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## Case Report

# Wandering spleen torsion in a patient with polysplenia syndrome ☆

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## ABSTRACT

Polysplenia Syndrome is a rare condition that refers to the presence of 2 or more spleens in association with other thoracoabdominal abnormalities. Here, we report a case of a 13-year-old girl who presented with acute lower abdominal pain and was diagnosed with polysplenia syndrome after obtaining a CT scan of her chest, abdomen and pelvis. Diagnostic imaging also revealed the presence of a wandering spleen hanging in the lower abdomen and upper pelvic cavity and showing signs of infarction. The patient underwent splenectomy afterward and splenic torsion was confirmed intraoperatively. To the best of our knowledge, this was the first reported case of wandering spleen torsion in a patient with polysplenia syndrome. Physicians should keep in mind the possibility of a wandering spleen torsion presenting in various locations when dealing with polysplenia syndrome patients complaining of abdominal pain.

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## Introduction

Heterotaxy is a very rare condition characterized by the abnormal arrangement of the thoracoabdominal organs resulting from a disruption in the normal left-right axis during early embryonic development [1]. Polysplenia syndrome (PSS) is a subtype of heterotaxy and refers to the presence of two or more spleens in association with other intra-abdominal and intra-thoracic abnormalities [2]. A wandering spleen is also a rare condition caused by an absence or underdevelopment of one or more of the ligaments that hold the spleen in its normal anatomical position in the left up-

per quadrant of the abdomen, increasing the risk of splenic torsion [3].

A wandering spleen in the setting of PSS has only been reported once [4], while splenic torsion in the context of PSS has so far been reported eight times in the English literature. Here, we present the first documented case of wandering spleen torsion in the setting of PSS.

## Case report

A 13-year-old girl presented with acute lower abdominal pain associated with nausea and fever. She had no recent history

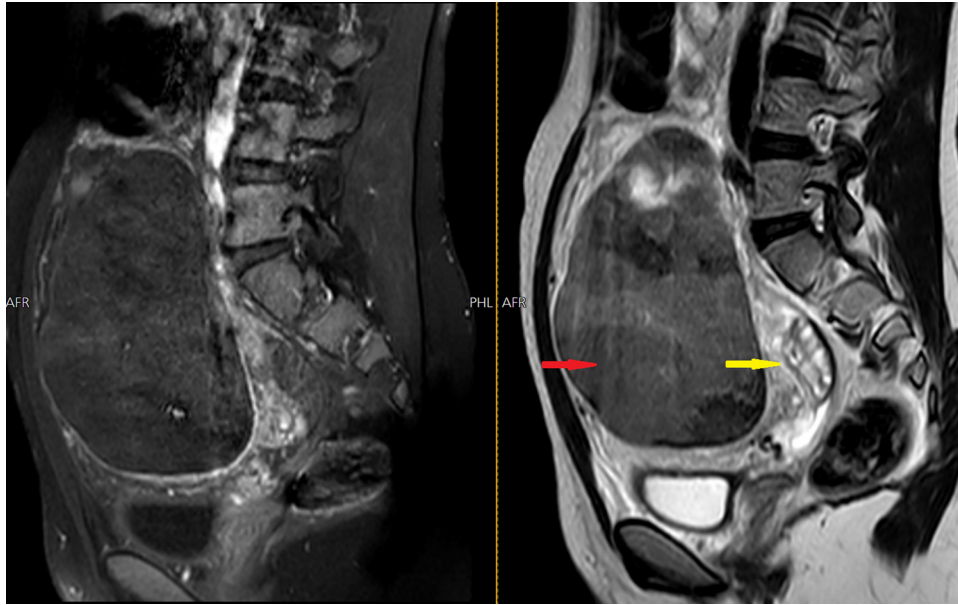
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**Fig. 1 – MRI obtained for suspected ovarian torsion. It shows a large mass (indicated by the red arrow) that has necrotic areas and that is clearly separate from the ovary (indicated by the yellow arrow).**

of trauma, and her past medical and family history was unremarkable. Abdominal examination revealed the presence of a tender lump in the right lower quadrant, but rebound tenderness was absent. Routine laboratory evaluation was normal except for an elevated lactate dehydrogenase level of 373. Ultrasonography of the abdomen and pelvis revealed 2 large hyperechoic masses with internal vascularity in the right side, while the right adnexa appeared to be normal. Neither the left adnexa nor the appendix could be visualized. Ovarian torsion was suspected, and so an MRI was ordered to confirm the diagnosis.

The MRI showed a large soft tissue mass (13 cm × 10 cm × 6 cm) located in the lower abdomen and upper pelvic cavity and was clearly separate from the ovaries and uterus (Fig. 1). It had a hilum, showed internal vascularization, had regular smooth margins, and showed no calcifications. It also showed heterogeneously low T1 and T2 WI signal intensity, and so wandering spleen torsion was suspected. Subsequently, a CT scan was performed to further clarify the previous findings and it demonstrated multiple spleens, along with other abnormalities typically seen in polysplenia syndrome (Fig. 2). The patient had bilobed lung fields bilaterally, her liver was enlarged and in mid-abdomen position, she had five spleens of variable size in the right side along with her stomach, she had intestinal malrotation, bicornuate uterus, and there was azygous continuation of the inferior vena cava. Moreover, the CT showed that the largest of the spleens did not demonstrate postcontrast enhancement (Fig. 3), and some abdominopelvic free fluid was seen as well, further validating the wandering spleen torsion diagnosis. An echocardiogram was then performed, but it showed no abnormalities.

Given these findings, it was decided to surgically remove the infarcted spleen. Laparotomy was performed under general anesthesia and through a lower midline incision, and

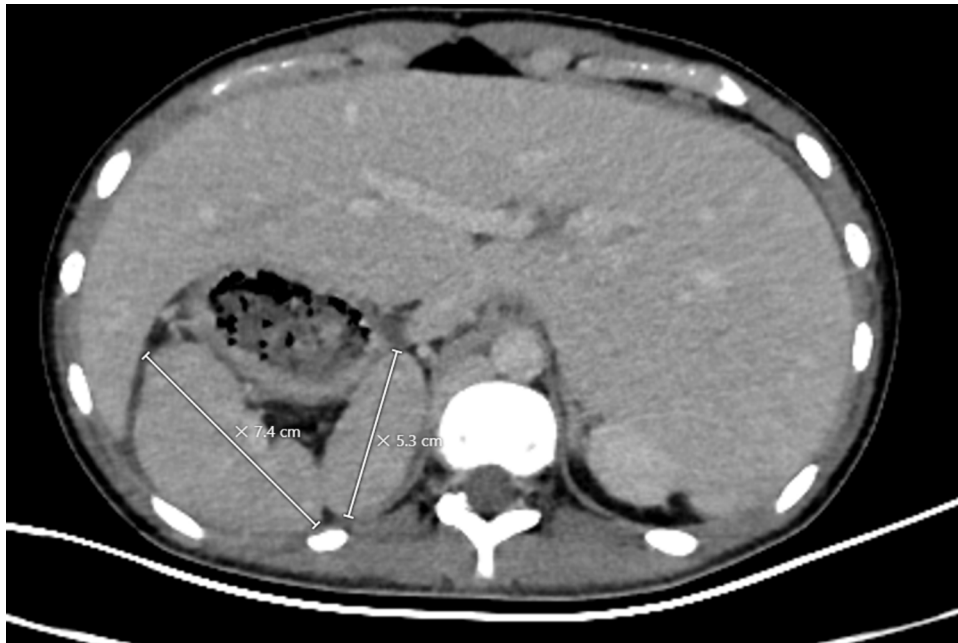
splenic infarction was seen intraoperatively in one of the five spleens. The rest of the spleens did not appear to be compromised and thus were left in place, while the infarcted spleen was excised (Fig. 4). Appendectomy was performed thereafter due to the abnormal position of the appendix in the left side of the abdomen as a result of intestinal malrotation. A pathology report was later produced and revealed normal appendiceal tissue and congested and hemorrhagic splenic tissue with multiple thrombosed blood vessels, confirming the splenic torsion diagnosis. The patient did not experience any postoperative complications and was discharged on the fourth postoperative day.

## Discussion

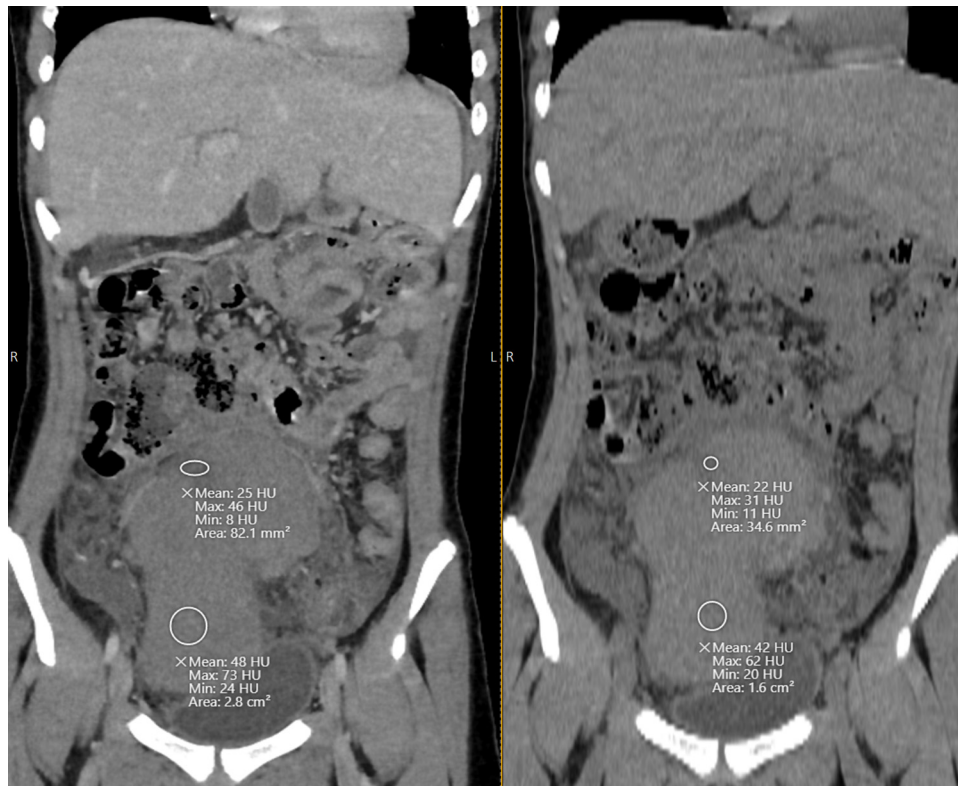
PSS was first described by Helwig in 1929. It is an extremely rare syndrome, with a reported incidence of just 1 per 250,000 live births [2]. In addition to having 2 or more spleens, PSS patients frequently have cardiac, pulmonary, gastrointestinal, and genitourinary anomalies [5]. The exact cause of PSS is still unknown. However, embryonic, genetic, and teratogenic components have all been implicated as causative factors [6].

Reports indicate that most PSS patients die before they reach five years of age, mostly due to cardiac defects. Nevertheless, about 5%-10% of patients might reach adulthood undiagnosed [5]. These patients might have their condition diagnosed incidentally or they might present with noncardiac complications, as in the case of our patient [7].

A wandering spleen is also a very rare condition, occurring at an incidence rate of less than 0.2% [8]. It is caused by an absence or underdevelopment of one or more of the ligaments that hold the spleen in its typical anatomical position in the



**Fig. 2 – Abdominal CT scan demonstrating PSS features. Two spleens can be seen on the right side along with the stomach, in addition to the liver being in mid-abdomen position.**



**Fig. 3 – Abdominal CT scan. The spleen did not demonstrate postcontrast enhancement.**



**Fig. 4 – Intraoperative photograph. It shows a congested and ischemic spleen (14 x 8 x 6 cm in size) after splenectomy.**

left upper quadrant of the abdomen, and so the spleen in these patients can be found in atypical abdominal or even pelvic positions. The laxity of the splenic ligaments and the abnormal length of the splenic vessels can lead to excessive splenic mobility, thus increasing the risk of splenic torsion [4].

Splenic torsion presents with nonspecific symptoms, such as abdominal pain, nausea, vomiting and fever, and so is often misdiagnosed with more common conditions, such as appendicitis and ovarian torsion. In our case, ovarian torsion was initially suspected, but the ultrasonography findings did not

**Table 1 – Previous case reports of splenic torsion in PSS patients.**

Author	Year	Age	Sex	Total number of spleens (number of infarcted spleens)	Treatment
Ackerman et al. [9]	1982	7 months	Female	3 (1)	Conservative
Griffiths et al. [10]	1984	23 years	Female	2 (2)	Laparotomy
Lachmann et al. [11]	2006	9 years	Female	Not mentioned (1)	Laparoscopy
Rasool et al. [12]	2011	2 days	Female	7 (1)	Laparotomy
Dash et al. [13]	2013	12 years	Male	5 (1)	Laparotomy
Fujiwara et al. [14]	2019	10 years	Female	2 (1)	Laparotomy
Kubo et al. [15]	2019	21 years	Female	3 (1)	Laparoscopy
Kennedy et al. [7]	2021	9 years	Female	7 (5)	Laparoscopy
Present case	2022	13 years	Female	5 (1)	Laparotomy

go well with the diagnosis. Obtaining MRI and CT imaging afterward was important in reaching the wandering spleen torsion diagnosis. Furthermore, obtaining a CT scan of the chest, abdomen and pelvis also allowed us to make the unexpected diagnosis of PSS. Preoperative imaging becomes even more important in the case of PSS given its abnormal abdominal anatomy, and so a careful assessment of visceral arrangement is needed before conducting the operation [7]. Additional considerations include obtaining an echocardiogram, as PSS is frequently associated with various cardiac anomalies, but our patient did not have any. Another consideration should be the administration of vaccines against encapsulated bacteria. However, since only one spleen had been removed while the others remained functional, there was no need for vaccination in our case.

Splenic torsion in the context of PSS has only been reported eight times in the English literature (Table 1). Wandering spleen torsion in the setting of PSS has never been reported before to the best of our knowledge, making this case the first of its kind.

Patients with PSS can present with various complaints. This case demonstrates the need to include splenic torsion in the differential diagnosis when dealing with this population, and since a wandering spleen is a possibility, physicians should still consider it even when the pain is in the lower abdomen. Having wandering spleen in mind can also facilitate the interpretation of imaging findings once the spleen is present in unexpected places, as this unexpectedness can be a great source of confusion for many physicians.

### Patient consent statement

Appropriate patient consent has been obtained for this case study.

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