METHODIST DEBAKEY CARDIOVASCULAR JOURNAL

Incidental Detection of Adult Polysplenia Syndrome With Situs Inversus Totalis, Interrupted Inferior Vena Cava, and Bronchiectasis

MULTIMODALITY
MUSEUM IMAGE

KANUPRIYA ARORA, MBBS
MANSI VERMA, DM
SUSHMA MAKHAIK, MD

*Author affiliations can be found in the back matter of this article

HOUSTON Methodist DEBAKEY HEART & VASCULAR CENTER

ABSTRACT

Incidental detection of polysplenia associated with situs inversus totalis in an adult is rarely reported in literature.[1,2] "Heterotaxy" refers to a wide spectrum of anomalies involving dysmorphism of thoracoabdominal organs across the right-left axis of the body. Polysplenia is observed in left isomerism along with bilateral bilobed bronchi and bilateral morphologic left atria. However, in this case, polysplenia was present without isomerism.

CORRESPONDING AUTHOR:
Mansi Verma, DM
Indira Gandhi Medical College, Shimla, India
mansiverma1608@gmail.com

KEYWORDS:
polysplenia; cardiac anomalies; bronchiectasis

TO CITE THIS ARTICLE:
Arora K, Verma M, Makhaik S. Incidental Detection of Adult Polysplenia Syndrome With Situs Inversus Totalis, Interrupted Inferior Vena Cava, and Bronchiectasis. Methodist DeBakey Cardiovasc J. 2025;21(1):6-9. doi: 10.14797/mdcvj.1537

A 32-year-old male was referred to our hospital for evaluation of exertional dyspnea and cough. Chest x-ray depicted reticular opacities in left middle and lower lung zones. Interestingly, there was dextrocardia, right-sided aortic arch, and gastric bubble beneath the right hemidiaphragm.

Contrast-enhanced computed tomography done for further evaluation revealed his liver on the left side and stomach on the right, suggesting abdominal situs inversus (Figure 1 A,B). There were multiple spleens on the right side of the upper abdomen. On lung window setting, the left bronchus was eparterial and right bronchus was hyparterial, suggesting bronchial situs inversus. There was dextrocardia with atrial and ventricular situs inversus (Figure 1). However, there was atrioventricular and ventriculoarterial concordance. Furthermore, the systemic drainage was anomalous. The intrahepatic portion of the inferior vena cava (IVC) was interrupted with azygous continuation. Hepatic veins were draining directly into the right atrium (Figure 2). Aortic arch was right sided with mirror image branching. The patient had no other cardiac abnormality. There was cystic bronchiectasis involving bilateral lungs. Noncontrast scan of the paranasal sinuses showed no evidence of sinusitis (Figure 3). The patient is presently in good medical condition with no significant symptoms.

Polysplenia syndrome results when multiple (usually two to six) similar-sized splenic masses are present along the greater curvature of the stomach. It is a rare congenital disorder generally diagnosed in early childhood and usually associated with a gamut of cardiovascular anomalies, often situs ambiguous and interrupted IVC

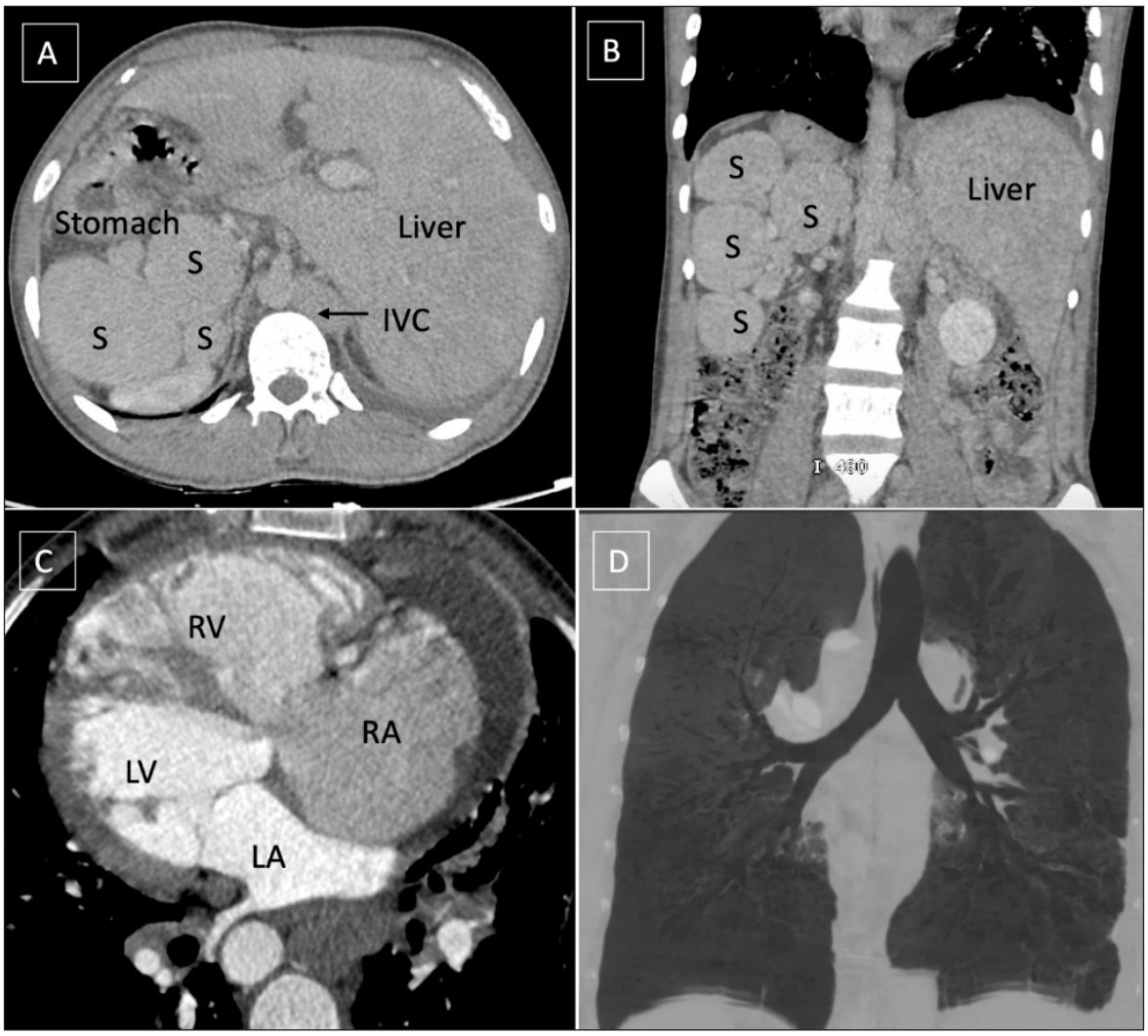

Figure 1 Computed tomography axial **(A)** and coronal **(B)** image depicting abdominal situs inversus with polysplenia axial image **(C)** demonstrating dextrocardia with right-sided cardiac chambers on left side and vice-versa. **(D)** Coronal image in minimum intensity projection showing left main bronchus to be shorter than right main bronchus. S: spleen; IVC: inferior vena cava; RA: right atrium; RV: right ventricle; LV: left ventricle

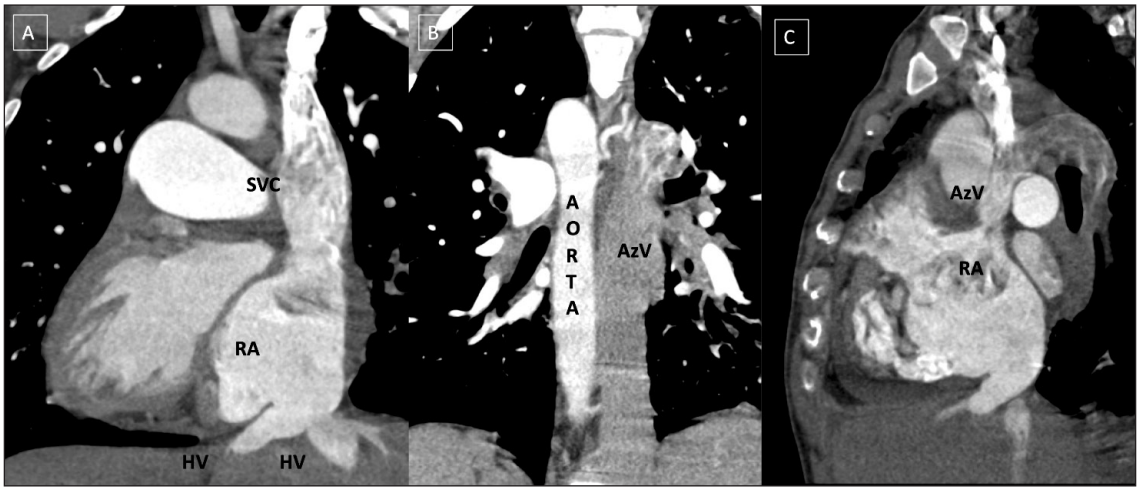

Figure 2 (A,B) Computed tomography coronal and sagittal view depicting drainage of hepatic veins into right atrium. Intrahepatic inferior vena cava was absent with azygous continuation and subsequent drainage into right atrium. RA: right atrium; HV: hepatic vein; SVC: superior vena cava; AzV: azygous vein

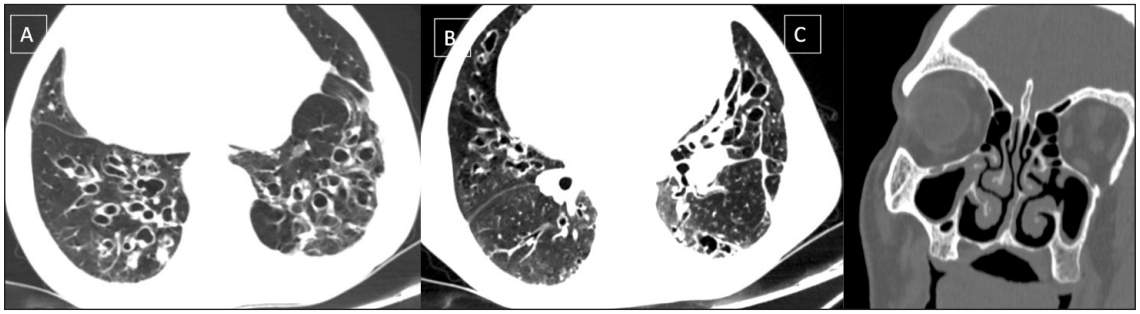

Figure 3 (A,B) Axial lung window image revealing cystic bronchiectasis in bilateral lungs. **(C)** Coronal noncontrast scan of paranasal sinuses revealed no evidence of sinusitis.

with azygous continuation. A majority of patients die by age 5 due to severe cardiovascular anomalies.[1] The gastrointestinal abnormalities associated with polysplenia include double ureters, renal agenesis, and short pancreas. Incidental detection of polysplenia associated with situs inversus totalis in an adult is rarely reported in literature.[1,2] "Heterotaxy" refers to a wide spectrum of anomalies involving dysmorphism of thoracoabdominal organs across the right-left axis of the body. Polysplenia is observed in left isomerism along with bilateral bilobed bronchi and bilateral morphologic left atria. However, in the present case, there was polysplenia without isomerism. It is critical to be aware of the various cardiovascular anomalies associated with polysplenia syndrome that will have clinical implications if a patient must undergo catheterization, bypass surgery, IVC filter placement, and temporary pacing through transfemoral approach.

COMPETING INTERESTS

The authors have no competing interests to declare.

AUTHOR AFFILIATIONS

Kanupriya Arora, MBBS orcid.org/0009-0003-8651-0334
Indira Gandhi Medical College, Shimla, India
Mansi Verma, DM orcid.org/0000-0002-3313-4350
Indira Gandhi Medical College, Shimla, India
Sushma Makhaik, MD orcid.org/0000-0001-8455-6329
Indira Gandhi Medical College, Shimla, India

REFERENCES

1. **El Mortaji H, Elatiqi K, El Hammaoui H, Alj S.** Polysplenia syndrome with situs ambiguous, common mesentery, and IVC interruption discovered incidentally in an adult. Radiol Case Rep. 2019 Jun 29;14(9):1072-1075. doi: 10.1016/j.radcr.2019.05.032
2. **Kwon SH, Shin SY.** Incidental adult polysplenia with situs inversus, interrupted inferior vena cava with azygos continuation, patent ductus arteriosus, and aortic branches variations: a case report. J Thorac Dis. 2018 Feb;10(2):E138-E141. doi: 10.21037/jtd.2018.01.128

TO CITE THIS ARTICLE:
Arora K, Verma M, Makhaik S. Incidental Detection of Adult Polysplenia Syndrome With Situs Inversus Totalis, Interrupted Inferior Vena Cava, and Bronchiectasis. Methodist DeBakey Cardiovasc J. 2025;21(1):6-9. doi: 10.14797/mdcvj.1537

Submitted: 21 December 2024 **Accepted:** 22 January 2025 **Published:** 06 March 2025