## Diagnostic Challenge

## Don't know much about embryology...

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### **CASE HISTORY**

A 51-year-old man presented to the emergency department with a 3-day history of dull, intermittent, right upper quadrant abdominal pain that radiated to his back and was associated with diaphoresis, shortness of breath, and vomiting. Over the past few months, he had experienced episodes with a similar cluster of symptoms, but the current episode was more intense and prolonged. The patient's medical history included correction of transposition of the great arteries as a neonate, adult-onset hypertension, and hypercholesterolemia. He reported being a former smoker. His family history was positive for coronary artery disease.

On physical examination, he appeared to be in discomfort, but his vital signs were normal and his cardiac rhythm was sinus. Auscultation of the chest revealed crackles in the lower lung fields bilaterally and a soft ejection systolic murmur in the pulmonary area of the precordium. He was tender in the right upper quadrant, and Murphy sign was positive. Laboratory investigations revealed a slight elevation in white blood cell count with left shift and mild elevation of liver function indices. Serum troponin and D-dimer were negative. Electrocardiography (ECG) and chest radiography were performed (Figure 1 and Figure 2, respectively).

### QUESTION

What is the most likely diagnosis?

- 1. Acute coronary syndrome
- 2. Acute pulmonary embolism
- 3. Acute right heart failure with acute hepatic congestion
- 4. Acute cholecystitis

For the answer to this challenge, see next page.

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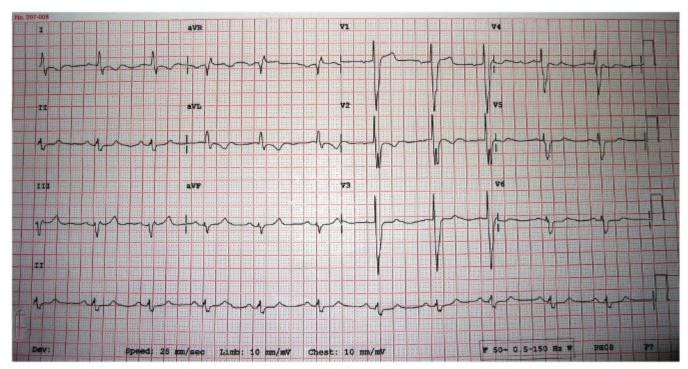


Figure 1. Twelve-lead electrocardiogram with conventionally placed precordial leads.



Figure 2. Posteroanterior chest x-ray.

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### **ANSWER**

The ECG showed sinus rhythm with right bundle branch block, and there was situs solitus with dextrocardia (suggested by the left-sided stomach bubble) on the chest x-ray. In a patient with dextrocardia and situs solitus, plus a history of congenital heart disease, the determining etiology of the patient's right upper quadrant pain is complicated. Acute coronary syndrome might be a possibility, but serial troponin measurements were negative. D-dimer was also negative, making a diagnosis of pulmonary embolism less likely. In decompensated cor pulmonale, one would expect to detect hepatomegaly and peripheral edema, but these signs were absent.

The patient had abdominal ultrasonography, which revealed cholelithiasis, gallbladder wall thickening, and pericholic fluid consistent with a diagnosis of acute cholecystitis.<sup>1</sup>

#### **DISCUSSION**

Situs describes the configuration of asymmetrical structures including the lung, liver, spleen, and cardiac chambers. The cardiac atria, pulmonary veins, and vena cavae develop simultaneously, such that their positions become fixed early on in the embryonic period, whereas the positioning of the ventricles does not. Therefore, atrial situs corresponds to the visceral situs.<sup>2</sup> Situs solitus is the normal arrangement of the viscera, with the morphologic right atrium to the right of the morphologic left atrium. Situs inversus is the mirror image of situs solitus. In situs ambiguous, the relationship between visceral and atrial anatomy is not maintained, and many of these patients are asplenic.<sup>2</sup>

Dextrocardia, mesocardia, and levocardia are positional anomalies of the heart in which the cardiac apex is located in the right, middle, or left side of the thorax, respectively. The terms do not refer to the orientation of the cardiac chambers. Normal arrangement of the cardiac atria and viscera with a right-sided heart is known as situs solitus with dextrocardia. In this situation, the sinoatrial node is located at the junction between the superior vena cava and right atrium, and atrial depolarization occurs in an inferior and leftward direction. This generates positive P waves in leads I and aV $_{\rm F}$  and negative P wave in lead aV $_{\rm R}$  on the ECG. In the precordial leads, R waves become progressively smaller. In situs inversus with dextrocardia, the P waves are negative in leads I and aV $_{\rm L}$  but positive in aV $_{\rm F}$  because the right atrium is situated on the left of left atrium. Situs solitus with dextrocardia is rare but is often associated with cardiovascular malformations such as transposition of the great arteries.

Situs solitus is associated with a right-sided gallbladder, whereas in situs inversus, the gallbladder is usually on the left.<sup>4</sup> Cardiac malpositions along with their situs variants provide an additional challenge in making the diagnosis in common medical conditions. Being able to pick up clues to these anatomic variants from the ECG and chest x-ray will help simplify the process.

Competing interests: None declared.

Keywords: cholecystitis, dextrocardia, situs solitus

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