

Case of a Middle-Aged Woman with Sick Sinus Syndrome and Cor Triatriatum Dexter

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This study was carried out at the Hunan Provincial People's Hospital (Hunan Normal University First Affiliated Hospital), Changsha, Hunan, People's Republic of China.

ABSTRACT

This case study involves a 51-year-old woman with a complex cardiovascular condition, namely cor triatriatum dexter, accompanied by sick sinus syndrome. She presented with bradycardia, dizziness, and amaurosis, and was admitted to the emergency department due to right limb numbness. Diagnostic examinations, including computed tomography and cardiac color ultrasound screening, revealed the presence of cor triatriatum dexter with an enlarged left atrium and ventricle.

Additional abnormalities, such as absent inferior vena cava and polysplenia syndrome, were also detected. Based on the findings, a double-chamber pacemaker surgery was recommended, supported by cardiac and thoracic-abdominal computed tomography angiography and three-dimensional vascular reconstruction.

Keywords: Cor Triatriatum. Sick Sinus Syndrome. Bradycardia. Heterotaxy Syndrome. Inferior Vena Cava, Heart Atria.

Abbreviations, Acronyms & Symbols

CTA	= Computed tomography angiography
CTD	= Cor triatriatum dexter

successful double-chamber pacemaker surgery and had a satisfactory recovery, achieving the desired therapeutic outcome. In summary, this 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.

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 seen 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, the research team successfully identified multiple cardiovascular anomalies in the patient, including an absent inferior vena cava and polysplenia syndrome. By utilizing cardiac and thoracic-abdominal computed tomography angiography (CTA) and three-dimensional vascular reconstruction, they were able to accurately evaluate the heart's structure and formulate an effective surgical approach. Ultimately, the patient underwent

CASE PRESENTATION

A 51-year-old woman with a 15-year history of bradycardia, dizziness, and amaurosis and a five-year history of bipolar disorder-manic attack presented to the emergency department due to right limb numbness. Her blood pressure was 129/71 mmHg, and heart rate was 40 bpm. On examination, she appeared conscious and articulate and had a negative neural localization sign. Computed tomography of the brain showed an emerging mild cerebral infarction in the prefrontal lobe. Based on her dynamic electrocardiogram examination, the average heart rate was 37 bpm, with the lowest recorded rate of 28 bpm. The longest sinus arrest during the day reached 4.55 seconds. The patient has also experienced an increased frequency of dizziness and syncope episodes, accompanied by new-onset numbness

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in the right limbs, which may indicate an acute cardiac event. Therefore, we considered the feasibility of temporary pacemaker implantation (Figure 1). Cardiac color ultrasound screening indicated a longitudinal septum in the middle of the right atrium accompanied by an enlarged left atrium and ventricle, while there was no abnormality detected in valves and the atrioventricular septum (Figure 2).

Through comprehensive multidisciplinary data synthesis, we determined to proceed with dual-chamber pacemaker implantation guided by cardiac and thoraco-abdominal CTA and three-dimensional vascular reconstruction.

Cardiac color ultrasound screening demonstrated a longitudinal septum in the right atrium, suggesting an extremely rare adult CTD, which was usually accompanied by other cardiovascular developmental abnormalities^[1]. The patient should be thoroughly examined for these abnormalities before selecting an appropriate venipuncture approach for pacemaker surgery. As expected, the cardiac and thoracic-abdominal CTA examination and three-dimensional vascular reconstruction showed uncommon abnormalities, including absent inferior vena cava, a giant azygos vein which passed the superior vena cava and merged into the right atrium, inverted abdominal organs, a midline-type liver, and multiple spleens but no master spleen (Figure 3).

Therefore, this patient was diagnosed with congenital CTD and polysplenia syndrome. Attention to the fact that there is a longitudinal diaphragm in the right atrium based on cardiac color ultrasound screening, which can predict the presence of other cardiovascular abnormalities and avoid failure of emergency temporary pacemaker surgery via femoral venipuncture and double-chamber pacemaker surgery via the right axillary venipuncture.

Surgical Technique

After determining the surgical plan and completing the necessary examinations, we performed local anesthesia on the patient

in the operating room while closely monitoring vital signs. We selected the left subclavian vein and successfully performed a venipuncture under ultrasound guidance. After that, a guidewire was inserted, and the sheath was placed over the guidewire into the vein to provide access for the subsequent implantation of the pacemaker lead.

The temporary pacing electrode was then advanced through the catheter and delivered into the right ventricle via the tricuspid valve from the right atrium. The right atrial electrode was implanted in the cardio ear clip, and the right ventricular electrode was implanted in the septum. The electrode was connected to the temporary pacemaker, and appropriate pacing parameters were established. During the procedure, we continuously monitored the electrocardiogram, observing pacing spikes and assessing the patient's ability to perceive heartbeats, ensuring that pacing was suspended during any intrinsic cardiac activity. Postoperatively, the patient reported significant improvement in symptoms of dizziness, syncope, and limb numbness. Monitoring indicated that the heart rate was maintained at 60 - 70 bpm following pacemaker implantation, demonstrating effective pacing and perception (Figure 4).

One week later, the patient had recovered and was discharged satisfactorily.

DISCUSSION

Cor triatriatum is anatomically defined by the fact that the atrium is divided into two cavities (the true atrium and an accessory atrium) by a fibrous or fibromuscular diaphragm^[2]. It includes left cor triatriatum and CTD. Cor triatriatum with other congenital cardiopathies mainly occurs in childhood with an incidence of 80%, but rarely reaches adulthood^[3,4]. Other than that, the patient in this case was also diagnosed with polysplenia syndrome with absent inferior vena cava. In fact, emergency temporary pacemaker surgery was performed preferentially, but ended in failure. The emergency temporary pacemaker failed to be placed due to

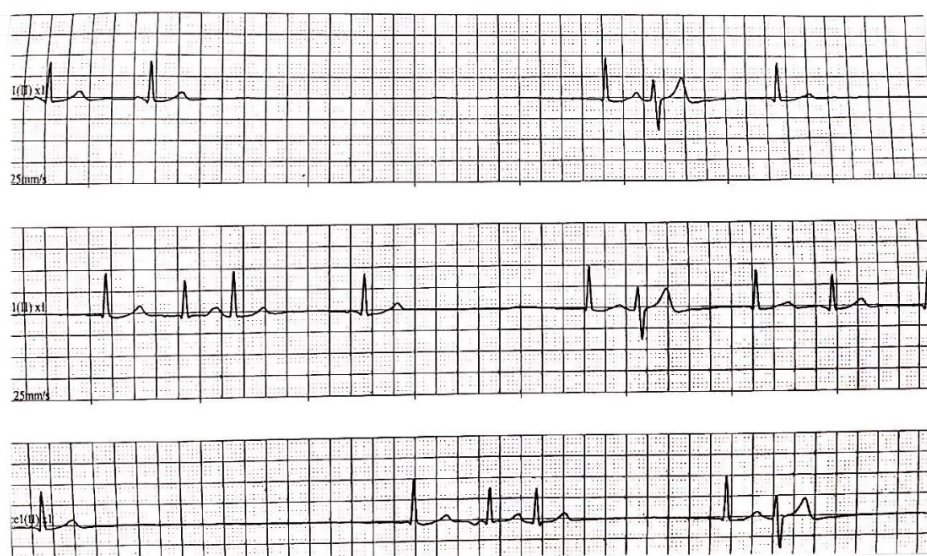


Fig. 1 - The dynamic electrocardiogram examination indicated an average heart rate of 37 bpm, the lowest heart rate of 28 bpm, and the longest sinus arrest during the day reaching 4.55 seconds.

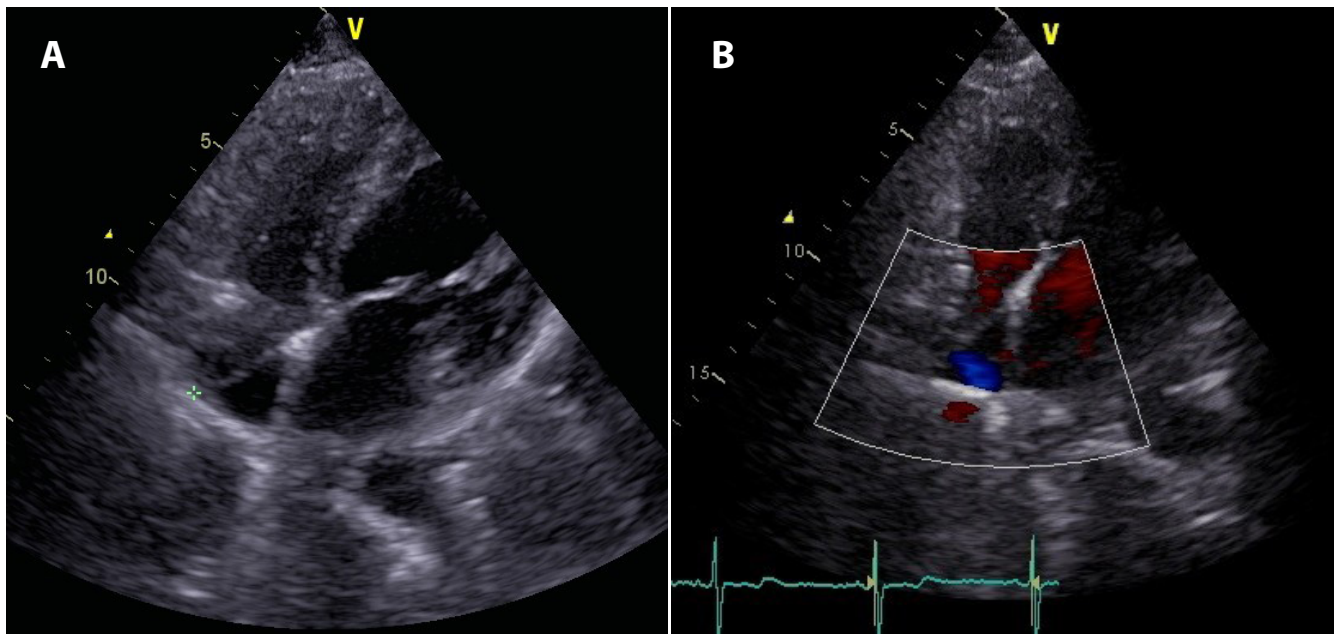


Fig. 2 - Color ultrasound examination of the patient's heart. (A) The orange arrow indicates a longitudinal septum in the middle of the right atrium. (B) The orange arrow indicates the red blood flow in the upper part of the right atrium, and the blue blood flow in the lower one cannot pass through the septum in the middle of the right atrium.

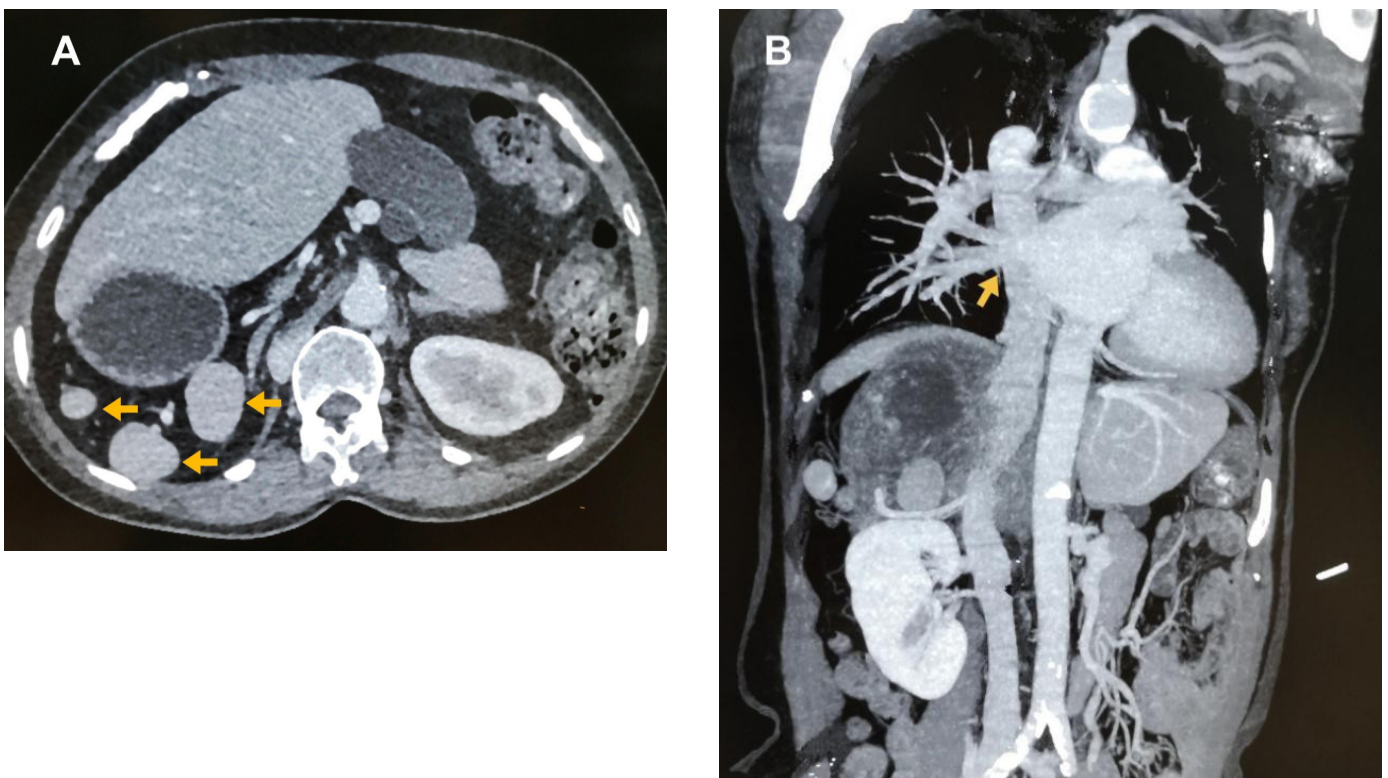


Fig. 3 - Enhanced computed tomography examination of the thorax and abdomen in the patient. (A) Visceral abnormalities were observed. The stomach and spleen were located in the right upper abdominal cavity, and the liver and gallbladder had been converted into a midline-type liver. The orange arrows represent multiple spleens without a master spleen. (B) Cardiac abnormalities were observed. The inferior vena cava appeared to be absent. The orange arrow denotes the continuation of the giant malformed azygos vein behind the atrium to the superior vena cava.

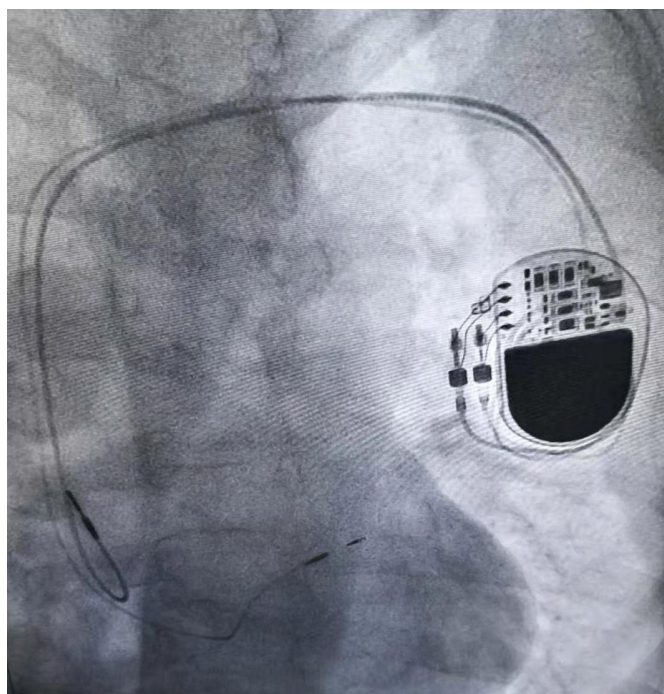


Fig. 4 - The patient was successfully implanted with a two-chamber pacemaker.

the existing limitations of two-dimensional imaging approach and inexperience on the part of the interventional cardiologist in the face of this rare case. After femoral venipuncture using X-ray imaging as a guide, the pacemaking electrode was inserted into the giant malformed azygos vein close to the right atrium, proving inaccessible entry with regard to the right ventricle. Hence, to overcome the limitation of two-dimensional imaging, three-dimensional reconstruction is necessary for patients with cardiovascular malformation.

Cor triatriatum can be diagnosed at any age and depends upon the symptoms^[5]. The size of the junction between the true atrium and the accessory atrium and other concomitant cardiovascular diseases can determine when symptoms arise^[6]. In this case, the absent inferior vena cava was a relatively minor cardiovascular abnormality, and there were large-sized junctions between the top of this diaphragm in the abnormal right atrium and the superior vena cava entry, so that this patient survived into adulthood. Interestingly, besides CTD with sick sinus syndrome, this patient had polysplenia syndrome and multiple cardiovascular developmental malformations, which were shown as the inferior vena cava abnormality, defects in the double superior vena cava, double aortic arch, interventricular septum or atrial septum, and single atrium abnormality. It is worth verifying whether the absence of the inferior vena cava is associated with cor triatriatum or polysplenia syndrome.

CONCLUSION

Adult CTD is a rare cardiovascular abnormality that is usually accompanied by other developmental abnormalities. It is crucial to conduct a comprehensive examination to identify associated

abnormalities before selecting the appropriate treatment approach. Two-dimensional imaging has limitations in certain complex cases like CTD. Three-dimensional reconstruction can provide a more accurate assessment of cardiovascular malformations, helping to guide 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 option.

The successful placement of the temporary pacemaker electrode in the right ventricle, leading to normal pacing and perception, highlights the importance of a multidisciplinary approach and the expertise of interventional cardiologists in managing complex cases like CTD.

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No conflict of interest.

Authors' Roles & Responsibilities

JW	Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; drafting the work or revising it critically for important intellectual content; final approval of the version to be published
YW	Drafting the work or revising it critically for important intellectual content; final approval of the version to be published
CZ	Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved; final approval of the version to be published

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