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Case Report

Internal Medicine Section

Congenitally Corrected Transposition of Great Arteries in a Young Adult: A Case Report

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ABSTRACT

Congenitally Corrected Transposition of the Great Arteries (ccTGA) is a rare congenital cardiac anomaly characterised by atrioventricular and ventriculoarterial discordance, comprising less than 0.05% of all congenital heart diseases. While often associated with abnormalities such as Ventricular Septal Defect (VSD), pulmonary obstruction, coarctation, and Ebstein anomaly, ccTGA can also occur as an isolated defect. Such cases may remain asymptomatic for decades and are sometimes discovered incidentally. We present a case of a 26-year-old asymptomatic male diagnosed with ccTGA during an evaluation for inguinal hernia surgery. The patient's only complaint was occasional palpitations. Electrocardiography revealed Complete Heart Block (CHB), which was confirmed by echocardiography and coronary Computed Tomography (CT) angiography, showing typical findings of ccTGA. The Right Ventricle (RV) had a reduced ejection fraction of 40-45%, with mild regurgitation of the tricuspid and aortic valves. Pacemaker implantation was recommended for the management of CHB. This case highlights the importance of early recognition of ccTGA-associated complications such as CHB and underscores the need for regular follow-up to monitor for Heart Failure (HF) and other late sequelae.

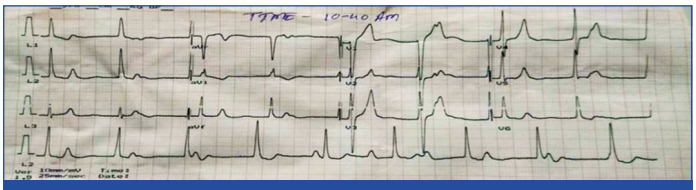
Keywords: Complete heart block, Congenital heart disease, Pacemaker implantation

CASE REPORT

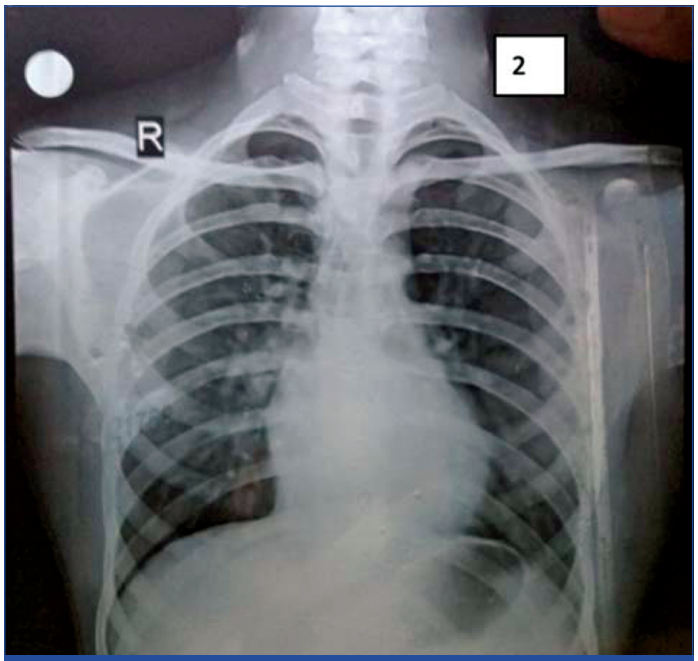
A 26-year-old unmarried male presented to our hospital with an inguinal swelling causing discomfort for the past two months. Upon routine preoperative work-up for surgical management of his inguinal hernia, he was found to have CHB on the Electrocardiogram (ECG). His medical history was significant for monthly administrations of injection Penidura (penicillin prophylaxis) for the past two years due to an apparent history of rheumatic heart disease; however, no previous medical records were available. Upon further inquiry, he reported occasional palpitations but denied experiencing chest pain, dizziness, or shortness of breath.

Physical examination revealed bradycardia (mean heart rate of 56 bpm) with an irregular pulse, and peripheral pulses were equal in both upper limbs. He was normotensive (mean blood pressure of 110/72 mmHg) with an oxygen saturation of 98% on room air. Cardiac examination showed a laterally displaced cardiac impulse and a grade 3/6 holosystolic murmur audible throughout the precordium. Apart from the inguinal hernia, the rest of the physical examination was unremarkable.

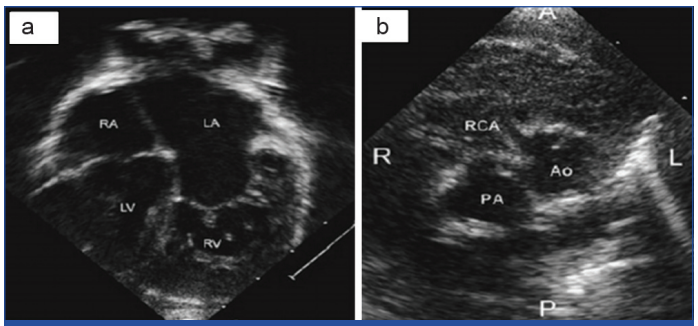
Routine surgical fitness evaluation included an ECG, which indicated CHB [Table/Fig-1]. A chest radiograph (posteroanterior view) showed a prominent left upper cardiac border with an unusual aortic knob and pulmonary trunk [Table/Fig-2]. Subsequent echocardiography revealed findings suggestive of ccTGA. In the five-chamber apical view, atrioventricular and ventriculoarterial discordance were evident. The morphological RV had a reduced ejection fraction of 40-45%, with mild regurgitation of the tricuspid and aortic valves. The Right Atrium (RA) connected to the Left Ventricle (LV) via the mitral valve [Table/Fig-3a,b]. There were no associated abnormalities, such as VSD, nor any valvular or subvalvular pulmonic stenosis.



[Table/Fig-1]: ECG indicating Complete Heart Block (CHB).

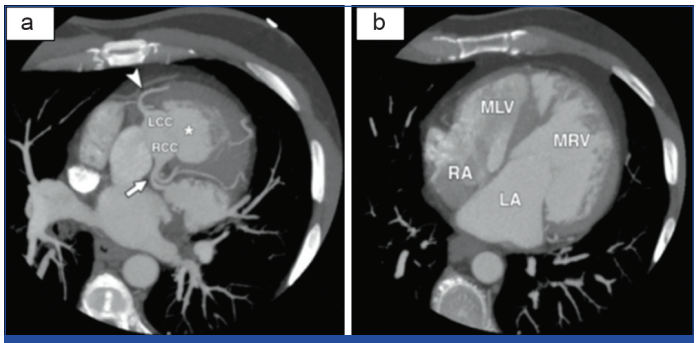


[Table/Fig-2]: A chest radiograph (posteroanterior view) showing a prominent left upper cardiac border with an unusual aortic knob and pulmonary trunk.



[Table/Fig-3a,b]: Echocardiogram showing Right Atrium (RA) connected to the Left Ventricle (LV) via the mitral valve.

To further assess cardiac morphology and coronary anatomy, coronary CT angiography was performed. It confirmed the diagnosis of ccTGA and showed good biventricular contraction. The ascending aorta was located anteriorly and to the left of the main pulmonary artery. Coronary-ventricular concordance was noted, with the Right Coronary Artery (RCA) being dominant [Table/Fig-4a,b].



[Table/Fig-4]: In CT scan coronary-ventricular concordance was noted, with the Right Coronary Artery (RCA) being dominant: a) Axial CT Maximum-Intensity-Projection (MIP) image shows anterior, right-sided aortic sinus giving rise to anterior descending coronary artery (arrowhead), which courses along anterior interventricular groove supplying morphologic LV. Posterior aortic sinus gives rise to Right Coronary Artery (RCA) (arrow), which courses along posterior atrioventricular groove between left atrium and morphologic Right Ventricle (RV) and, in turn, gives rise to infundibular and marginal branches supplying morphologic RV. LCC: Left coronary cusp, RCC: Right coronary cusp, star= morphologic right ventricular outflow tract; b) Axial CT MIP image shows four chambers of heart. RA: Right atrium, LA: Left atrium, MRV: Morphologic right ventricle, MLV: Morphologic left ventricle. Of note, moderator bandlike structure in morphologic LV most likely represents anomalous papillary muscle.

The patient was counseled about his condition, emphasising the importance of recognising warning symptoms related to acute cardiac decompensation. He underwent permanent pacemaker implantation within three weeks for the management of CHB, in accordance with the joint guidelines set by the American College of Cardiology (ACC), the American Heart Association (AHA), and the Heart Rhythm Society (HRS) [1]. After discharge, the patient continues to attend regular outpatient follow-ups every six months for lifelong surveillance. At the most recent follow-up, one year after pacemaker implantation, the patient is asymptomatic, and the ECG is unremarkable.

DISCUSSION

ccTGA is a rare congenital cardiac anomaly characterised by atrioventricular and ventriculoarterial discordance, accounting for less than 0.05% of all congenital heart diseases. It frequently occurs alongside associated abnormalities such as VSD, pulmonary or subpulmonary obstruction, coarctation, and Ebstein anomaly, although it may also present as an isolated cardiac defect [2]. In a study by Marler AT et al., a patient was treated for an atrial septal defect with an occluder device before the diagnosis of ccTGA was considered [3]. While congenital heart diseases typically manifest during infancy or childhood, patients with isolated ccTGA may remain asymptomatic for decades [4].

Most cases of ccTGA are associated with one or more concomitant structural abnormalities, with VSD being the most common. In cases where ccTGA is associated with a large VSD or severe Tricuspid Regurgitation (TR), patients often present with congestive heart failure in infancy or childhood [5]. In a study by Obongonyinge B et al., 80% of their cases had VSD and TR [6]. However, isolated ccTGA, observed in only 1% of cases [7], may remain haemodynamically stable and symptomatically silent for many years, as seen in our patient.

Due to the systemic role of the RV, patients are at high risk for heart failure-related morbidity and mortality [8]. The decline in RV function may be attributed to longstanding pressure overload, myocardial ischaemia, TR, mechanical dyssynchrony, or arrhythmias [9]. CHB in ccTGA patients develops mainly due to the abnormal disposition of the atrioventricular conduction system. Its incidence increases with age, estimated at 30-38% in older children and adults with ccTGA [10]. Ajmera P et al., reported a case of a 42-year-old presenting with dizziness and loss of consciousness as a manifestation of CHB secondary to ccTGA [11]. Our patient developed CHB at the age of 26 years. Additional late complications may include progressive TR (systemic AV valve), left ventricular outflow tract obstruction, and, rarely, ventricular tachycardia. Life expectancy is reduced for these patients, with 50% of those with isolated ccTGA surviving to age 60 [8].

This case underscores the need to recognise potential complications of ccTGA, such as CHB and heart failure. Regular monitoring is crucial, as CHB occurs at an approximate rate of 2% per year [12], usually manifesting after the age of 40 years. In our patient, CHB was the primary manifestation, with occasional palpitations as the only symptom. Prompt pacemaker implantation is essential to ensure adequate cardiac rhythm and prevent further complications. Although our patient showed no signs of right-sided heart failure, vigilant monitoring and appropriate management of heart failure are necessary, especially as the risk increases with age and the presence of regurgitant cardiac lesions [13]. Ensuring regular follow-up and optimal management can significantly improve outcomes and quality of life for these patients.

CONCLUSION(S)

ccTGA is a rare congenital defect where atrioventricular and ventriculoarterial discordance results in physiologically corrected circulation. However, long-term complications arise due to the morphologic RV functioning as the systemic ventricle. This case highlights the importance of early recognition of ccTGA-associated complications, such as CHB, and underscores the need for regular follow-up to monitor for heart failure and other late sequelae.

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