When the organs are not in their correct position. Imaging findings in situs ambiguous with polysplenia

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Learning objectives

1. To review situs anomalies (situs solitus, situs inversus and situs ambiguous)

2. To illustrate the hepatic, splenic, pancreatic, gastrointestinal tract, thoracic and vascular findings associated with situs ambiguous with polysplenia.
Background

The term *situs* refers to the position of the heart and viscera relative to midline.

*Situs solitus* represents the normal position of the heart and abdominal viscera, with the cardiac apex, spleen, stomach, and aorta located on the left and the liver and inferior vena cava (IVC) located on the right. Congenital heart disease occurs in less than 1% of individuals with situs solitus.

*Situs inversus* indicates mirror image location of the viscera relative to situs solitus. There are two major subcategories of situs inversus:

- *situs inversus with dextrocardia*: is more common. Is characterized by mirror image location of the heart and viscera relative to situs solitus. Congenital heart disease occurs in 3-5% of cases

- *situs inversus with levocardia*: is an extremely rare variant that is characterized by mirror image location of the viscera relative to situs solitus and a left-sided cardiac apex. Nearly all affected individuals have congenital heart disease.

*Situs ambiguous or heterotaxia* is defined as the abnormal arrangement of organs and vessels as opposed to the orderly arrangement typical of situs solitus and situs inversus. Is not characterized by a single set of abnormalities but by a spectrum of abnormalities. The two major subcategories of situs ambiguous are:

- *situs ambiguous with asplenia*: Also known as *right isomerism*. Is characterized by ambiguous location of the abdominal organs and absence of the spleen. Is associated with severe congenital heart disease in 99-100% of cases.

- *situs ambiguous with polysplenia*: Also known as *left isomerism*. Is characterized in general by midline or ambiguous location of the majority of the abdominal organs and multiple spleens. However, there is no single anomaly that is pathognomonic for this condition. Affected patients have a lower prevalence of congenital heart disease (50-90%) and less severe defects than do those with situs ambiguous with asplenia.
Imaging findings OR Procedure details

SITUS AMBIGUOUS WITH POLYSPLENIA:

This anomaly does not have a fixed set of characteristics that are present in all cases. A sufficient number of associated findings occur in the majority of patients to allow the diagnosis to be established.

Thorax and mediastinum:

There are bilateral bilobed lungs and both main bronchi may be seen inferior to the corresponding pulmonary arteries, indicating bilateral left lung anatomy (left isomerism) (Fig. 1 on page 6).

In majority patients there is levocardia. The most common cardiac anomalies in patients with polysplenia are partial anomalous pulmonary venous return, atrial septal defect and atrioventricular canal (the most frequent anomaly).

Vascular anomalies:

Besides being associated with duplication of leftsided structures such as the spleen, situs ambiguous with polysplenia is associated with the absence of right-sided structures such as a portion of the IVC. IVC interruption with azygous or hemiazygous continuation (Fig. 2 on page 6) is the most specific sign of polysplenia. Useful signs on chest X-ray film of this IVC anomaly are:

- convexity in the right tracheobronchial angle on the posteroanterior view (Fig. 3 on page 7)
- and an abscence of the IVC shadow on the lateral view.

Preduodenal portal vein (PPV) (Fig. 4 on page 8) is an unusual congenital anomaly in association with polysplenia syndrome. The presence of PPV causes technical difficulties at the time of upper abdominal surgery. Correct preoperative diagnosis of PPV is critical to avoid potential hazards during surgery.

In majority patients the aorta is located to the left of midline.

Viscera:
In majority patients there are multiple discrete spleens (Fig. 5 on page 9) of variable size and number. May be located in either the left or right (60%) side of the abdomen, always on the same side as the stomach. Nevertheless, some studies report patients who have a single, lobulated spleen (Fig. 5 on page 9) or even a normal spleen.

In majority patients there is a centrally located liver (Fig. 6 on page 10). Some studies report patients who have a right-sided liver or a left-sided liver (less common) (Fig. 6 on page 10).

**Anular pancreas** has been described in large series of patients with polysplenia. The association of polysplenia with short pancreas (Fig. 7 on page 11) has been reported too. There is absence of the pancreatic body and tail with a normal-sized pancreatic head located to the right of midline.

**Stomach** may be located in either the left or right side of the abdomen or in the midline (Fig. 8 on page 11).

**Malrotation of the bowel** is a frequent finding (Fig. 9 on page 12).
Fig. 0

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CT scan of a polysplenic patient with an interrupted inferior vena cava and azygous continuation. Azygous (az); inferior vena cava (IVC); azygous arch (aa).

Fig. 0

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**Fig. 0:** A posteroanterior chest X-ray film shows a round mass in the right superior mediastinum, which was shown to represent a dilated azygos arch.

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Situs ambiguous with polysplenia:

Preduodenal portal vein (arrows) that can be confused with gallstone.

Fig. 0

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Situs ambiguous with polysplenia:

Single, lobulated spleen on the same side as the stomach in a patient with polysplenia.

Multiple spleens in a patient with polysplenia.

Left-sided liver in a patient with polysplenia. An uncommon finding.

Centrally located liver in a patient with polysplenia.

Fig. 0

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Situs ambiguous with polysplenia:

CT scan of a polysplenic patient shows a rounded portion of a short pancreas (arrows).

Fig. 0

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Fig. 0: Stomach is located in right side of the abdomen

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Situs ambiguous with polysplenia and malrotation of the bowel in a 14 year old boy who came to emergency room with a chief complaint of abdominal pain. Finally this patient was diagnosed with appendicitis by CT. An appendicolith can be seen (arrowhead). Coronal CT images with contrast show the inversion of superior mesenteric vein (SMV) – superior mesenteric artery (SMA) with the SMA (red arrow) rotating from the left of SMV (blue arrow) to the right of the SMV. Note that there is a right-side small bowel and that the large bowel lays on the left side.

Fig. 0

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Conclusion

- Situs ambiguous with polysplenia is not characterized by a single set of abnormalities but by a spectrum of abnormalities.

- Interrupted IVC with azygous or hemiazygous continuation is classically described as the most specific sign of situs ambiguous with polysplenia. This finding should alert radiologists to the possibility of polysplenia syndrome.
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