

# Heterotaxia associated with polysplenia

Idil Gunes Tatar,<sup>1</sup> Onur Ergun,<sup>1</sup> Hale Altunoglu,<sup>1</sup> Tolga Tatar<sup>2</sup>

<sup>1</sup>Department of Radiology, Diskapi Research Hospital, Ankara, Turkey

<sup>2</sup>Department of Cardiovascular Surgery, Gazi University, Ankara, Turkey

## Correspondence to

Dr Idil Gunes Tatar,  
idiltr@yahoo.com

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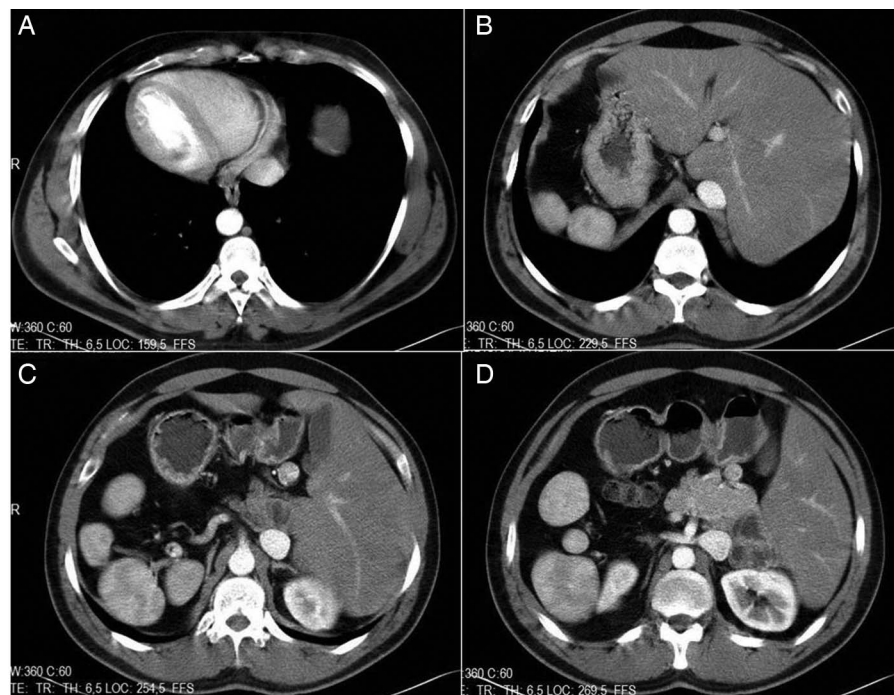
## DESCRIPTION

A 31-year-old man underwent abdominal CT for blunt abdominal trauma. CT scan demonstrated cardiac apex (figure 1A), stomach (figure 1B, C) and multiple splenic nodules situated on the left of the midsagittal plane; liver and inferior vena cava on the right and abdominal aorta in the midline (figure 1B–D). There were no signs of laceration or free peritoneal fluid. He was diagnosed with heterotaxia associated with polysplenia.

Heterotaxia (situs ambiguus) is the disturbance of the common right and left side localisation of thoracoabdominal viscera, which does not demonstrate the typical pattern of either situs solitus or inversus. The diagnosis is crucial since patients are strikingly under the risk of immunodeficiency; cardiac complications including congenital heart disease, atrial or ventricular arrhythmia, mitral or tricuspid valve insufficiency, heart failure, pulmonary thromboembolism, pericardial effusion, endocarditis; extracardiac complications such as pulmonary nodules, bronchitis, cyanosis due to pulmonary arteriovenous shunts or systemic venous shunt to the left heart, pleural effusion, liver dysfunction, cirrhosis, benign and malignant masses of

liver and kidney, diabetes, renal failure, protein-losing enteropathy, malrotation, ascites, fertility problems in women, oedema and coagulopathies.<sup>1,2</sup> Renal cysts and hepatic haemangiomas are very common lesions without clinical significance. Adrenal lesions in patients without a history of prior carcinomas are usually if not always benign. However, incidental solid renal masses should raise suspicion since they usually represent renal cell carcinomas. Pulmonary nodules need close follow-up depending on the risk status of the patient.

Heterotaxia typically occurs with either polysplenia or asplenia. Polysplenia is defined as multiple splenic nodules in the absence of a history of splenectomy associated with other findings of heterotaxia.<sup>1</sup> Although heterotaxia with asplenia (right-sided isomerism or Ivemark syndrome) is usually mortal due to severe congenital heart disease and immunodeficiency, heterotaxia with polysplenia (left-sided isomerism) can be asymptomatic and detected incidentally.<sup>3</sup> Yet it is important to recognise the complex anatomy due to the long-term clinical significance as well as to plan surgical and interventional procedures. This striking congenital disease demands close clinical follow-up.



**Figure 1** Arterial phase IV contrast-enhanced thoracoabdominal CT cardiac apex (A), stomach (B and C) and multiple splenic nodules (B–D) are located on the right of the midsagittal plane; liver and inferior vena cava are located on the left (B–D) and aorta in the midline (B–D).



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## Learning points

- ▶ Polysplenia is defined as multiple splenic nodules in the absence of a history of splenectomy associated with the presence of other findings of left-sided isomerism.
- ▶ The diagnosis of heterotaxia is important since patients with heterotaxy syndrome are at risk of immunodeficiency, cardiac and extracardiac pathologies.
- ▶ Although heterotaxia and polysplenia are asymptomatic conditions detected incidentally, it is important to recognise the complex anatomy due to long-term clinical significance as well as to plan surgical and interventional procedures.

**Competing interests** None.

**Patient consent** Obtained.

**Provenance and peer review** Not commissioned; externally peer reviewed.

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