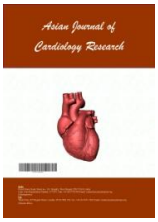

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Pacemaker Implantation in Incidentally Detected Case of Dextrocardia with Congenitally Corrected Transposition of the Great Arteries with Situs Solitus: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Complex congenital heart disease involving Congenitally Corrected Transposition of the Great Arteries (CCTGA) with dextrocardia presenting with complete heart block (CHB) possess great difficulty in placing permanent pacemaker (PPI). We had a 52 -year-old female admitted with breathlessness on exertion for 20 days. Electrocardiogram (ECG) showed complete Atrioventricular block with ventricular rate of 40/min. Chest xray (CXR) showed dextrocardia. Echocardiogram revealed a congenitally corrected transposition of the great arteries. Angiography guided technique used for dual chamber pacemaker implantation.

Keywords: Pacemaker Implantation; dextrocardia; congenital heart disease; angiography.

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ABBREVIATIONS

CCTGA : Congenital Corrected Transposition of Great Arteries
ECG : Electrocardiogram
ECHO : Echocardiography
PPI : Permanent Pacemaker
TPI : Temporary Pacemaker
ASD : Atrial Septal Defect
AO : Aorta
PA : Pulmonary Artery

1. INTRODUCTION

Since diagnosis and survival of congenital heart disease has improved over period so these patients are also seen and managed by adult cardiologists. In patient with CCTGA with dextrocardia presenting with CHB infrequently we face difficulties in device implantation because of change in relationship of atria and ventricle and ventricle to great arteries and also there is altered fluoroscopic orientation makes permanent pacemaker lead placement very challenging. There are very few literatures describing PPI in complex congenital heart disease. We describe a middle-aged patient with CCTGA who present with complete atrioventricular block. We inserted a temporary pacemaker (TPI) via right femoral route. Permanent dual chamber pacemaker (DDDR, Medtronic) later implanted with angiography guided technique.

2. CASE REPORT

A 52 -year-old female presented to JJ Hospital Mumbai, Maharashtra in March 2021 with history of breathlessness, chest pain and giddiness on exertion for 20 days. Her Blood pressure was 100/64 mmHg and heart rate was 40/min. Apex beat and heart sounds appreciated on right side and gastric symphony noted on left side. The ECG (Fig. 1) showed complete heart block with ventricular beat of 40/min with right axis deviation, absent R wave progression, positive QRS in aVR lead suggestive of dextrocardia. CXR (Fig. 2) was suggestive of mild cardiomegaly with dextrocardia. Temporary pacemaker was inserted via right femoral vein which is seen to course along right sided inferior vena cava which further confirm atrial situs as solitus (Fig. 3). Echocardiogram (Fig. 4A, 4B, 4C) revealed Visceroatrial situs solitus (VASS), dextrocardia with AV discordance present. Left atrium connected to morphological RV and is left to morphological LV which is connected to right atrium. Ostium secundum atrial septal defect (OS ASD) of 15 mm also seen. Ventriculo-atrial (VA) discordance present, aorta arising from morphological right ventricle (RV) forming arch. Pulmonary artery arising from morphological left ventricle (LV) and is bifurcating into two branches of pulmonary artery (PA). CT cardiac and abdomen done showed situs solitus and finding in CT were correlated with Echocardiography. DDD pacemaker was implanted via right subclavian vein. We used Dual chamber permanent pacemaker programmed in DDDR mode for our patient. In anteroposterior (AP) fluoroscopic view, under local anaesthesia right subclavian vein identified and access obtained. We noted difficulty in passing guidewire as there was acute angulation between subclavian vein and superior vena cava that was preventing guidewire to enter right atrium so a short guidewire (Terumo) exchanged over dilator and 3,5 JR (Judkins right) coronary catheter was used to pass guidewire into right atrium. Then ventricular lead advanced through subclavian vein to morphological right atrium to morphological left ventricle and advanced into pulmonary artery. Then slowly withdrawn to morphological left ventricle and fixed to a position that is seen towards lateral in right anterior oblique (RAO) view. Lead was screwed to apex. Parameters obtained here were threshold: 0.7 V, sensed R wave of 12.5mV with lead impedance of 720 ohm. Then atrial lead was advanced and positioned into right atrial appendage. Pacing parameter here obtained were threshold 0.6 V, sensed P wave of 2.1 mV and impedance of 630ohm. Both leads attached to pacemaker battery that placed in prepectoral area and wounds closed in layers. After pacemaker implantation, the patient did well with no postoperative complications. Post procedure 12 lead ECG showed RBBB pacing pattern (Fig. 6).

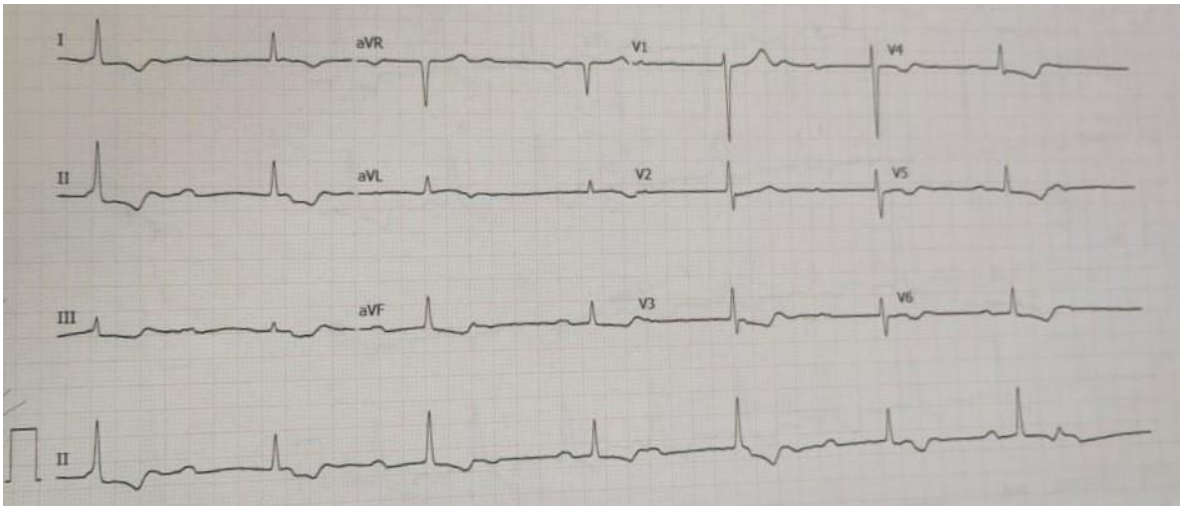

Fig. 1. ECG at the time of admission. Preprocedural ECG showing sinus rhythm with AV dissociation complete heart block. Note the normal p wave axis, the decreasing amplitude of R waves across leads V2–V6, and the absence of septal q waves suggestive of dextrocardia

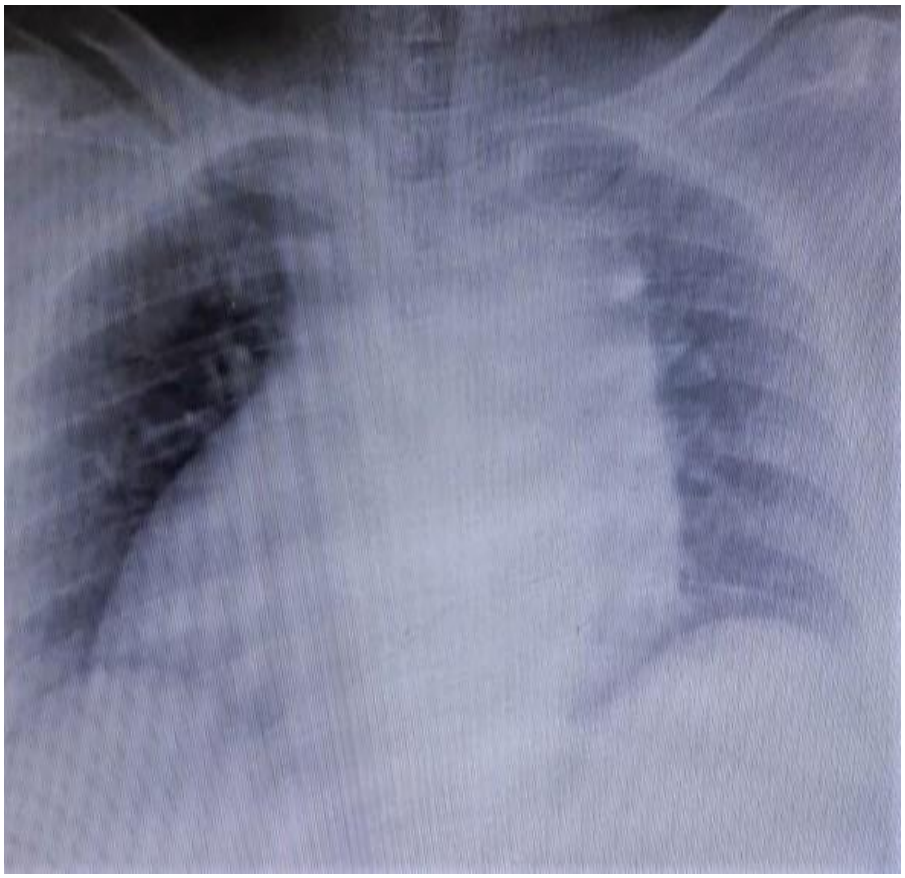

Fig. 2. CXR PA view. Note here apex clearly seen on right side suggestive of dextrocardia. Liver also seen on right side suggestive of situs solitus

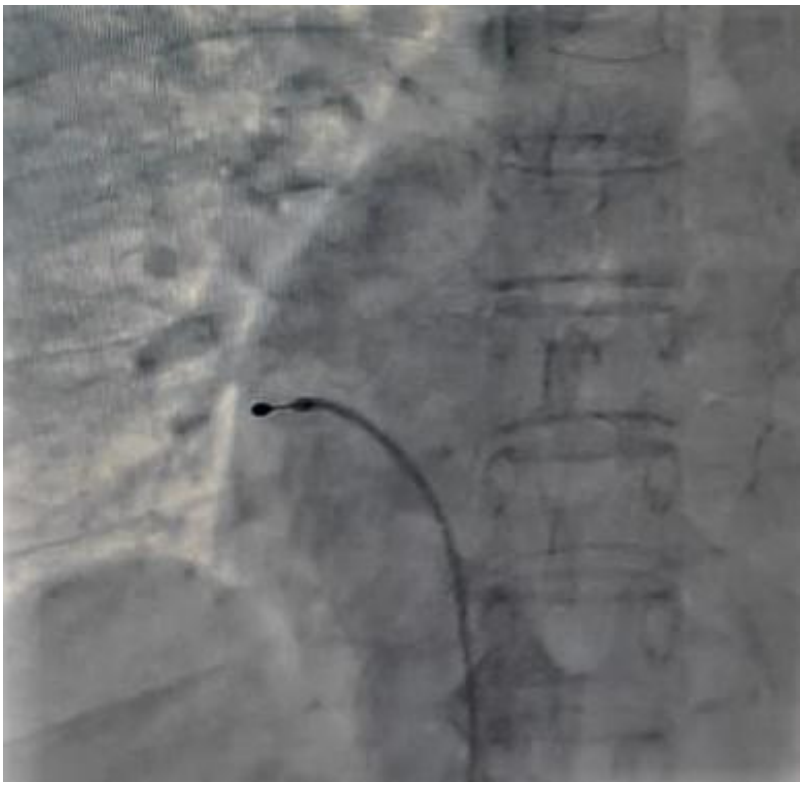

Fig. 3. Temporary pacemaker inserted soon after diagnosing complete heart block via right femoral route and paced into ventricle. Note temporary pacemaker lead is seen right to spine which further confirms the situs solitus. Here lead can be seen in morphological left ventricle

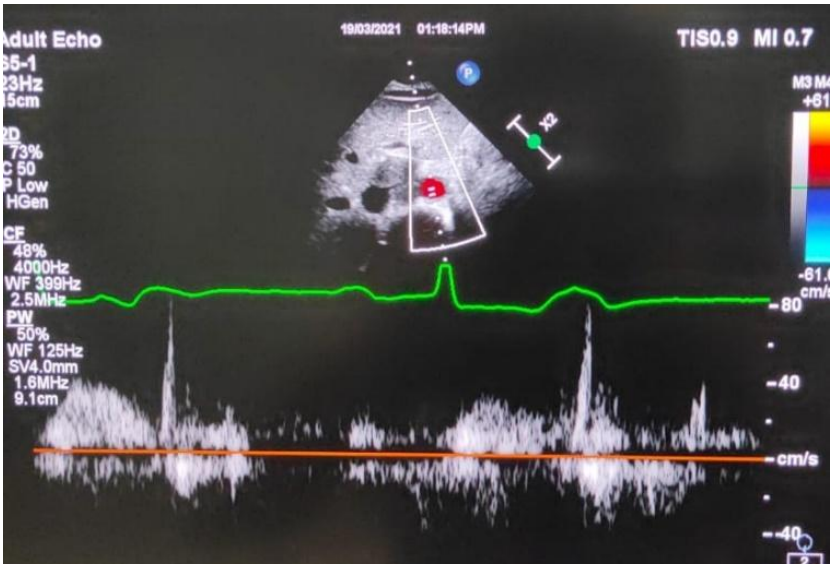

Fig. 4A. Echocardiography of our patient. Showing aorta on left side and inferior vena cava on right side suggestive of situs solitus

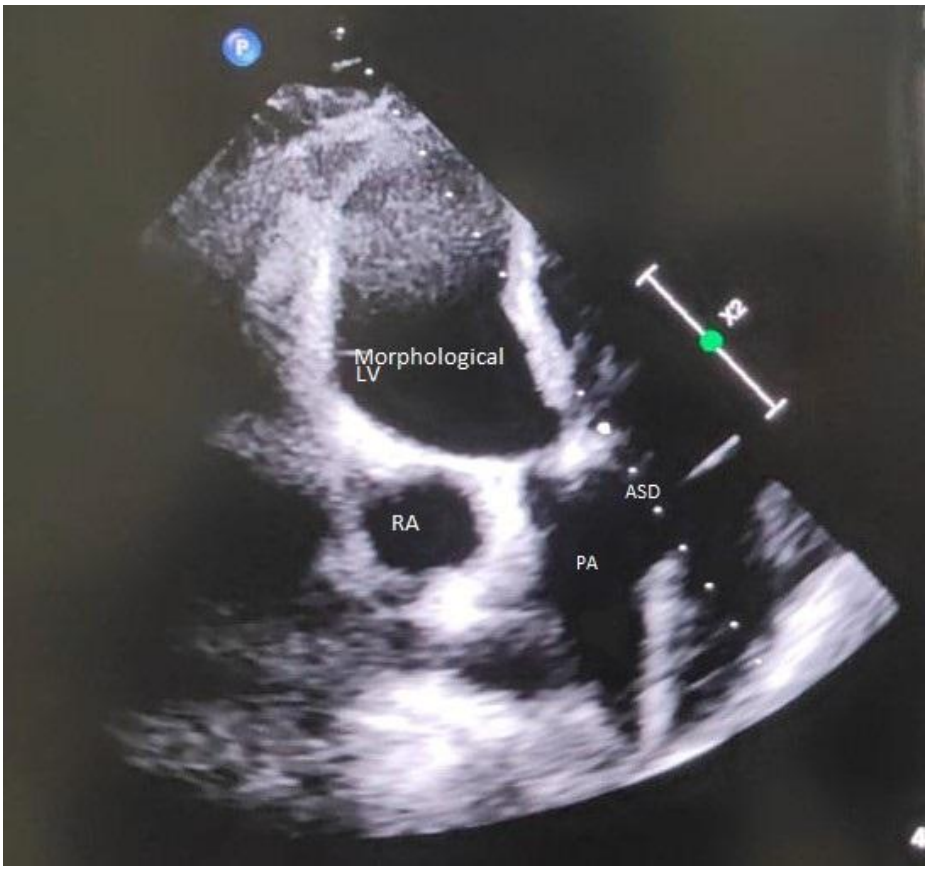

Fig. 4B. Right atrium (RA) connecting to morphological left ventricle (LV). Pulmonary artery and bifurcation seen arising from morphological left ventricle. Ostium secundum atrial septal defect (ASD) of 15 mm can be seen

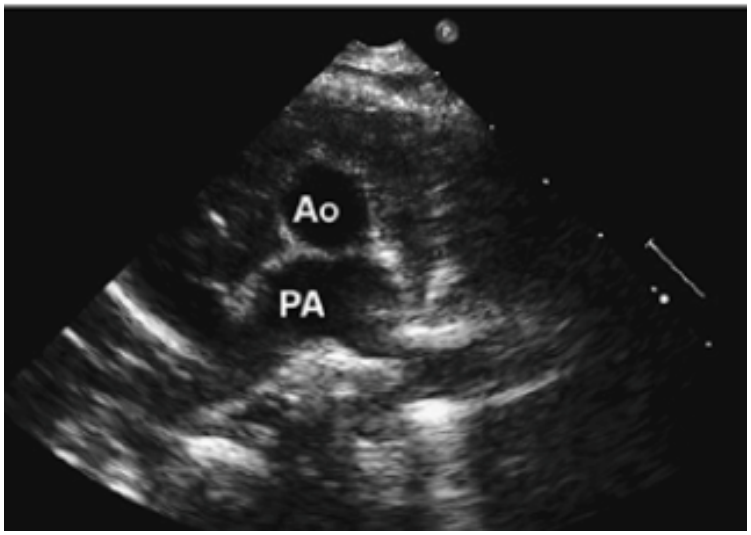

Fig. 4C. Short axis view showing aorta (AO) is anterior and left to the pulmonary artery (PA)

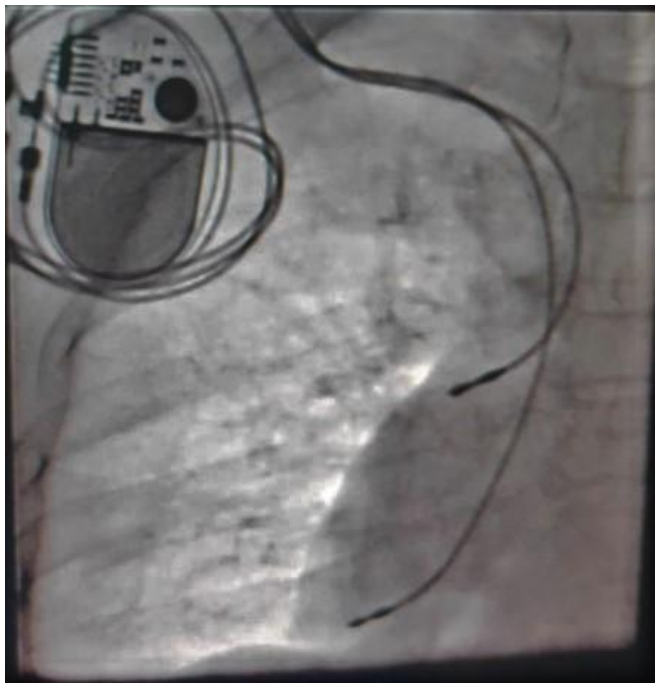

Fig. 5. RAO view. Post permanent pacemaker implantation. After stabilisation patient underwent permanent pacemaker implantation (DDDR mode). Note: Atrial lead can be seen placed into morphological right atrium appendage. And ventricle can be seen lead placed in morphological LV and screwed to apex

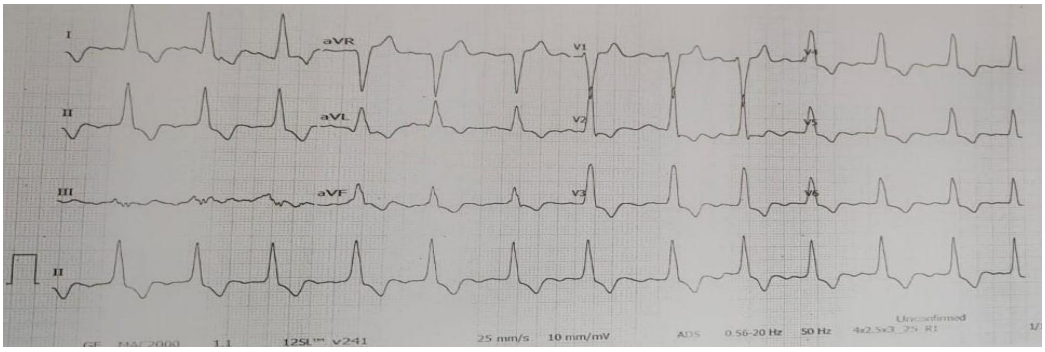

Fig. 6. Post Permanent pacemaker implantation ECG. Note here ECG showing atrial sense and ventricle pacing rhythm. ECG showing RBBB pattern here as ventricle lead is placed in morphological left ventricle

3. DISCUSSION

In CCTGA there occurs embryonic malrotation of the atria, ventricles, and great arteries resulting in atrioventricular (AV) and ventriculoarterial discordance (VA) [1]. CCTGA has a prevalence of 0.4–0.6% of all congenital heart disease cases [2]. The hallmark of this complex disease is “double discordance” or a combination of atrioventricular and ventriculoarterial (VA) discordances. This can occur in the presence of situs solitus or situs inversus [3]. The majority of cases is associated with additional cardiac defects, such as a OS Atrial septal defect,ventricular septal defect (VSD), pulmonary outflow tract obstruction, tricuspid valve (TV) anomalies, dextro-/mesocardiac, aortic arch anomalies and rhythm disturbances/atrioventricular block [4-7]. In contrast, the presence of extracardiac or

chromosomal anomalies is low. Usually, the diagnosis is made for the first time because of a heart murmur or incidentally when an ECG, chest X-ray, or echocardiogram is performed for other reasons [8]. The common presentations are symptoms of congestive heart failure due to systemic AV valve regurgitation if there is no associated lesion. Patients with CCTGA have an increased incidence of both supraventricular and ventricular arrhythmias with a concomitant increased risk of sudden cardiac death [9]. AV conduction disturbances are found in almost half of the patients, the commonest being a prolonged PR interval. Complete heart block develops in up to 30% of patients with CCTGA [10]. CCTGA is associated with displacement of the AV node away from Koch's triangle to an anterior/superior position within the right atrium due to the congenitally malpositioned conduction system with an elongated AV bundle, which has to take a much longer route to reach the ventricles [11]. Some authors reported that conduction system fibrosis was recognized to contribute to a complete AV block [12]. In CCTGA, there are both AV and ventriculoarterial discordance such that the right atrium drains into the morphologic left ventricle, which is connected to the pulmonary artery. This results in unusual orientation of both AV valves and the cardiac chambers, increasing the complexity of device implantation procedures [13]. The highlights of this unique case are: CCTGA is a rare cardiac anomaly where many patients will remain asymptomatic for much of their lives. CCTGA patients have reduced tolerance for exercise and have reported reduced quality of life compared to a general population. Prompt diagnosis and early recognition of signs of cardiac failure are essential in preventing mortality and morbidity in such cases.

4. CONCLUSION

Patient presenting with dextrocardia with situs solitus one should suspect possibility of CCTGA. Permanent pacemaker implantation may be needed in 30% of such cases as they present with CHB at any time of life. So having this knowledge, these patients need to follow up regularly with ECG and Echocardiography. Interventional cardiologists and electrophysiologists should be familiar with this condition. Very few cases report available on such patients so good knowledge of cardiac anatomy may help pacemaker implantation in this population.

CONSENT AND ETHICAL APPROVAL

As per international standard or university standard guideline patients consent and ethical approval has been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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