Situs Revisited: Imaging of the Heterotaxy Syndrome

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Situs anomalies present a diagnostic challenge to radiologists because of the overlapping spectrum of findings commonly seen in asplenia and polysplenia. In a series of 21 patients with a diagnosis of heterotaxy syndrome, all 11 asplenic patients and seven of 10 polysplenic patients had congenital heart disease. Although there was a variety of complex congenital heart disease, the most common type in both patient groups was a common atrioventricular canal. In both groups, the laterality of the aorta and stomach was quite variable, but intestinal malrotation was a constant feature. In 11 asplenic patients, the most frequent findings were a bridging liver (10 cases), absent spleen (10 cases), and left-sided inferior vena cava (nine cases). Only seven of these patients had an aorta ipsilateral to the inferior vena cava, contrary to previous thought that this finding was specific for asplenia. In the 10 polysplenic patients, bridging livers were less frequent (five cases), single (six cases) or multiple (four cases) spleens were seen, and azygous continuation with interruption of the inferior vena cava was usually present (eight cases). Although not pathognomonic of polysplenia, inferior vena cava interruption with azygous or hemiazygous continuation is the most common anatomic finding of this condition. Although the terms asplenia and polysplenia are helpful in suggesting the typical anatomy, both syndromes encompass an overlapping spectrum that needs to be described individually and may best be called heterotaxy syndrome.
INTRODUCTION

Situs anomalies are rare, complex, and confusing. Situs ambiguous, or heterotaxy, implies a disordered organ arrangement in the chest or abdomen. This unpredictable anatomy differs from the orderly arrangement of the truncal organs in either the typical anatomy (situs solitus) or the mirror image of it (situs inversus).

Situs anomalies present a diagnostic challenge to the radiologist. Attempts at classification are difficult and usually oversimplify cases, since many patients do not demonstrate the expected, classical findings. We believe it is essential to understand situs abnormalities and their appearances on radiologic studies in order to recognize which children are at increased risk of congenital heart disease, immune deficiency (due to splenic absence), and catastrophic volvulus with malrotation.

In an attempt to clarify an imaging approach for evaluating children with abnormal situs or suspected cardiосplenic anomalies, we reviewed the recent literature and the medical records of a series of 21 children with heterotaxy syndrome (asplenia or polysplenia). In this article, we briefly discuss the terminology and embryologic development of heterotaxy syndrome, describe our proposed classification approach and radiologic evaluation, and present our clinical experience within the context of the radiology literature.

TERMINOLOGY

The term situs means “position, site, or location” and, in the context of congenital heart disease, refers to the position of the atria and viscera relative to the midline. The atrium whose appendage is broad-based and receives blood from the inferior vena cava may best be called the systemic or right atrium. The atrium with the smaller, narrower appendage and that receives blood from the pulmonary veins is called the pulmonary or left atrium (1). The right atrium has both terminal crest muscles not present in the left atrium and coarser pectinate muscles (seen at surgery or autopsy).

Situs solitus is the usual arrangement of organs and vessels within the body. The systemic atrium is on the right with a right-sided trilobed lung, liver, gallbladder, and inferior vena cava. The pulmonary atrium is on the left with a left-sided bilobed lung, stomach, single spleen, and aorta. The cardiac apex is on the left. The incidence of congenital heart disease in patients with situs solitus and levocardia is only 0.6%-0.8% (2).

Situs inversus refers to an anatomic arrangement that is the mirror image of situs solitus. The systemic atrium is on the left with a left-sided trilobed lung, liver, gallbladder, and inferior vena cava. The pulmonary atrium is on the right with a right-sided bilobed lung, stomach, single spleen, and aorta. The cardiac apex is on the right. Situs inversus is seen in 0.01% of the population (2), and the incidence of congenital heart disease in patients with situs inversus is 3%-5% (2). Atrioventricular discordance and transposition are the most common abnormalities, with the aortic arch being right-sided in 80% of patients (3). Kartagener syndrome (which consists of situs inversus, nasal polyposis with chronic sinusitis, and bronchiectasis) is present in 20% of all patients with situs inversus. The bronchiectasis is caused by compromised mucociliary transport secondary to dynein protein structural and functional abnormalities (ie, the immotile cilia syndrome) (4). Fifty percent of patients with immotile cilia syndrome have situs solitus and 50% have situs inversus.

Situs ambiguous, or heterotaxy, refers to visceral malposition and dysmorphism associated with indeterminate atrial arrangement (5). This abnormal arrangement of body organs is different from the orderly arrangement seen in situs solitus or situs inversus (3). The complexity of this syndrome is reflected in the various terms used to subclassify it, including asplenia syndrome, double right-sidedness, right isomerism, or Ivemark syndrome and polysplenia syndrome, double left-sidedness, or left isomerism. The incidence of congenital heart disease in patients with heterotaxy is very high, ranging from 50% to nearly 100% (1,3,6). In asplenia (ie, right isomerism or bilateral right-sidedness), both lungs have three lobes and eparterial bronchi (Fig 1). The main bronchus is located superior to the ipsilateral main pulmonary artery on each side. In polysplenia (ie, left isomerism or bilateral left-sidedness), both lungs have two lobes and hyparterial bronchi (Fig 2). In this situation, the reverse is seen: The main bronchus passes inferior to the ipsilateral main pulmonary artery on each side (Figs 3, 4). In children, especially with thymic tissue obscuring hilar structures, it may be difficult to identify this anatomy.
Figures 1, 2. (1) Drawing shows the typical anatomic features of classic asplenia: trilobed lungs with bilateral minor fissures and eparterial bronchi, bilateral systemic atria, midline liver, absent spleen, and variable location of the stomach. (2) Drawing shows the typical anatomic features of classic polysplenia: bilobed lungs with bilateral hyparterial bronchi, bilateral pulmonary atria, midline liver, and multiple spleens located along the greater curvature of the stomach, which occurs in variable locations.

Figure 3. Axial computed tomographic (CT) scans (lung windows; a at a higher level than b) of a patient with heterotaxy syndrome (polysplenia) show the characteristic bilateral hyparterial bronchi and bilobed lungs.
Figure 5. Drawing depicts the early atrial development at the 17-somite embryo stage. At this age, the embryo shows incomplete cardiac chamber septation. The atria are new formations, without separation, which have the appearance of a common atrium, a congenital heart anomaly seen in many patients with heterotaxy syndrome.

Other terms frequently used in discussions of situs inversus or heterotaxy are cardiac malposition, dextrocardia, levocardia, and mesocardia. Cardiac malposition is used to describe both the location of the heart anywhere other than the left hemithorax in a patient with situs solitus and the location of the heart in the left hemithorax when other organs are ambiguous in position. Dextrocardia, levocardia, and mesocardia indicate the position of the cardiac apex only and do not describe intracardiac or visceral anatomy (6).

■ EMBRYOLOGIC DEVELOPMENT

Although the exact timing is not known, most of the abnormalities in the asplenia syndrome can be linked to horizon XIII, a developmental stage of the embryo that corresponds to approximately 28 days gestation and 28 somites (2,7). It is during the period of 20–30 days gestation that the primitive heart and venous connections form (Fig 5). Disruption of this early embryologic event, when the cardiac chambers are incompletely septated, helps explain the preponder-
ance of common atria, single ventricles, abnormal pulmonary venous connections, and conotruncal anomalies observed in heterotaxy syndrome (Figs 6, 7).

Hutchins et al (8) believed that minor alterations in the embryonic body curvature can help explain the spectrum of anatomy seen in the heterotaxy syndrome. The pattern of altered development in patients with heterotaxy syndrome most likely results from a primary defect in lateralization, which leads to a failure of normal, asymmetric development. Although early reports suggested that asplenia and polysplenia were distinct entities (9), in 1983, Arnold et al (10) suggested that these two conditions were part of the same wide spectrum.

Modes of inheritance proposed for heterotaxy include autosomal dominant, autosomal recessive, and X-linked recessive. The last mode may help in part to explain the male preponderance of this syndrome (9). Overall, though, careful genetic study supports a multifactorial inheritance (11).

Figure 7. (a) Axial T1-weighted MR image of a patient with heterotaxy syndrome (asplenia) shows the common atroventricular canal with a common atrium. (b) Axial T1-weighted MR image obtained caudad to a demonstrates the ipsilateral inferior vena cava and aorta, which is a common although not a pathognomonic finding in asplenia.

Figure 6. Four chamber view echocardiogram of a patient with heterotaxy syndrome (asplenia) shows a common atrium. The arrow points to the septal “strand,” an expected echocardiographic finding representing a portion of the rudimentary atrial septum. LV = left ventricle, RV = right ventricle.
PROPOSED CLASSIFICATION OF HETEROXY SYNDROME: THE INDIVIDUALIZED APPROACH

The nomenclature used to describe the congenital abnormalities and cardiac malformations seen in heterotaxy has evolved over centuries. Until the 1970s, it was common to describe the multiple malformations of heterotaxy as discrete syndromes entitled either asplenia or polysplenia (3). However, because of the almost infinite number of combinations of possible malformations, more recent methods have used the segmental approach described by Van Praagh and Vlad (12).

We also propose a segmental or individualized approach for describing patients with heterotaxy syndrome, such that all suspected cases are labeled heterotaxy syndrome (not asplenia, polysplenia, or isomerism) followed by a description of the patient’s specific known anatomy in parentheses. For example, a radiologic dictation may read “heterotaxy syndrome (bilateral bilobed lungs, levocardia, left-sided malrotated stomach).” Although cumbersome, this terminology would accurately reflect the patient’s unique anatomy and would not presume that he or she has all features of classic asplenia or classic polysplenia syndrome. In addition, such a specific label would serve as a reminder of each patient’s anatomy to clinicians who may not be familiar with these complex syndromes and their terminology. Use of specific labels has important clinical implications. Knowledge of a patient’s possible malrotation and risk of intestinal volvulus, congenital heart disease, atypical placement of major abdominal organs, or altered immune status (particularly if asplenic) will enhance patient care.

RADIOLOGIC EVALUATION

The critical structures to be evaluated with imaging in determining situs are (a) position of the atria; (b) position of venous drainage below the diaphragm relative to midline; (c) position of the aorta relative to midline; (d) position of the stomach and presence of malrotation; (e) position of the liver and gallbladder; (f) position of the cardiac apex; (g) presence, appearance, and number of spleens; and (h) presence of tri- or bilobed lungs, including presence or absence of bilateral minor fissures.

These anatomic structures may be evaluated with chest radiography, ultrasonography (US), CT, MR imaging, and angiocardiography, although the last study is now rarely necessary for diagnosis. With chest radiography, one can determine the presence or absence of normal situs (ie, situs solitus). Normal situs is inferred when the aortic arch, cardiac apex, and stomach bubble are all located on the left. When these structures are positioned on the right or reversed, situs inversus is present (Fig 8). When any other situation is seen, an indeterminate situs or heterotaxy is inferred (Figs 9–11). The position of the atria may be inferred from the venous drainage below the diaphragm in a
Figures 9–11. (9) Chest radiograph of a patient with heterotaxy syndrome shows mesocardia and a right-sided stomach bubble. The patient also had a double-outlet right ventricle, common atrioventricular canal, pulmonary stenosis, and asplenia. (10) Chest radiograph of a patient with heterotaxy syndrome shows dextrocardia, a mildly enlarged heart, and a left-sided stomach bubble. The patient also had a single ventricle and total anomalous pulmonary venous drainage, which are typical congenital cardiac anomalies in asplenic patients. (11) Frontal chest radiograph of a patient with heterotaxy syndrome shows a left cardiac apex and a right-sided stomach bubble (therefore, situs ambiguous). The patient also had asplenia and congenital heart disease consisting of a single ventricle, common atrioventricular canal, and pulmonary stenosis. The increased soft tissues seen at the right mediastinum represent total anomalous pulmonary venous return via the right superior vena cava. Total anomalous pulmonary venous return is a common finding in asplenia, whereas partial anomalous pulmonary venous return is common in polysplenia.
However, in the presence of heterotaxy, even the echocardiographer may not be able to identify the appendages. The stomach may be displaced from its normal location directly under the hemidiaphragm by a bridging (midline) liver in either asplenic or polysplenic patients (Figs 12, 13). Further imaging with abdominal US usually allows identification of the spleen(s) and of the aorta and inferior vena cava. If the patient is large or if overlying gas-filled bowel obscures the upper abdominal anatomy at US, CT can be used to define these structures. MR imaging and angiocardiography may be used for surgical planning in patients with congenital heart disease.

**CLINICAL EXPERIENCE AND REVIEW OF THE LITERATURE**

Using the proposed segmental approach, we reviewed the medical records and imaging studies of 21 patients with heterotaxy syndrome referred to the Pediatric Cardiology Clinic from June 1994 to January 1997. Locations of the cardiac apex, inferior vena cava, aorta, stomach, liver, gallbladder, and spleen were determined. Azygous continuation, pulmonary anatomy, and the presence or absence of intestinal malrotation were assessed. Results were tabulated and compared with those in the literature. In the Table, we have used the “classic” asplenia and polysplenia classifications to demonstrate the variability of presentation of our 21 cases.
### Anatomic Characteristics of 21 Patients with Heterotaxy Syndrome

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Figure 15. Chest radiograph of a patient with asplenia and a common atrioventricular canal shows bilateral minor fissures (arrows) and, therefore, bilateral trilobed lungs.

linked to the understandably poor prognosis of this group of patients. Death occurs in the first year of life in up to 80% of cases (3).

As emphasized by Freedom and Fellows (13) in their radiographic-pathologic study in 1973, conventional radiographs do not necessarily demonstrate the patterns of visceral symmetry found in patients with autopsy-proved asplenia. In a second series of 51 autopsy cases of heterotaxy, 16 of 32 asplenic patients had either situs solitus or situs inversus and did not show typical features of bilateral right-sidedness (14). Seven of these asplenic patients had rudimentary spleens. Freedom and Fellows (13) noted that the most reliable indicator of asplenia was the ipsilateral position of the inferior vena cava and aorta.

In our retrospective series, 11 cases (six female and five male patients) were best classified as asplenia. We were able to determine definite bronchial branching patterns in only one of 11 cases. The radiologists reviewing the chest radiographs in this study were unable to accurately predict the position of the left and right pulmonary arteries relative to their respective bronchi. These relationships have been used to identify the eparterial bronchi in asplenia and
Figures 16, 17. (16) Transverse US image of an asplenic patient demonstrates a midline aorta and left-sided inferior vena cava. (17) Venogram of a patient with asplenia demonstrates a left-sided inferior vena cava that crosses the midline at the diaphragm to enter the right side of a common atrium.

Figure 18. Transverse US image of an asplenic patient demonstrates an ipsilateral right-sided aorta and inferior vena cava. There is pulmonary venous drainage below the diaphragm (solid arrow) into the portal vein (open arrow).

patients (Fig 15). Chest radiographs showed abnormal position of the cardiac apex, stomach, or both in nine of 11 patients. Two of 11 patients had dextrocardia and a left-sided stomach bubble, one had mesocardia, and eight had levocardia. The position, size, and configuration of the cardiomedistinal silhouette were all highly variable (Figs 9–12, 15).

In the 11 asplenic cases in our series, bridging livers were seen in 10 and spleens were absent in 10. Most of these patients were infants, and US helped determine splenic absence in all cases. A left-sided inferior vena cava was a frequent finding (nine of 11 cases) (Figs 16, 17). Only six of 11 patients had an aorta ipsilateral to the inferior vena cava, a finding that was previously thought to be specific for asplenia (Fig 18). No patients had inferior vena cava interruption with azygous continuation, and all had congenital heart disease.

Because patients with asplenia are at greater risk of sepsis, careful search for the spleen is important and US will be successful most of the time. MR imaging or CT may be helpful in large patients. Nuclear medicine studies are rarely indicated.
Heterotaxy Syndrome with Polysplenia
Classic left isomerism or bilateral left-sidedness (Fig 2) implies that patients have bilateral bilobed lungs, bilateral pulmonary atria, a centrally located liver, a stomach in indeterminate position, and multiple spleens. Interruption of the inferior vena cava with azygous or hemiazygous continuation (Fig 19) may be suggested from the chest radiograph and is the most consistent finding seen in heterotaxy with polysplenia. In fact, the short intrahepatic segment of the inferior vena cava is present in these cases. However, Winer-Muram and Tonkin (6) emphasized that “there is no single pathognomonic anomaly seen” in patients with polysplenia.

Polysplenia is more common in females and has more variable clinical manifestations and prognosis. Fewer polysplenic patients (vs asplenic patients) present with cyanosis and more present with symptoms of congestive heart failure from left-to-right shunts. Absence of the inferior vena cava on the lateral chest radiograph and the azygous continuation on the frontal chest radiograph with discordance of the apex and abdominal viscera help suggest the diagnosis (Figs 20–22). In general, cardiac anomalies are less common in polysplenic patients and not as complex as those in asplenic patients (the most common cardiac anomalies in this group are partial anomalous pulmonary venous return, atrial septal defect, and atroventricular canal). In an autopsy series of 146 patients with polysplenia, heterotaxy of abdominal viscera was seen in 56%; bilateral bilobed lungs, in only 55%; and cardiac anomalies, in over 50% (15). Many patients have no or mild congenital heart disease. Several cases of adult patients have been described in which the patients had bilateral bilobed lungs, normal abdominal situs, and no cardiac abnormality (6). In some children, splenic function is not normal. The spleen or spleens are always on the same side as the stomach, typically along the greater curvature (5). Some of these patients will have biliary atresia. Death occurs in the first year of life in 60% of cases (3).

In our retrospective series, 10 cases (nine female patients and one male patient) were best classified as polysplenia. Three of our ten patients had bilateral bilobed lungs on chest radiographs. Nine of 10 patients had levocardia, one
Figures 20–22. (20) Frontal chest radiograph of a polysplenic patient with an interrupted inferior vena cava shows the azygous continuation as a sharply defined, right paraspinal soft-tissue stripe and a prominent azygous arch (arrow). The patient has a left cardiac apex and left-sided stomach bubble and, therefore, apparently normal situs. (21) Lateral chest radiograph shows absent inferior vena cava stripe, which may be seen in polysplenic patients with inferior vena cava interruption and azygous continuation. (22) CT scans (a at a higher level than b) of a polysplenic patient demonstrate the inferior vena cava interruption with azygous continuation (arrow in a), the right-sided stomach and spleen (always ipsilateral), and midline liver (predominantly left-sided).
had dextrocardia, and none had mesocardia. The stomach occurred in variable positions, being on the right in five cases, the left in three, the medial left in one, and the medial right in one. Eight of 10 polysplenic patients in our study demonstrated azygous continuation with interruption of the inferior vena cava. Seven of 10 patients had congenital heart disease. Although CT and MR imaging helped confirm the presence of the spleen(s), US showed the location and number of spleens in all cases (Figs 23–25). Six patients had a single spleen, and four had multiple spleens (maximum, four spleens).

Chest radiography is useful for identifying situs ambiguous but not for categorizing an individual’s anatomy. US of the abdomen will clarify the venous anatomy below the diaphragm, the position of the liver and gallbladder, and the presence of the spleen(s).

**Malrotation**
Malrotation of the bowel is a frequent finding in heterotaxy syndrome. Ditchfield and Hutson (16) noted that in 25 patients with heterotaxy, three had normal rotation, six had incomplete or nonrotation, four had reverse rotation, and two presented with midgut volvulus. In another review of 23 cases of heterotaxy, seven patients presented with intestinal obstruction caused by
midgut volvulus, intraluminal membrane, annular pancreas, jejunal atresia, or preduodenal portal vein (17).

In our series, five of the 11 asplenic and three of the 10 polysplenic patients underwent an upper gastrointestinal series, and all of them had malrotation (Figs 26, 27). One of the asplenic patients had a midline microgastria and severe gastroesophageal reflux. At our institution, all infants with heterotaxy that survive the first few months of life undergo a prophylactic Ladd procedure to prevent the occurrence of midgut volvulus. One patient underwent a Ladd procedure when he developed vomiting suggestive of intermittent volvulus, although at surgery volvulus was not present.

**Figure 25.** Axial T1-weighted MR images (a at a higher level than b) of the upper abdomen reveal multiple splenules on the left, a left-sided stomach (St), and midline liver in a patient with heterotaxy syndrome (polysplenia, interrupted inferior vena cava with azygous-hemiazygous continuation).

**Figures 26, 27.** (26) Abdominal radiograph of a patient with heterotaxy syndrome (asplenia, left-sided stomach) and intestinal malrotation shows the duodenum and jejunum on the patient’s right side. However, the duodenal-jejunal junction is not located on the left at the level of the bulb, as would be expected in normal bowel rotation. (27) Abdominal radiograph of a patient with heterotaxy syndrome (polysplenia, right-sided stomach) and intestinal malrotation shows the duodenal-jejunal junction, which 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 the duodenal bulb.
CONCLUSIONS

Review of the literature, recent breakthroughs in embryologic understanding, and our own experience illustrate the complexity and spectrum of abnormalities seen in patients with the heterotaxic syndromes. We propose that the term heterotaxy syndrome be used to described these patients and that the patient’s individual anatomy be described, rather than using the classic but imprecise terms isomerism, asplenia, or polysplenia. For example, heterotaxy syndrome (dextrocardia, atrioventricular canal, polysplenia, malrotation) is much more descriptive than polysplenia. When a patient is noted to have congenital heart disease as part of the heterotaxy syndrome, or if an atypical position of organs is noted at imaging evaluation, we recommend that the patient undergo the following examinations: chest radiography, echocardiography, abdominal US, and upper gastrointestinal series.

These studies should help clarify the specific anatomy in these patients. Echocardiography will help define the intracardiac anatomy and cardiovascular connections. Abdominal US will help identify the spleen(s) and position of the aorta and inferior vena cava in most cases. When anatomic questions remain, further evaluation with CT, MR imaging, or angiography may be necessary. In this way, the unique anatomy of each of these patients can be delineated.

We believe it is essential to understand situs abnormalities and their wide variety of appearances on radiologic studies. A more accurate classification approach to each patient with heterotaxy is the most practical way to recognize those children who are at increased risk of congenital heart disease, immune deficiency (splenic absence), and catastrophic volvulus with malrotation.

REFERENCES