A Case of Successfully Implanted Dual Chamber Pacemaker in a Young Patient with Dextrocardia and Sick Sinus Syndrome

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ABSTRACT: Dextrocardia is a rare congenital anomaly, whereas its association with sick sinus syndrome in young adults without accompanying heart abnormalities is exceptional. We report a case of a 38 year old female patient who was admitted to our hospital due to syncope as a consequence of sinus pauses up to 4.9 seconds. She was also diagnosed with situs inversus totalis, with mirror image dextrocardia. Pacemaker implantation was indicated. Under local anesthesia, from the left subclavian vein, the guide wire was passed through superior vena cava to the right atrium and ventricle without any obstacles. In conclusion, situs inversus totalis with mirror image dextrocardia may present in combination with sick sinus syndrome as early as the fourth decade of life. Implantation of pacemaker leads from the left subclavian vein appears accessible and safe in patients with dextrocardia with situs inversus (mirror image).

KEYWORDS: Dextrocardia, sick sinus syndrome, DDD pacemaker, young adult

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Introduction

Dextrocardia is a rare congenital abnormality with an estimated incidence of 1 in 8000 to 25 000 live births. There are 3 configurations of dextrocardia: situs solitus, situs inversus, and situs ambiguous. Situs inversus or situs inversus totalis, L-loop ventricles and inverted great vessels, known as "mirror-image dextrocardia" is the most common form of dextrocardias in general population. Mirror-image dextrocardia and structurally normal heart is usually an incidental finding on physical exam and/or chest X-ray.

Among the associated abnormalities, Kartagener's syndrome is seen in 25% of patients who have "mirror-image dextrocardia" and is characterized by the presence of situs inversus totalis, paranasal sinusitis, bronchiectasis, ciliary dysmotility, and infertility. Other cardiac abnormalities are rare in mirror-image dextrocardia.[1]

Case Report

We report a case of 38 year old female patient who was admitted to our Cardiology Clinic due to an episode of syncope 1 week prior to admission. Following the syncope, a 24 hour ECG Holter monitoring was performed, which demonstrated signs of sick sinus syndrome, with several sinus arrest episodes, where the longest pause registered was 4.9 seconds (Figure 1). The average heart rate was 57 beats per minute, with minimum heart rate of 30 beats per minute, and maximum heart rate of 124 beats/minute. A 3 hour episode of atrial fibrillation was also recorded. These findings indicated hospital admission.

At admission, physical examination showed a blood pressure of 100/70 mmHg and a heart rate of 52 beats per minute. On physical examination of the thorax, dextrocardia was evidenced, as cardiac dullness, and apex cordis were to the right of the sternum. Whereas, hepatic dullness was demonstrated on the left side of the abdomen.

Laboratory findings were within reference range, including electrolytes and thyroid tests.

ECG showed negative P and T wave and predominantly negative QRS complex in lead I, whereas the R wave was of low voltage in precordial leads V3 to V6. The rhythm of the ECG at admission was junctional escape rhythm, with heart rate of approximately 55 beats per minute.

Chest X ray showed the heart silhouette and the apex in the right hemithorax, descending aorta on the right side of the sternum and the left hemidiaphragm slightly elevated.

Echocardiography window was attained from the right parasternal and apical approach. All cardiac chambers were within reference values, and the left and right ventricular function were normal. No intracardiac shunts or valve abnormalities were detected. No anomalies of systemic venous return were detected by echocardiography.

Abdominal ultrasound and computed tomography of the abdomen were performed, which showed that the liver was located on the left side, while there were multiple spleens on the right side (polysplenia syndrome). A left ovarian cyst of 4 cm was detected.

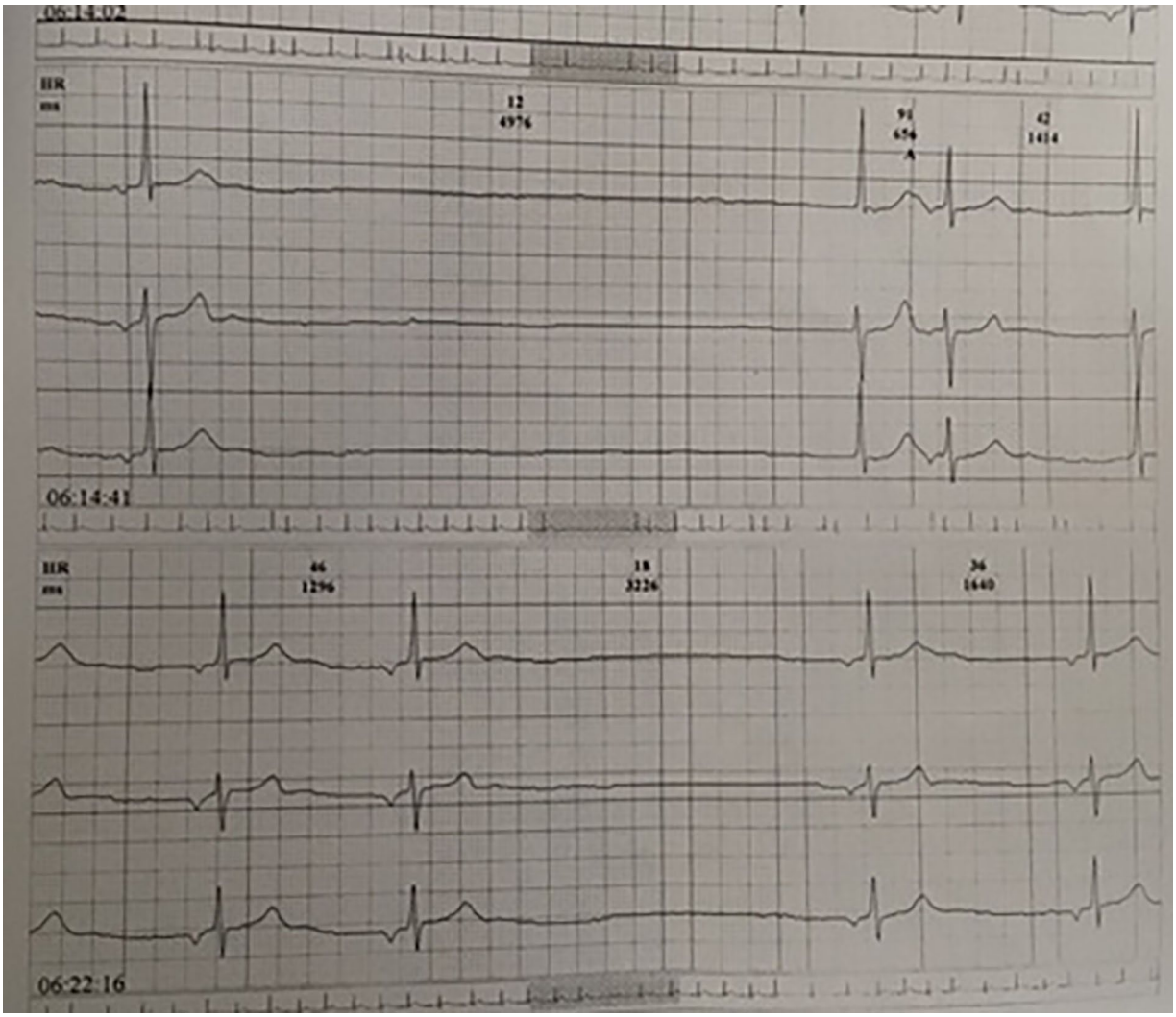

Figure 1. ECG Holter demonstrating sick sinus syndrome, with a 4.9 seconds pause.

Based on patient's symptoms and examinations it was concluded that the patient had situs inversus totalis, with mirror image dextrocardia and sick sinus syndrome. Pacemaker implantation was indicated.

Based on echocardiography findings, the operator decided not to perform angiography prior to the procedure. The approach site was chosen left subclavian vein. Under local anesthesia, from the left subclavian vein, the guide wire was passed through superior vena cava to the right atrium, and ventricle without any obstacles. The passive right ventricular electrode was positioned adequately and subsequently the atrial electrode was positioned and fixed. Chest X ray demonstrated correct placement of the atrial and ventricular leads (Figure 2).

Discussion

Possible causes of sick sinus syndrome are: degenerative fibrosis, ischemia, cardiomyopathies, infiltrative heart diseases, congenital abnormalities, medications, hypothyroidsm, electrolyte abnormalities, autonomic dysfunction, etc. Sick sinus syndrome usually occurs in older patients, although it can be found in all

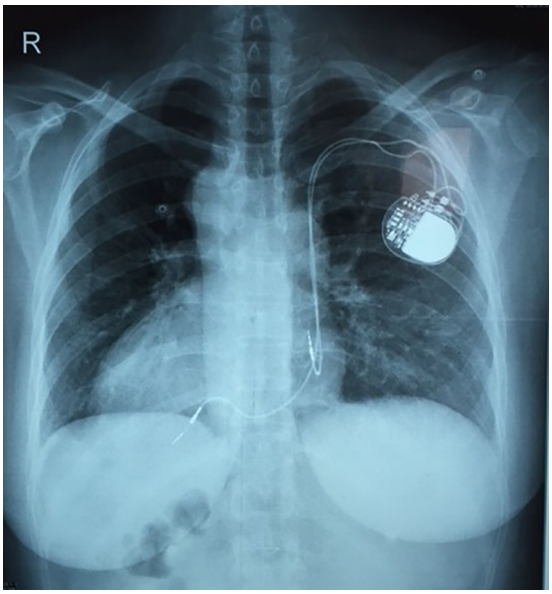

Figure 2. Chest X ray showing the heart silhouette and the apex in the right hemithorax and correct placement of atrial and ventricular pacemaker leads.

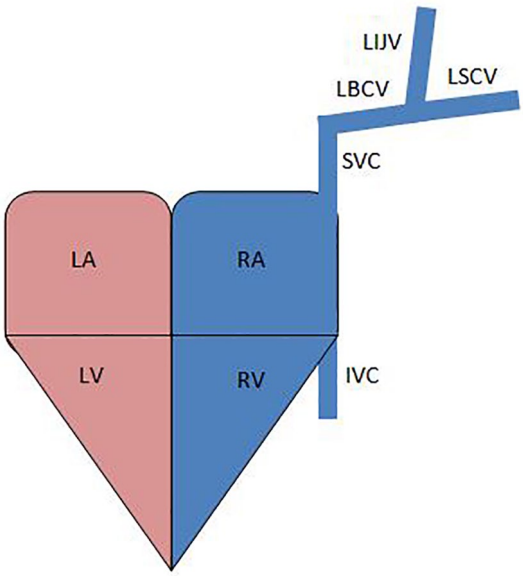

Figure 3. Schematic presentation of the heart anatomy in relation to right atrial venous draining and left subclavian vein access in our patient with situs inversus dextrocardia.
Abbreviations: IVC, inferior vena cava; LA, left atrium; LBCV, left brachiocephalic vein; LIJV, left internal jugular vein; LSCV, left subclavian vein; LV, left ventricle; RA, right atrium; RV, right ventricle; SVC, superior vena cava.

age groups.[2] Congenital abnormalities are among possible causes of sick sinus syndrome. Aside from dextrocardia, our patient did not show any other cardiac congenital anomalies on echocardiography. However, there are a few reported cases in the literature revealing the association between sick sinus syndrome and dextrocardia.[3-6] Nevertheless, to our best knowledge our patient is the youngest adult patient reported in the medical literature with dextrocardia and sick sinus syndrome. Kay et al in a large retrospective study showed that of 1484 pacemakers placed at their institution during a 10 year period, there were 18 (1.2%) patients between the ages 20 and 40 years who were not postoperative congenital heart disease cases. Twelve of these 18 patients had primary sick sinus syndrome. This large study confirms that sick sinus syndrome requiring pacemaker implantation is rare in young adults.[7]

The necessity for pacemaker implantation in patients with dextrocardia, as reported in the literature, could be either due to sick sinus syndrome and high grade or complete atrio-ventricular block.[8] The first report of pacemaker implantation in a patient with dextrocardia was described in 1976.[3] Implantation of permanent pacemaker in dextrocardia may present difficulties for the operator due to distorted anatomy of the heart as well as possible venous alterations. Therefore, authors of the few reported cases of pacemaker implantation in patients with dextrocardia recommend to perform venous angiography prior to the procedure in order to make a strategy regarding the site of the venous approach as well as other maneuvers that may be needed to facilitate pacemaker lead engagement in the right heart chambers. The first case report suggests that angiogram should always be used to guide pacemaker implantation,[3] whereas Shenthar et al, who reported the largest series of pacemaker implantations in dextrocardia, recommend angiography only in challenging implantation cases, particularly in patients with dextrocardia and situs solitus.[8] Furthermore, in this series of 6 patients, the site of venous approach was always from the right side, as it was in most of other reports.[8-10] However, there was one report where right venous access through subclavian vein failed due to anomalous venous drain in the right atrium and it succeeded from the left side following angiography which provided useful information regarding the venous anatomy.[5] Our team, based on the data from echocardiography and the fact that our patient had situs inversus dextrocardia, which is less likely to have additional vessel anomalies, decided to proceed from the left subclavian vein without prior angiography. The pacemaker leads reached the right ventricle and atrium and were adequately positioned without any difficulties. It needs to be emphasized that the proximity of superior vena cava to the left subclavian vein, in case of mirror image dextrocardia without associated venous anomalies, makes the left venous access a straightforward approach (Figure 3). Alternatively, dextrocardia that may be associated with venous anomalies, such as persistent vena cava, which empty to the right atrium through the coronary sinus are far more challenging for lead implantation.[11]

Conclusions

Situs inversus totalis, with mirror image dextrocardia may present in combination with sick sinus syndrome as early as the fourth decade of life. Implantation of the pacemaker leads from the left subclavian vein appears accessible and safe in patients with dextrocardia with situs inversus (mirror image).
Any underlying material can be accessed.

Author Contributions

AB: managed the patient while hospitalized, main contributor in designing and preparing the manuscript; **IJ**: designed the strategy and performed the procedure, contribution in providing literature regarding challenging pacemaker implantations; **XK**: data interpretation, mayor contribution in writing the manuscript; **LS**: patient referral, supervised the writing of the manuscript. All authors have read and approved the final version of the manuscript.

Informed Consent

Informed consent was obtained from the patient for publication of the images and the case report.

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