modality. Therefore, radiologists must be aware of their existence and be able to recognize them to avoid surgical complications and major intraoperative injuries.

Conflict of interest

None.

References:

1. Akbulut S, Ulku A, Senol A et al: Left-sided appendicitis: Review of 95 published cases and a case report. World J Gastroenterol, 2010; 16(44): 5598–602
2. Banerjee A, Kumar IA, Tapadar A, Pranay M: Morphological variations in the anatomy of caecum and appendix – a cadaveric study. National Journal of Clinical Anatomy, 2012; 1: 30–35
3. Pickhardt PJ, Bahlla S: Intestinal malrotation in adolescents and adults: Spectrum of clinical and imaging features. Am J Roentgenol, 2002; 179(6): 1429–35
4. Ben Ely A, Gorelik N, Cohen-Sivan Y et al: Appendicitis in adults with incidental midgut malrotation: CT findings. Clin Radiol; 2013; 68(12): 1212–19

Indexed in: [PMC] [PubMed] [Emerging Sources Citation Index (ESCI)] [Web of Science by Clarivate]

5. Kim S, Lee YS, Jung JH: Anomalies of abdominal organs in polysplenia syndrome: Multidetector computed tomography findings. J Korean Soc Radiol, 2016; 74(2): 114–22
6. Fulcher AS, Turner MA: Abdominal manifestations of situs anomalies in adults. Radiographics, 2002; 22(6): 1439–56
7. Anand U, Chaudhary B, Priyadarshi RN, Kumar B: Polysplenia syndrome with preduodenal portal vein. Ann Gastroenterol, 2013; 26(2): 182
8. Varga I, Babala J, Kachlik D: Anatomic variations of the spleen: Current state of terminology, classification, and embryological background. Surg Radiol Anat, 2017 [Epub ahead of print]
9. Maier M, Wiesner W, Mengiardi B: Annular pancreas and agenesis of the dorsal pancreas in a patient with polysplenia syndrome. Am J Roentgenol, 2007; 188(2): W150–53
10. Burton EC, Olson M, Rooper L: Defects in Laterality with emphasis on heterotaxy syndromes with asplenia and polysplenia: An autopsy case series at a single institution. Pediatr Dev Pathol, 2016; 17(4): 250–64
11. Seymour NE, Andersen DK: Laparoscopic treatment of intestinal malrotation in adults. JSLS, 2005; 9(3): 298–301
12. Hunrington JT, Lopez JJ, Mahida JB et al: Comparing laparoscopic versus open Ladd's procedure in pediatric patients. J Pediatr Surg, 2017; 52(7): 1128–31
13. Yu DC, Thiagarajan RR, Laussen PC et al: Outcomes after the Ladd procedure in patients with heterotaxy syndrome, congenital heart disease, and intestinal malrotation. J Pediatr Surg, 2009; 44: 1089–95

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