

## Wandering spleen in the past ten years: A single-institution experience in Croatia

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### Abstract

In this article we present a rare pathology of the spleen, namely the wandering spleen. Wandering spleen is defined as a spleen without peritoneal attachments, only attached with its pedicle. Wandering spleen is a rare condition in which spleen is located anywhere in the abdomen. The syndrome may be diagnosed during infancy or childhood, but it is not infrequently an incidental finding in adults.

In the international literature, there are two methods of treating ectopic spleen - splenopexy and splenectomy. If there are no associated anomalies, surgical treatment of excess spleen is necessary in case of complications, such as torsion or places where there is a possibility of common trauma.

We report our three Croatian patients with wandering spleen, and in all of these patients we performed splenectomy. All the three patients had recurrent pains in the lower parts of abdomen. In all patients we found torsion of the ectopic spleen. In one of the patients we performed open splenectomy, in another one we conducted laparoscopy splenectomy, whereas in the other patient we tried to do laparoscopic surgery, which we completed with conversion.

Wandering spleen is an uncommon problem with quite rare clinical presentations. Methods of treatment depend on clinical presentation and findings at operative surgery.

**Keywords:** children, Croatia, surgical procedure, wandering spleen.

## Introduction

The spleen is almost entirely surrounded by peritoneum, which is firmly adherent to its capsule. It is held in position by two folds of this membrane. The spleen is attached with suspensory ligaments - phrenicolienal ligament, gastrolienal ligament and phrenicocolic ligament (1).

Wandering spleen is a rare condition in which the spleen is located anywhere in the abdominal cavity other than in its usual place. Abnormal location of the spleen may be attributed to malformation or total agenesis of the splenic suspensory ligaments (2,3).

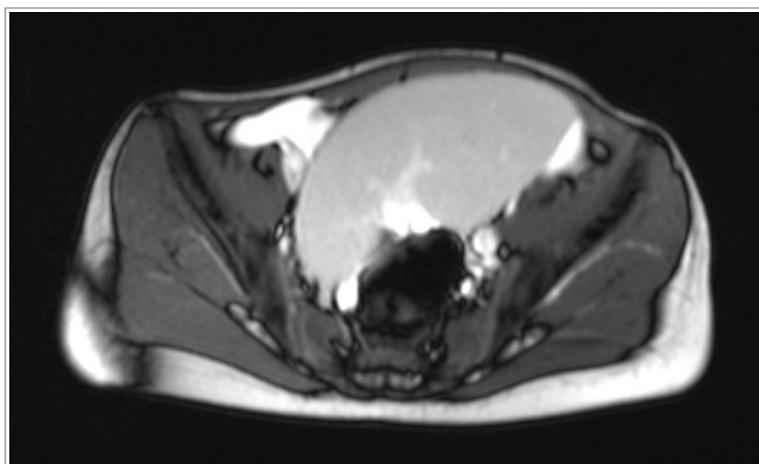
Polysplenia is a very rare entity and typical feature of this syndrome is the existence of the multiple small spleens in the abdominal cavity with absence of the normally located spleen (4). Polysplenia is rarely encountered in the elderly people because a number of associated congenital anomalies involving the cardiovascular system and the viscera require surgical treatment in early childhood. The differential diagnosis of these intra-abdominal masses can sometimes be difficult and can be confused with lymphadenopathy or metastases (5,6). Only 5%-10% of infants may have no associated anomalies (7). The diagnosis of polysplenia can be established in most instances by  $^{99}\text{Tc}^m$  labelled heat-denatured red blood cell selective spleen scintigraphy without biopsy (8).

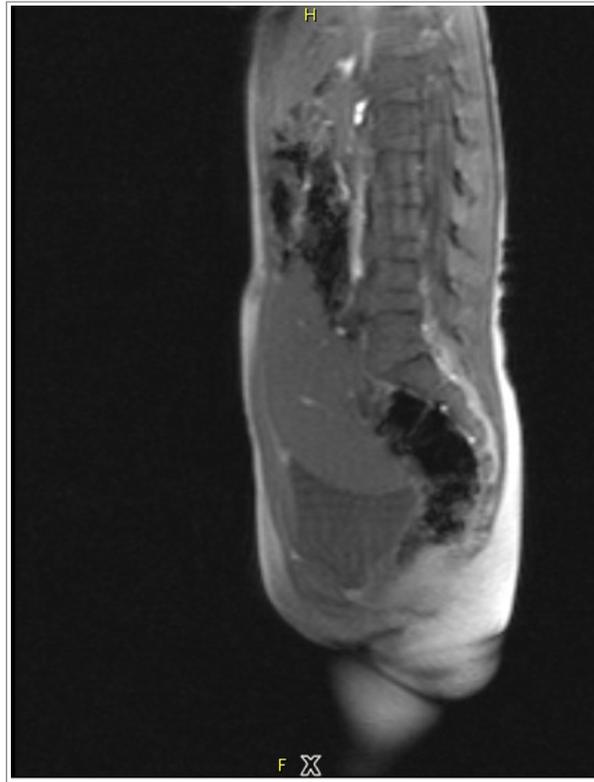
In this article we report three cases of wandering spleen, one of whom presented with torsion. One

patient had polysplenia without associated anomalies. In all three children we had to perform splenectomy. Splenectomy was performed in all three children for spleen torsion.

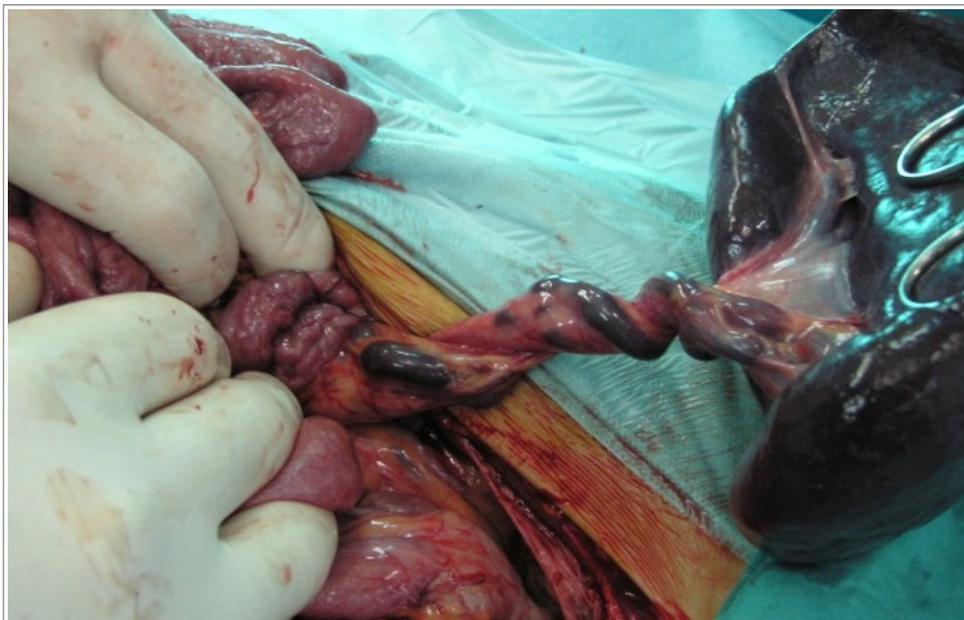
In the last ten years we have had in our hospital in Zagreb, Croatia, only three patients who exhibited a wandering spleen. The first patient was an eight years old girl who showed up at the surgical clinic complaining of pain in the abdomen that was most pronounced in the middle section below the umbilicus. In the left lower abdomen, it was palpable a lesion that was hard, inert and slightly sensitive. Abdominal ultrasound examination showed the ectopic spleen, which was located in the lower left abdomen, mostly in the pelvis behind the urinary bladder. Spleen size was 15x7 cm. Ultrasound examination showed ectopic spleen, but for further treatment we needed a more detailed analysis of the vascularization of the spleen and its blood supply. Magnetic resonance provided a better assessment of the vascularity of the spleen and localization (Figure 1 and Figure 2). We performed also CT angiography, which did not show pathological blood supply of the spleen. Instead, splenic blood vessels had a starting point on the anatomical location, but they were elongated and thrombosed. Upon MRI, there was evidence of splenic infarction, which was established as a consequence of acute torsion of the blood vessels.

Figure 1. MR-spleen in the pelvic



**Figure 2. MR-spleen in the pelvis**

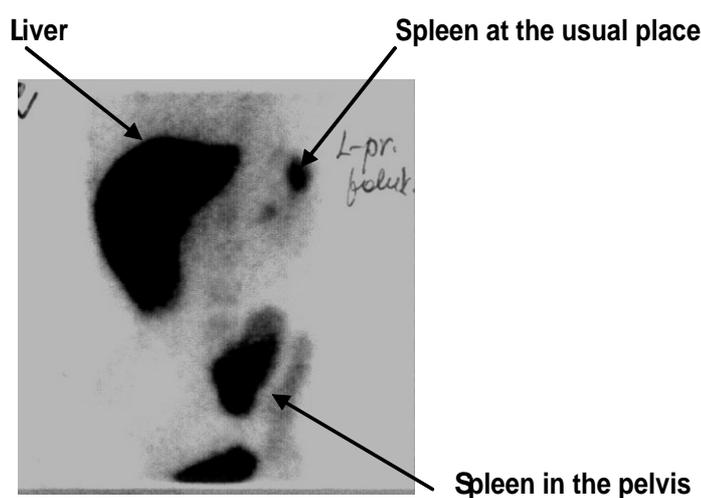
A surgical intervention was indicated. We found a large and dark spleen in the left lower part of the abdomen. The spleen was on the long vascular loop, which was rotated for 9000 [5x1800] (Figure 3).

**Figure 3. Torsion of the spleen vessels with infarction of spleen**

After derotation of the spleen and vascular pedicle, we noticed thrombosis of blood vessels and damage of the spleen tissue. Therefore, we indicated splenectomy. Splenectomy was necessary because

after derotation we noticed vascular thrombosis and splenic infarction. Upon abdominal exploration, we noticed an accessory spleen with a diameter about 2 cm that was not surgically removed (Figure 4).

**Figure 4. Scintigraphy with ectopic spleen**



The second patient was a ten year-old boy who came to our hospital for frequent and recurrent pain. During hospitalization, a routine abdominal ultrasound was done and we found in the normal position parenchymatous organ, which was about 5 cm in diameter. In pelvis on the left side, we found other parenchymal lesions and immediately we thought that it was a multiple spleen. We thought that it could be a polysplenia. Immediately, we started with the diagnosis of the patient. Polysplenia is a congenital disease manifested by multiple small spleens with a normally located spleen and very frequent associations with congenital anomalies. We did ultrasound examination, cardiac and pulmological examination and genetic analysis. Also, the patient underwent abdominal CT and scintigraphy with Tc-99m. We did not find any associated anomaly. Only 5%-10% of infants may have no associated anomalies. We indicated operative treatment. We tried to do laparoscopic exploration of the abdomen. We wanted to perform laparoscopic splenectomy, for a spleen which was located in the pelvis. In the

normal position of the spleen, we found the spleen tissue in a diameter of about 5 cm. Another spleen was located in the pelvis below the bifurcation of the aorta, which was the size of about 8 cm in diameter. The spleen in the pelvis did not have a normal circulation, with blood supply from radicular mesentery and intestinal circulation. Because of the disturbed anatomical relationships and pathologic circulation of the spleen, we had to do the conversion and perform an open splenectomy. The surgical procedure and postoperative course were uneventful (Figure 4).

The third patient was a six year-old boy who was admitted for frequent and occasional pains in the abdomen. On several occasions in the last two years, he had been inspected by a competent pediatrician, but no detailed analysis was performed. In this painful attack the boy was vomiting frequently, and the pain did not stop. Immediately, we made abdominal US and CT and noticed a large ectopic spleen in the pelvis on the left side with the absence of blood circulation through the spleen. It was necessary to perform an urgent

surgical intervention and the spleen was found on the long vascular pedicle, which was not rotated but vein and artery were thrombosed. It was indicated an operative treatment and laparoscopy splenectomy was done.

## Discussion

Wandering spleen is an unusual entity, which usually occurs in children and another peak is between 20-40 years of age in both sexes. Clinical diagnosis is difficult due to lack of the symptoms, unless splenic torsion has occurred. The increased mobility of the spleen results from absence or laxity of supporting ligaments (gastrosplenic and splenorenal ligaments) that normally anchor the spleen in its normal position (9). The first successful splenectomy for the wandering spleen was done in 1878 by Martin and marked the beginning of surgical treatment for this condition (10). In recent years, the laparoscopic splenectomy has been employed successfully because this kind of operative treatment offers more benefits. In our hospital in Zagreb, Croatia, we are also doing laparoscopic splenectomy whenever possible. In the case of rotation of the vascular pedicle we always make open laparotomy and splenectomy (11,12).

Sometimes, one can find an accessory spleen. The accessory spleen is an embryological anomaly found in about 16% of the children that undergo splenectomy for different reasons. In adults, an accessory spleen is located among 10%-30% of splenectomized patients. The accessory spleen is a smaller part of the spleen tissue outside the spleen, most

often in the hilus and close to the pancreatic tail, along the upper and lower part of the pancreas, the large curvature of the stomach, in the great omentum, and rarely can be present anywhere in the abdominal cavity. The accessory spleen is quite often difficult to observe, because it may be similar to the enlarged lymph nodes. In splenectomized patients, the accessory spleen can be enlarged and it may take a larger part of the function of the spleen. Therefore, it must be conducted a very careful and thorough search of the abdominal cavity after splenectomy for hypersplenism to prevent recurrence in patients who undergo surgery (13).

The splenectomized patient should be vaccinated to decrease the risk of overwhelming postsplenectomy sepsis (OPSS) due to organisms such as *Streptococcus pneumoniae*, *Haemophilus influenzae* type B, and *Neisseria meningitidis*. The vaccine against these microorganisms should be given two weeks before the elective splenectomy, or immediately after an emergency splenectomy. It is also necessary to carry out antibiotic prophylaxis with penicillin preparations especially in children younger than five years (14-16).

In conclusion, the wandering spleen is an uncommon condition with quite rare clinical presentations. For diagnosis, ultrasound is the initial choice of examination, but a CT scan and liver-spleen radionuclide scan are excellent methods when diagnosis remains questionable. When wandering spleen is diagnosed, splenopexy is the method of choice in asymptomatic and symptomatic patients without necrosis. If splenic necrosis is present, splenectomy is required.

**Conflicts of interest:** None declared.

## References

1. Jalšovec D. Systematic and topographical human anatomy [Croatian]. Zagreb: Školska KNJIGA, 2005:210-1.
2. Fiquet-Francois C, Belouadah M, Ludot H, Defauw B, Mcheik JN, Bonnet JP, et al. Wandering spleen in children: multicenter retrospective study. *J Pediatr Surg* 2010;45:1519-24.
3. Di Crosta I, Inserra A, Gil CP, Pisani M, Ponticelli A. Abdominal pain and wandering spleen in young children: the importance of an early diagnosis. *J Pediatr Surg* 2009;44:1446-9.
4. Lachmann R, Loff S, Düber C, Neff KW. Visceral heterotaxia with polysplenia syndrome and haemorrhagic splenic

- infarction as a rare cause of the acute paediatric abdomen. *Pediatr Radiol* 2006;36:572-3.
5. Gupta NA, Abramowsky C, Pillen T, Redd D, Fasola C, Heffron T, et al. Pediatric hepatopulmonary syndrome is seen with polysplenia/ interrupted inferior vena cava and without cirrhosis. *Liver Transpl* 2007;13:680-6.
  6. Gürses D, Ülger Z, Levent E, Ozyürek AR. A very rare case of polysplenia syndrome with congenital diffuse pulmonary arteriovenous fistulas. *Turk J Pediatr* 2006;48:96-9.
  7. Varga I, Galfiova P, Adamkov M, Danisovic L, Polak S, Kubikova E, et al. Congenital anomalies of the spleen from an embryological point of view. *Med Sci Monit* 2009;15:RA269-76.
  8. Abut E, Akkaya L, Uysal U, Arman A, Güveli H, Bölükbas C, et al. Selective spleen scintigraphy in the diagnosis of polysplenia syndrome. *Br J Radiol* 2004;77:698-700.
  9. Krmpotić NJ, Marušić A. *Human Anatomy [Croatian]*. Zagreb, Medicinska naklada, Sveučilišni udžbenici, 2007.
  10. Martin A. A successful case of splenectomy. *Br J Med* 1878;61:191-2.
  11. Carmona J, Lugo VH. Laparoscopic splenectomy for infarcted splenoptosis in a child: a case report. *Bol Asoc Med P R* 2010;102:47-9.
  12. Rescorla FJ, West KW, Engum SA, Grosfeld JL. Laparoscopic splenic procedures in children: experience in 231 children. *Ann Surg* 2007;246:683-8.
  13. Impellizzeri P, Montalto AS, Borruto FA, Antonuccio P, Scalfari G, Arena F, et al. Accessory spleen torsion: rare cause of acute abdomen in children and review of literature. *J Pediatr Surg* 2009;44:e15-8.
  14. Webb CW, Crowell K, Cravens D. Clinical inquiries. Which vaccinations are indicated after splenectomy? *J Fam Pract* 2006;55:711-2.
  15. Aguilar RB, Keister KJ, Russell AC. Prevention of sepsis after splenectomy. *Dimens Crit Care Nurs* 2010;29:65-8.
  16. Coignard-Biehler H, Lanternier F, de Montalembert M, Mahlaoui N, Suarez F, Lecuit M, et al. Infections in splenectomized patient. *Rev Prat* 2008;58:2209-14.