"Missing organs": Where did they go?

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Learning objectives

Recognize situs anomalies in adults. Discuss the challenge of diagnosing disease processes in patients with situs anomalies.

Background

Situs anomalies are often detected incidentally in adults during imaging evaluation for unrelated conditions. Technical advances in sonography, computed tomography (CT), and magnetic resonance (MR) imaging have greatly enhanced our ability to detect and characterize these anomalies. Situs anomalies are rare, complex and confusing. More common are ectopic organs with no relation to any heterotaxy syndrome.

Imaging findings OR Procedure details

In this exhibit, we briefly discuss the terminology and classification of situs anomalies and present our clinical experience within the context of the radiology literature. The authors also present different examples of ectopic organs with no relation to the more important heterotaxy syndromes, which are frequently associated with an increased risk of congenital heart disease, immune deficiency and catastrophic volvulus with malrotation.

Images for this section:


"MISSING ORGANS": WHERE DID THEY GO?
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Fig. 1: 1

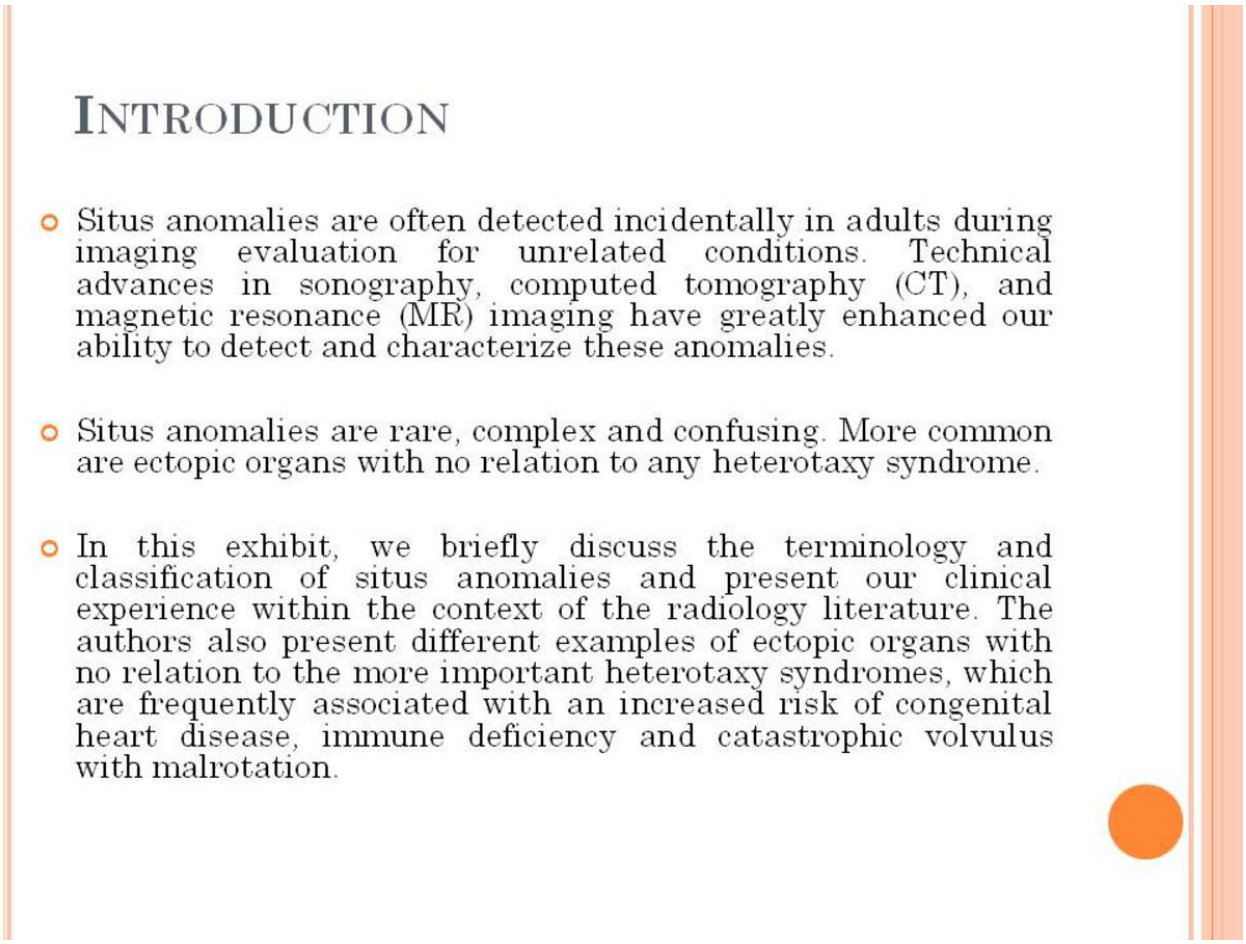

INTRODUCTION
Situs anomalies are often detected incidentally in adults during imaging evaluation for unrelated conditions. Technical advances in sonography, computed tomography (CT), and magnetic resonance (MR) imaging have greatly enhanced our ability to detect and characterize these anomalies.
Situs anomalies are rare, complex and confusing. More common are ectopic organs with no relation to any heterotaxy syndrome.
In this exhibit, we briefly discuss the terminology and classification of situs anomalies and present our clinical experience within the context of the radiology literature. The authors also present different examples of ectopic organs with no relation to the more important heterotaxy syndromes, which are frequently associated with an increased risk of congenital heart disease, immune deficiency and catastrophic volvulus with malrotation.

Fig. 2: 2

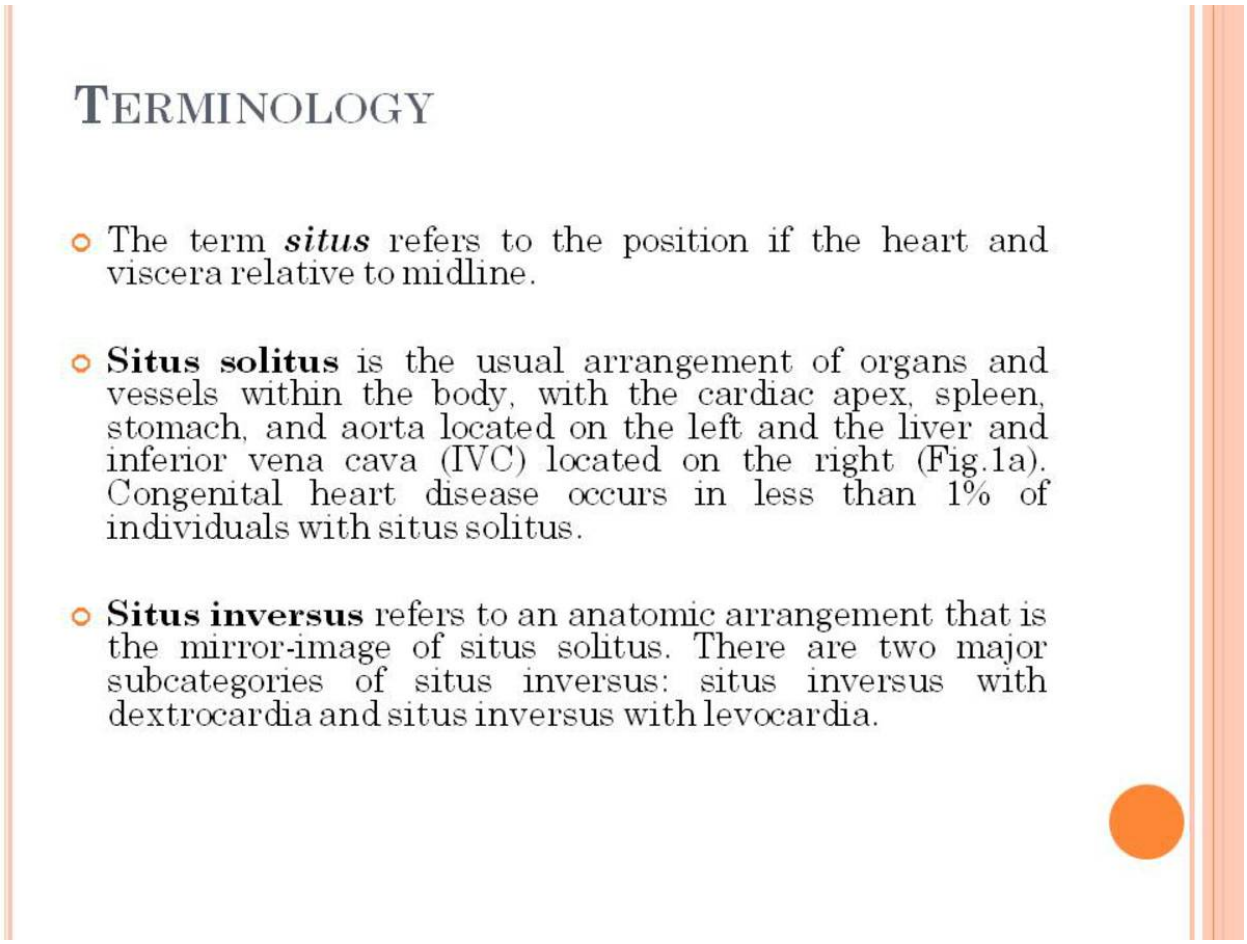

TERMINOLOGY
The term *situs* refers to the position if the heart and viscera relative to midline.
Situs solitus is the usual arrangement of organs and vessels within the body, with the cardiac apex, spleen, stomach, and aorta located on the left and the liver and inferior vena cava (IVC) located on the right (Fig.1a). Congenital heart disease occurs in less than 1% of individuals with situs solitus.
Situs inversus refers to an anatomic arrangement that is the mirror-image of situs solitus. There are two major subcategories of situs inversus: situs inversus with dextrocardia and situs inversus with levocardia.

Fig. 3: 3

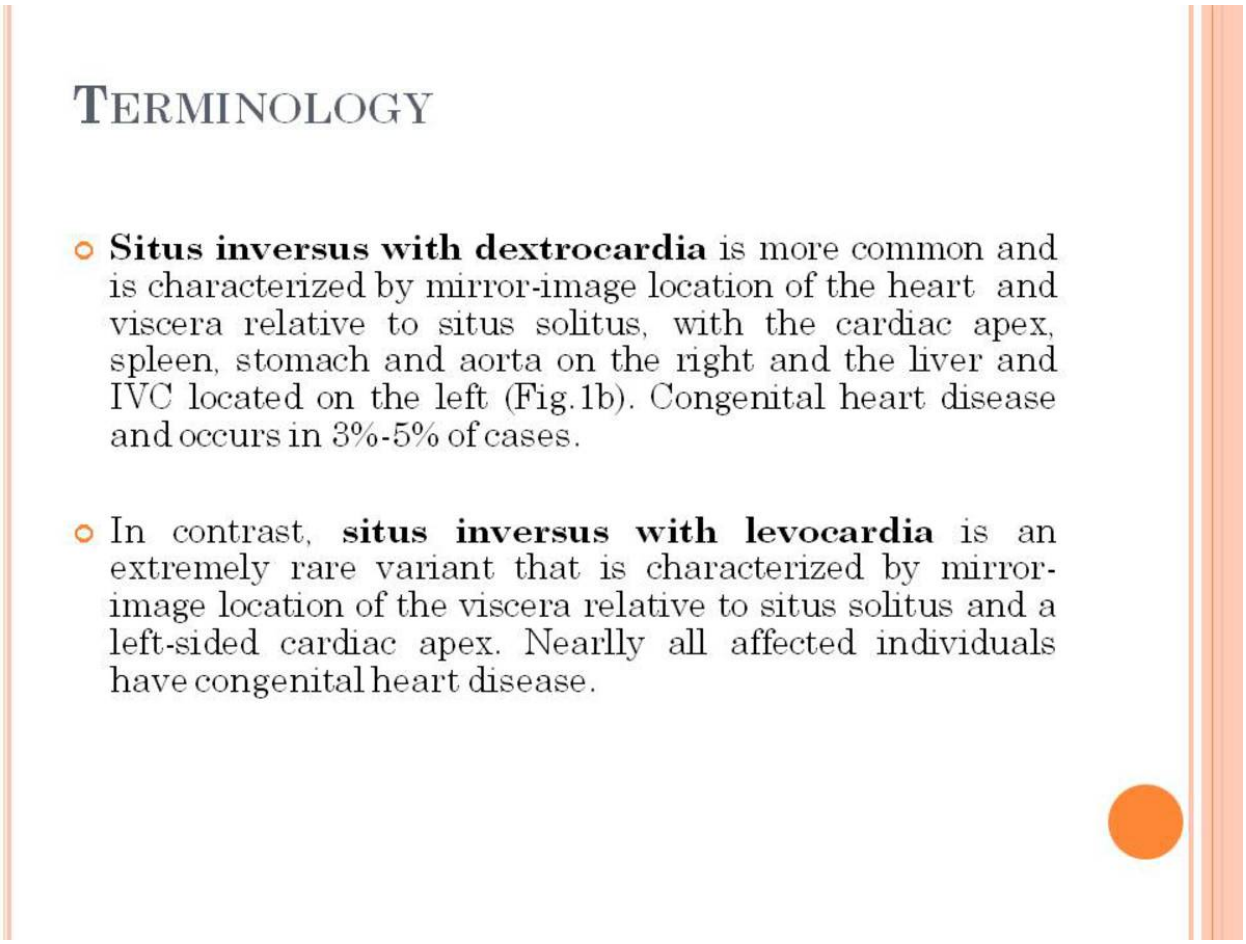

TERMINOLOGY
Situs inversus with dextrocardia is more common and is characterized by mirror-image location of the heart and viscera relative to situs solitus, with the cardiac apex, spleen, stomach and aorta on the right and the liver and IVC located on the left (Fig.1b). Congenital heart disease and occurs in 3%-5% of cases.
In contrast, 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. Nearlly all affected individuals have congenital heart disease.

Fig. 4: 4

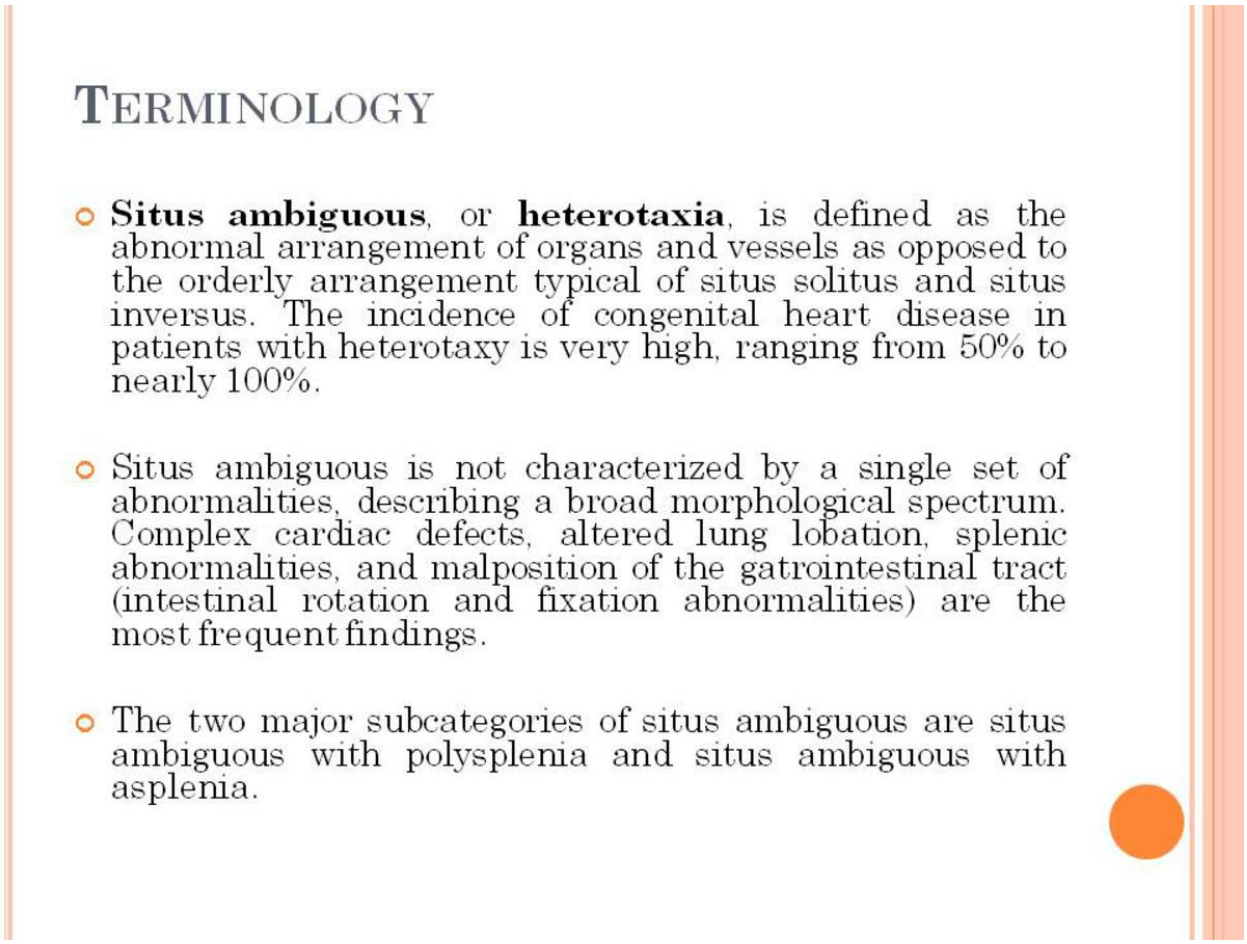

TERMINOLOGY
Situs ambiguous, or heterotaxia, is defined as the abnormal arrangement of organs and vessels as opposed to the orderly arrangement typical of situs solitus and situs inversus. The incidence of congenital heart disease in patients with heterotaxy is very high, ranging from 50% to nearly 100%.
Situs ambiguous is not characterized by a single set of abnormalities, describing a broad morphological spectrum. Complex cardiac defects, altered lung lobation, splenic abnormalities, and malposition of the gatrointestinal tract (intestinal rotation and fixation abnormalities) are the most frequent findings.
The two major subcategories of situs ambiguous are situs ambiguous with polysplenia and situs ambiguous with asplenia.

Fig. 5: 5

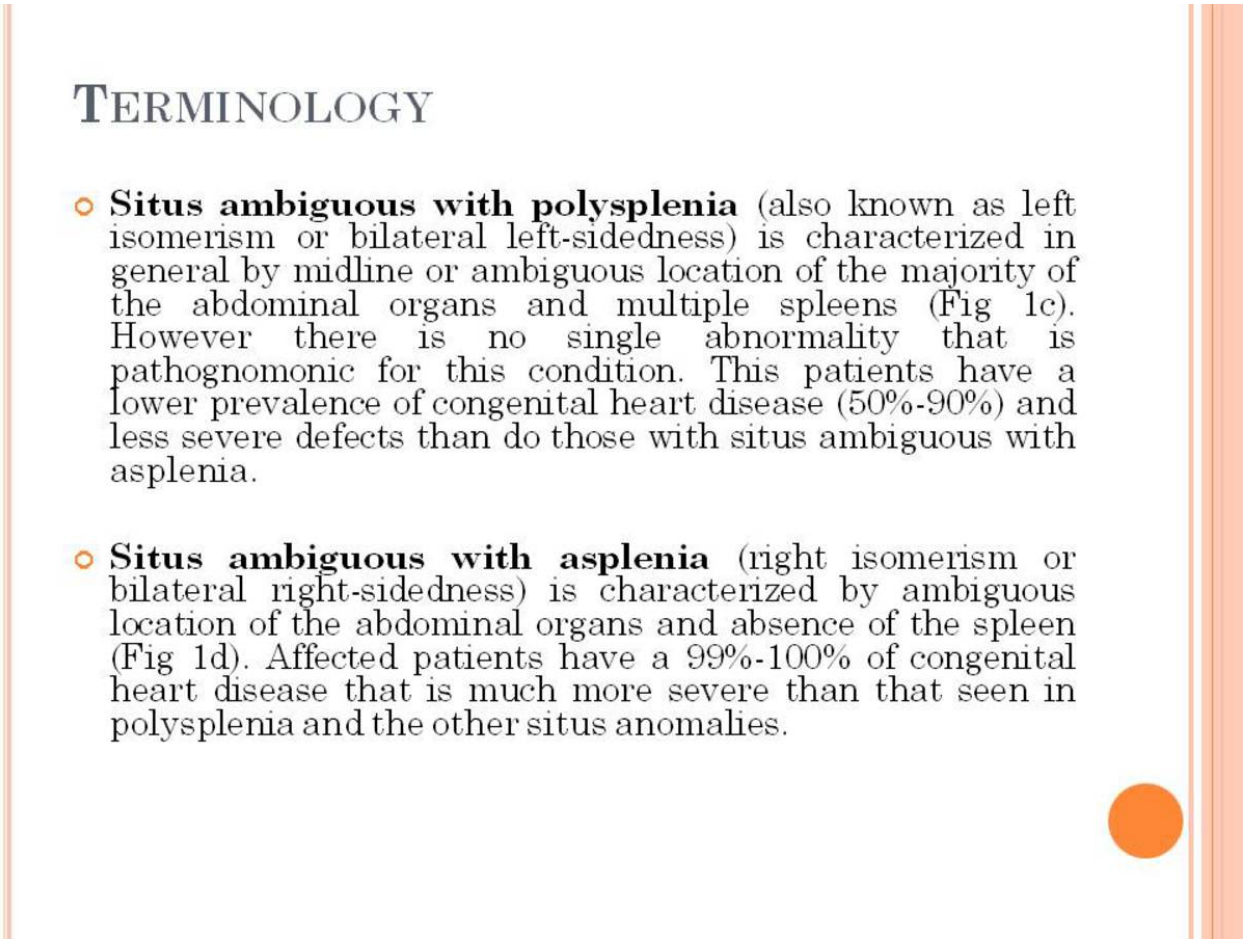

TERMINOLOGY
Situs ambiguous with polysplenia (also known as left isomerism or bilateral left-sidedness) is characterized in general by midline or ambiguous location of the majority of the abdominal organs and multiple spleens (Fig 1c). However there is no single abnormality that is pathognomonic for this condition. This patients have a lower prevalence of congenital heart disease (50%-90%) and less severe defects than do those with situs ambiguous with asplenia.
Situs ambiguous with asplenia (right isomerism or bilateral right-sidedness) is characterized by ambiguous location of the abdominal organs and absence of the spleen (Fig 1d). Affected patients have a 99%-100% of congenital heart disease that is much more severe than that seen in polysplenia and the other situs anomalies.

Fig. 6: 6

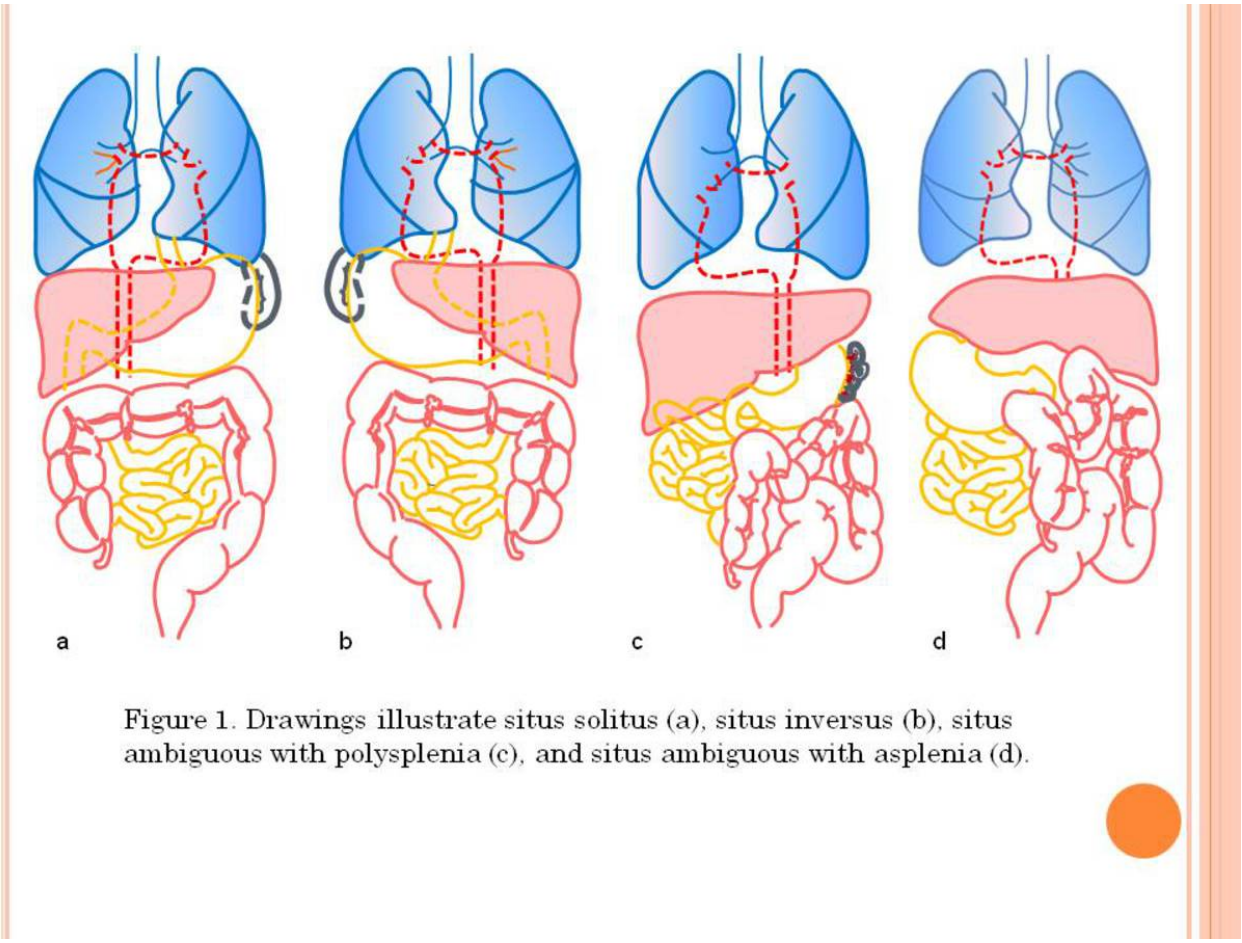


Figure 1. Drawings illustrate situs solitus (a), situs inversus (b), situs ambiguous with polysplenia (c), and situs ambiguous with asplenia (d).

Fig. 7: 7

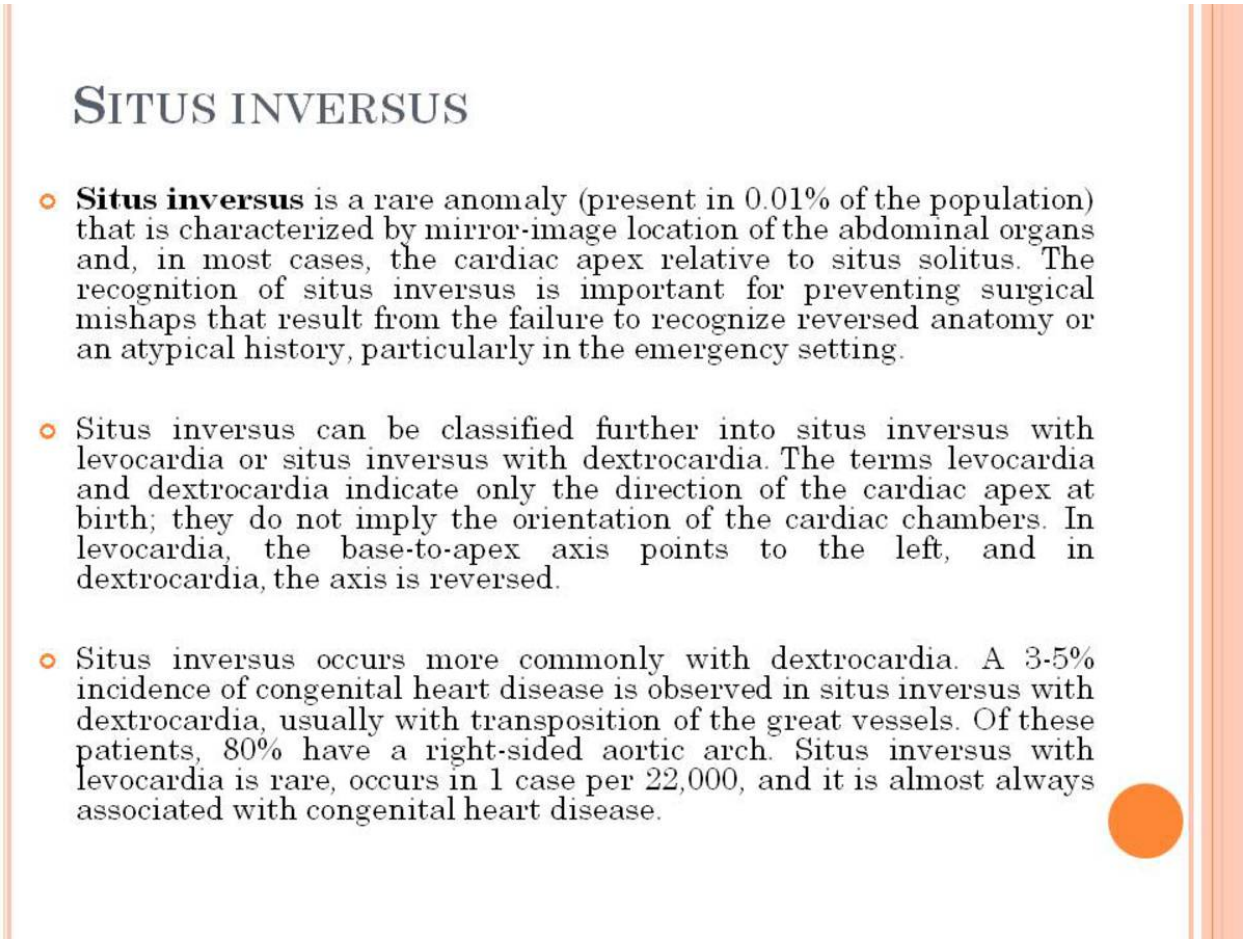

SITUS INVERSUS
Situs inversus is a rare anomaly (present in 0.01% of the population) that is characterized by mirror-image location of the abdominal organs and, in most cases, the cardiac apex relative to situs solitus. The recognition of situs inversus is important for preventing surgical mishaps that result from the failure to recognize reversed anatomy or an atypical history, particularly in the emergency setting.
Situs inversus can be classified further into situs inversus with levocardia or situs inversus with dextrocardia. The terms levocardia and dextrocardia indicate only the direction of the cardiac apex at birth; they do not imply the orientation of the cardiac chambers. In levocardia, the base-to-apex axis points to the left, and in dextrocardia, the axis is reversed.
Situs inversus occurs more commonly with dextrocardia. A 3-5% incidence of congenital heart disease is observed in situs inversus with dextrocardia, usually with transposition of the great vessels. Of these patients, 80% have a right-sided aortic arch. Situs inversus with levocardia is rare, occurs in 1 case per 22,000, and it is almost always associated with congenital heart disease.

Fig. 8: 8

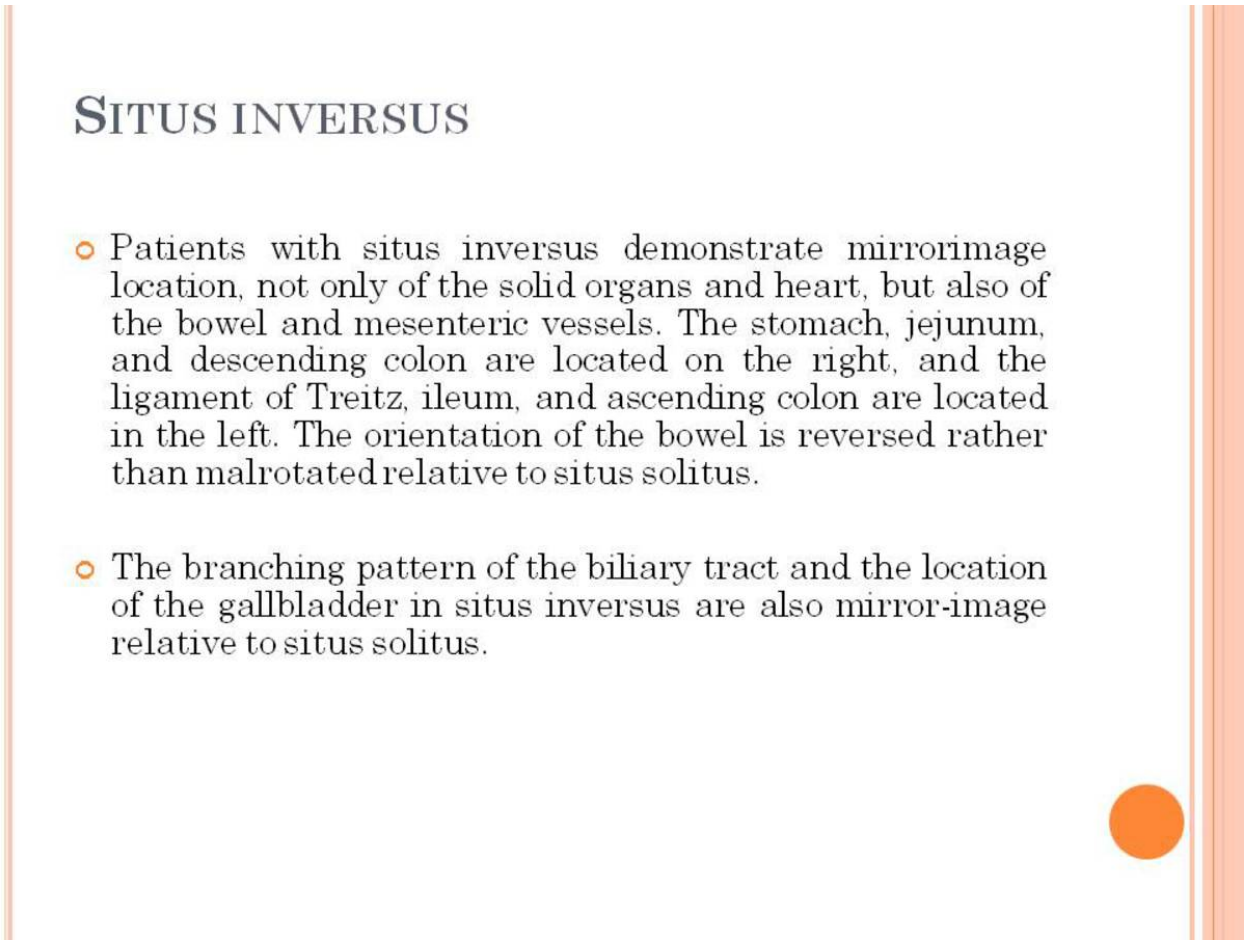
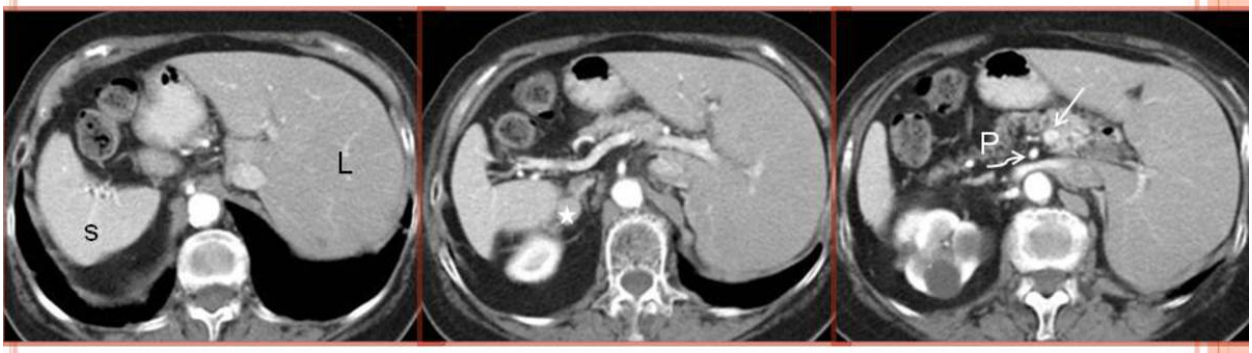

SITUS INVERSUS
Patients with situs inversus demonstrate mirrorimage location, not only of the solid organs and heart, but also of the bowel and mesenteric vessels. The stomach, jejunum, and descending colon are located on the right, and the ligament of Treitz, ileum, and ascending colon are located in 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 also mirror-image relative to situs solitus.

Fig. 9: 9



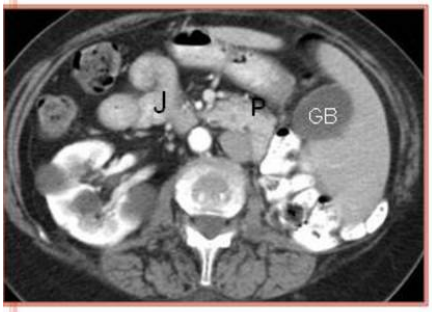


Fig.2. Situs inversus in a 79-year-old woman. Post-contrast CT scan of the abdomen demonstrates mirror-image location of the abdominal structures relative to situs solitus. The liver (*L*) is located in the left upper quadrant, whereas the spleen (*S*) and the stomach are located in the right upper quadrant. There is a left-sided gallbladder (*GB*) and a right-sided pancreatic tail (*P*) in the region of the splenic hilum. There is an acessory spleen (*asterisk*) near the posterior aspect of the spleen. One can see inversion of the SMV (*arrow*) and SMA (*curved arrow*). The jejunum (*J*) is in the right side of the abdomen. The pancreatic head (*P*) and uncinate process lie to the left of midline.

Fig. 10: 10

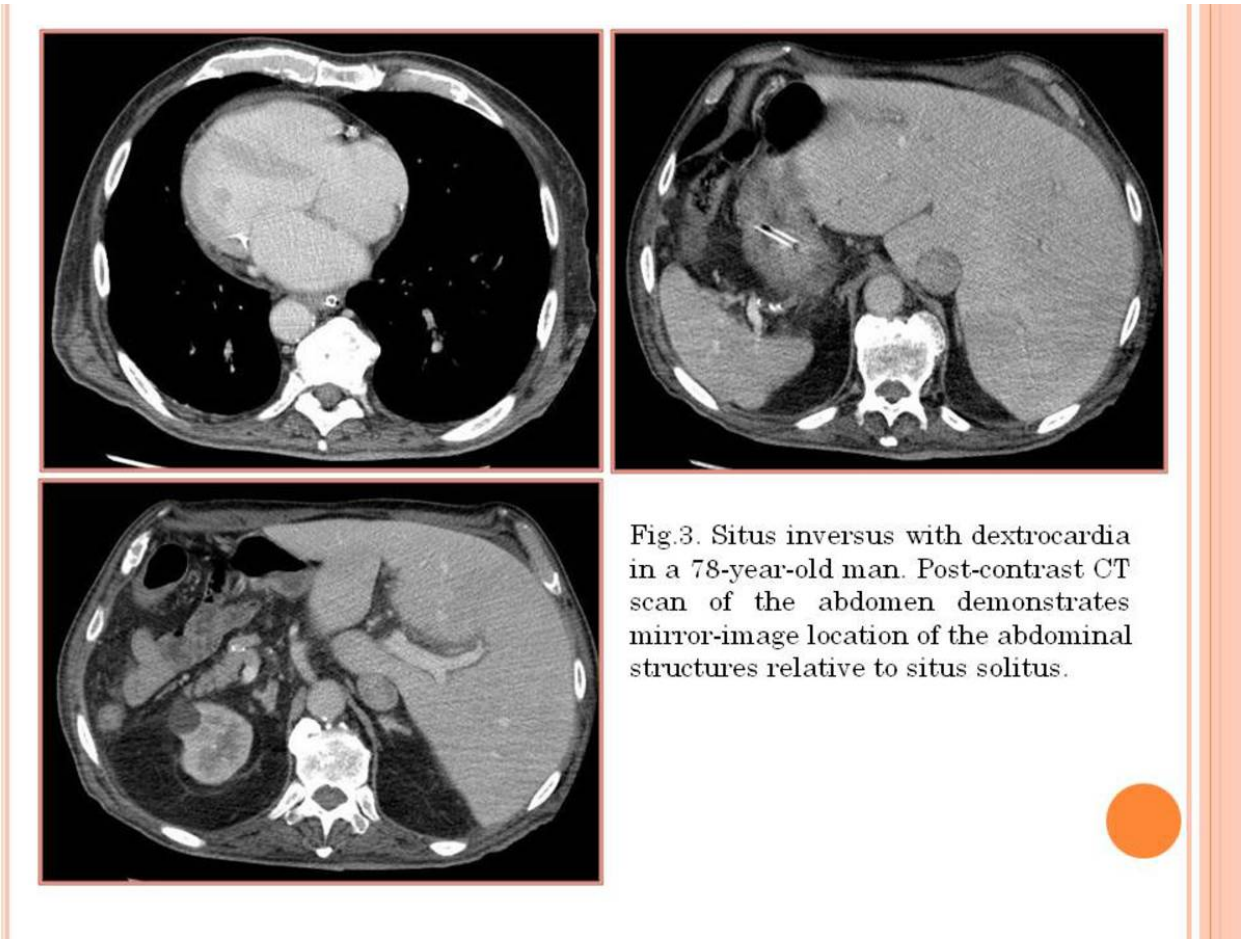


Fig.3. Situs inversus with dextrocardia in a 78-year-old man. Post-contrast CT scan of the abdomen demonstrates mirror-image location of the abdominal structures relative to situs solitus.

Fig. 11: 11

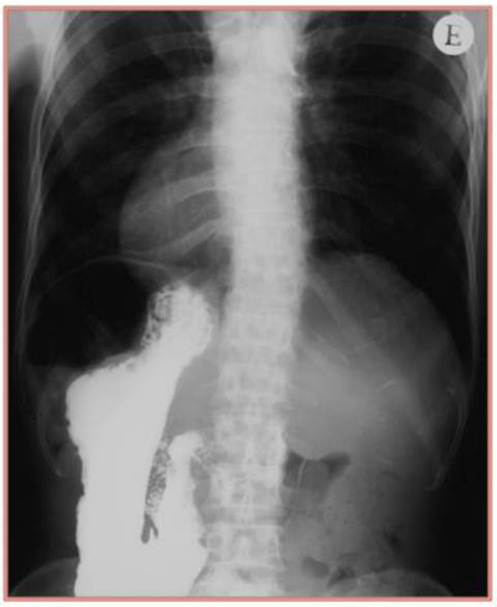


Fig.4. Situs inversus with dextrocardia in a 73-year-old man. Radiograph from an upper gastrointestinal study shows that the stomach is located in the right side of the abdomen.

Fig. 12: 12

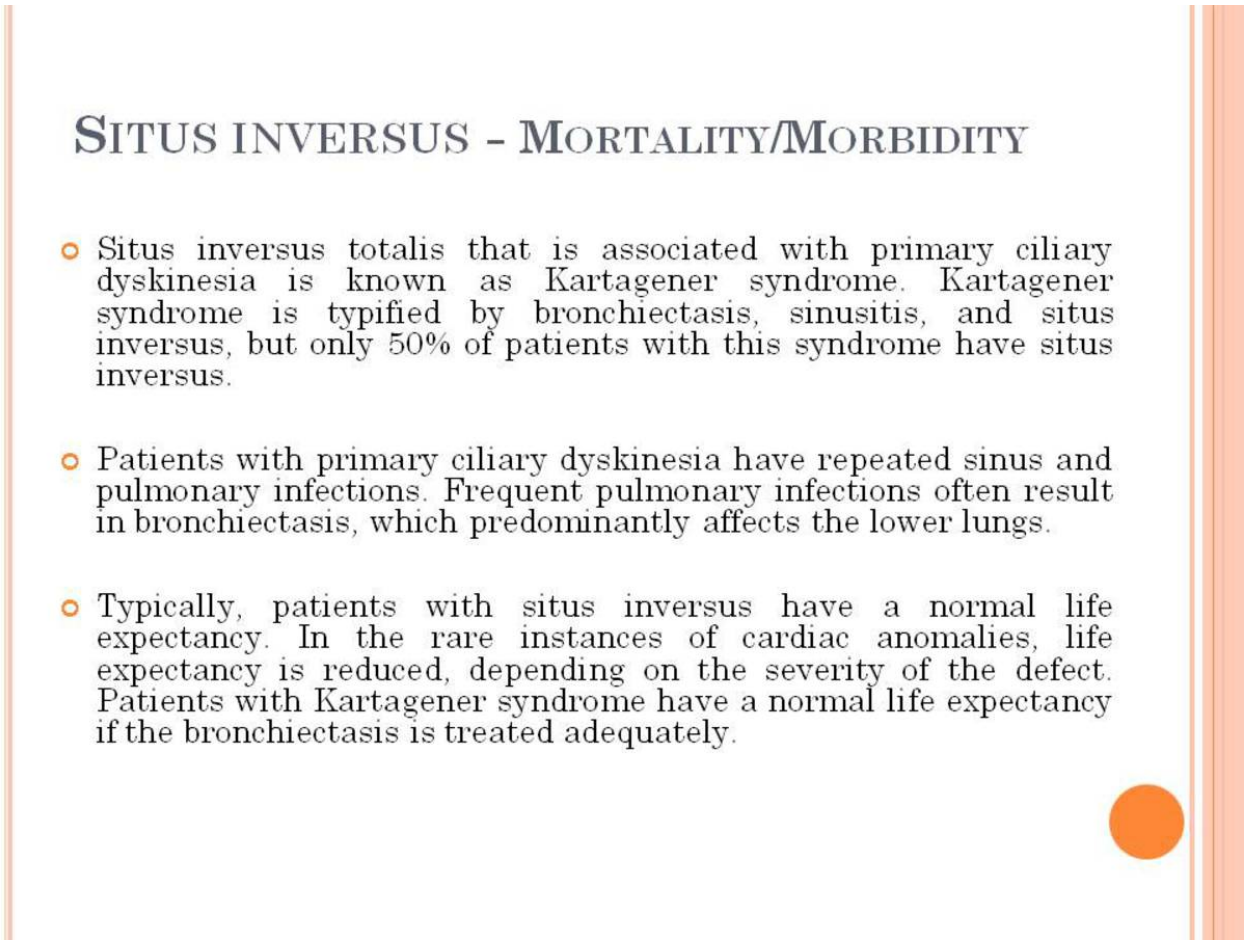

SITUS INVERSUS - MORTALITY/MORBIDITY
Situs inversus totalis that is associated with primary ciliary dyskinesia is known as Kartagener syndrome. Kartagener syndrome is typified by bronchiectasis, sinusitis, and situs inversus, but only 50% of patients with this syndrome have situs inversus.
Patients with primary ciliary dyskinesia have repeated sinus and pulmonary infections. Frequent pulmonary infections often result in bronchiectasis, which predominantly affects the lower lungs.
Typically, patients with situs inversus have a normal life expectancy. In the rare instances of cardiac anomalies, life expectancy is reduced, depending on the severity of the defect. Patients with Kartagener syndrome have a normal life expectancy if the bronchiectasis is treated adequately.

Fig. 13: 13

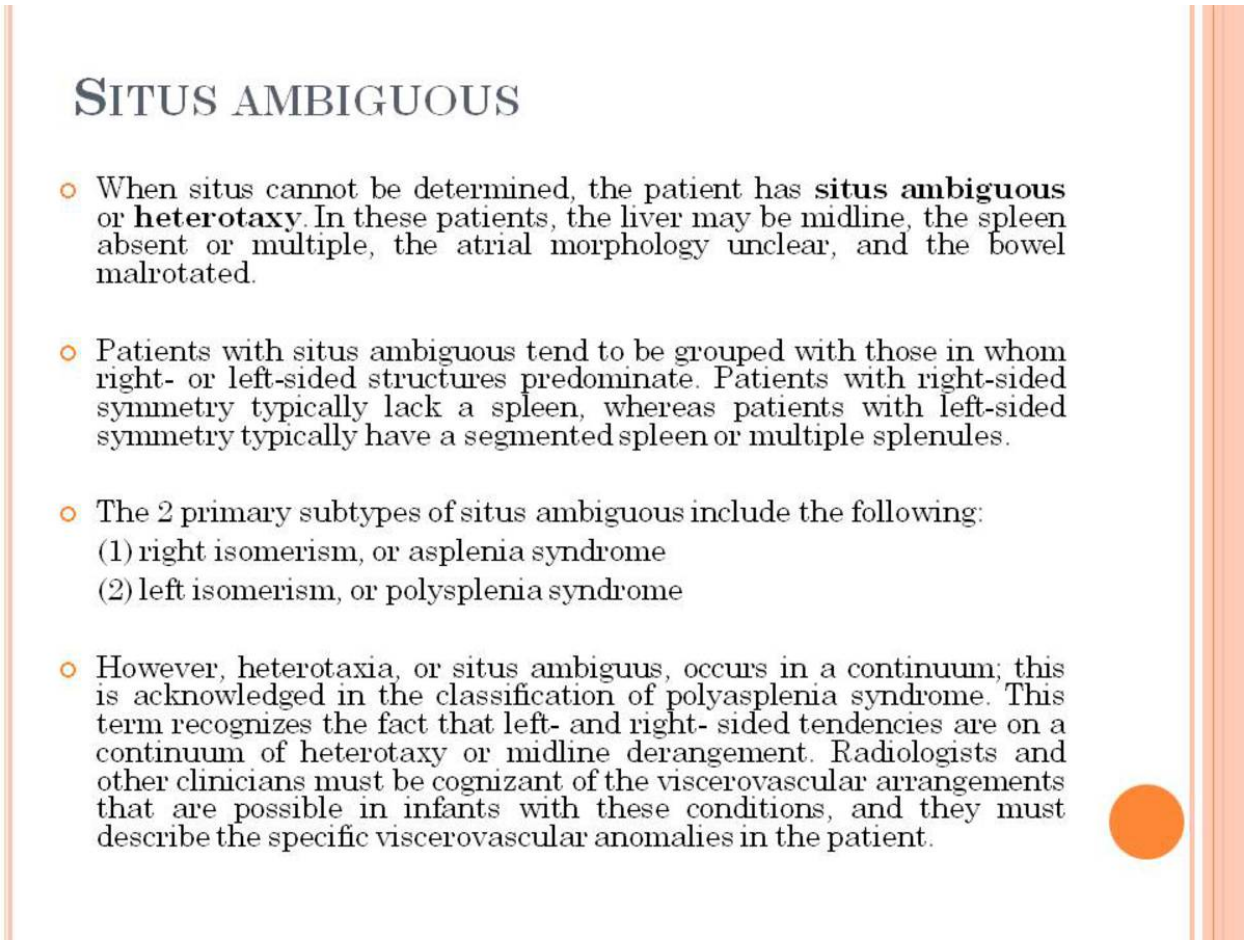

SITUS AMBIGUOUS
When situs cannot be determined, the patient has situs ambiguous or heterotaxy. In these patients, the liver may be midline, the spleen absent or multiple, the atrial morphology unclear, and the bowel malrotated.
Patients with situs ambiguous tend to be grouped with those in whom right- or left-sided structures predominate. Patients with right-sided symmetry typically lack a spleen, whereas patients with left-sided symmetry typically have a segmented spleen or multiple splenules.
The 2 primary subtypes of situs ambiguous include the following:
(1) right isomerism, or asplenia syndrome
(2) left isomerism, or polysplenia syndrome
However, heterotaxia, or situs ambiguus, occurs in a continuum; this is acknowledged in the classification of polyasplenia syndrome. This term recognizes the fact that left- and right- sided tendencies are on a continuum of heterotaxy or midline derangement. Radiologists and other clinicians must be cognizant of the viscerovascular arrangements that are possible in infants with these conditions, and they must describe the specific viscerovascular anomalies in the patient.

Fig. 14: 14

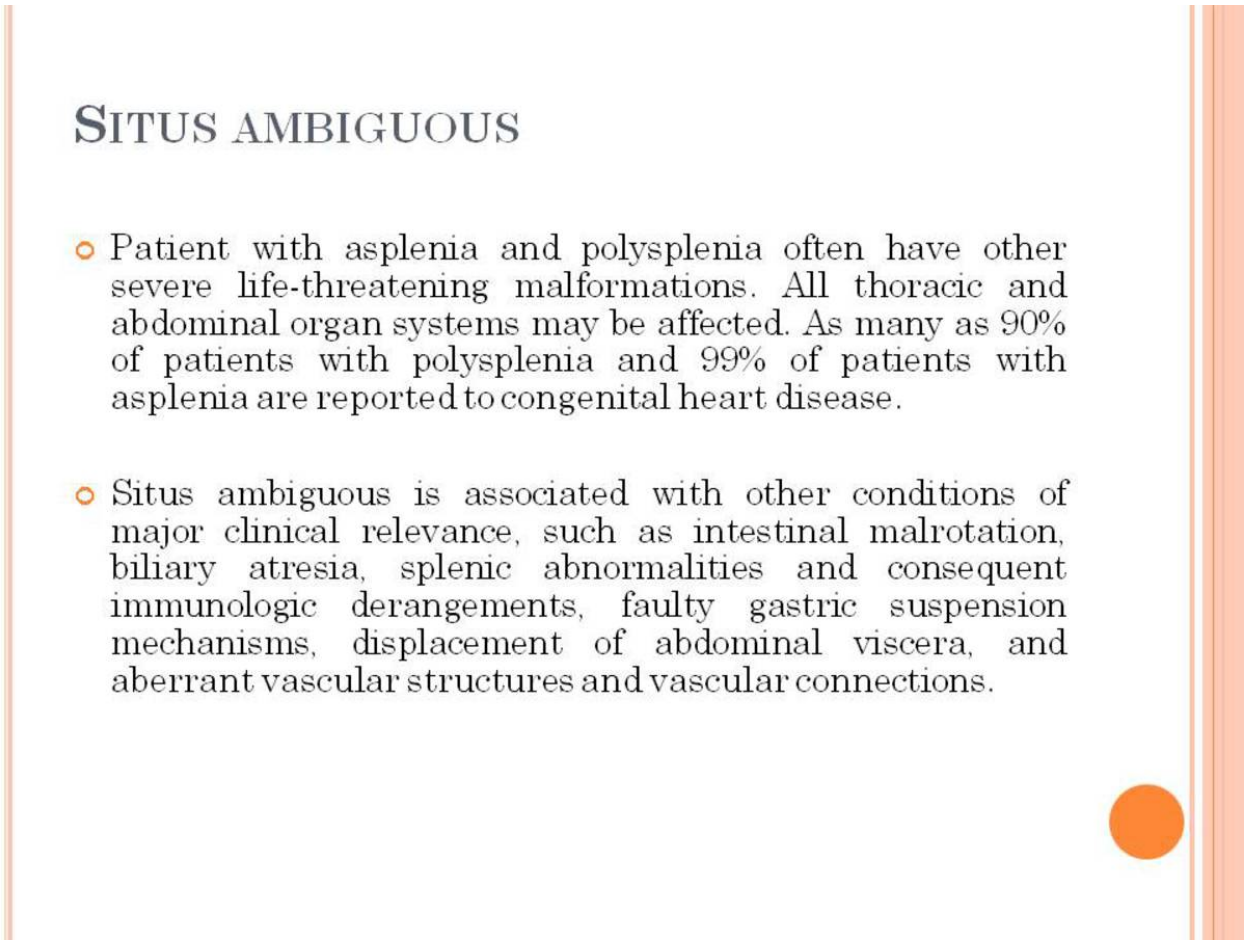

SITUS AMBIGUOUS
Patient with asplenia and polysplenia often have other severe life-threatening malformations. All thoracic and abdominal organ systems may be affected. As many as 90% of patients with polysplenia and 99% of patients with asplenia are reported to congenital heart disease.
Situs ambiguous is associated with other conditions of major clinical relevance, such as intestinal malrotation, biliary atresia, splenic abnormalities and consequent immunologic derangements, faulty gastric suspension mechanisms, displacement of abdominal viscera, and aberrant vascular structures and vascular connections.

Fig. 15: 15

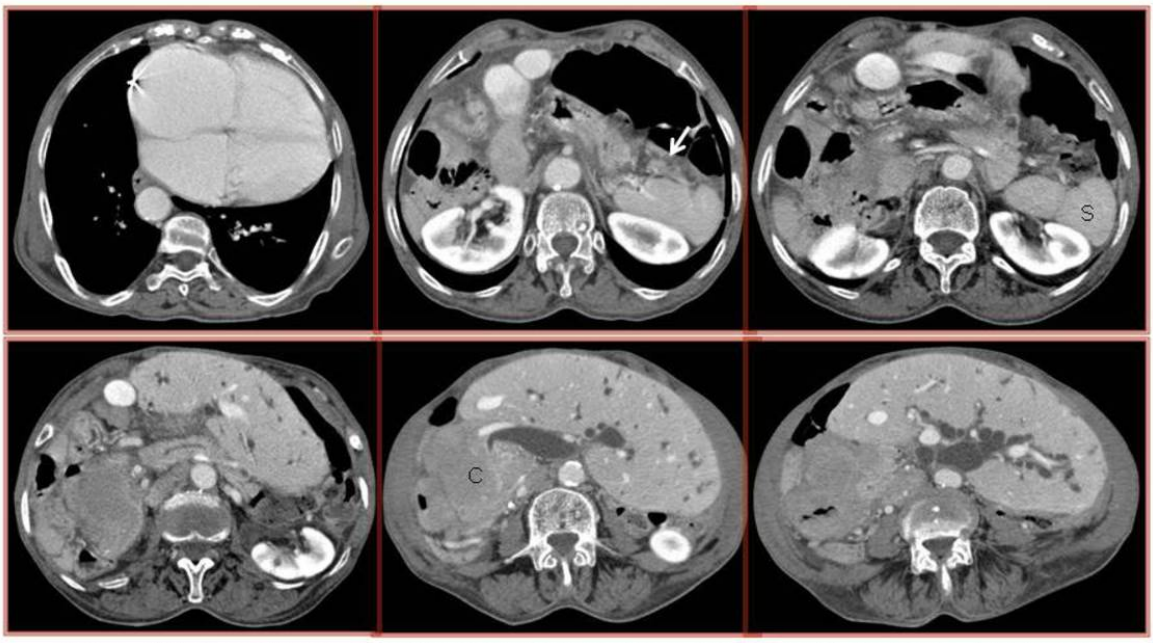


Fig.5. Situs ambiguous with polysplenia in a 85-year-old woman. Contrast-enhanced TC scan of the lower chest reveals levocardia. CT scan of the upper abdomen reveals a predominantly left-sided liver, with bile duct dilatation secondary to a carcinoma (C) of the pancreatic head, which is right-sided. The stomach and the spleen (S) are in the left upper quadrant, with a few small acessory spleens (arrow) near the splenic hilum. The aorta is located at the left of the midline and the IVC at the right.

Fig. 16: 16

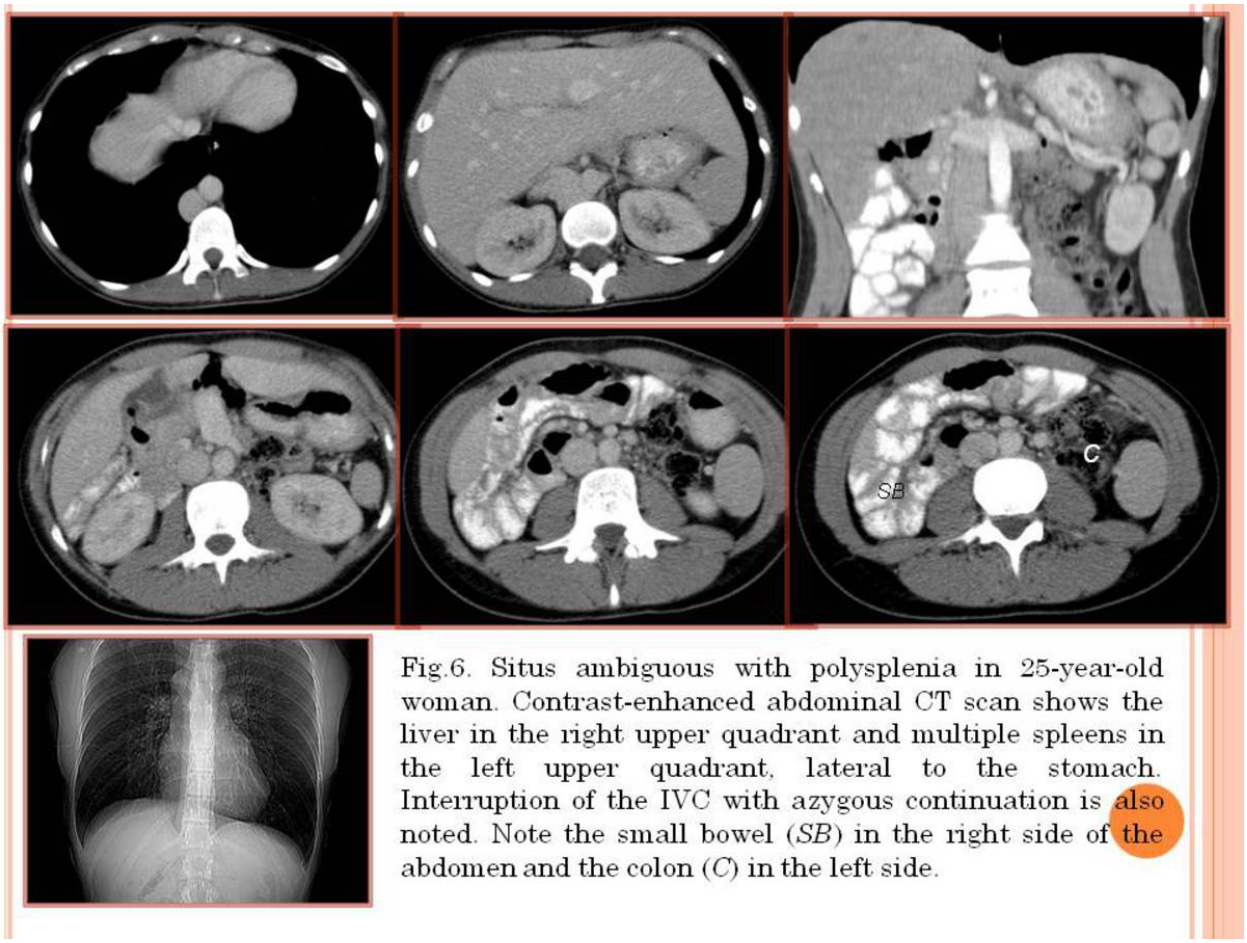


Fig.6. Situs ambiguous with polysplenia in 25-year-old woman. Contrast-enhanced abdominal CT scan shows the liver in the right upper quadrant and multiple spleens in the left upper quadrant, lateral to the stomach. Interruption of the IVC with azygous continuation is also noted. Note the small bowel (*SB*) in the right side of the abdomen and the colon (*C*) in the left side.

Fig. 17: 17

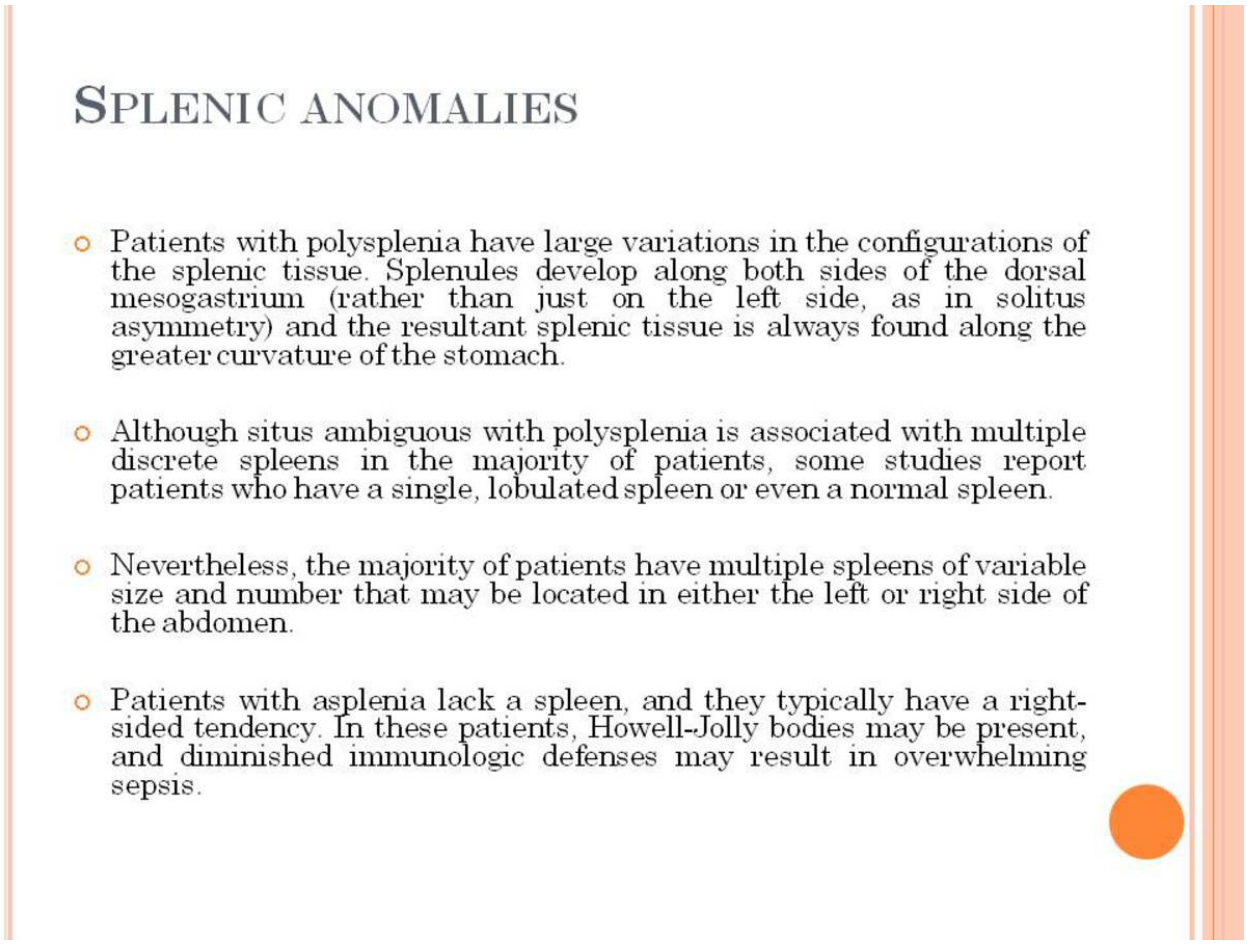

SPLENIC ANOMALIES
Patients with polysplenia have large variations in the configurations of the splenic tissue. Splenules develop along both sides of the dorsal mesogastrium (rather than just on the left side, as in solitus asymmetry) and the resultant splenic tissue is always found along the greater curvature of the stomach.
Although situs ambiguous with polysplenia is associated with multiple discrete spleens in the majority of patients, some studies report patients who have a single, lobulated spleen or even a normal spleen.
Nevertheless, the majority of patients have multiple spleens of variable size and number that may be located in either the left or right side of the abdomen.
Patients with asplenia lack a spleen, and they typically have a right-sided tendency. In these patients, Howell-Jolly bodies may be present, and diminished immunologic defenses may result in overwhelming sepsis.

Fig. 18: 18

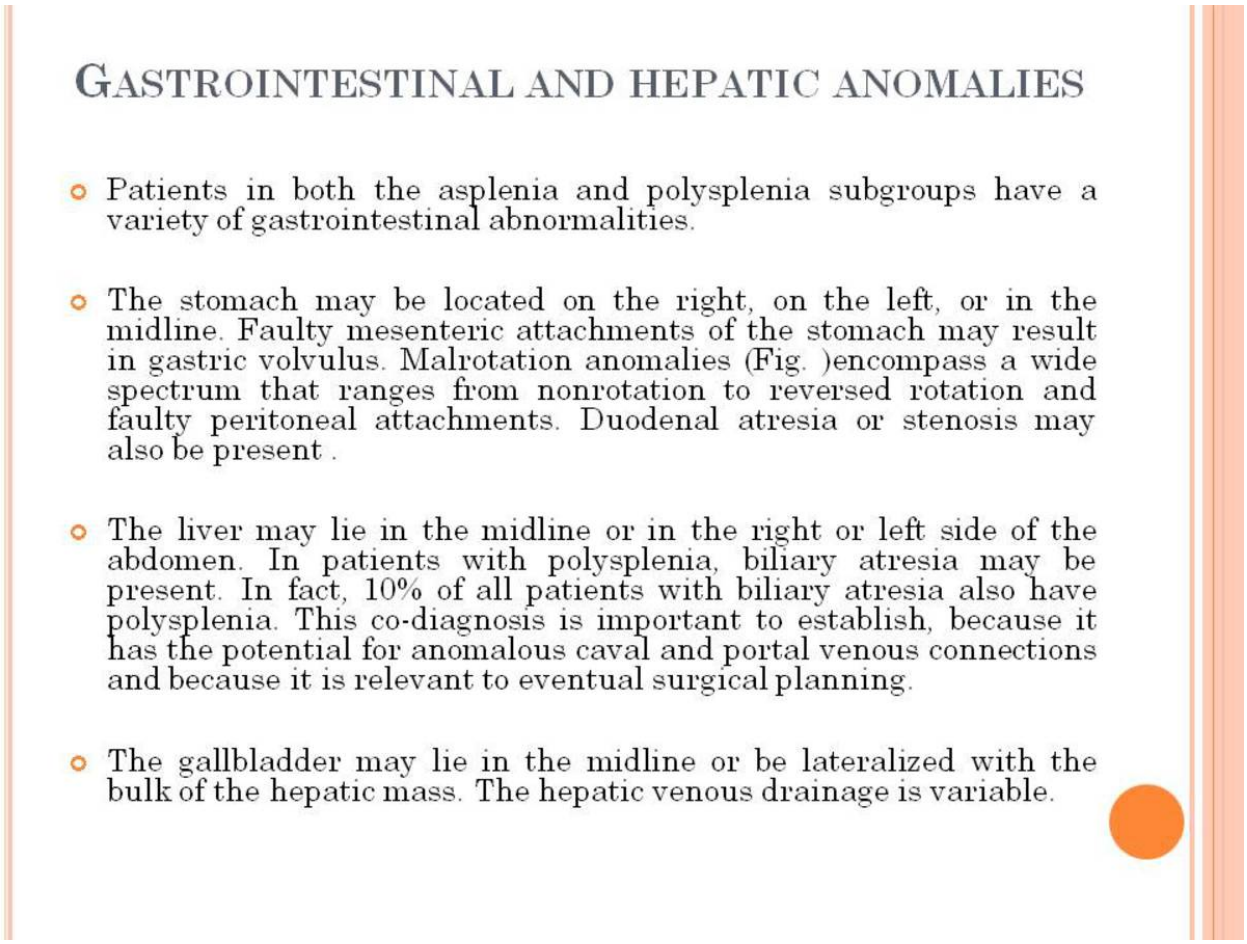

GASTROINTESTINAL AND HEPATIC ANOMALIES
Patients in both the asplenia and polysplenia subgroups have a variety of gastrointestinal abnormalities.
The stomach may be located on the right, on the left, or in the midline. Faulty mesenteric attachments of the stomach may result in gastric volvulus. Malrotation anomalies (Fig.)encompass a wide spectrum that ranges from nonrotation to reversed rotation and faulty peritoneal attachments. Duodenal atresia or stenosis may also be present .
The liver may lie in the midline or in the right or left side of the abdomen. In patients with polysplenia, biliary atresia may be present. In fact, 10% of all patients with biliary atresia also have polysplenia. This co-diagnosis is important to establish, because it has the potential for anomalous caval and portal venous connections and because it is relevant to eventual surgical planning.
The gallbladder may lie in the midline or be lateralized with the bulk of the hepatic mass. The hepatic venous drainage is variable.

Fig. 19: 19

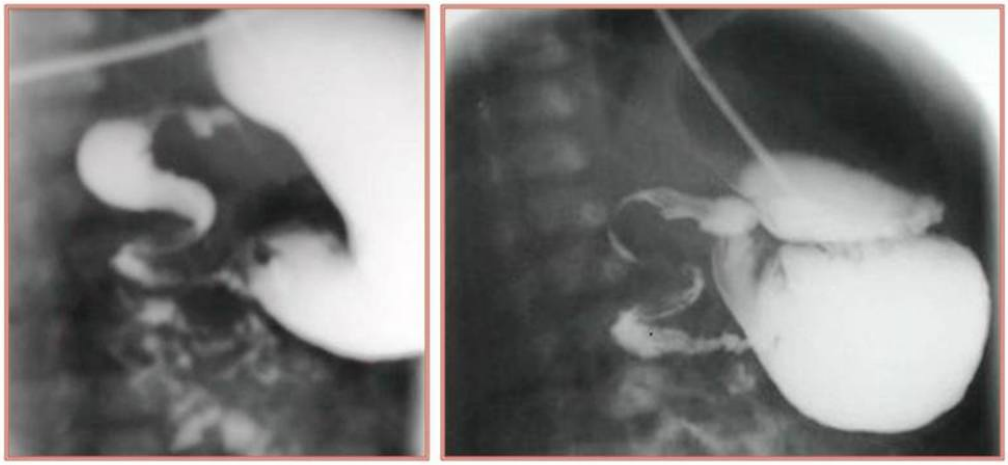


Fig.7. Malrotation with midgut volvulus. In this upper GI series the duodenum does not cross the midline and the contrast tapers in a corkscrew pattern.

Fig. 20: 20

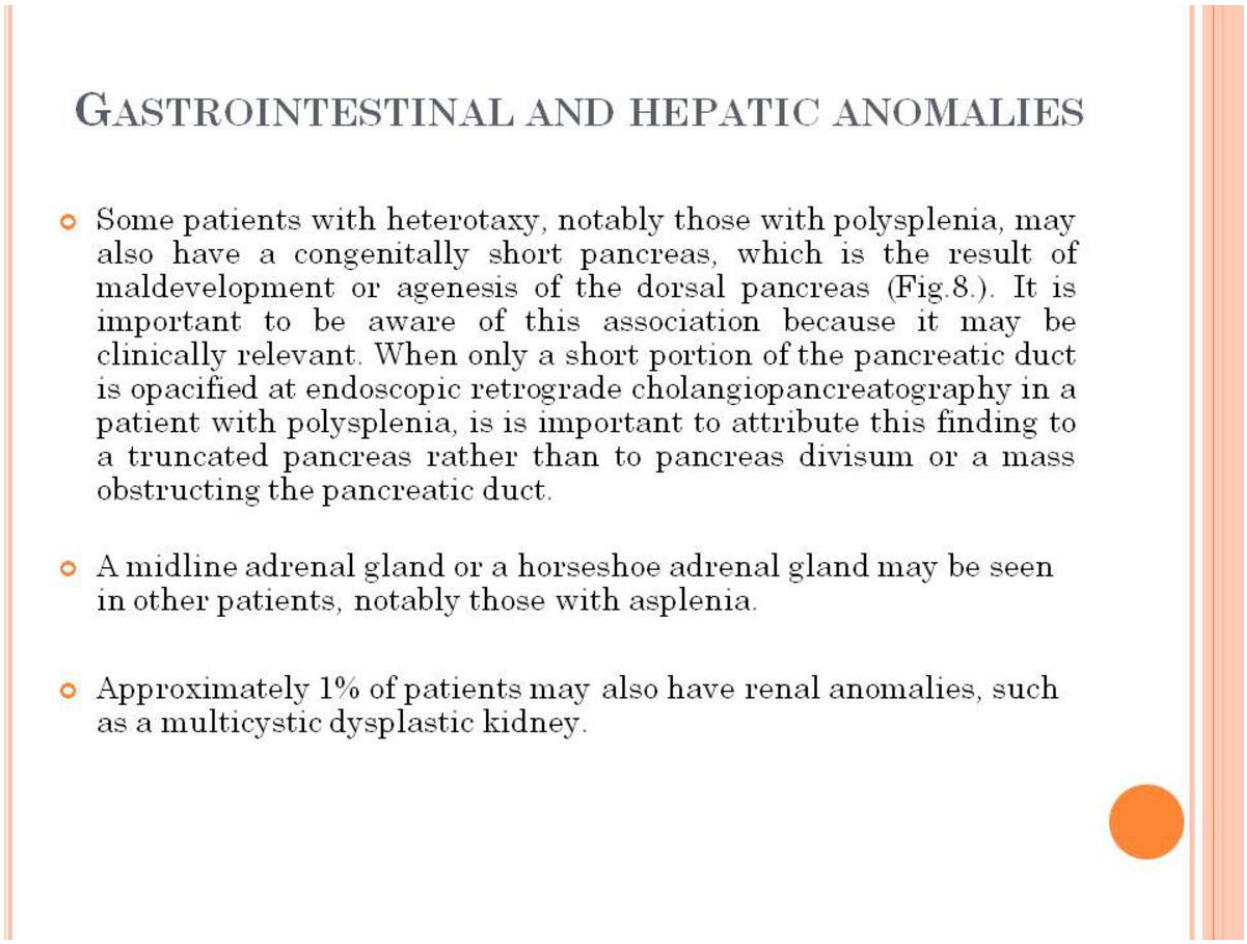

GASTROINTESTINAL AND HEPATIC ANOMALIES
Some patients with heterotaxy, notably those with polysplenia, may also have a congenitally short pancreas, which is the result of maldevelopment or agenesis of the dorsal pancreas (Fig.8.). It is important to be aware of this association because it may be clinically relevant. When only a short portion of the pancreatic duct is opacified at endoscopic retrograde cholangiopancreatography in a patient with polysplenia, is is important to attribute this finding to a truncated pancreas rather than to pancreas divisum or a mass obstructing the pancreatic duct.
A midline adrenal gland or a horseshoe adrenal gland may be seen in other patients, notably those with asplenia.
Approximately 1% of patients may also have renal anomalies, such as a multicystic dysplastic kidney.

Fig. 21: 21

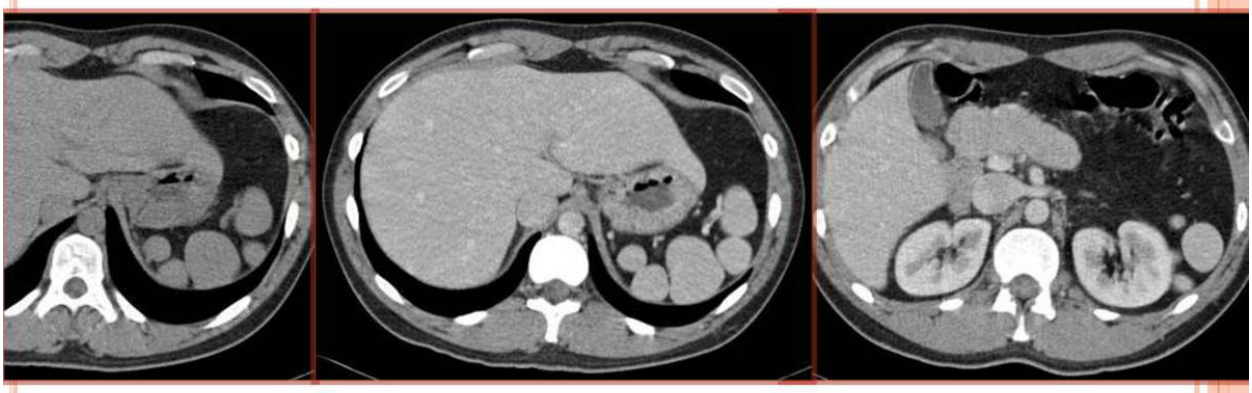


Fig.8. Polysplenia syndrome. Axial pre and post-contrast CT images show multiple round, soft tissue masses of different sizes in the left upper quadrant – splenules. A short pancreas ("truncated pancreas"), consisting of the head and a small portion of the body, is seen.

Fig. 22: 22

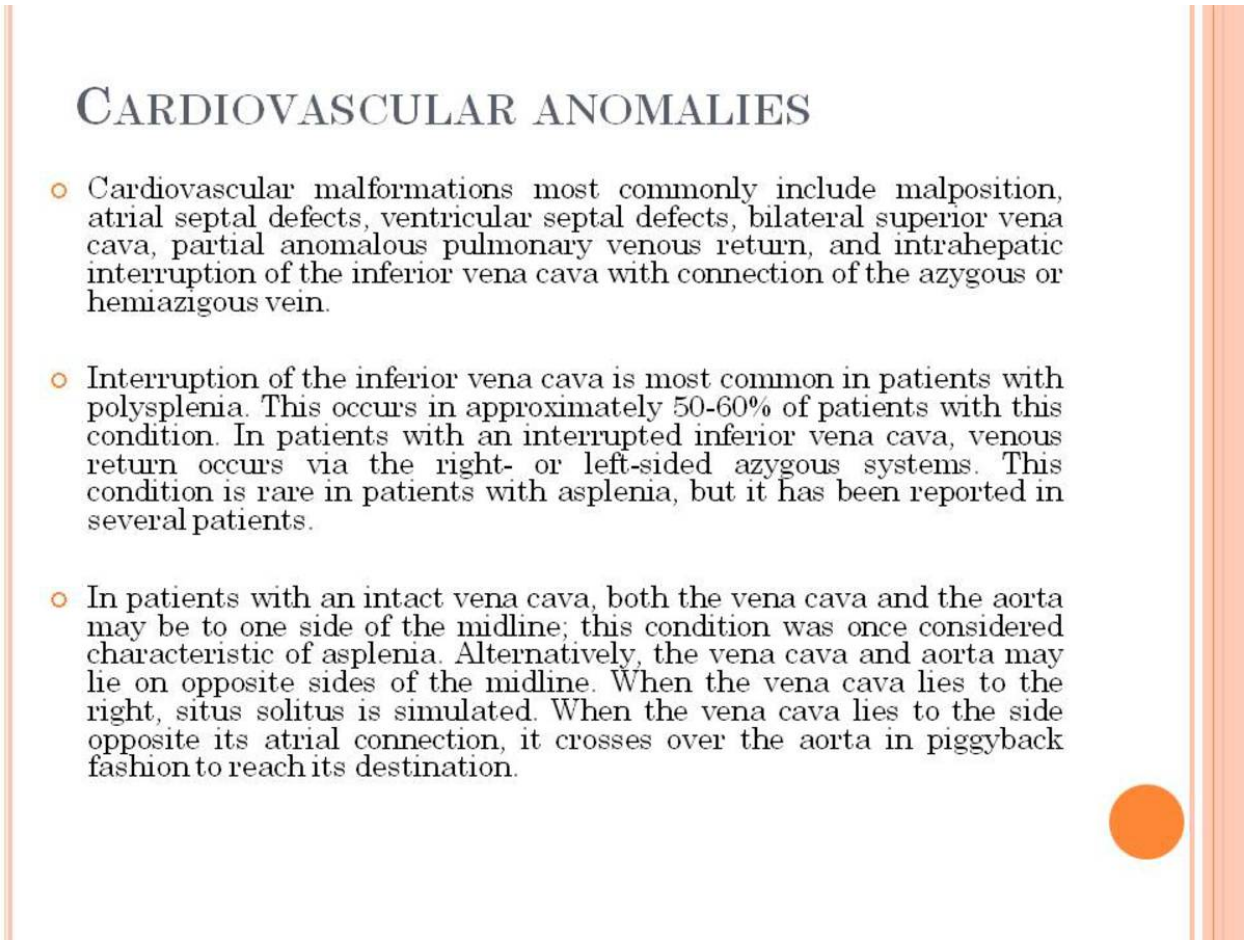

CARDIOVASCULAR ANOMALIES
Cardiovascular malformations most commonly include malposition, atrial septal defects, ventricular septal defects, bilateral superior vena cava, partial anomalous pulmonary venous return, and intrahepatic interruption of the inferior vena cava with connection of the azygous or hemiazigous vein.
Interruption of the inferior vena cava is most common in patients with polysplenia. This occurs in approximately 50-60% of patients with this condition. In patients with an interrupted inferior vena cava, venous return occurs via the right- or left-sided azygous systems. This condition is rare in patients with asplenia, but it has been reported in several patients.
In patients with an intact vena cava, both the vena cava and the aorta may be to one side of the midline; this condition was once considered characteristic of asplenia. Alternatively, the vena cava and aorta may lie on opposite sides of the midline. When the vena cava lies to the right, situs solitus is simulated. When the vena cava lies to the side opposite its atrial connection, it crosses over the aorta in piggyback fashion to reach its destination.

Fig. 23: 23

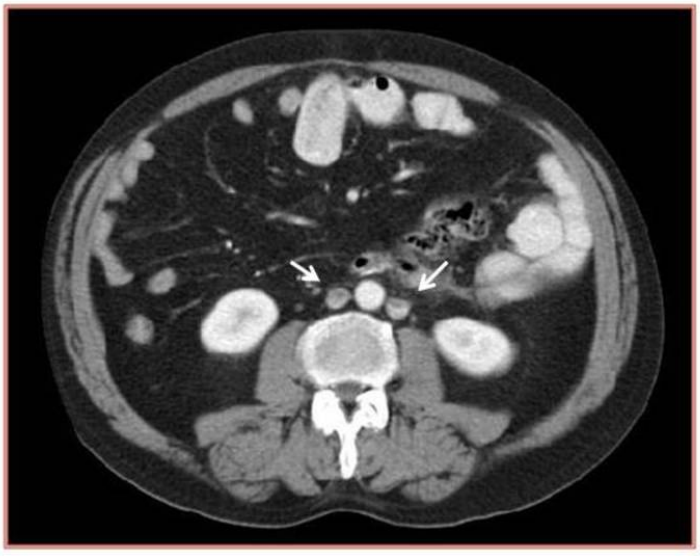


Fig.9. Post-contrast CT scan shows duplication of the infrarenal inferior vena cava in a situs ambiguous.

Fig. 24: 24

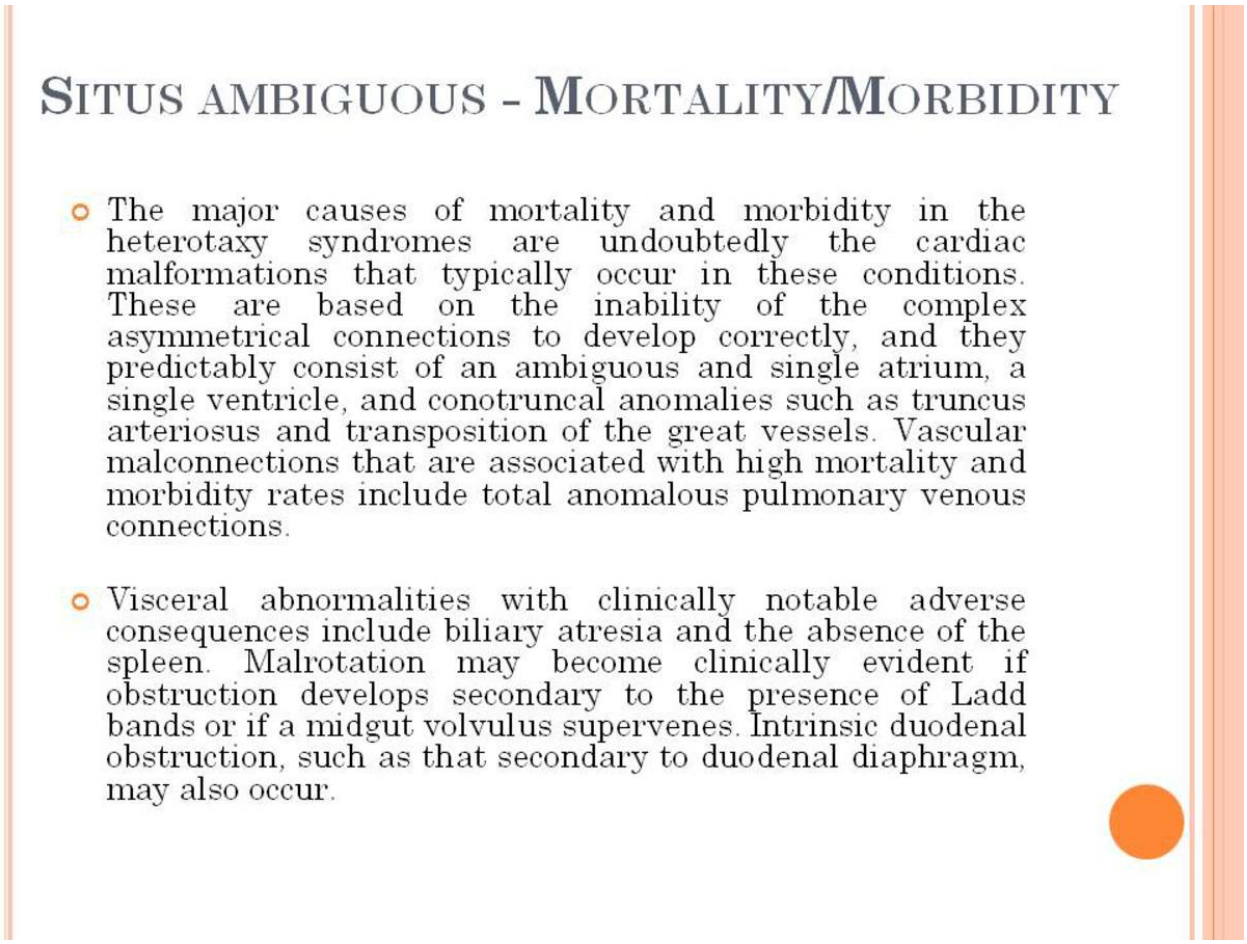

SITUS AMBIGUOUS - MORTALITY/MORBIDITY
The major causes of mortality and morbidity in the heterotaxy syndromes are undoubtedly the cardiac malformations that typically occur in these conditions. These are based on the inability of the complex asymmetrical connections to develop correctly, and they predictably consist of an ambiguous and single atrium, a single ventricle, and conotruncal anomalies such as truncus arteriosus and transposition of the great vessels. Vascular malconnections that are associated with high mortality and morbidity rates include total anomalous pulmonary venous connections.
Visceral abnormalities with clinically notable adverse consequences include biliary atresia and the absence of the spleen. Malrotation may become clinically evident if obstruction develops secondary to the presence of Ladd bands or if a midgut volvulus supervenes. Intrinsic duodenal obstruction, such as that secondary to duodenal diaphragm, may also occur.

Fig. 25: 25

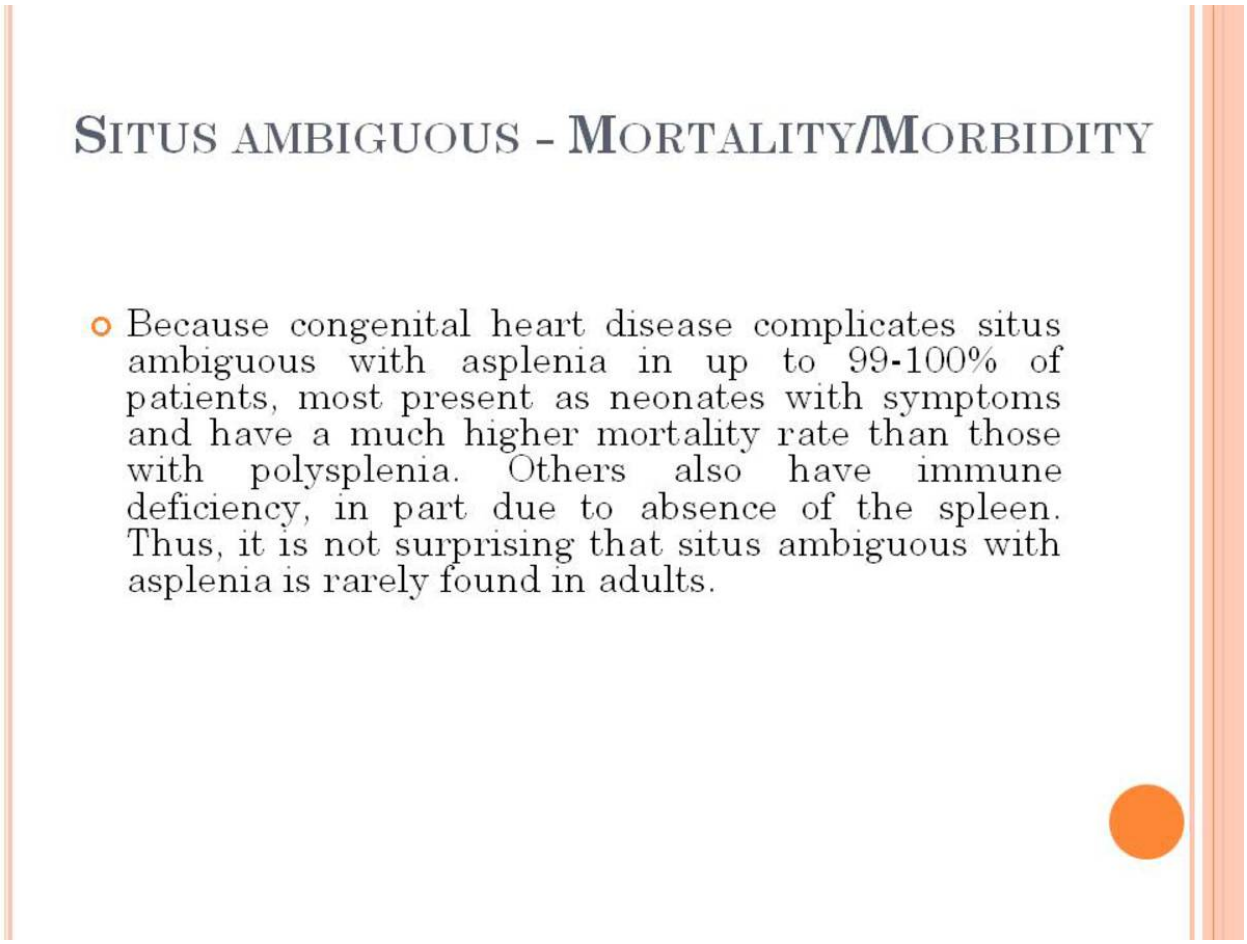

SITUS AMBIGUOUS - MORTALITY/MORBIDITY
Because congenital heart disease complicates situs ambiguous with asplenia in up to 99-100% of patients, most present as neonates with symptoms and have a much higher mortality rate than those with polysplenia. Others also have immune deficiency, in part due to absence of the spleen. Thus, it is not surprising that situs ambiguous with asplenia is rarely found in adults.

Fig. 26: 26

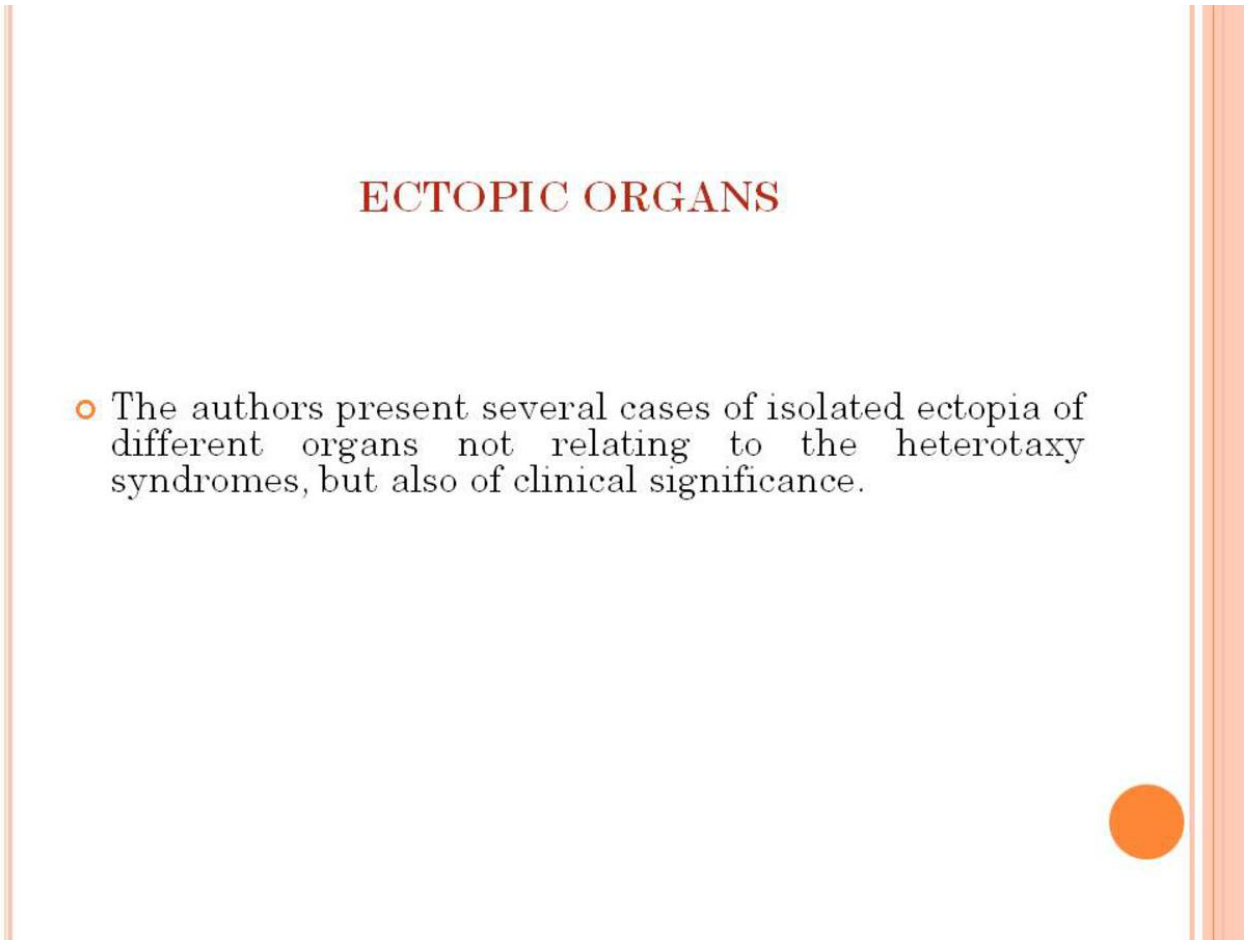

ECTOPIC ORGANS
The authors present several cases of isolated ectopia of different organs not relating to the heterotaxy syndromes, but also of clinical significance.

Fig. 27: 27

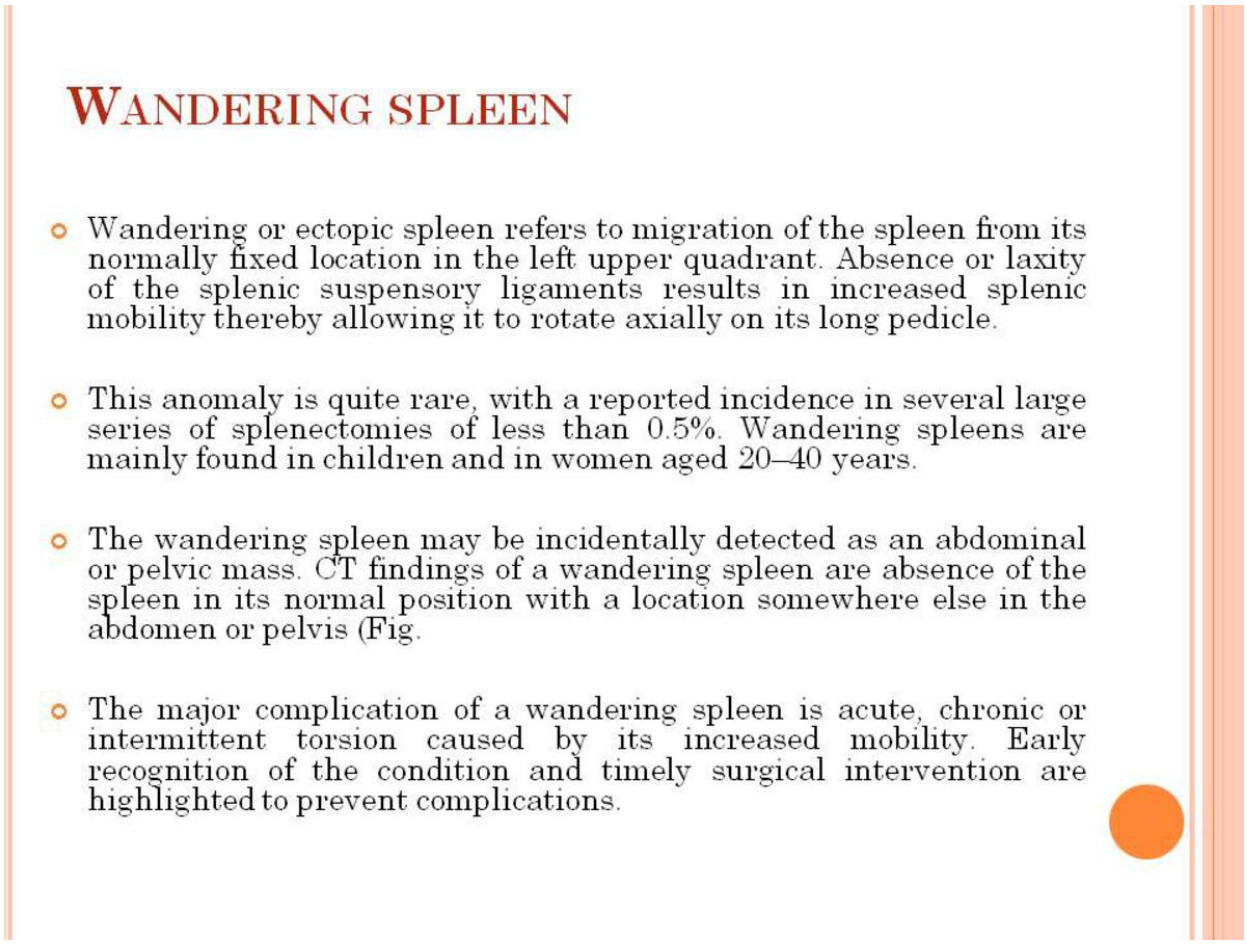

WANDERING SPLEEN
Wandering or ectopic spleen refers to migration of the spleen from its normally fixed location in the left upper quadrant. Absence or laxity of the splenic suspensory ligaments results in increased splenic mobility thereby allowing it to rotate axially on its long pedicle.
This anomaly is quite rare, with a reported incidence in several large series of splenectomies of less than 0.5%. Wandering spleens are mainly found in children and in women aged 20–40 years.
The wandering spleen may be incidentally detected as an abdominal or pelvic mass. CT findings of a wandering spleen are absence of the spleen in its normal position with a location somewhere else in the abdomen or pelvis (Fig.
The major complication of a wandering spleen is acute, chronic or intermittent torsion caused by its increased mobility. Early recognition of the condition and timely surgical intervention are highlighted to prevent complications.

Fig. 28: 28

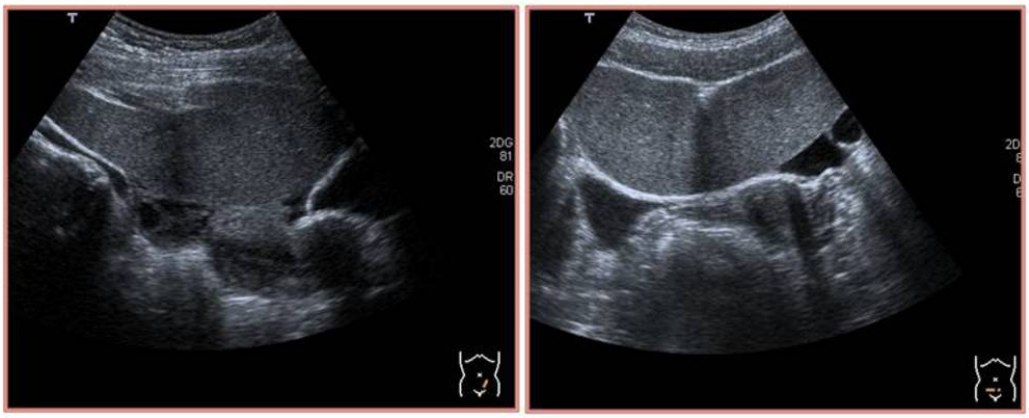


Fig.10. Wandering spleen. Ultrasound images show the spleen in an abnormal position, in the pelvis, adjacent to the uterus and left ovary.

Fig. 29: 29

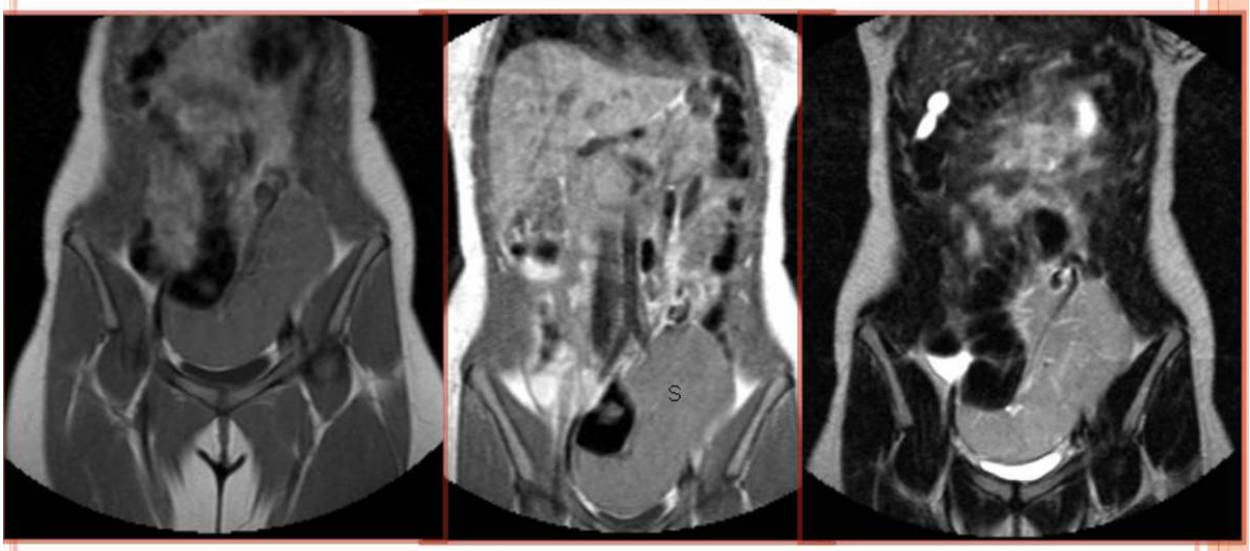


Fig.11. Wandering spleen. Coronal T1- and T2-wheighted MR images show the spleen (*S*) located at the pelvis.

Fig. 30: 30

ECTOPIC SPLEEN

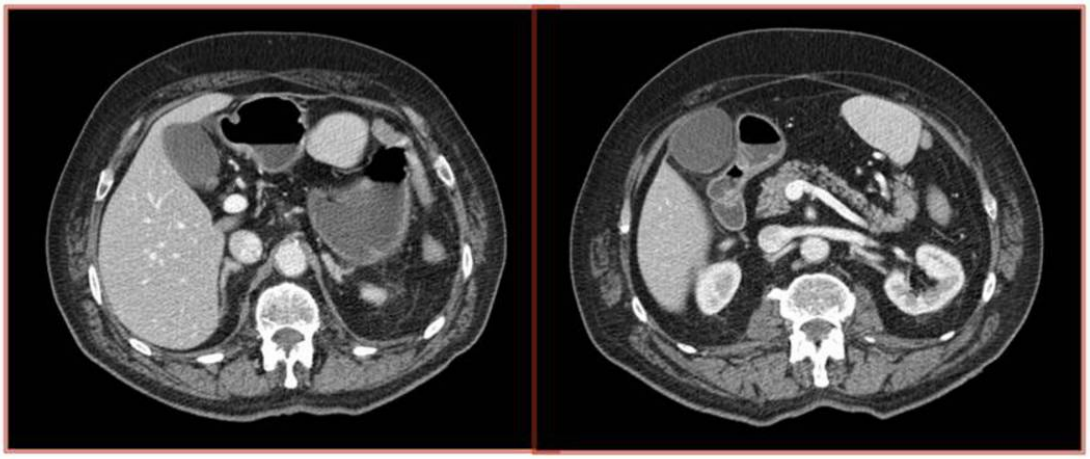


Fig.12. Enhanced CT scan shows the spleen in an unusual topography, posterior to the anterior abdominal wall, to the left of the midline and superiorly bounded by the gastric wall.

Fig. 31: 31

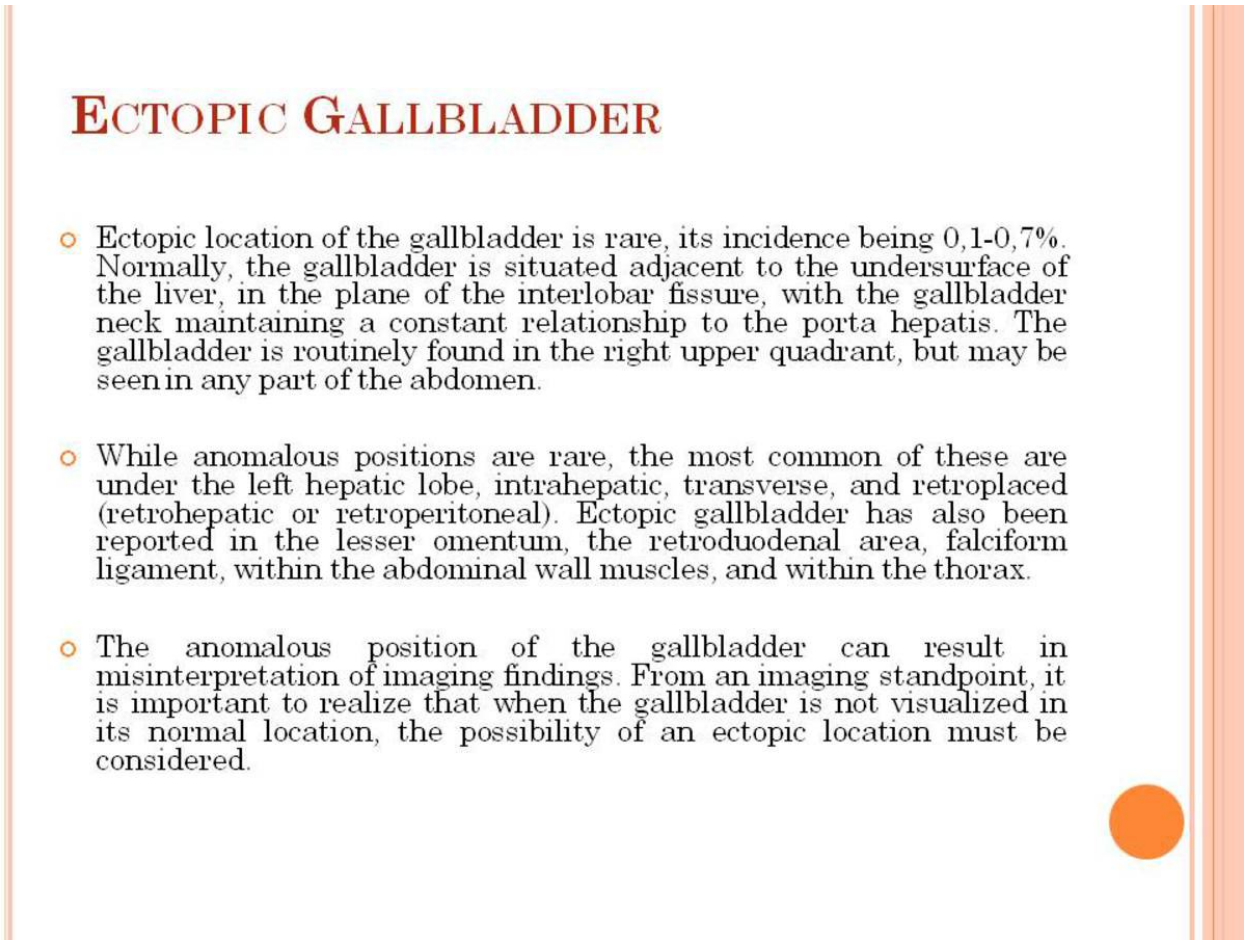

ECTOPIC GALLBLADDER
Ectopic location of the gallbladder is rare, its incidence being 0,1-0,7%. Normally, the gallbladder is situated adjacent to the undersurface of the liver, in the plane of the interlobar fissure, with the gallbladder neck maintaining a constant relationship to the porta hepatis. The gallbladder is routinely found in the right upper quadrant, but may be seen in any part of the abdomen.
While anomalous positions are rare, the most common of these are under the left hepatic lobe, intrahepatic, transverse, and retroplaced (retrohepatic or retroperitoneal). Ectopic gallbladder has also been reported in the lesser omentum, the retroduodenal area, falciform ligament, within the abdominal wall muscles, and within the thorax.
The anomalous position of the gallbladder can result in misinterpretation of imaging findings. From an imaging standpoint, it is important to realize that when the gallbladder is not visualized in its normal location, the possibility of an ectopic location must be considered.

Fig. 32: 32

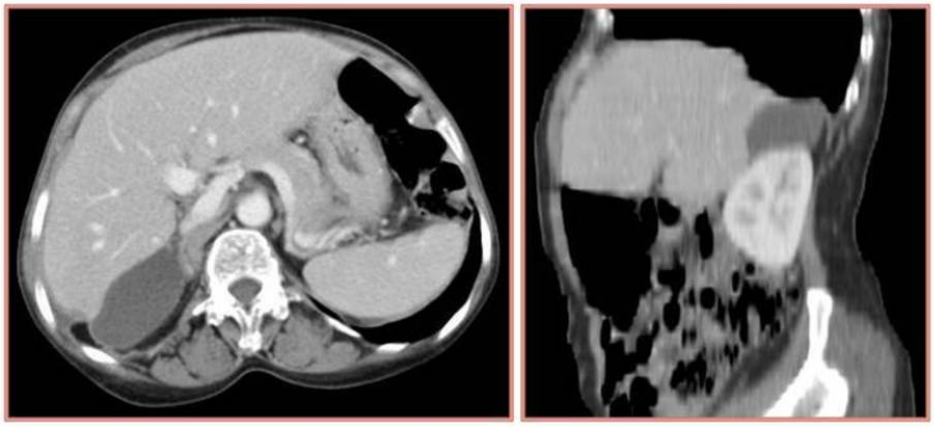


Fig.13. Axial and sagital CT images show an ectopic location of the gallbladder, situated posterior to the right hepatic lobe and superior to the right kidney.

Fig. 33: 33

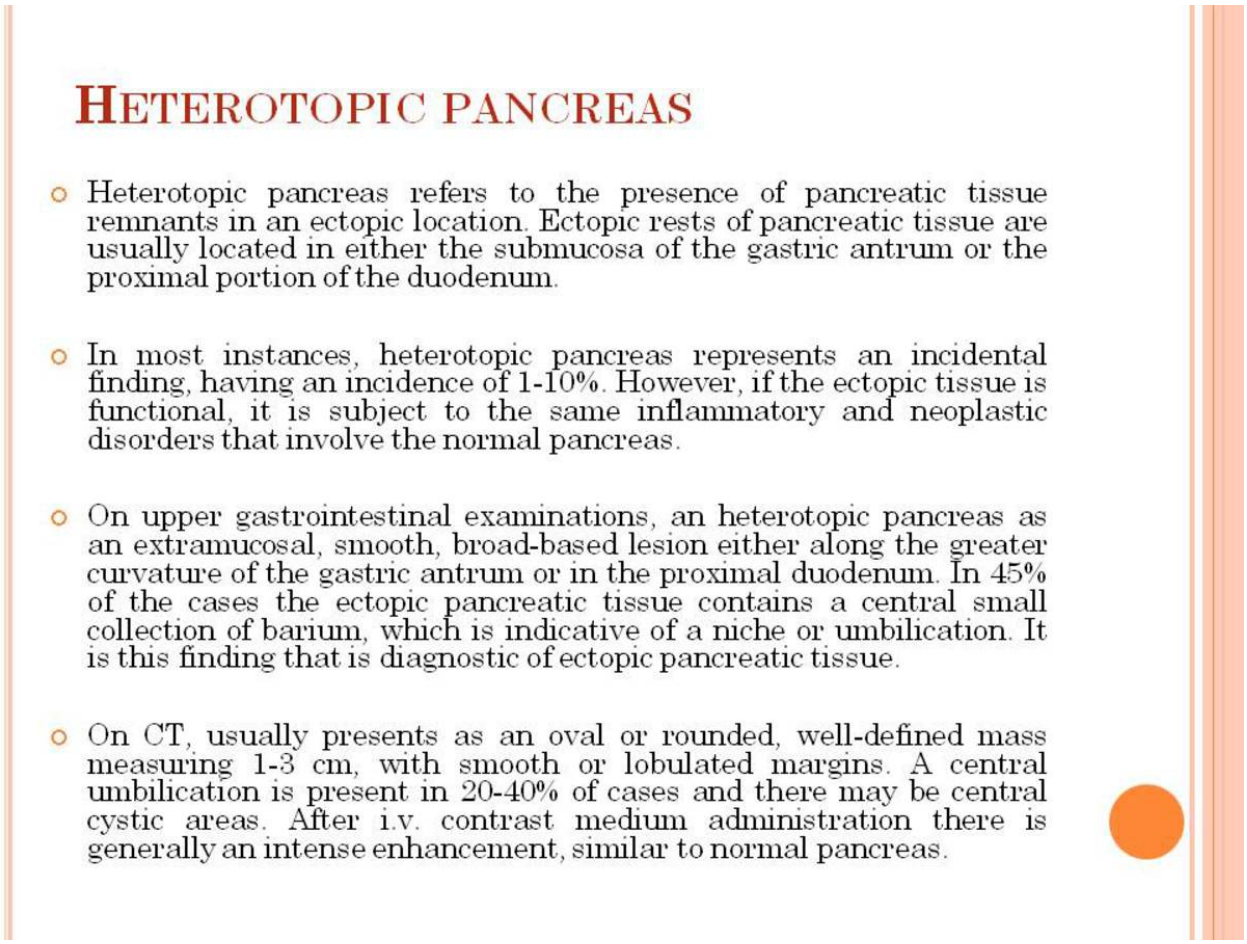

HETEROTOPIC PANCREAS
Heterotopic pancreas refers to the presence of pancreatic tissue remnants in an ectopic location. Ectopic rests of pancreatic tissue are usually located in either the submucosa of the gastric antrum or the proximal portion of the duodenum.
In most instances, heterotopic pancreas represents an incidental finding, having an incidence of 1-10%. However, if the ectopic tissue is functional, it is subject to the same inflammatory and neoplastic disorders that involve the normal pancreas.
On upper gastrointestinal examinations, an heterotopic pancreas as an extramucosal, smooth, broad-based lesion either along the greater curvature of the gastric antrum or in the proximal duodenum. In 45% of the cases the ectopic pancreatic tissue contains a central small collection of barium, which is indicative of a niche or umbilication. It is this finding that is diagnostic of ectopic pancreatic tissue.
On CT, usually presents as an oval or rounded, well-defined mass measuring 1-3 cm, with smooth or lobulated margins. A central umbilication is present in 20-40% of cases and there may be central cystic areas. After i.v. contrast medium administration there is generally an intense enhancement, similar to normal pancreas.

Fig. 34: 34

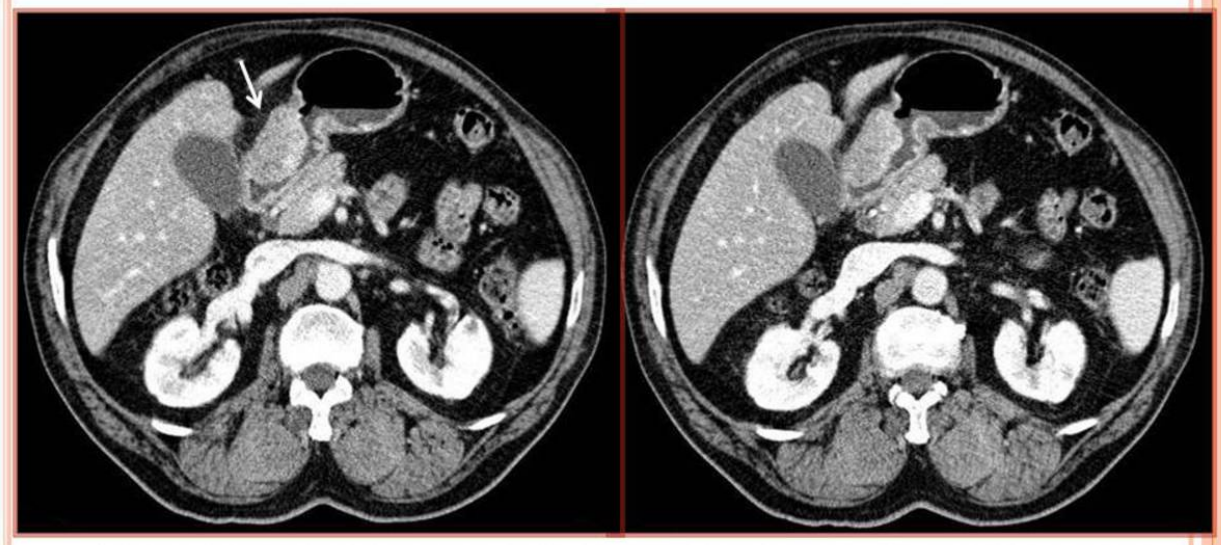


Fig.14. Heterotopic pancreas. Axial contrast enhanced CT images show a focal nodular thickening of the anterior wall of the gastric antrum. Biopsy was performed and pathology revealed an adenocarcinoma in an heterotopic pancreas.

Fig. 35: 35

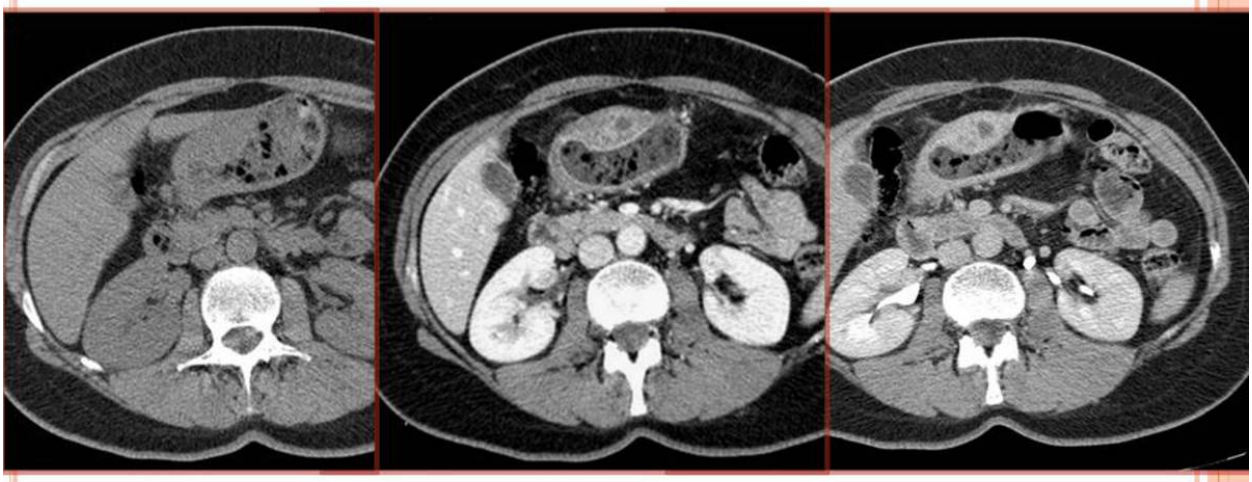


Fig.15. Heterotopic pancreas with signs of inflammation. Pre-contrast CT (a) revealed a biconvex thickening of the anterior wall of the gastric antrum. Post-contrast CT in the portal phase (b) shows heterogenous enhancement of the lesion, with small low-attenuation central areas. In the late phase (c), there is persistent enhancement.

Fig. 36: 36

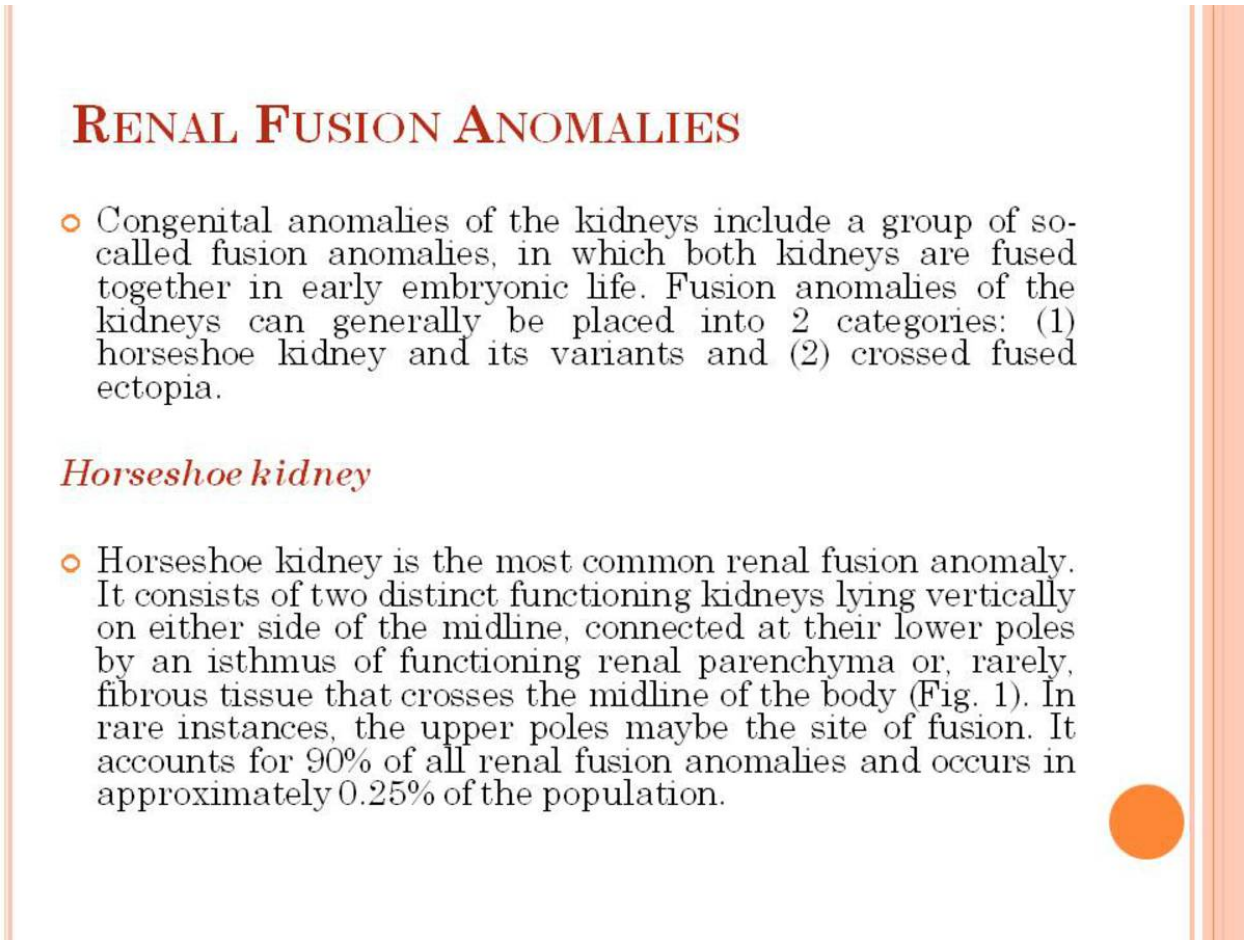

RENAL FUSION ANOMALIES
Congenital anomalies of the kidneys include a group of so-called fusion anomalies, in which both kidneys are fused together in early embryonic life. Fusion anomalies of the kidneys can generally be placed into 2 categories: (1) horseshoe kidney and its variants and (2) crossed fused ectopia.
Horseshoe kidney
Horseshoe kidney is the most common renal fusion anomaly. It consists of two distinct functioning kidneys lying vertically on either side of the midline, connected at their lower poles by an isthmus of functioning renal parenchyma or, rarely, fibrous tissue that crosses the midline of the body (Fig. 1). In rare instances, the upper poles maybe the site of fusion. It accounts for 90% of all renal fusion anomalies and occurs in approximately 0.25% of the population.

Fig. 37: 37

HORSESHOE KIDNEY

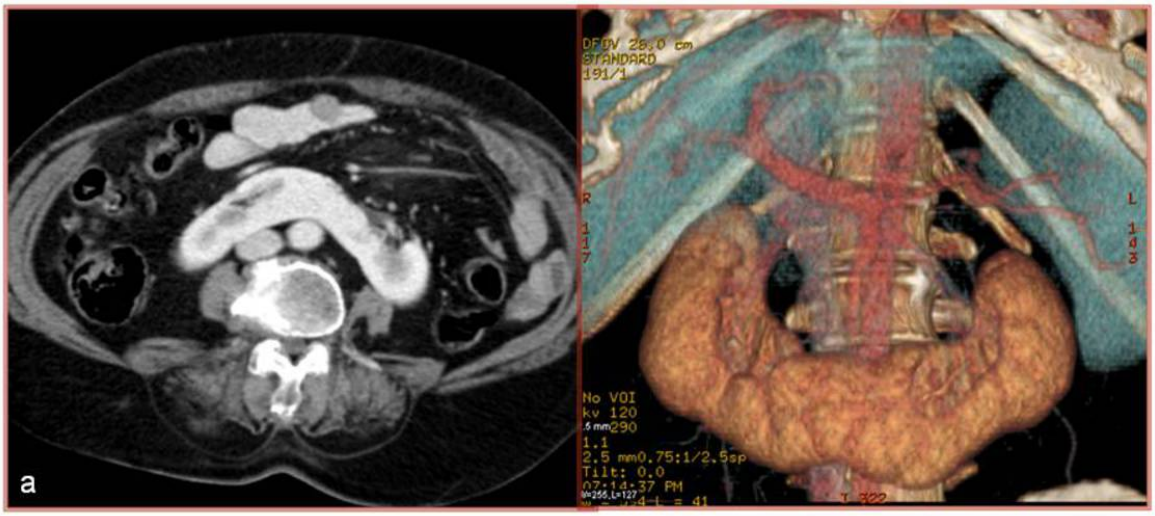


Fig.16. a, b. Axial CT (a) and coronal volume rendering (b) images show horseshoe kidneys that are fused with functioning renal parenchyma at their lower poles. The isthmus lies anterior to the aorta.

Fig. 38: 38

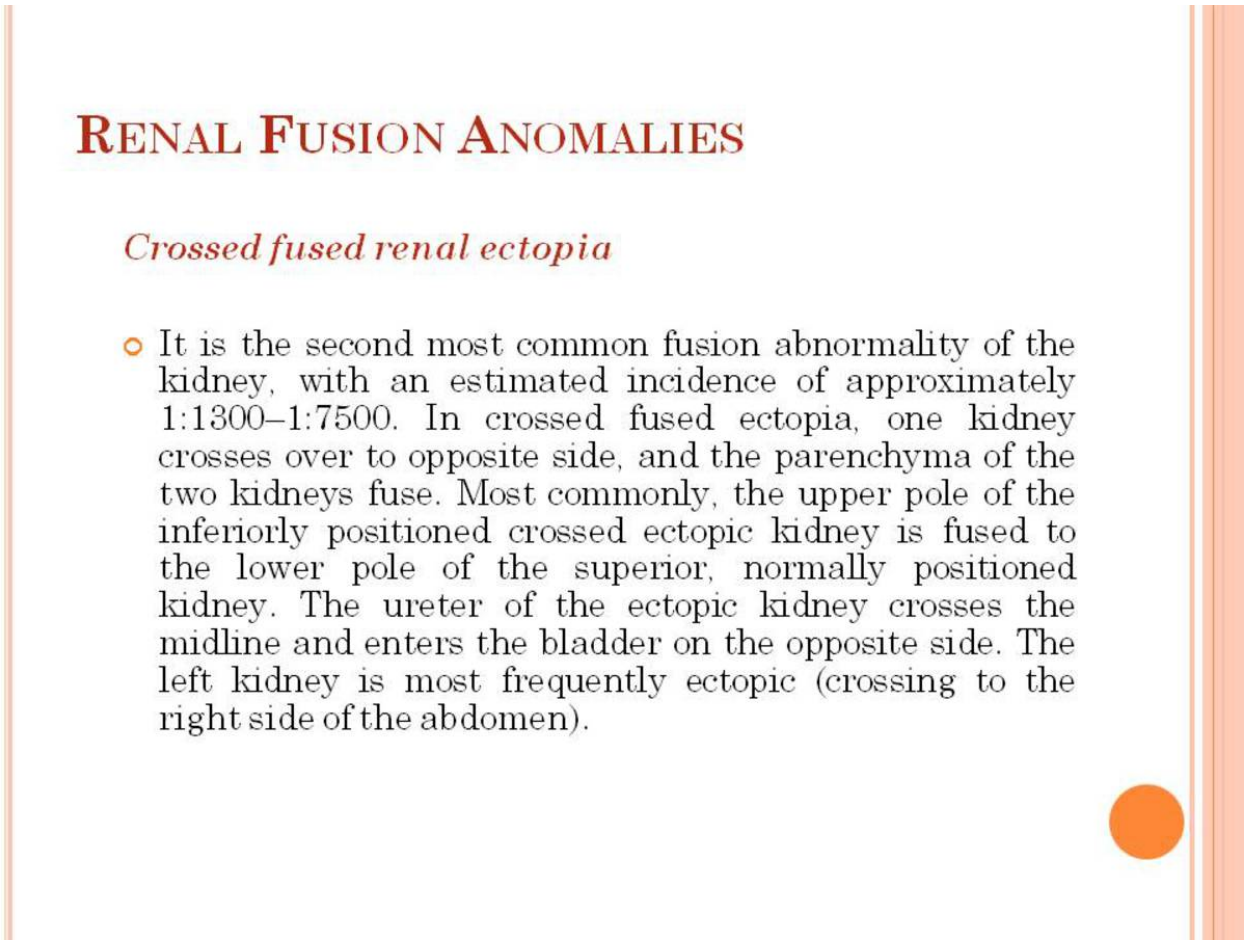

RENAL FUSION ANOMALIES
Crossed fused renal ectopia
It is the second most common fusion abnormality of the kidney, with an estimated incidence of approximately 1:1300–1:7500. In crossed fused ectopia, one kidney crosses over to opposite side, and the parenchyma of the two kidneys fuse. Most commonly, the upper pole of the inferiorly positioned crossed ectopic kidney is fused to the lower pole of the superior, normally positioned kidney. The ureter of the ectopic kidney crosses the midline and enters the bladder on the opposite side. The left kidney is most frequently ectopic (crossing to the right side of the abdomen).

Fig. 39: 39

CROSSED FUSED RENAL ECTOPIA

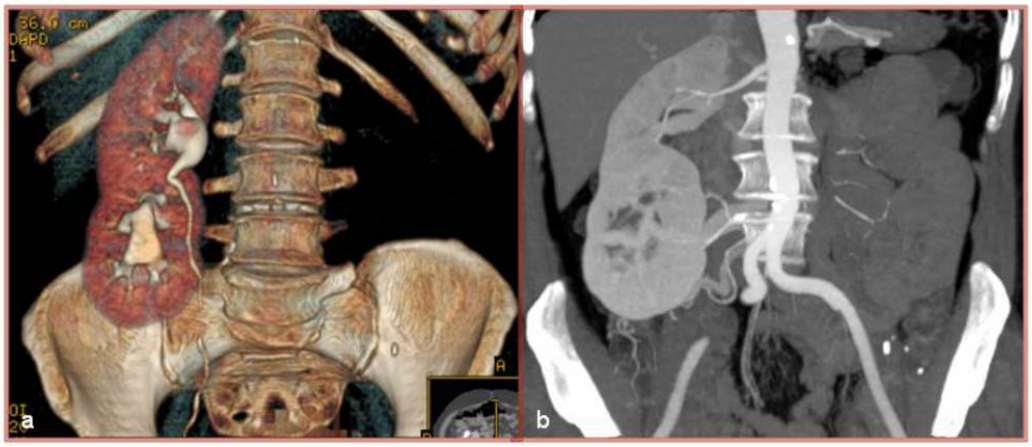


Fig.17. a, b. Coronal volume rendering (a) and maximum intensity projection (b) CT images show crossed fused renal ectopia on the right side.

Fig. 40: 40

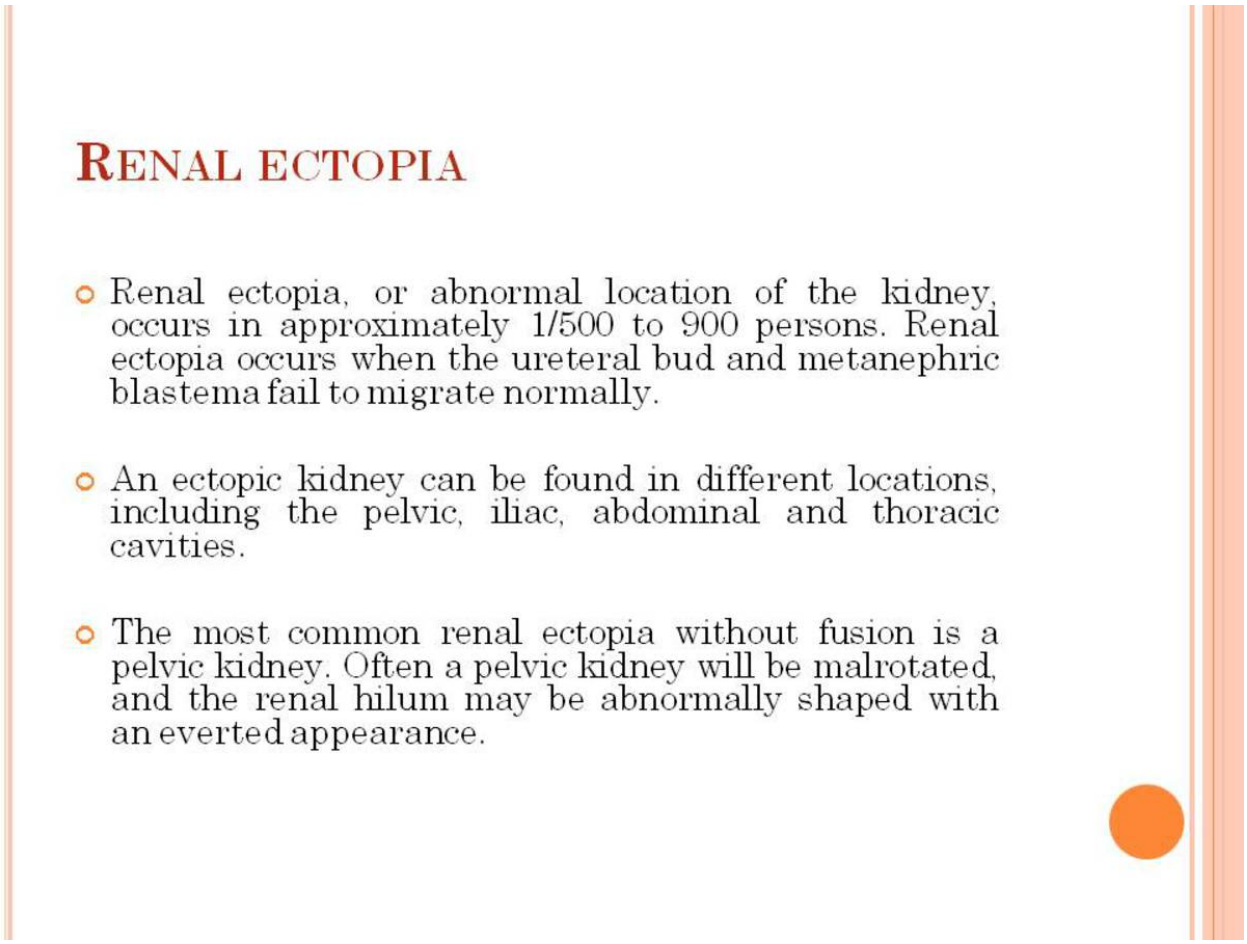

RENAL ECTOPIA
Renal ectopia, or abnormal location of the kidney, occurs in approximately 1/500 to 900 persons. Renal ectopia occurs when the ureteral bud and metanephric blastema fail to migrate normally.
An ectopic kidney can be found in different locations, including the pelvic, iliac, abdominal and thoracic cavities.
The most common renal ectopia without fusion is a pelvic kidney. Often a pelvic kidney will be malrotated, and the renal hilum may be abnormally shaped with an everted appearance.

Fig. 41: 41

SOLITARY PELVIC KIDNEY

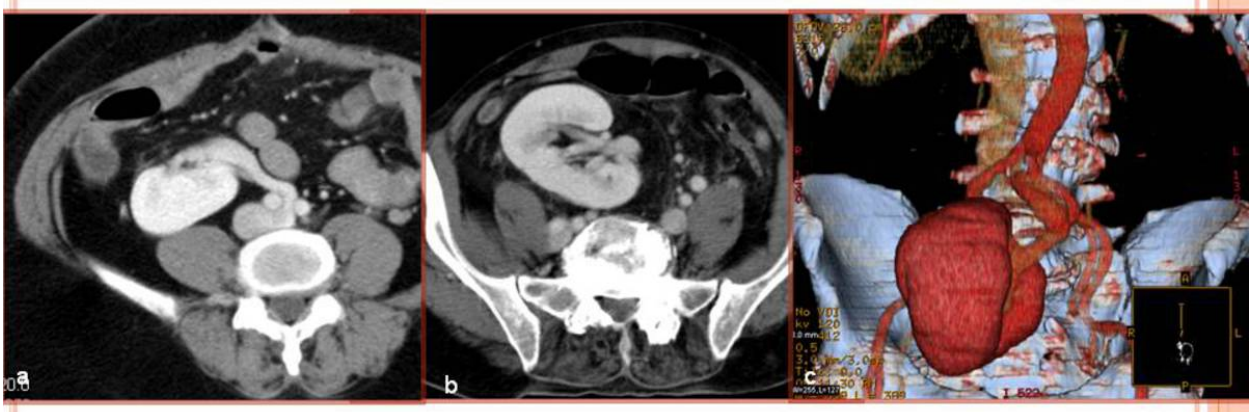


Fig.18. Contrast-enhanced CT (a,b) and coronal oblique volume rendering (c) images show a distorted, malrotated solitary pelvic kidney.

Fig. 42: 42

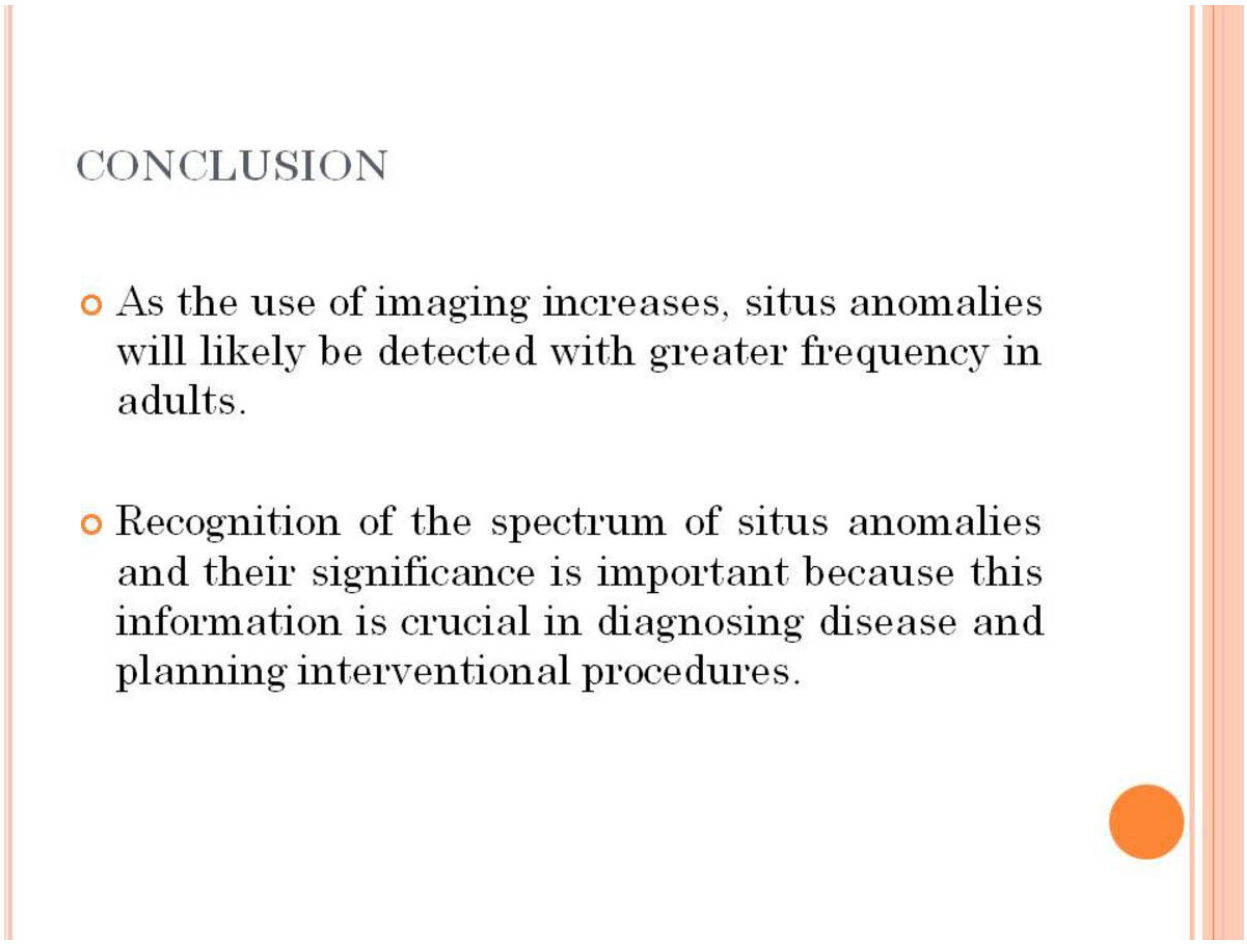

CONCLUSION
As the use of imaging increases, situs anomalies will likely be detected with greater frequency in adults.
Recognition of the spectrum of situs anomalies and their significance is important because this information is crucial in diagnosing disease and planning interventional procedures.

Fig. 43: 43

Conclusion

Recognition of the spectrum of situs anomalies and their significance is important because this information is crucial in diagnosing disease and planning interventional procedures.

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