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## Case Report

# Polysplenia syndrome with situs ambiguous, common mesentery, and IVC interruption discovered incidentally in an adult

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### ABSTRACT

Polysplenia syndrome associates multiple spleens to other malformations, most frequently cardiac, vascular, visceral, and biliary malformations. Polysplenia has been described mainly in childhood owing to critical anatomic malformations related to cardiac defects or biliary atresia. We present a case of polysplenia syndrome found in a 58-year-old man. Only those with mild anatomical abnormalities reach adulthood without a diagnosis. Radiological examinations are helpful to the diagnosis by ascertaining the location and number of spleens, location of other organs in the chest and abdomen, and identification of other associated anomalies.

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## Introduction

Most thoracic and abdominal organs are asymmetric and lateralized. Situs solitus refers to the usual arrangement of those organs. Situs inversus totalis refers to exactly mirror-image of the usual arrangement. On the other hand, situs ambiguous or heterotaxy is abnormal arrangement of organs and vessels as opposed to the orderly arrangement typical of situs solitus and situs inversus [1]. Heterotaxy is generally classified into 2 major syndromes, polysplenia and asplenia syndromes [1,2].

Polysplenia syndrome is a rare congenital subtype of heterotaxia syndrome associated with various visceral and vascular abnormalities. It is characterized by presence of 2 or more spleens and anomalies of other asymmetric organs [1].

Technical advances in sonography, computed tomography, and magnetic resonance imaging have greatly enhanced our ability to detect and characterize these anomalies [2].

## Case report

A 56-year-old male patient was admitted in our radiology department with a history of hemolytic anemia and vaguely located abdominal pain. On examination, he has a pale skin and a normal abdominal examination.

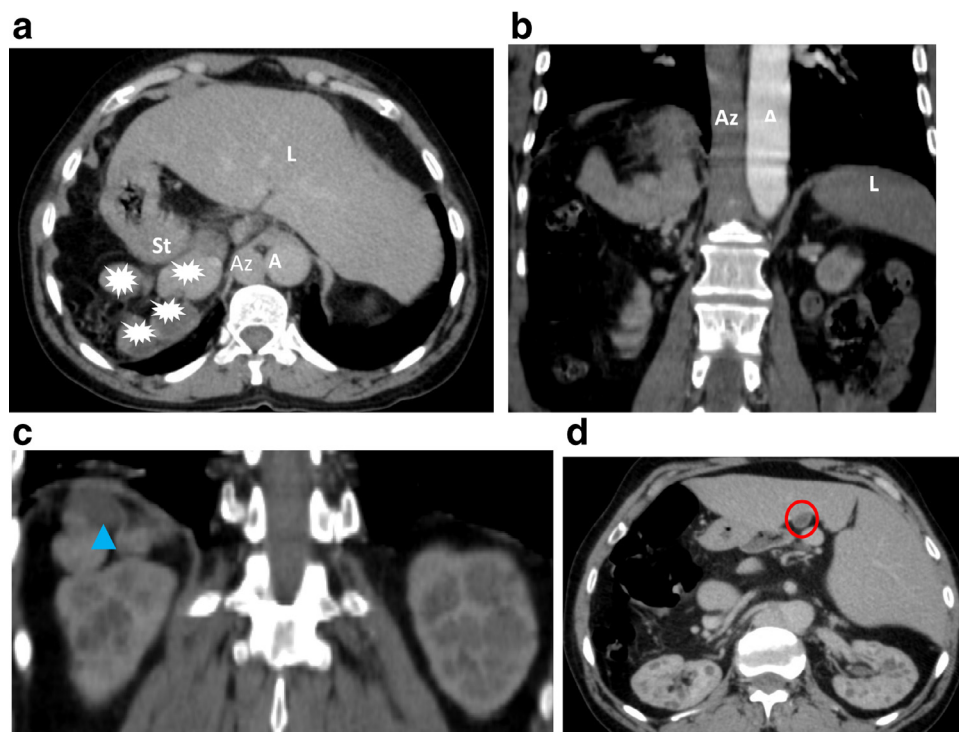
Ultrasonography was first performed by his family doctor showing cystic formations in the right hypochondriac region. Then a computed tomography was performed showing

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**Fig. 1 – Polysplenia syndrome in an adult with situs ambiguous, common mesentery, and IVC interruption. (a) Axial CT image of the abdomen at a upper level showing the liver (L) mainly on the left side with. The spleens (asterisks) are on the right side, the same side of the stomach (St). (b) Coronal CT image: the suprarenal segment of the IVC is absent with enlarged azygous vein (Az) on the right side. The descending aorta (A) is on the left. (c) Coronal CT image showing a cystic formation on one of the spleens (triangle). (d) Axial CT image of the abdomen showing multiple renal cysts. Note that the gall bladder (circle) is located on the left.**

the evidence of liver, gallbladder, and common bile duct on the left side of the abdomen. Stomach and multiples spleens (6 in total) were on the right side. It has also showed a common mesentery with a reversed incomplete intestinal rotation. An Inferior vena cava (IVC) interruption with azygous continuation has also been with a normal atrial situs and levocardia. The 2 cystic formations discovered on the ultrasonographic exam belonged to the multiple spleen on the CT scan. The diagnosis of polysplenia syndrome was made. In addition to the discovery of this entity in the adulthood, the peculiarity of our case is that splenic cysts were never been reported in the literature as part of polysplenia syndrome.

## Discussion

Polysplenia syndrome refers to the association of 2 or more multiple spleens with multiple congenital abnormalities in abdomen and chest. However, some cases of polysplenia syndrome have been described with a single bilobed spleen or a single normal splenic gland. In all cases, the spleens are still located on the same side of the stomach along the greater curvature due to the development of embryonic splenic tissue in the posterior mesogastrum (future gastrosplenic ligament) [1].

It is a rare congenital disease initially described by Helwig in 1929 [2,3]. Since then, few cases were described in the literature with an incidence of 1/250,000 live births [4].

The exact cause of polysplenia has not been clearly defined. Embryonic, genetic, and teratogenic components have all been implicated as causative factors in polysplenia [5].

Symptomatic patients presented with atrioventricular septal defects and may have anomalous of the inferior vena cava or other gastrointestinal structural abnormalities including partial or complete agenesis of the dorsal pancreas. These findings may have clinical relevance with a possible increased risk for intestinal volvulus, diabetes mellitus, or pancreatitis. In asymptomatic patients, this congenital anomaly may be found incidentally during abdominal surgery or radiological examination [6].

## Spleens

The splenic mass is usually divided into fairly equalized masses, varying in number from 2 to 6 and ranging from 1 to 6 cm in diameter. The location of the spleens is in either the left or right upper quadrant, along the greater curvature of the stomach [6]. Splenic cysts were never reported in the literature.

## Venous anomalies

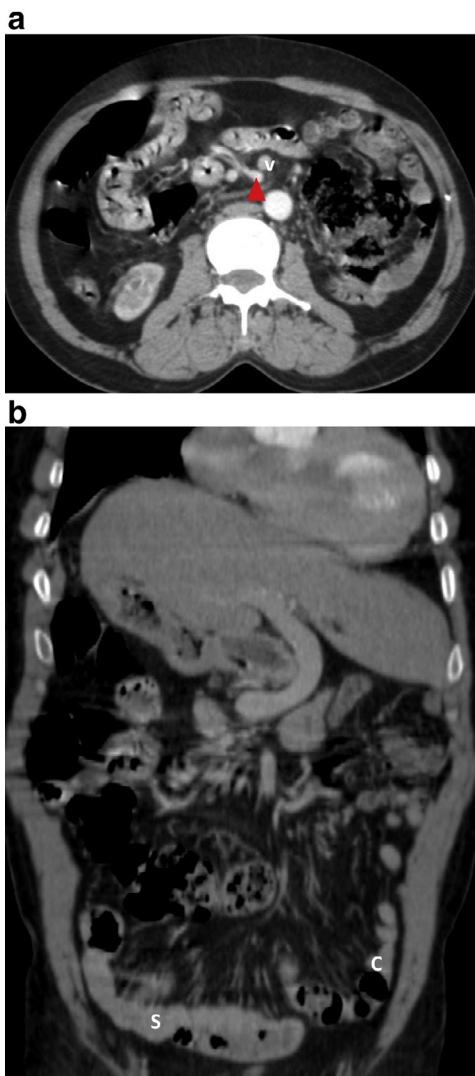
Inferior vena cava interruption with azygous continuation is the second most common abnormality observed in polysplenia patients after multiple spleens [1]. To understand this anomaly, it should be noted that the embryologic development of IVC is complex, and the normal IVC is composed of 4 segments: hepatic, suprarenal, renal, and infrarenal. The hepatic segment develops from the vitelline vein. The suprarenal segment is formed by the right subcardinal vein and subcardinal-hepatic anastomosis. The renal segment develops from the right supra-subcardinal and postsubcardinal anastomoses. The infrarenal segment derives from the right supracardinal vein [7]. Interruption of the IVC results from failure of the right subcardinal-hepatic anastomosis with consequent atrophy of the subcardinal vein (suprarenal IVC) and continuation of the infrarenal IVC as the azygous vein [7]. The suprahepatic segment of the IVC is usually present and drains separately into the right atrium [8].

## Intestinal malrotation

Polysplenia is commonly associated with anomalies of intestinal rotation, especially in patients with abdominal heterotaxy [1]. Intestinal rotation abnormalities include nonrotation (the small bowel is entirely on the right of the spine and the colon on the left), incomplete rotation (the appearance is between normal and nonrotation), and the rare reversed complete or incomplete rotation. In nonrotation and incomplete rotation, the mesenteric attachment is very narrow which predisposes to occurrence of midgut volvulus [9]. Radiologic identification of intestinal malrotation is important even if there are no symptoms because prediction of patients who are at risk of midgut volvulus is not possible. Surgical correction is advised by many authorities for all surgically fit patients with malrotation, regardless of age and symptoms [9,10].

## Preduodenal portal vein

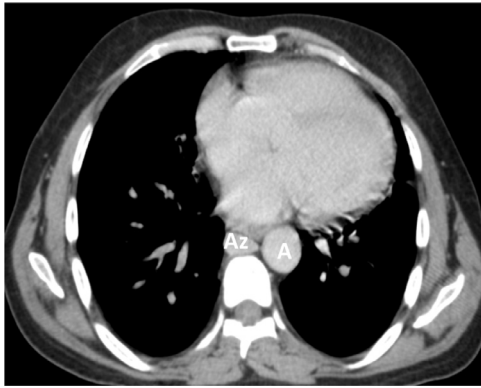
A preduodenal portal vein was reported as one of the anomalies in association with polysplenia. Embryologically, the portal venous system is derived from the vitelline veins which drain the primitive gastrointestinal tract. The 2 vitelline veins are connected by 3 interconnecting veins, cranial (in the liver), middle (behind the duodenum), and caudal (in front of the duodenum), simulating a figure of 8. Preduodenal portal vein occurs due to variation in this process where the middle and cranial interconnecting veins and the left vitelline vein atrophy resulting in an L-shaped preduodenal portal vein [11]. This anomaly is usually an incidental finding of no clinical importance except if biliary or hepatic surgical procedures are planned [12].



**Fig. 2 – (a) Common mesentery with transposition of the superior mesenteric artery (triangle) and vein (v). (b) Coronal CT image showing the sigmoid colon (s) on the right while the cecum (c) is located on the left. Note that the hepatic hilum is located on the left too.**

## Pancreas anomalies

Anomalies of the pancreas have been described in polysplenia syndrome. Normal pancreas formation occurs from fusion of ventral and dorsal pancreatic buds. The ventral pancreatic bud gives rise to the uncinate process and the head, while the dorsal pancreatic bud gives rise to the body and tail. The development of both dorsal pancreatic bud and spleen occur in the dorsal mesogastrium. Consequently, anomalies in both these organs can be expected in patients with polysplenia syndrome [13]. Most case series report a high incidence of short or truncated pancreas, where only the pancreatic head with or without a small portion of the pancreatic body is present. The clinical relevance includes an increased incidence of pancreatitis and diabetes mellitus [1] (Figs. 1–3).



**Fig. 3 – Axial CT image at the level of the heart showing normal atrial situs and levocardia. Note the enlarged azygous vein (Az) on the right side of the descending aorta (A).**

### Cardiac anomalies

Heterotaxy syndromes are known to be associated with increased incidence of complex cardiac anomalies. Cardiac anomalies are generally less common in polysplenia syndrome than asplenia [14]. The various cardiovascular anomalies that may be encountered are atrial septal defect, ventricular septal defect, bilateral superior vena cava, right-sided aortic arch, partial anomalous pulmonary venous return, transposition of the great arteries, pulmonary valvular stenosis, and subaortic stenosis [15].

Note that with increased use of cross-sectional imaging, the number of reported patients with polysplenia and normal hearts who are incidentally discovered during adulthood has increased.

### Visceroatrial situs

The relationship between polysplenia and visceroatrial situs is not well described in the literature. The classic term (bilateral sidedness) implies the presence of bilateral bilobed lungs with hyperarterial bronchi and bilateral pulmonary atria. However, it should be noted that these findings were not consistently described in association with polysplenia. In fact, we believe that the incidence of bilateral left-sidedness is much less in adult patients with polysplenia who usually have no cardiac anomalies, and therefore are not expected to show bilateral left-sidedness which is commonly associated with cardiac anomalies such as partial anomalous pulmonary venous return or atrial septal defect [1,16].

In conclusion, polysplenia syndrome is a complex syndrome with a broad spectrum of abnormalities, the most common of which are multiple spleens and IVC interruption with azygous continuation. As the use of imaging increases, situs anomalies will likely be detected with greater frequency in adults. Therefore, it is important for radiologists to become familiar with these anomalies. The finding of splenic cysts

should make us think of the possibility of another anomaly related to the polysplenia syndrome.

### Conflict of interest

None.

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