Heterotaxy pattern associated with Sinus Node Dysfunction in an adult: A case report.

naman shah[1], sankalp acharya[2], apporva tripathi[2], himanshi bisht[2], maitri shah[1], aayushi pareek[3], asmita gera[3], Abhigan Shrestha[4], and Vikash Jaiswal[5]

[1]GMERS Medical College and Hospital
[2]BJ Medical College
[3]RUHS College of Dental Sciences
[4]M Abdur Rahim Medical Medical College Hospital
[5]AMA School of Medicine

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Authors : Naman Shah MBBS[1], Sankalp Acharya[2], Apoorva Tripathi MBBS[2], Himanshi Bisht MBBS[2], Maitri Shah MBBS[1], Aayushi Pareek MBBS[3], Asmit Gera MBBS[4], Abhigan Babu Shrestha, MBBS[5], Vikash Jaiswal MD[6]

1. GMERS Medical college Sola, Ahmedabad, India
2. B. J. Medical College, Ahmedabad, India
3. RHUS College of Medical Science, Jaipur, Rajasthan
4. JCCR Cardiology Research, Varanasi, India
5. M Abdur Rahim Medical College, Dinajpur, Bangladesh
6. AMA School of Medicine, Makati, Philippines

Corresponding Author

Abhigan Babu Shrestha

M Abdur Rahim Medical College, Dinajpur, Bangladesh

Abigan17@gmail.com

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Abstract:

This case report presents a 26-old aged male patient brought to the medical ward with an experience of repeating blackout for a year. The patient was diagnosed with sick sinus syndrome, and further investigations revealed left isomerism, polysplenia, and no congenital heart defects. Holter monitoring, ultrasonography, electrocardiography, and computed tomography were used to confirm the diagnosis. The patient underwent DDDR pacemaker implantation for the treatment of SA node dysfunction. The report highlights the variability of anatomical findings associated with polysplenia pattern and the various types of heartbeat disruptions that may occur in the atrial appendages of the left side isomerism.

Key Clinical Message:

- A 26-old male patient admitted to the hospital ward with experience of repetitive syncopes for a year.
- The patient was diagnosed with sick sinus syndrome.
- The aim of this clinical report is to highlight the variability of anatomical findings associated with polysplenia pattern .

Keywords: Polysplenia Syndrome, SA node dysfunction, Pathology

INTRODUCTION

Heterotaxy pattern, also called Situs ambiguous, is a medical condition characterised by an atypical arrangement of the internal organs in the Thorax and abdomen. This disease can lead to complicated congenital deformities that affect various organ systems, such as the circulatory, pulmonary, digestive, reproductive and the urinary system systems. Situs inversus totalis is a rare congenital condition in which the internal organs of the body are asymmetrically placed, with the heart located on the opposite side of the body from its normal position. However, other congenital defects are also known to cause serious health problems. These include abnormal connections between major blood vessels and the heart, which can lead to altered blood flow. Pulmonary system is also affected, lungs have different numbers of lobes than normal anatomical lungs, and the size of bronchi is also affected. Furthermore, it is also possible for this medical condition to lead to absence of spleen or the condition may result in the formation of numerous small and inefficient spleens (polysplenia) in the abdomen. The liver is located in the center of the body rather than its usual spot on the right side of the abdomen. The condition also varies in severity. Some individuals may experience mild health problems while others may face potentially life-threatening risks even with treatment during infancy or childhood (1).

The mortality rate for most patients diagnosed with polysplenia syndrome is alarmingly high, with many not surviving beyond the age of five. This is largely attributed to the presence of severe cardiac abnormalities. Yet, a miniscule fraction (5.0-10.0 %) of people with this syndrome have normal hearts or minor cardiac abnormalities and can mature without exhibiting any significant signs. This case presentation is an example of such an occurrence (2).

Heterotaxy syndrome is classified into two main categories: right isomerism and left isomerism. Left Isomerism is a condition in which paired organs are located on the left side of the body, whereas in Right Isomerism, paired organs are positioned along the right side, and there may be an absence of organs on the left. Both versions are correlated with complicated heart malfunctions. In left isomerism, one can expect to find left atrial isomerism with bilateral morphologic left atrial appendages, viscerocardiac heterotaxy, and multiple cardiac malformations such as congenital heart block, atrioventricular septal defects and pulmonary stenosis, multiple polysplenia, bilobed lungs on the left side with hyparterial bronchi, intestinal malrotation, non-random genitourinary malformations, and an interruption of the inferior vena cava (IVC) accompanied by azygos continuation. (3). Hereby, we report Sick sinus syndrome in a 26-year-old man who was admitted to our clinic with complaints of syncope (sick sinus syndrome) and found to have left isomerism on the investigation.

Case presentation

A 26-year- old age male patient presented to our medical facility with a history of multiple episodes of fainting scattered throughout the previous year. Upon further inquiry, the patient mentioned that they had experienced three episodes of fainting within a period of one year. On examination, her pulse was at 40/min with an irregular rhythm. Her blood pressure (BP) reading registered as 140/90 mmHg. The cardiovascular system examination showed no abnormalities. Further investigations showed that the full blood count and sugar levels were in the normal range. Additionally, the kidney function tests, serum electrolytes and liver tests all yielded normal results. The electrocardiography (ECG) results showed the presence of sinus node arrest accompanied by a junctional escape that was recorded at a rate of 35 to 40 beats per minute **(Figure 1).** Holter monitoring was performed to verify the diagnosis and identify any presence of AV blockage or alternate rhythm disturbances that might result in syncope. The test results confirmed Sick Sinus Syndrome, which showed pauses and arrest in sinus rhythm along with junctional escape beats/rhythm **(Figure 2).** An ultrasonography revealed a midline liver that was more towards the left side, with a leftward hilum and normal hepatic veins and inferior vena cava on the right side. Additionally, no spleen was identified on the left side, in accordance with situs ambiguous abdominis. Echocardiography revealed normal left ventricular ejection fraction and no congenital heart defects. An HRCT chest with upper abdomen was conducted on a 128-slice dual source dual energy scanner. The scan results revealed the presence of liver on the left side of the abdomen and in the midline stomach. (Figure 3A). The abdomen contained numerous accessory spleens, at least 7. A bilateral trilobed lung with hyparterial bronchus is seen (Figure 3B). Situs ambiguous with polysplenia syndrome was diagnosed on CT. (Figure **3C).** The patient received a DDDR (dual-pacing dual-sensing dual-response rate-adaptive) pacemaker for sinus node dysfunction. The patient responded well, with improvement in symptoms and findings at follow-up.

Discussion

Polysplenia is a syndrome that is characterized by various anatomical findings, and it can be quite complex. In cases where there is isomerism of the left atrial appendages, individuals may experience one of three types of heart rhythm disturbances. These include atrioventricular block (4,5), dual atrioventricular nodal pathways and sinus node dysfunction (6). Patients with Polysplenia syndrome have been found to exhibit different mutations, such as LEFTYA3, Z1C3 etc. (6) Our patient, on the other hand, did not exhibit or undergo any study for such mutations.

The reported patient was an adult who had been diagnosed with polysplenia syndrome, had a structurally normal heart, and suffered from sinus node dysfunction. It is worth noting that the occurrence of isolated conduction disorder in patients with polysplenia syndrome and a structurally normal heart is quite rare and unusual (5).

When it comes to patients who have sinus node dysfunction, using AAI C/R and DDD C/R pacemakers for electrical stimulation yields a better outcome due to the reduced incidence of atrial fibrillation, thromboembolic events, and heart failure. This is supported by research findings mentioned in the source (3).

IVC interruption with azygous continuation is seen in left isomerism and affects the course of catheter or TPI (thrombolytic predictive instrument) lead while performing temporary pacemaker implantation from femoral vein access. Alternatively, one can choose for internal jugular vein access for TPI.

While doing TPI, one must be aware of the presence of left sided SVC and pacemaker lead course accordingly.

Conclusion

Left isomerism is an important etiology of Sinus node dysfunction in young patients and should be evaluated thoroughly. Diagnosis of left isomerism in sick sinus syndrome will affect the management of patients with regards to the mode and access of pacemaker implantation.

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Conflict of Interest: None

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Figure Legend:

Figure 1: Electrocardiography shows sinus node arrest with a junctional escape at 35 – 40 bpm.

Figure 2 : Holter shows sinus pause and sinus arrest with junctional escape beats/rhythm, confirming the diagnosis of sick sinus syndrome).

Figure 3A: CT scan (HRCT) of the chest with upper Abdomen shows evidence of liver on the left side of the abdomen and midline stomach.

Figure 3B: CT scan of the chest (HRCT) with upper Abdomen shows Bilateral trilobed lung with hyparterial bronchus.**Figure 3C:** CT scan on axial view shows evidence of liver on the left side of the abdomen and midline stomach. Multiple accessory spleens (at least 7) are seen in the abdomen.

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