Situs at the mirror: from situs inversus to situs ambiguus

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Learning objectives

- Define the three types of situs (solitus, inversus and ambiguus or heterotaxy), approaching their anatomic features and related terminology in a comprehensible way.

- Understand that although some generalizations may be made about the expected anatomic features in asplenic and polysplenic patients, these are somewhat arbitrary groups, as heterotaxy occurs in a continuum, being important to describe the patient’s specific anatomy in the radiology report.

- Recognize that situs ambiguus is associated with other conditions of major clinical relevance, such as congenital heart disease, intestinal malrotation with catastrophic volvus and immune deficiency (due to splenic absence).
Background

Although symmetry is a characteristic of the external mammalian phenotype and of some internal organs, notably the genitourinary system, much of our internal anatomy, namely the cardiovascular, pulmonary, and gastrointestinal systems, is asymmetrical.

The asymmetry is specific and originates in the genetic and molecular identity of the embryonic midline developmental field complex. This normal asymmetrical arrangement of the viscerovascular anatomy is called situs solitus.

WHAT'S IN A NAME?

Few topics engender more discussion, more passion between proponents of particular nomenclatures and more potential confusion than the description of variations in situs.

We list the definitions of the most relevant terms in the context of situs anomalies.

Situs: site or position; describes the position of the cardiac atria and viscera.

Solitus: normal or usual.

Situs solitus: normal position; describes the normal anatomy.

Situs inversus: the mirror image of situs solitus.

Situs ambiguous or heterotaxy: the word heterotaxy stems from greek "heteros" meaning other and "taxis" meaning arrangement. This term describe a group of disorders that involves abnormal left-right relations of the abdominal and thoracic organs. This abnormal arrangement of body organs is different from the orderly arrangement seen in situs solitus or situs inversus. The two primary subtypes of situs ambiguous include right isomerism or asplenia syndrome and left isomerism or polysplenia syndrome.

Isomerism: from the Greek "isos," meaning equal, and "meros," meaning part. The similarity of bilateral structures that are normally dissimilar, such as right and left bronchi and right and left lungs.
**Right isomerism**: bilateral structures with morphologic right characteristics, such as bilateral morphologic right bronchi and bilateral trilobed lungs.

**Left isomerism**: bilateral structures with morphologic left characteristics, such as bilateral morphologic left bronchi and bilateral bilobed lungs.

**Asplenia**: congenital absence of the spleen.

**Polysplenia**: multiple spleens, each of which is appreciably smaller than a normal-sized spleen.

**Kartagener syndrome**: triad of situs inversus, chronic sinusitis and bronchiectasis. It is present in 20% of all patients with situs inversus.

**Cardiac position**: the intrathoracic position of the heart as left sided, right sided, or midline (i.e. levocardia, dextrocardia or mesocardia). Levocardia, dextrocardia and mesocardia indicate the position of the cardiac apex only and not describe intracardiac or visceral anatomy.

**Eparterial bronchus**: eparterial means situated above an artery; the right superior lobar bronchus that passes above the right pulmonary artery.

**Hyparterial bronchus**: hyparterial means situated below an artery; the left main bronchus passes below the left pulmonary artery.

**SITUS SOLITUS**

Situs solitus is the normal anatomic arrangement, with the right atrium, liver and inferior vena cava on the right side; the left atrium, the aorta, stomach and spleen on the left side; a right-sided trilobed lung with an early origin of the upper lobe bronchus from the right main stem bronchus; and a left-sided bilobed lung with a more distal origin of the upper lobe bronchus. The right pulmonary artery lies in front of the right bronchus (eparterial bronchial position), and the left pulmonary artery crosses above the left bronchus (hyparterial bronchial position). *(Fig. 1 on page 10)*

**SITUS SOLITUS WITH DEXTROCARDIA**
In situs solitus with a right thoracic heart, the lungs and abdominal viscera are normally positioned, the ascending aorta and aortic knuckle are normally located, the descending aorta runs its normal course along the left side of the vertebral column, but the major cardiac shadow lies to the right of midline. The base-to-apex axis points to the right, so the right hemidiaphragm is lower than the left hemidiaphragm.

Determination of thoracoabdominal situs is not directly dependent on the presence or absence of levocardia, dextrocardia or mesocardia. (Fig. 2 on page 10)

**SITUS INVERSUS**

Situs inversus has incidence of 1 in 10000 live births and can be classified further into situs inversus with dextrocardia (also termed situs inversus totalis) or situs inversus with levocardia, an extremely rare variant of situs inversus, almost always associated with congenital heart disease (CHD) (Fig. 3 on page 11).

Patients with situs inversus totalis demonstrate mirror-image location, not only of the solid organs, bronchial tree (Fig. 4 on page 12) and heart, but also of the bowel and mesenteric vessels. The stomach, jejunum, and descending colon are located on the right, and the ligament of Treitz, ileum, and ascending colon are located on the left. The orientation of the bowel is reversed rather than malrotated relative to situs solitus. The branching pattern of the biliary tract and the location of the gallbladder in situs inversus are mirror image relative to situs solitus. (Fig. 5 on page 13, Fig. 6 on page 14)

**KARTAGENER´S SYNDROME**

Dyskinetic cilia syndrome (DCS) is characterized by abnormal ciliary structure and movement, resulting in abnormal mucociliary clearance and chronic infection. Bronchiectasis and sinusitis are common manifestations. About half of patients with DCS also have situs inversus. The combination of bronchiectasis, sinusitis and situs inversus is called Kartagener´s syndrome. It is present in 20% of all patients with situs inversus.

Symptoms of recurrent bronchitis, pneumonia and sinusitis often date from childhood. Appropriate antibiotic treatment is associated with a normal life expectancy.

Radiographs and CT typically show bilateral bronchiectasis with basal (lower and middle lobe) predominance. Cylindrical bronchiectasis are most common (Fig. 7 on page 15).

**SITUS AMBIGUUS OR HETEROTAXY**
Failure of normal lateralization results in abnormal bilateral symmetry of normally asymmetric viscera and duplication of either right- or left-sided structures. These conditions present with indeterminate situs or situs ambiguous and are often referred to as isomerism or heterotaxy syndromes.

The broad spectrum of abnormalities can be complex and includes right and left isomerism. This is often associated with asplenia (Ivermark syndrome) and polysplenia, respectively. There may be levocardia, dextrocardia or mesocardia.

**Understanding situs ambiguous** - The venous return

Some generalizations could be made about the venous return in situs ambiguous, although the features of situs ambiguous are inconsistent.

The right atrium develops as an extension of the inferior vena cava (IVC) and hepatic veins and is usually located on the same side as the IVC and liver.

In left atrial isomerism, the systemic veins are unable to drain normally into the morphologic right atrium. The IVC may be absent, or interrupted, with azygos or hemiazygous continuation of the infrarenal segment of the IVC, with drainage into the right or left superior vena cava. The hepatic veins drain into the azygos system or directly into the atria.

In right isomerism, there is no left atrium to receive pulmonary venous drainage, and total anomalous pulmonary venous drainage into a systemic vein is seen in 50% of cases. The IVC and hepatic veins are related to the morphologic right atrium and will therefore drain into either side. Right atrial isomerism is usually associated with severe cyanotic heart disease in infancy.

**Situs ambiguous with polysplenia**

Situs ambiguous with polysplenia, often referred to as left isomerism or bilateral left-sidedness, is characterized by duplication of left-sided structures, with bilateral bilobed lungs, hyparterial bronchi and bilateral left atria.

It has a slight female predominance and a milder course than asplenia. One quarter of patients do not have significant cardiac anomalies. CHD commonly associated with polysplenia includes endocardial cushion defects, double-outlet right ventricle and left heart obstruction, such as coarctation of the aorta.

Abnormalities of systemic venous drainage are common and include interruption of the intrahepatic IVC with azygos/hemiazygous continuation, duplication of the superior vena cava (SVC) and partial anomalous pulmonary venous return.
Abdominal anomalies include multiple small rounded spleens, a symmetric or transverse liver, biliary atresia, a truncated pancreas and malrotation of the bowel. The multiple spleens are typically adjacent to the greater curvature of the stomach, which is usually right sided, but can be left sided (Fig. 8 on page 16, Fig. 9 on page 17, Fig. 10 on page 18, Fig. 11 on page 19, Fig. 12 on page 20, Fig. 13 on page 21, Fig. 14 on page 22).

**SITUS AMBIGUUS WITH ASPLENIAS**

Situs ambiguous with asplenia is characterized by an absent spleen and duplication of right-sided structures (bilateral right-sidedness), affecting males twice as commonly as females.

Bilateral right-sidedness anomalies include bilateral trilobed lungs with eparterial bronchi.

Cardiac anomalies associated with asplenia are usually severe, are present at an early age, and have a poor prognosis. A single ventricle or large ventricular septal defect and pulmonic stenosis or atresia commonly occurs, resulting in undercirculation and cyanosis.

Both systemic and pulmonary venous drainage may be anomalous. Characteristically, the IVC and the abdominal aorta have a common course, with the abdominal aorta being juxtaposed to the IVC, and together they traverse the midline just below the diaphragm to enter a common atrium. Total anomalous pulmonary venous return and malposition of the great arteries are frequently associated with asplenia. Bilateral SVC drain into a common atrium with features of bilateral right atria.

Abdominal anomalies include an absent spleen and transverse liver, with the stomach on either side. Bowel malrotation, gallbladder agenesis, imperforate anus, horseshoe kidneys and urethral valves are associated with asplenia.

**Imaging heterotaxy - Key features**

Although some generalizations may be made about the expected anatomic features in asplenic and polysplenic patients, these are somewhat arbitrary groups, as heterotaxy occurs in a continuum (Fig. 15 on page 23, Fig. 16 on page 24).

The features of situs ambiguous are inconsistent therefore these cases are challenging and require thorough evaluation of the viscera. Eight key features should be described in a heterotaxy syndrome (Fig. 17 on page 25).

**MINOR FORMS OF SITUS AMBIGUUS**
Minor forms of situs ambiguous include M-anisosplenia or third syndrome and F-anisosplenia or fourth syndrome. They are characterized by a bifurcated spleen and bronchial symmetry.

**M-anisosplenia** is more common in males and resembles a mild form of asplenia with bilateral eparterial bronchi. Associated cardiac anomalies include anomalous pulmonary venous return, common atrium, pulmonary stenosis and bilateral SVC.

**F-anisosplenia** is more common in females and resembles a severe form of polysplenia with bilateral hyparterial bronchi. Associated cardiac anomalies include double-outlet right ventricle, bilateral SVC and azygos continuation of the IVC.

**DISEASES PROCESSES IN SITUS ANOMALIES**

**Congenital heart disease**

A 3-5% incidence of CHD is observed in situs inversus with dextrocardia, usually with transposition of the great vessels. Situs inversus with levocardia is very rare and it is almost always associated with CHD.

The severe cardiac anomalies and abnormal immune status explain the poor prognosis of patients with heterotaxy syndrome with asplenia. Death occurs in the first year of life in up 80% of cases.

Heterotaxy syndrome with polysplenia has a milder course than asplenia. One quarter of patients do not have significant cardiac anomalies (Fig. 18 on page 26).

**Bowel malrotation**

Malrotation of the bowel is a frequent finding in heterotaxy.

Infants with heterotaxy that survive the first few months of life can undergo a prophylactic Ladd procedure (surgical correction for the congenital bowel malrotation) in an attempt to eliminate the risk of midgut volvulus, bowel ischemia and infarction (Fig. 19 on page 27, Fig. 20 on page 28).

**Immune deficiency**

An absent spleen results in life threatening infections at an early age. Careful search for the spleen is therefore important and ultrasound will be successful most of the time.
Others disease processes in situs anomalies

Although situs anomalies do not in themselves usually cause symptoms in adults, their presence often creates a confusing clinical picture, especially in the setting of diseases such as appendicitis, cholecystitis, and splenic infarction when the patient's pain does not correlate with the expected locations of the appendix, gallbladder, and spleen. Likewise, situs anomalies may lead to diagnostic dilemmas on imaging examinations if radiologists are not aware of the spectrum of findings associated with these anomalies (Fig. 21 on page 29).
Fig. 1: Situs solitus. Axial CT images show a left-sided descending aorta (DA) (A), normal geographic cardiac chamber position (B), a right sided liver (L), a left-sided stomach (St) and spleen (S) (C) and a right-sided inferior vena cava (IVC) (D). AA, ascending aorta; LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle.

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**Fig. 2:** Situs solitus with dextrocardia, in a 20-year-old-man, without cardiac anomalies besides dextrocardia. Unenhanced axial CT images shows dextrocardia (A), a right-sided liver (L), a left-sided stomach (St) and a left-sided spleen (S) (B).

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**Fig. 3:** Schematic illustrations of the 4 basic cardiac positions (normal and 3 malpositions) and the relations of the descending aorta (Desc. ao.), cardiac apex, stomach, and liver as viewed on a frontal plain chest x-ray.

In situs solitus, the morphologic right bronchus (RB) is short, wide, and straight, and the morphologic left bronchus (LB) is long, thin, and curved (left). In situs inversus (right), the morphologic right bronchus is left-sided, and the morphologic left bronchus is right-sided.

Fig. 4

**Fig. 5:** Situs inversus in a 46-year-old man. Axial CT images show a right aortic arch (RAA) (A), a right-sided descending aorta (DA) (B) and reversal of the normal geographic cardiac chamber position (C). Lung window view shows mirror-image bronchial anatomy, with left-sided bronchus intermedius (BI) (D). AA, ascending aorta; LV, left ventricle; RV, right ventricle.

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Fig. 6: Situs inversus in a 46-year-old man. Axial CT images of the abdomen demonstrate mirror-image location of the abdominal structures relative to situs solitus. The liver (L) is located in the left upper quadrant, whereas the spleen (S) and the stomach (St) are located in the right upper quadrant (E). Note the left-sided gallbladder (GB) and a right sided pancreatic tail (P) in the region of the splenic hilum. The gallbladder has a drainage catheter (F). Inversion of the superior mesenteric vein (*) and superior mesenteric artery (G). Left-sided inferior vena cava (IVC) (H).

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Fig. 7: Kartagener’s syndrome in a 35-year-old male patient. Posteroanterior chest radiograph is consistent with situs inversus totalis (dextrocardia, right-sided aortic arch, left-sided liver and the hemidiaphragm is lower on the side of the right cardiac apex). There is left-sided basilar tram-tracking compatible with bronchiectasis (A). Axial chest CT demonstrates left middle lobe cylindrical bronchiectasis (which in patients with situs inversus has the characteristic segmentation of the right lung) (B). Axial head CT reveals mucosal thickening of the maxillary (arrow) (C) and sphenoidal (arrow) (D) sinuses, suggests rhinosinusitis.

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**Fig. 8:** Situs ambiguous with polysplenia and azygous continuation of IVC, in a 55-year-old woman with CHD. Chest CT scanogram shows dextrocardia and cardiomegaly (A). Unenhanced chest CT scan shows large azygos arch (arrows) (B). Unenhanced chest CT scan near diaphragm shows large azygos vein (Az) (C). Coronal reformatted unenhanced CT scan demonstrates azygous continuation of inferior vena cava (arrow) (D).

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**Fig. 9**: Situs ambiguus with polysplenia and azygous continuation of IVC, in a 55-year-old woman with CHD. Unenhanced CT scan of upper abdomen reveals several small rounded spleens (I - IV) on the left. Liver (L) is left-sided and stomach (St) lies in midline. Note azygous (Az) continuation of inferior vena cava; enlarged azygous vein is similar in caliber and just to right of descending aorta (E).

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**Fig. 10:** Situs ambiguous with polysplenia and azygous continuation of IVC, in a 15-year-old male patient. Chest CT scanogram shows levocardia and a midline liver (A). Contrast-enhanced chest CT scan shows large azygos arch (arrow)(B) and large azygos vein (Az) (C). Contrast-enhanced upper abdomen CT scan reveals a bridging or midline liver (L), right-sided stomach (St), large azygos vein (Az) and a normal sized hemiazygous vein (arrow) (D).

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**Fig. 11:** Situs ambiguus with polysplenia and azygous continuation of IVC, in a 15-year-old male patient. More caudal axial CT scan shows several small rounded spleens (I - IV) on the right, posterior to right-sided stomach (E). Abdomen CT scan obtained at the level of renal hila shows a truncated pancreas (arrows). Note the inversion of the superior mesenteric vein (*) and superior mesenteric artery. There is a midline gallbladder (GB) (F). Coronal reformatted CT scan shows multiple small spleens (I-V) in the right upper quadrant (G).

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**Fig. 12:** Situs ambiguous with polysplenia and azygous continuation of IVC in a 51-year-old woman with CHD. Axial MR image shows azygous (Az) continuation of IVC (A). Coronal MR image reveals a right (RSVC) and left superior vena cava (LSVC), cardiomegaly and a midline liver (L) (B). Axial MR image demonstrates interventricular communication (IVc) and a single atrium (SA) (C).

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**Fig. 13:** Situs ambiguous with polysplenia and azygous continuation of IVC in a 51-year-old woman with CHD. Sagittal MR image shows the hepatic veins (HV) draining directly into the single atrium (D). Coronal MR image shows both main bronchi passing inferior to the ipsilateral main pulmonary artery on each side (bilateral left or hyparterial bronchi) (E).

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Fig. 14: Situs ambiguous with polysplenia and azygous continuation of IVC in a 51-year-old woman with CHD. Axial MR image of the upper abdomen reveal two splenules on the left (I, II) (F).

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Fig. 15: Fig 15

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*Fig. 16:* Fig 16

EIGHT KEY IMAGING FEATURES THAT SHOULD BE DESCRIBED IN HETEROTAXY

(a) position of the atria
(b) position of the cardiac apex
(c) presence of tri/bilobed lungs and presence/absence of bilateral minor fissures
(d) position of venous drainage below the diaphragm relative to midline
(e) position of the aorta relative to midline
(f) position of stomach and presence of malrotation
(g) position of the liver and gallbladder
(h) presence, appearance and number of spleens

Fig. 17: Fig 17
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### RELATION OF CARDIAC POSITION TO CHD

<table>
<thead>
<tr>
<th>Visceroatrial Situs</th>
<th>Frequency of Association with CHD (%)</th>
</tr>
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<tbody>
<tr>
<td><strong>Situs solitus</strong></td>
<td></td>
</tr>
<tr>
<td>With levocardia</td>
<td>&lt;1</td>
</tr>
<tr>
<td>With dextrocardia</td>
<td>95</td>
</tr>
<tr>
<td><strong>Situs inversus</strong></td>
<td></td>
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<tr>
<td>With dextrocardia</td>
<td>3–5</td>
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<tr>
<td>With levocardia</td>
<td>100</td>
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<tr>
<td><strong>Situs ambiguus</strong></td>
<td></td>
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<tr>
<td>With left isomerism (polysplenia)</td>
<td>90</td>
</tr>
<tr>
<td>With right isomerism (asplenia)</td>
<td>99–100</td>
</tr>
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</table>

**Fig. 18:** Fig 18

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Fig. 19: Situs ambiguous with levocardia, polysplenia, azygous continuation of IVC and bowel malrotation, in a male child. Posteroanterior chest radiograph demonstrated a left cardiac apex and right-sided stomach bubble. The left hemidiaphragm is lower than the right hemidiaphragm. Note the prominent azygous arch (arrow) (A). Unenhanced chest CT scan shows azygous (Az) continuation of IVC (B). Lung window view from chest CT shows normal bronchial anatomy, with right-sided bronchus intermedius (BI) and left-sided lingular bronchus (LB) (C). More caudal chest CT scan reveals several small rounded spleens (I - IV) on the right, posterior to right-sided stomach (St). Liver (L) is midline (D).

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Fig. 20: Situs ambiguous with levocardia, polysplenia, azygous continuation of IVC and bowel malrotation, in a male child. Abdominal radiograph from an upper gastrointestinal study shows a right-sided stomach and bowel malrotation as the duodenal-jejunal junction does not cross the midline. It should be located on the same side of the body as the stomach and approximately at the level of duodenal bulb.

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**Fig. 21:** Situs inversus in a 46-year-old-man with acute cholecystitis. Pre-operative cholangiogram shows the right hepatic duct (RHD) and left hepatic duct (LHD) as well as the extrahepatic bile duct in the left upper quadrant. The cystic duct inserts in the extrahepatic bile duct. Note the gallbladder drainage catheter.

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Imaging findings OR Procedure details

This educational exhibit illustrates and discusses the imaging findings of situs anomalies, with emphasis on computer tomography and chest radiography.
Conclusion

Situs abnormalities are very rare but as the use of imaging becomes widespread is expected that they will be increasingly identified.

Radiologists should recognize the viscerovascular arrangements that are possible in these conditions and describe the specific viscerovascular anomalies in the patient. This information is crucial in planning and performing surgical, radiologic and endoscopic interventions.
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