

# A Case of A Middle-Aged Woman with Cor Triatriatum Dexter and Sick Sinus Syndrome: Comprehensive Cardiovascular Evaluation

Jun Wang, Yongqin Wang, Changqing Zhong\*

Department of Hunan Provincial People's Hospital Hunan Normal University First Affiliated Hospital, Changsha 410000, Hunan Province, China

\*Corresponding author: Changqing Zhong, zchqfly@126.com

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**Abstract:** This case study discusses a 51-year-old woman with a complex cardiovascular condition, cor triatriatum dexter, complicated by sick sinus syndrome. The patient presented with a history of bradycardia, dizziness, and amaurosis, and was admitted to the emergency department due to numbness in her right limb. Diagnostic examinations, including computed tomography (CT) and cardiac color-ultrasound screening, revealed cor triatriatum dexter along with an enlarged left atrium and ventricle. Additional findings included the absence of the inferior vena cava and polysplenic syndrome. Based on these results, double-chamber pacemaker surgery was recommended, supported by cardiac and thoracic-abdominal CT angiography and three-dimensional vascular reconstruction. This case underscores the importance of comprehensive examinations in identifying associated cardiovascular abnormalities.

**Keywords:** Bradycardia; Double-chamber pacemaker surgery; Cardiovascular abnormalities; Polysplenic syndrome; Adult congenital heart disease

**Online publication:** October 28, 2024

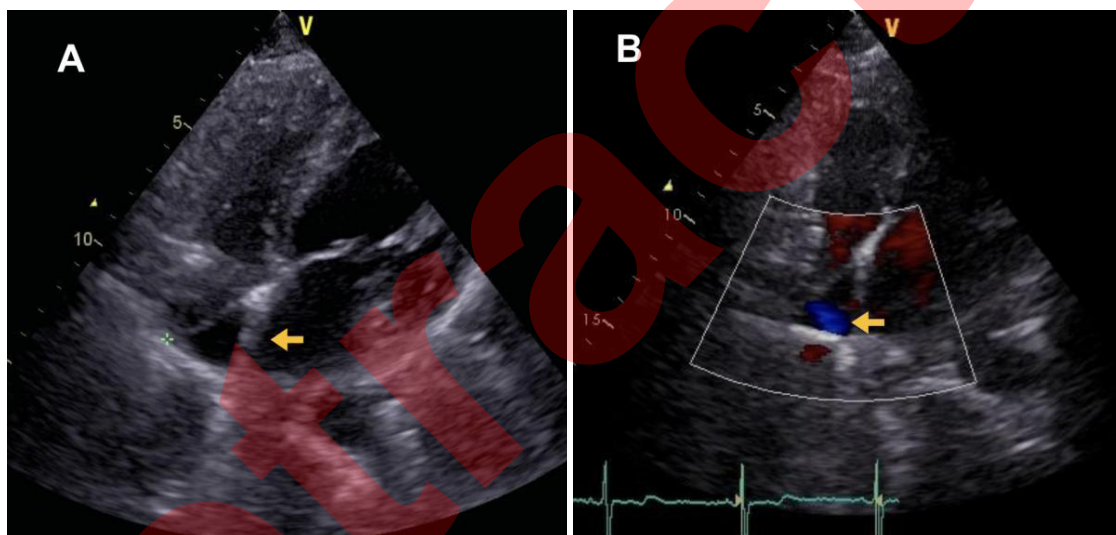
## 1. Introduction

Cor triatriatum dexter (CTD) is a rare congenital heart defect characterized by the division of the atrium into two chambers by a fibrous or fibromuscular diaphragm. This condition is more commonly observed in children and is often associated with other cardiovascular developmental abnormalities. Sick sinus syndrome refers to the malfunction of the heart's sinoatrial node, leading to bradycardia and symptoms such as dizziness and fainting. In this report, multiple cardiovascular anomalies, including an absent inferior vena cava and polysplenic syndrome, were successfully identified in the patient. By utilizing cardiac and thoracic-abdominal computed tomography (CT) angiography and three-dimensional vascular reconstruction, the heart's structure was accurately evaluated, leading to the formulation of an effective surgical approach. Ultimately, the patient

underwent successful double-chamber pacemaker surgery and had a satisfactory recovery, achieving the desired therapeutic outcome. In summary, the article successfully met its main objectives by employing comprehensive imaging studies and a personalized surgical plan, effectively managing this complex case and providing valuable clinical insights for similar patients.

## 2. Case report

A 51-year-old woman with a 15-year history of bradycardia, dizziness, and amaurosis, and a five-year history of bipolar disorder with manic episodes, presented to the emergency department two days prior due to right limb numbness. Her blood pressure was 129/71 mmHg, and her heart rate was 40 bpm. On examination, she was conscious and articulate, with no signs of neural localization. CT of the brain revealed a mild cerebral infarction in the prefrontal lobe. Dynamic electrocardiogram examination showed an average heart rate of 37 bpm, a lowest heart rate of 28 bpm, and a longest daytime sinus arrest of 4.55 seconds. Cardiac color-ultrasound screening indicated a longitudinal septum in the middle of the right atrium, accompanied by an enlarged left atrium and ventricle, with no abnormalities detected in the valves or atrioventricular septum (**Figure 1**).

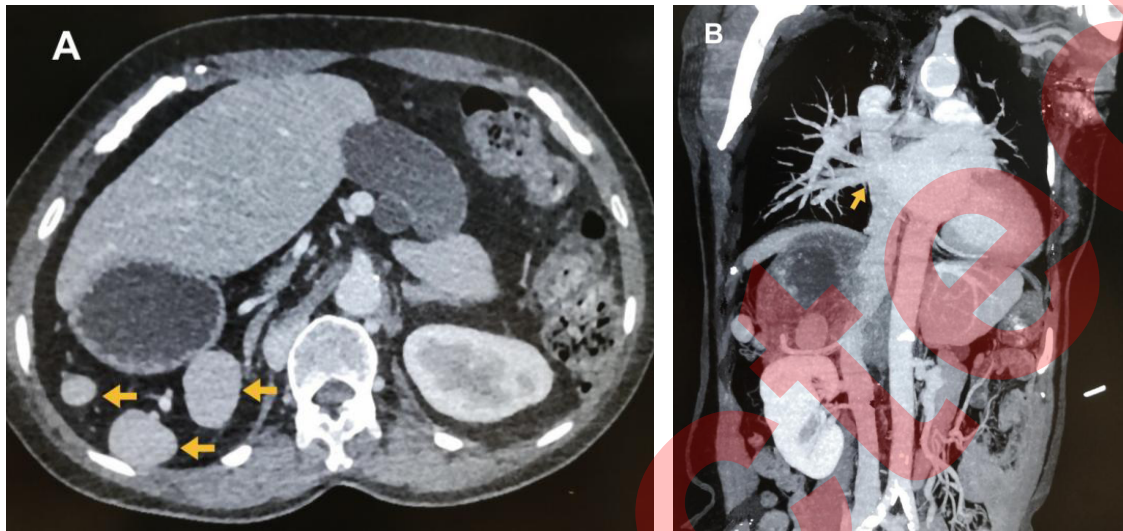


**Figure 1.** Color ultrasound examination of the heart in the patient. (A) The orange arrows indicate the presence of a longitudinal septum in the middle of the right atrium. (B) The orange arrows indicate red blood flow in the upper part of the right atrium and blue blood flow in the lower part, unable to pass through the septum in the middle of the right atrium.

Based on the diagnosis, the following options were suggested: (1) Order cerebral infarction therapy; (2) Order emergency temporary pacemaker surgery via femoral venepuncture; (3) Order temporary pacemaker surgery via femoral venepuncture followed by double-chamber pacemaker surgery via the right axillary venepuncture; (4) Order double-chamber pacemaker surgery based on cardiac and thoracic-abdominal CT angiography (CTA) and three-dimensional vascular reconstruction.

In this case, Option 4 (order double-chamber pacemaker surgery based on cardiac and thoracic-abdominal CTA and three-dimensional vascular reconstruction) was opted for. Cardiac color-ultrasound screening demonstrated a longitudinal septum in the right atrium, suggesting an extremely rare adult case of CTD, which is often accompanied by other cardiovascular developmental abnormalities<sup>[1]</sup>. The patient should be thoroughly

examined for these abnormalities before selecting an appropriate venepuncture approach for pacemaker surgery. As expected, cardiac and thoracic-abdominal CTA and three-dimensional vascular reconstruction revealed uncommon abnormalities, including an absent inferior vena cava, a giant azygos vein passing the superior vena cava and merging into the right atrium, inverted abdominal organs, a midline-type liver, and multiple spleens without a dominant spleen (**Figure 2**).



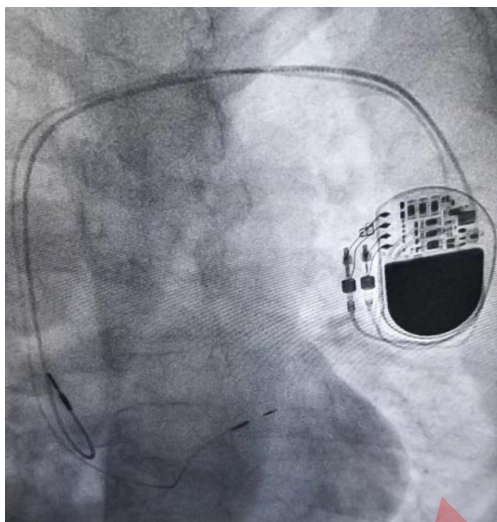
**Figure 2.** Enhanced CT examination of the thorax and abdomen in the patient. **(A)** Visceral abnormalities were observed, with the stomach and spleen located in the right upper abdominal cavity, and the liver and gallbladder converted into a midline-type liver. The orange arrows represent multiple spleens without a dominant spleen. **(B)** Cardiac abnormalities were observed, with the inferior vena cava absent. The orange arrows indicate the continuation of the giant malformed azygos vein behind the atrium into the superior vena cava.

This patient was therefore diagnosed with congenital CTD and polysplenic syndrome. The presence of a longitudinal diaphragm in the right atrium, as identified by cardiac color-ultrasound screening, can predict other cardiovascular abnormalities and help avoid the failure of emergency temporary pacemaker surgery via femoral venepuncture (Choice 2) or double-chamber pacemaker surgery via the right axillary venepuncture (Choice 3).

### 3. Patient outcome

A vagina vasorum was inserted into the right internal jugular vein, and the temporary pacing electrode was guided into the right ventricle through the tricuspid valve, resulting in effective pacing and sensing (**Figure 3**).

The punctured left subclavian vein was successfully catheterized, and the pacing catheter was eventually delivered to the right atrium and ventricle. The right atrial electrode was implanted in the auricle, and the right ventricular electrode was implanted in the septum, achieving normal pacing and sensing. One week later, the patient had recovered and was discharged satisfactorily.



**Figure 3.** The patient was successfully implanted with a two-chamber pacemaker

#### 4. Discussion

Cor triatriatum is anatomically defined by the division of the atrium into two cavities (the true atrium and an accessory atrium) by a fibrous or fibromuscular diaphragm [2]. It includes both left cor triatriatum and CTD. Cor triatriatum, often accompanied by other congenital cardiopathies, predominantly occurs in childhood with an incidence of 80% but rarely persists into adulthood [3,4]. In this case, the patient was also diagnosed with polysplenic syndrome and an absent inferior vena cava. Initially, emergency temporary pacemaker surgery (Choice 2) was attempted but failed. The failure was attributed to the limitations of the two-dimensional imaging approach and the interventional cardiologist's inexperience in handling this rare condition. After femoral venepuncture, guided by X-ray imagery, the pacing electrode was mistakenly inserted into the giant malformed azygos vein near the right atrium, preventing successful entry into the right ventricle. Thus, three-dimensional reconstruction is essential for patients with cardiovascular malformations to overcome the limitations of two-dimensional imaging.

Cor triatriatum can be diagnosed at any age, with symptom onset depending on the specific characteristics of the condition [5]. The size of the junction between the true atrium and the accessory atrium, along with any concomitant cardiovascular diseases, influences when symptoms emerge [6]. In this case, the absence of the inferior vena cava was a relatively minor cardiovascular abnormality. Additionally, the large junction between the diaphragm in the abnormal right atrium and the superior vena cava entry likely allowed the patient to survive into adulthood. Interestingly, besides CTD and sick sinus syndrome, the patient had polysplenic syndrome and multiple cardiovascular developmental malformations, including the inferior vena cava abnormality, defects in the double superior vena cava, a double aortic arch, and abnormalities in the interventricular or atrial septum and single atrium. It remains to be verified whether the absence of the inferior vena cava is associated with cor triatriatum or polysplenic syndrome.

#### 5. Conclusion

Adult CTD is a rare cardiovascular abnormality, often accompanied by other developmental anomalies.

Comprehensive examination is crucial for identifying associated abnormalities and selecting the appropriate treatment approach. Two-dimensional imaging has limitations in complex cases like CTD, and three-dimensional reconstruction can offer a more accurate assessment of cardiovascular malformations, guiding surgical interventions. In this case, double-chamber pacemaker surgery, based on cardiac and thoracic-abdominal CTA and three-dimensional vascular reconstruction, was the recommended treatment. The successful placement of the temporary pacemaker electrode in the right ventricle, leading to normal pacing and sensing, underscores the importance of a multidisciplinary approach and the expertise of interventional cardiologists in managing complex cases like CTD.

## **Funding**

Hunan Province Natural Science Foundation (Grant No. 2020JJ8061)

Hunan Province Science and Technology Innovation Program Project (Project No. 2020SK50921)

## **Disclosure statement**

The authors declare no conflict of interest.

## **Ethical approval and consent to participate**

The participant provided consent to publish the study. This case report was approved by the Institutional Review Board of Hunan Provincial People's Hospital, The First Affiliated Hospital of Hunan Normal University, prior to publication. All identifying information about the patient has been removed or anonymized to protect their privacy.

## **Consent for publication**

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images.

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