

Situs ambiguous and Hepatic Vein Anomaly: A Case Report

Situs ambiguous y Anomalía de la Vena Hepática: Informe de un Caso

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SUMMARY: *Situs ambiguous* is the placement of vessels and organs in the thoracoabdominal space that are anatomically located outside its normal position in a certain order. This condition is a broad definition that includes many variations. In this case report, we reported a patient with *Situs ambiguous* with an abnormal hepatic vein who was diagnosed incidentally during medical imaging (computed tomography, sonography and MRI study).

KEY WORDS: *Situs ambiguous*; Left isomerism; Hepatic vein anomaly.

INTRODUCTION

Situs solitus describes the condition in which the heart and viscera are in their normal position. *Situs inversus* determines the situation in which organs are placed in a mirror image relative to *situs solitus* (Ghosh *et al.*, 2009). This term points out that the heart is located on the right side of the thorax, the stomach and spleen on the right side of the abdomen and the liver of the left side. *Situs ambiguous*, is a condition in which the different organs in the thoracoabdominal cavity can be abnormal or malpositioned. It is a broad definition that includes many variations. *Situs ambiguous* occurs around the 28th day of pregnancy as a result of primary lateralization defect and deviation from the normal position of the internal organs (O'Rahilly *et al.*, 1987; Strife *et al.*, 1998).

CASE REPORT

A 40-year-old female patient who applied to Çukurova University Emergency Department with abdominal pain, nausea and shortness of breath had a history of Diabetes mellitus, Hypertension and a 23-year-old ventricular septal defect (VSD) operation. The patient was admitted to the Cardiology Service. Thoracic computed tomography (CT) angiography was performed on the patient due to suspicion of pulmonary embolism. Thoracic CT angiography of the

patient revealed no obvious filling defects compatible with pulmonary embolism. The heart was located in the left hemithorax, and its apex was on the left. The patient had cardiomegaly, and dilatation was observed in all heart chambers. Both main bronchi branched under the pulmonary artery and had a hyperarterial appearance (Figs. 1-2). There were bilateral "left" (bilobed) lungs and both main bronchial branches had a similar appearance (Fig. 3). The patient's carina angle was wide (Fig. 4). Abdominal ultrasonography and contrast-enhanced abdominal MRI were performed to investigate additional anomalies, since the patient had considered *Situs ambiguous* and left isomerism. V. hepatica dexter and v. hepatica intermedia was opened directly to the atrium cordis dextrum (Fig. 5). The vena hepatica sinister joins the vena cava inferior (Figs. 5-6). The vena cava inferior was located to the left of the aorta. The stomach and spleen were located in the upper right quadrant. The liver showed bridging in the middle line and stretched along the right and left sides. (Fig. 7). The spleen was multilobule-shaped, folded in a row, and there was an accessory spleen with a diameter of 1 cm in the spleen superior (Fig. 8). There is a short, round, pancreatic head and pancreatic neck adjacent to the duodenum, which is compatible with dorsal pancreatic hypoplasia and absence of a retromesenteric (retro-peritoneal) D3 segment of the duodenum (Fig. 9), as well as intestinal rotation anomaly (the large bowel was located at left of the midline and the small bowel at right).

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Fig. 1. Arrow heads on the Coronal CT of Thorax show both main bronchi. PA: Pulmonary artery

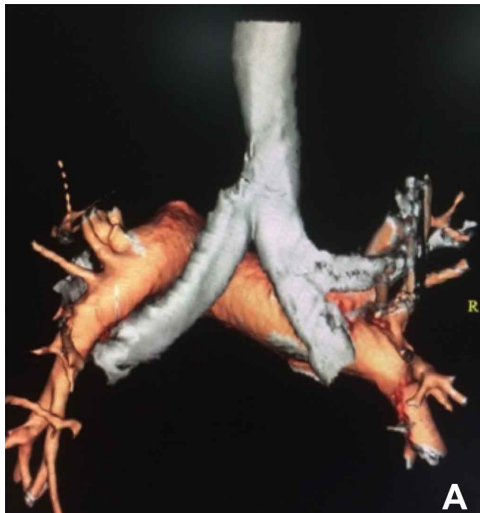


Fig. 2. A. When the 3D reconstruction image taken from the patient with situs solitus is viewed from behind (since the right upper bronchus is superimposed with the pulmonary artery when viewed from the front side) upper right bronchus branches above the right pulmonary artery. The left main bronchus, on the other hand, has a hypoarterial structure and branches below the pulmonary artery. B. In our patient with left isomerism, when the 3-D reconstruction image is viewed from behind, both main bronchi branch below the pulmonary artery.

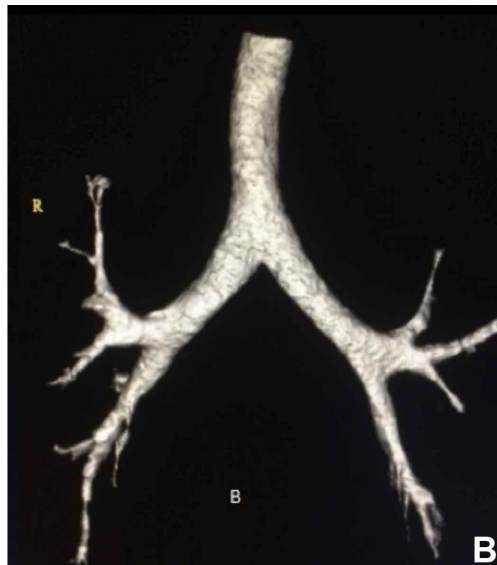


Fig. 3. A. A 3D reconstruction image of the trachea and main bronchi in a patient with situs solitus. B. In the left isomerism, there is an expansion in the carina angle, and both main bronchi are symmetrical to each other, and the main bronchi are shorter and horizontal in structure.

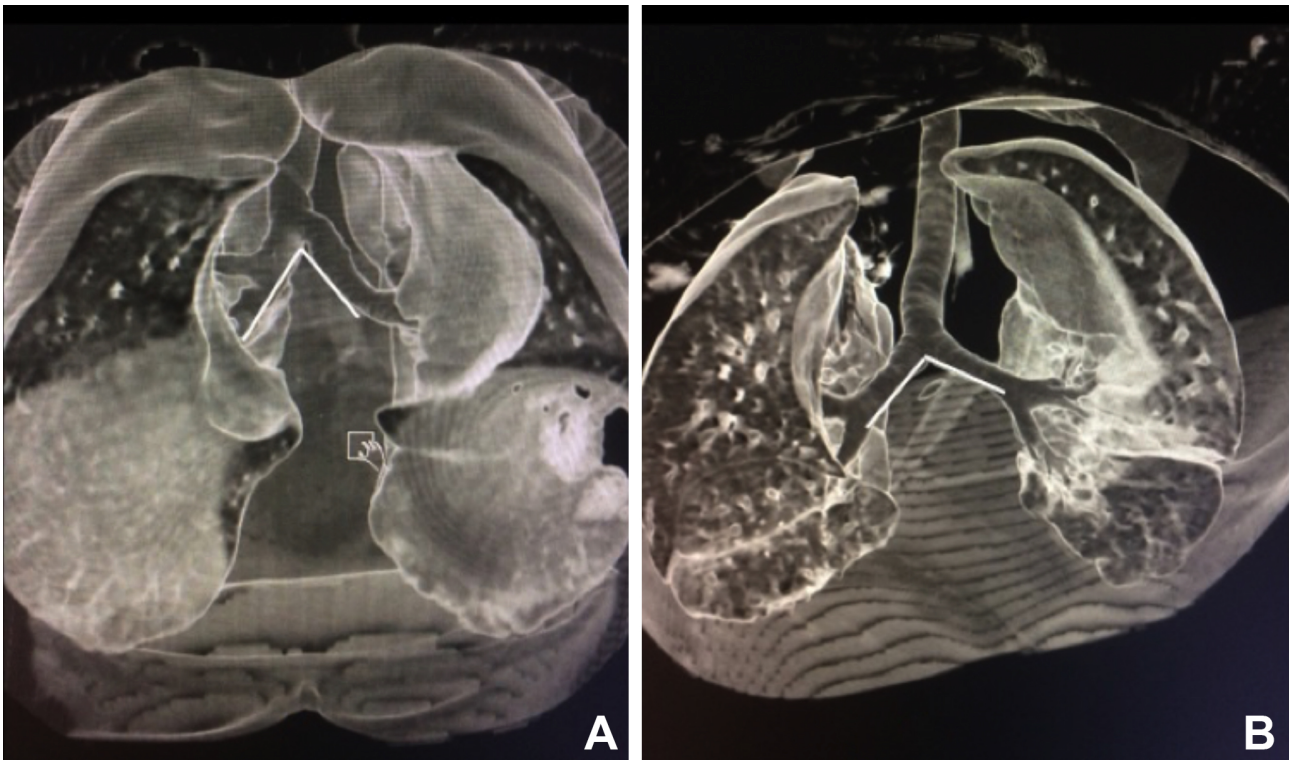


Fig. 4. A. 3D reconstruction image of both lungs and trachea in a patient with situs solitus. B. In left isomerism, the carina angle is wider.

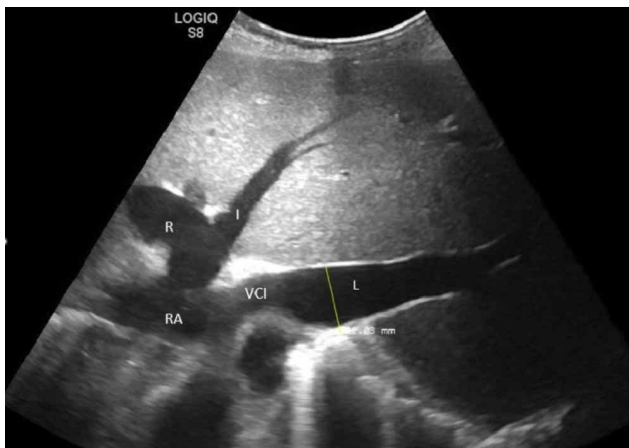


Fig. 5. The left hepatic vein joins the vena cava inferior, while the middle and right hepatic veins open directly into the right atrium. RA: Right atrium, L: left hepatic vein R: right hepatic vein I: intermediate hepatic vein, VCI: vena cava inferior.



Fig. 6. On a contrast-enhanced Coronal Abdominal MRI, the white arrow shows the inferior vena cava, and the black arrow shows the left hepatic vein. The aorta is located to the right of the inferior vena cava.

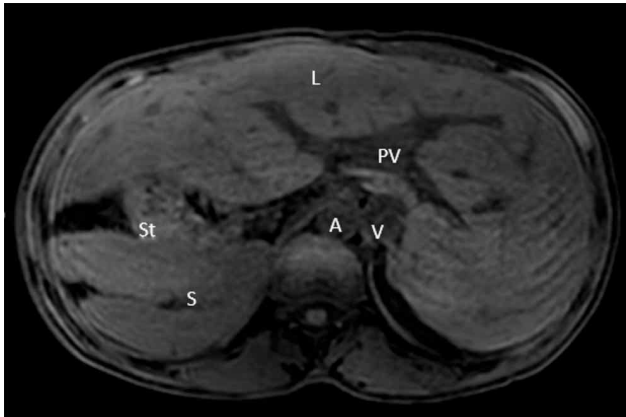


Fig. 7. L: Liver , PV: Portal Vein , St: Stomach , S: Spleen , A: Aorta , V: Vena cava inferior



Fig. 8. The white arrow indicates the superior mesenteric artery (SMA), the black arrow indicates the superior mesenteric vein (SMV), and the SMV was located to the left of SMA.



DISCUSSION

Insights from vertebrate model organisms show that heterotaxia is genetically highly heterogeneous with monogenic and polygenic or multifactorial causes (Peeters & Devriendt, 2006). In addition to genetic causes, it has been shown that environmental factors and teratogens can lead to laterality defects. It is known that maternal diabetes increases the rates of left isomerism (Splitt *et al.*, 1999). Exposure to retinoic acid from vitamin A derivatives in the intrauterine period has been shown to cause laterality defects in some vertebrates (Van Keuren *et al.*, 1991).

Situs ambiguous occurs at about 1 in 10,000 live births and the ratio of male to female is 2: 1 (Lin *et al.*, 2000). *Situs ambiguous* is accompanied by a wide spectrum of complex anomalies called right and left isomerism or heterotaxy syndromes. Heterotaxy syndromes (HS) are often associated with congenital heart diseases (CHD) and spleen abnormalities (Winer-Muram & Tonkin, 1989; Applegate *et al.*, 1999). The HS is generally classified as right and left atrial isomerism or asplenia and polysplenia syndrome. HS with asplenia implies that the patient has bilateral trilobed lungs with bilateral minor fissures and eparterial bronchi, bilateral systemic atria, a centrally located liver, and a stomach in an indeterminate position. Cardiac anomalies associated with asplenia are usually severe, are present at an early age, and have a poor prognosis. Ninety percent (90 %) of these patients have complex congenital heart anomalies (Sadler, 2018). Whereas, in HS with polysplenia, the patient has bilateral bilobed lungs, bilateral pulmonary atria, a centrally located liver, a stomach in an indeterminate position, and multiple spleens either in the left or right upper quadrant, along the greater curvature of the stomach. In the literature, VCI anomalies related to left isomerism are common again, the opening of the hepatic veins directly to the right atrium is common in the left isomerism. In our patient, v. hepatica intermedia and v. hepatica dexter were opening directly into the right atrium. HS with polysplenia is more common in females and has more variable clinical manifestations and prognosis (Winer-Muram & Tonkin, 1989; Sadler, 2018). Patients with left isomerism, in general, have less severe cardiac malformations than those with right isomerism. However, some may have complex cyanotic heart disease, frequently associated with a univentricular atrioventricular connection (Peeters & Devriendt, 2006). Our patient had the characteristic feature of polysplenia or left atrial isomerism, but the complete endocardia cushion defect

Fig. 9. D: Duodenum P: Pancreas white arrow: shows the pancreatic duct. There is absence of a retromesenteric (retro-peritoneal) D3 segment of the duodenum and the pancreatic body and tail are hypoplastic with preservation of the pancreatic head.

and duplicated right renal vein was unique for this case. Other clinically important anomalies seen in left atrial isomerism, include bilateral bilobed lungs, midline liver, extrahepatic biliary atresia, shortened pancreas and intestinal malrotation (Gilljam *et al.*, 2000).

In our case contrast-enhanced abdominal MRI shows an inversion in the superior mesenteric artery (SMA) / superior mesenteric vein (SMV) relationship, with the SMA on the right and the SMV on the left (Fig. 8). Although this has classically been advocated as a useful sign for suspecting or excluding intestinal malrotation (Taylor *et al.*, 2011). A more useful sign on for ruling out intestinal malrotation is the demonstration the retro-mesenteric D3 segment of the duodenum, where the horizontal (D3) segment of the duodenum should be seen in a transverse plane between the superior mesenteric vessels and the aorta (Yousefzadeh, 2009).

CONCLUSION

Although *Situs ambiguous* has been described in literature several times. Comprehensive knowledge of the variations in the origin of the SLA is paramount for every medical profession. *Situs ambiguous* may not always cause symptoms or medical problems in adults. As in our case, *Situs ambiguous* can be detected as an incidentally in medical imaging. The anatomical variations observed in *Situs ambiguous* can cause confusion in the diagnosis and problems during the invasive procedure. Recognition of these anomalies is important in case of a need for surgery in order to prevent vascular damage or in gaining access to the target organ. It is important to know the radiological clues that will make the correct diagnosis, to detect the situs anomaly in the patient, to perform appropriate surgical and interventional procedures.

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RESUMEN: El *Situs ambiguous* es la colocación de vasos y órganos en el espacio toracoabdominal que anatómicamente se encuentran fuera de su posición normal en un cierto orden. Esta condición es una definición amplia que incluye muchas variaciones. En este reporte de caso, reportamos un paciente con *Situs ambiguous* con una vena hepática anormal que fue diagnosticado incidentalmente durante un estudio de imagen médica (tomografía computarizada, ecografía y resonancia magnética).

PALABRAS CLAVE: *Situs ambiguous*; Isomería izquierda; Anomalía de la vena hepática.

REFERENCES

- Applegate, K. E.; Goske, M. J.; Pierce, G. & Murphy, D. Situs revisited: imaging of the heterotaxy syndrome. *Radiographics*, 19(4):837-52, 1999.
- Ghosh, S.; Yarmish, G.; Godelman, A.; Haramati, L. B. & Spindola-Franco, H. Anomalies of viscerotaxial situs. *AJR Am. J. Roentgenol.*, 193(4):1107-17, 2009.
- Gilljam, T.; McCrindle, B. W.; Smallhorn, J. F.; Williams, W. G. & Freedom, R. M. Outcomes of left atrial isomerism over a 28-year period at a single institution. *J. Am. Coll. Cardiol.*, 36(3):908-16, 2000.
- Lin, A. E.; Ticho, B. S.; Houde, K.; Westgate, M. N. & Holmes, L. B. Heterotaxy: associated conditions and hospital-based prevalence in newborns. *Genet. Med.*, 2(3):157-72, 2000.
- O'Rahilly, R. & Müller, F. *Developmental Stages in Human Embryos, including a Review of Streeter's "Horizons" and a Survey of the Carnegie Collection*. Washington (D.C.), Carnegie Institution of Washington, 1987. pp.141-58.
- Peeters, H. & Devriendt, K. Human laterality disorders. *Eur. J. Med. Genet.*, 49(5):349-62, 2006.
- Sadler, T. W. *Langman's Medical Embryology*. 13th ed. New York, Wolters Kluwer, 2018.
- Splitt, M.; Wright, C.; Sen, D. & Goodship, J. Left-isomerism sequence and maternal type-1 diabetes. *Lancet*, 354(9175):305-6, 1999.
- Strife, J. L.; Bisset, G. S. & Burrows, P. E. *Cardiovascular System*. In: Kirks, D. R. (Ed.). *Practical Pediatric Imaging: Diagnostic Radiology of Infants and Children*. 3rd ed. Philadelphia, Lippincott-Raven, 1998. pp.524-7, 588-90.
- Taylor, G. CT appearance of the duodenum and mesenteric vessels in children with normal and abnormal bowel rotation. *Pediatr. Radiol.*, 41(11):1378-83, 2011.
- Van Keuren, M. L.; Layton, V. M.; Jacob, R. A. & Kurnit, D. M. Situs inversus in the developing mouse: proteins affected by the iv mutation (genocopy) and the teratogen retinoic acid (phenocopy). *Mol. Reprod. Dev.*, 29(2):136-44, 1991.
- Winer-Muram, H. T. & Tonkin, I. L. The spectrum of heterotaxial syndromes. *Radiol. Clin. North Am.*, 27(6):1147-70, 1989.
- Yousefzadeh, D. K. The position of the duodenojejunal junction: the wrong horse to bet on in diagnosing or excluding malrotation. *Pediatr. Radiol.*, 39 Suppl. 2:S172-7, 2009.

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