

# INCIDENTALLY DETECTED SITUS AMBIGUOUS IN ADULTS

JAE-GYUNG KIM, MD, HO-JOONG YOUN, MD, PHD, GEE-HEE KIM, MD, MI-HEE PARK, MD, JOON HUR, MD, JIN-SOK YU, MD, SOO-YEON JUNG, MD AND SOE-HEE AN, MD

*DIVISION OF CARDIOLOGY, DEPARTMENT OF INTERNAL MEDICINE, SCHOOL OF MEDICINE, THE CATHOLIC UNIVERSITY OF KOREA, SEOUL, KOREA*

Situs ambiguous is rare congenital anomaly in adults. In 2 adult patients who admitted for different cardiac problems, situs ambiguous with polysplenia was detected. A 42-year-old male admitted for radio frequent catheter ablation of atrial fibrillation, and he had left-sided inferior vena cava (IVC), hepatic segment of IVC interruption with hemiazygos continuation, multiple spleens and intestinal malrotation. And in a 52-year-old female case who was hospitalized due to infective endocarditis after implanting pacemaker for sick sinus syndrome, multiple spleens, left-sided stomach, bilateral liver with midline gallbladder, and left-sided IVC were found. Those findings were consistent with situs ambiguous with polysplenia, but their features were distinctive.

**KEY WORDS:** Situs ambiguous · Polysplenia syndrome · Adult.

## INTRODUCTION

Situs solitus represents the normal position of heart and viscera relative to midline, and situs inversus indicates mirror-image location of the viscera relative to situs solitus.<sup>1)</sup> Situs ambiguous (SA) is defined as an abnormality which can be considered to be present when the thoracic and abdominal organs are not clearly lateralized.<sup>2)</sup>

SA is typically associated with complex cardiovascular malformations. Also, splenic abnormalities and intestinal malrotation are common. Thus SA is usually categorized either as splenic morphology - polysplenia (bilateral left-sidedness, usually with multiple spleens, left isomerism, namely, polysplenia syndrome) or as asplenia (bilateral right-sidedness, with absence of spleen, right isomerism, namely, asplenia syndrome).<sup>2)</sup>

SA with polysplenia (SAP) is considerably rarely found in adults because of its high mortality rate with severe anomalies.<sup>3)</sup> However, patients with minor cardiac deformities can survive to adulthood.<sup>1)</sup> We report 2 cases of incidentally detected SAP.

## CASE

### CASE 1

A 42-year-old male was admitted for radiofrequency abla-

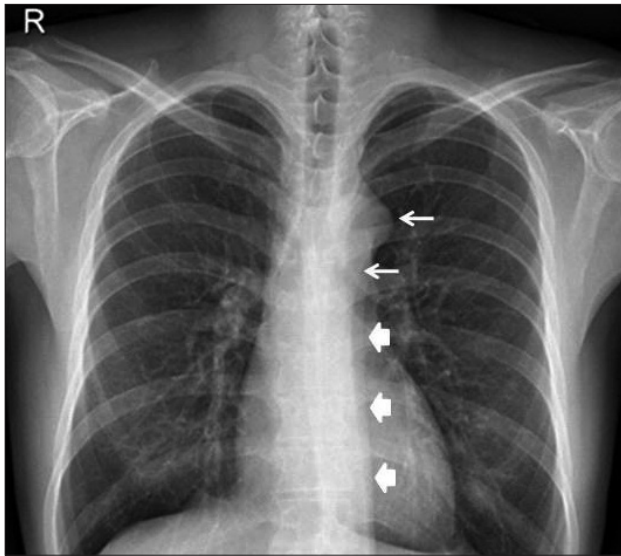
tion of atrial fibrillation (AF). He was diagnosed as AF 4 years ago and took anti-arrhythmic agent, beta blocker and anticoagulant. He had left side weakness due to cerebral infarction of right middle cerebral artery territory 3 years ago. He had a history of hypertension. His family had no history of diseases or congenital abnormality. He had no other symptoms but intermittent palpitation. His heart sound was irregular, but nothing particular was revealed on other physical and laboratory examinations. Double the shadow of thoracic aorta and widening state of superior mediastinum were shown in chest X-ray, but there were no other remarkable matters (Fig. 1). Initial electrocardiogram showed AF with moderate ventricular response (average 60-80 beats/min). On transthoracic and transesophageal echocardiography, no structural cardiac abnormalities were revealed. It seemed that hepatic vein was connected to right atrium through inferior vena cava (IVC) as usual. There was no pulmonary hypertension. We checked coronary multidirectional computed tomography (MDCT) to identify the anatomical variations of the patient's coronary vessels and heart before the ablation procedure. There was a tubular structure which was paralleling with descending thoracic aorta. It was supposed to be an IVC interruption with hemiazygos continuation (Fig. 2A). Hepatic veins were drained to right atrium. Abdomen computed tomography (CT) was

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• Address for Correspondence: Ho-Joong Youn, Division of Cardiology, Department of Internal Medicine, School of Medicine, The Catholic University of Korea, 222 Banpo-daero, Seocho-gu, Seoul 137-701, Korea Tel: +82-2-2258-1301, Fax: +82-2-591-1506, E-mail: younhj@catholic.ac.kr

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performed to evaluate other combined abnormality. Multiple and round soft tissue densities were detected around the

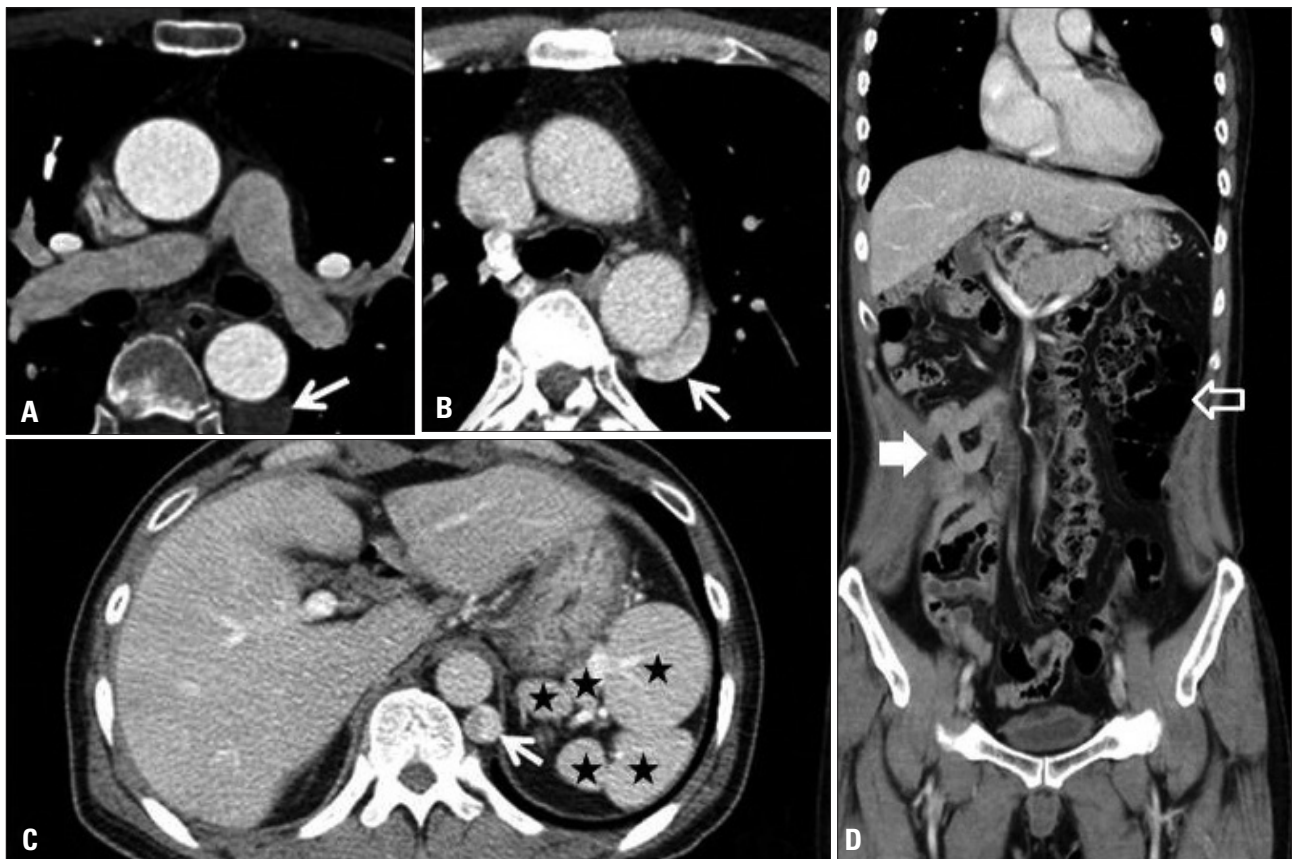


**Fig. 1.** Double the shadow of thoracic aorta (bold arrows) and widening state of superior mediastinum (sharp arrows) were shown in chest X-ray.

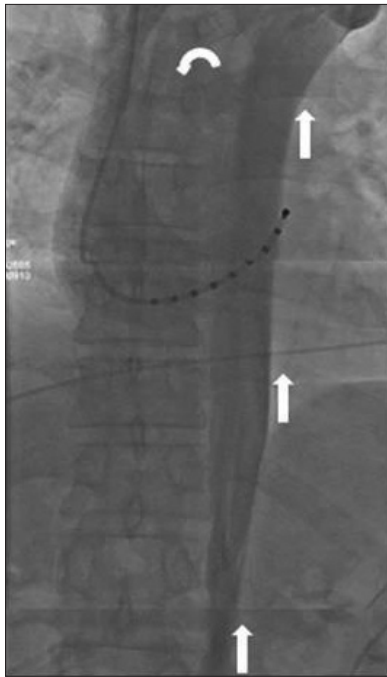
spleen, which were enhanced at the same degree of the spleen. Left-sided colon and right-sided small bowels indicated intestinal malrotation. IVC was located at the left side of aorta, and the hepatic segment of IVC was absent (Fig. 2B-D). By means of venography of IVC through right femoral vein, the interruption of the thoracic IVC with hemiazygos continuation along with aortic arch was confirmed (Fig. 3). All those findings were compatible with SAP.

## CASE 2

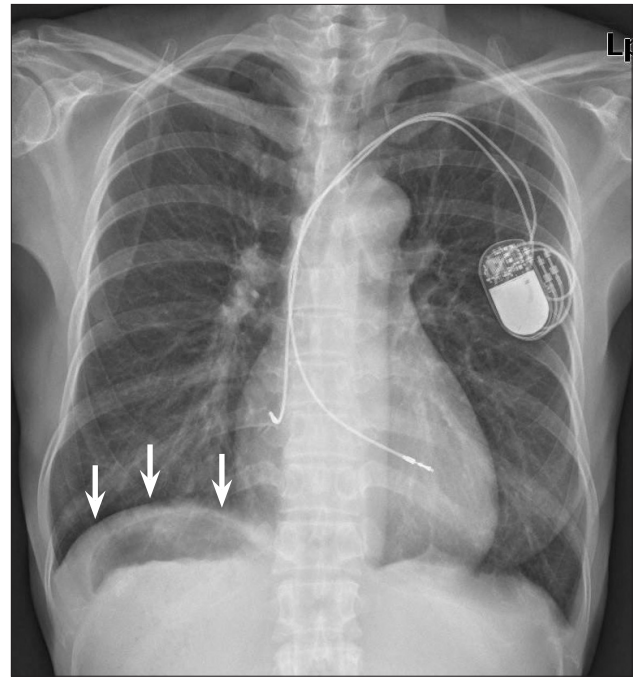
A 52-year-old female was presented with a history of intermittent fever for a month. She had been DDDR-type pacemaker insertion state for last 8 months due to sick sinus syndrome. Her family history was non-specific. She had no other symptoms and signs of fever. Nothing specific was shown on her physical and laboratory examinations. Her chest X-ray showed no significant lesions, except gastric air detected under the right side of diaphragm and hepatic shadow in the left side abnormally (Fig. 4). Liver dynamic CT was checked to identify the anatomy of her abdominal organs. The symmetric liver and gallbladder with multiple sandy stones were mid-line. Multiple spleens and stomach were located at the right



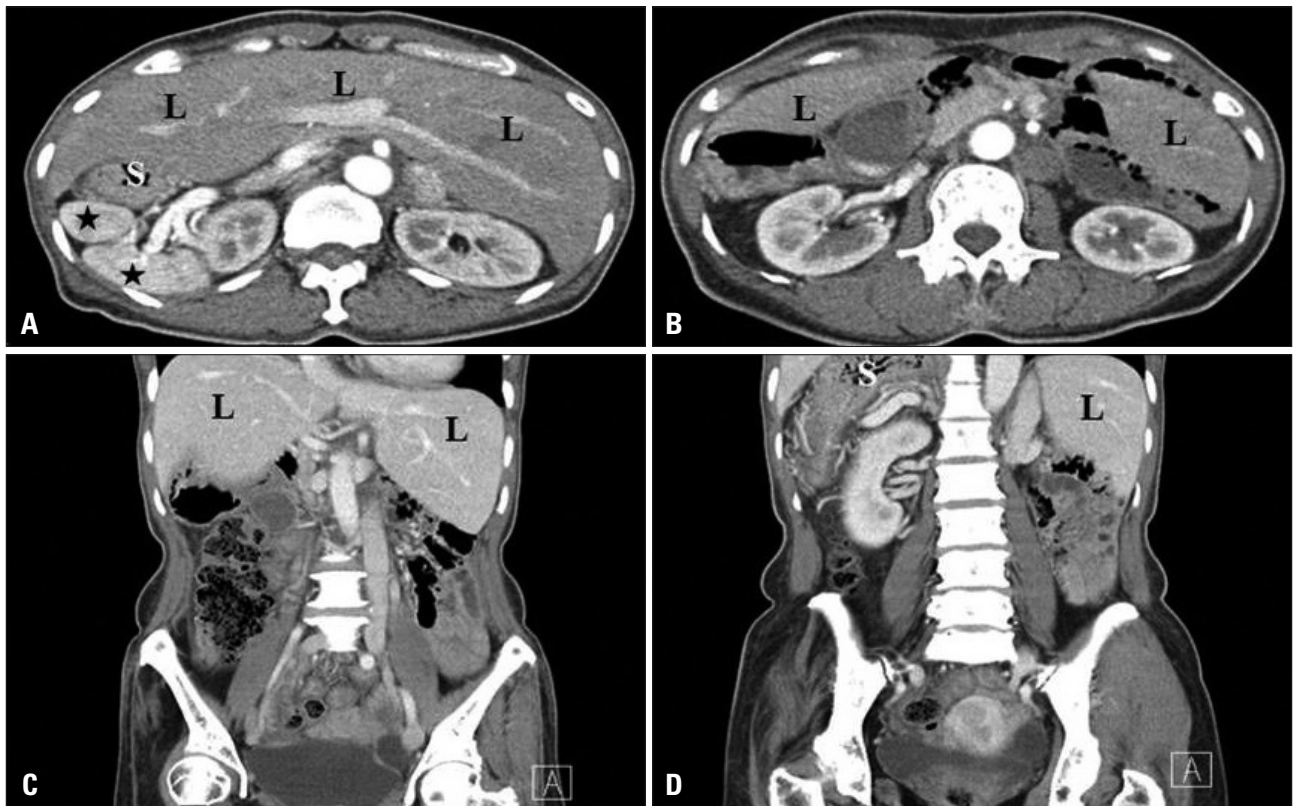
**Fig. 2.** (A) Coronal multidirectional computed tomography: There was a 1.7 cm sized, round, tubular structure which was paralleling with descending thoracic aorta (white arrows). (B-D) On abdomen computed tomography: (B) A dilated hemiazygos vein runs posterior to the descending aorta. (C) Multiple, well-defined round soft tissue densities were detected around the spleen (black stars). IVC was located at the left side of aorta, and hepatic segment of IVC was absent. (D) Left-sided colon (hollow arrow) and right-sided small bowels (bold arrow) indicated gastrointestinal malrotation. IVC: inferior vena cava.



**Fig. 3.** Venography of IVC through right femoral vein: Interruption of the thoracic IVC with hemiazygos continuation (arrows) along with aortic arch was confirmed. Enlarged hemiazygos vein drained into left brachiocephalic vein and then to superior vena cava. Infra-hepatic segment of IVC was intact. IVC: inferior vena cava.



**Fig. 4.** The chest X-ray of 52-year-old woman showed gastric air under the right side of diaphragm (arrows), and hepatic shadow in the left side abnormally.



**Fig. 5.** Liver dynamic computed tomography. A: There were midline symmetric liver (L) and multiple spleens (black stars) and stomach (S) are located at the right side of abdomen. B: Multiple sandy stones in midline gallbladder. Superior mesenteric vein was unusually located anterior to the superior mesenteric artery. C: The left sided inferior vena cava was crossing the aorta at the level of diaphragm and was drained to right atrium. D: Liver was observed in both sides relative to midline.



side of abdomen. Superior mesenteric vein was unusually located anterior to the superior mesenteric artery. The left-sided IVC was crossed the aorta at the level of diaphragm and drained into right atrium (Fig. 5). There was no intraabdominal lesion to develop fever. During hospitalization, methicillin resistant staphylococcus epidermidis was repeatedly incubated on blood cultures. She was referred us for an echocardiographic examination to find any evidence of infection in her heart. Echocardiogram revealed that large multiple mobile vegetations which were attached on the right ventricular pacemaker lead. The vegetations were prolapsed through the tricuspid valve, and the largest diameter of them was 20 mm. Coronary MDCT for the anatomical confirmation of vascular structure was checked before heart surgery. There was left-sided IVC, but no IVC interruption. She got surgery for removal of infected pacemaker lead and vegetation on tricuspid valve. After 4 weeks of antibiotics therapy, there was no longer pathogen growth in blood culture.

## DISCUSSION

Rose et al.<sup>4)</sup> estimated the minimal incidence of SA 1/40,000 live births. However Gatrads et al.<sup>5)</sup> obtained 1/24,000 affected in an English population and 1/2,700 in a highly inbred Asian population. The overall prognosis of children with SA had fatally poor prognosis in their early age, which result from the degree of congenital heart diseases.<sup>6)</sup>

SAP have congenital heart disease in 50-100% of cases.<sup>1)</sup> In a study of the spectrum of cardiac abnormalities in the 170 fetes with SAP, complete atrio-ventricular septal defect (68%), complete heart block (38%), double outlet right ventricle (23%), right ventricular outflow tract obstruction (21%) and total anomalous pulmonary vein (5%) drainage were detected, and only 58% of 170 survived.<sup>7)</sup> Therefore, the case reports of SA in middle aged adults are extremely rare. The abnormal arrangement of the abdominal organs was present up to 50% in a review of 146 autopsied cases of SAP.<sup>1)</sup> But, such cases without congenital cardiac defects in SAP was also rarely reported previously.

There were 5 case reports of adults' SAP in Korea from 1997 to 2010. These cases were different from one another in their compositions of organ arrangement and the mean age of these cases was 25.5 years. Three cases had congenital disease, non-compaction of the ventricular myocardium,<sup>8)</sup> coarctation of aorta with bicuspid aortic valve<sup>9)</sup> and intraluminal duodenal diverticulum,<sup>10)</sup> which were too minor to be detected until adulthood. In 2 cases, they were incidentally detected while respectively evaluating dyspnea due to congestive heart failure with atrial fibrillation<sup>11)</sup> and constipation.<sup>12)</sup>

Among venous malformation, the congenital interruption of the IVC is usually found in about 80% of patients SAP.<sup>13)</sup> The most typical IVC interruption is complete absence of the hepatic segment of the IVC with azygos continuation.<sup>14)</sup> Among the 5 Korean adults mentioned above, the IVC interruption

was presented in 4 cases, IVC was drained to hemiazygous vein in just one case.<sup>8-11)</sup> Venous malformations in SAP can be accompanied by various types which have usually no functional problems. But they do in some special circumstances as the 2 followings. In first case, a patient was found to be interrupted IVC while inserting catheter into right femoral vein for radiofrequency ablation of atrio-ventricular nodal reentrant tachycardia, only to give the procedure up.<sup>15)</sup> Secondly, a pacemaker was failed to be inserted in general approach because of persistent left superior vena cava which can be accompanied by SAP.<sup>16)</sup>

Consequently, the congenital abnormality does not always cause symptoms or medical problems in adults. However, these anatomical misarrangements can cause confusion in diagnosis and can bring problems during invasive procedure. Therefore, careful analysis of systemic anomalies is necessary to be done in medical approaching, especially for all patients expected surgical or medical interventions.<sup>14)</sup>

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