

Congenitally Corrected Transposition of the Great Arteries with Situs Solitus: Pacemaker Implantation in a Septuagenarian

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ABSTRACT

Pacemaker implantation in congenitally corrected transposition of the great arteries can be challenging due to distorted cardiac anatomy that in some cases it may be accompanied by other anomalies. The present report outlines a rare case of congenitally corrected transposition of the great arteries, asymptomatic until the seventh decade of life.

Keywords: congenital heart disease, congenitally corrected transposition of the great arteries, double discordance, levocardia, permanent pacemaker, situs solitus

INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare cardiac entity identified by double discordance of the atrioventricular (AV) and ventriculoarterial connections. Such abnormalities have an occurrence of 1 in every 33,000 live births and contribute approximately 0.05% of all congenital heart malformations. [1] Complete heart block is frequently encountered in patients with aberrant conduction system anatomy. These patients eventually require pacemaker implantation over time. [2] Here, we describe a case of ccTGA with subsequent permanent pacemaker implantation due to AV dissociation in a 70-year-old male.

CASE REPORT

A 70-year-old male presented to our cardiology department with chief complaints of giddiness and presyncope

since one day. His past history was unremarkable as no history of chest pain, shortness of breath, diaphoresis, convulsive activity, fever, neurological, genitourinary or gastrointestinal complaints was observed. The patient did not smoke or consume alcohol and led a sedentary lifestyle. He also did not have significant drug history. Physical examination revealed irregular heart rate (34 bpm) and blood pressure of 130/80 mmHg in the right arm in sitting position. Electrocardiography (ECG) revealed atrioventricular (AV) dissociation with ventricular rate of 30 bpm and left axis deviation ([Figure 1](#)). Laboratory investigations included complete blood count, renal function tests, liver function tests, lipid profile, and serum electrolyte tests. All tests were normal. A 24-hour Holter monitor recorded minimum heart rate of 38 bpm and maximum heart rate of 80 bpm with intermittent AV dissociation ([Figure 2a, b, & c](#)).

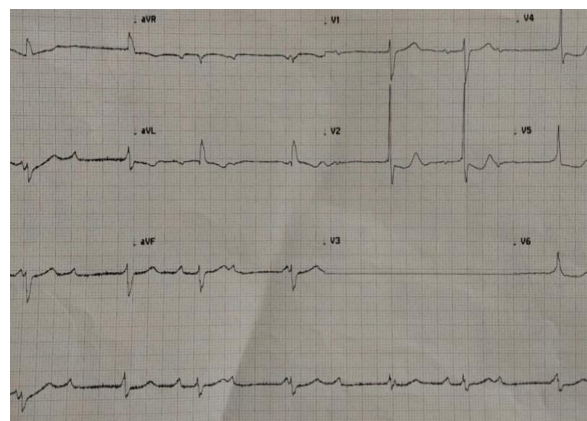


Figure 1: ECG showing left axis deviation and atrioventricular dissociation.

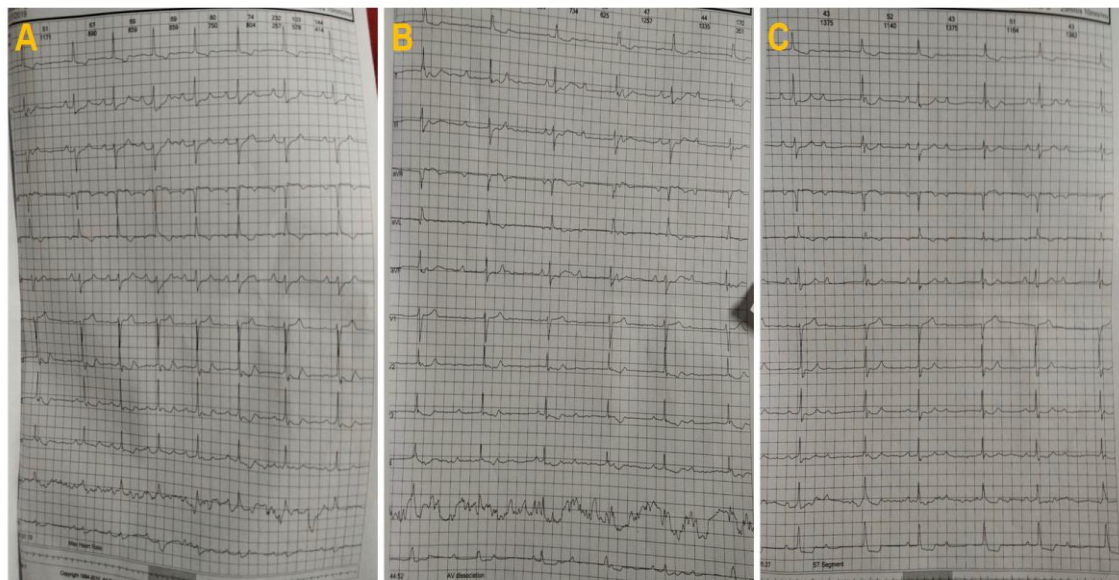


Figure 2: Holter monitor showing: (A) first degree heart block; (B) atrioventricular dissociation; and (C) atrioventricular dissociation.

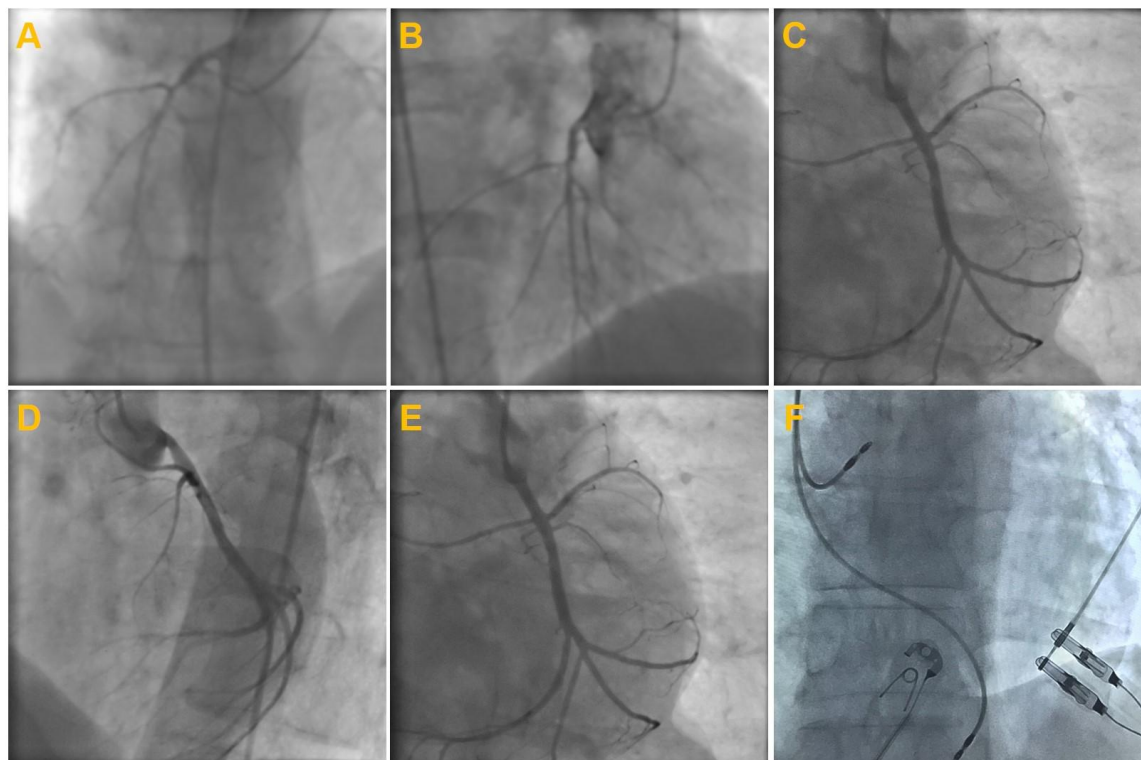


Figure 3: Coronary angiography showing: (A) left main coronary artery giving rise to left anterior descending coronary artery & left circumflex artery; (B) left coronary artery arising from right coronary ostia; (C) artery arising from left coronary ostia supplying systemic right ventricle; (D) right coronary artery supplying systemic ventricle right; (E) large right coronary artery; (F) pacemaker lead position in right atrium and sub pulmonary left ventricle

Echocardiography revealed congenitally corrected transposition of great arteries (ccTGA). The patient had situs solitus with levocardia. There were two atria and two ventricles with atrioventricular and ventriculoarterial discordance. The inferior and superior vena cavae drained into the

right atrium and three pulmonary veins drained into the left atrium. Right-sided left ventricular function was good and left-sided right ventricular function was moderate with ejection fraction of 30%. The left-sided AV valve was apically displaced with mild left AV valve regurgitation. Coronary

angiography was done to rule out any associated coronary artery disease as his history was very short of one day only. It showed classical pattern of coronary arteries in ccTGA; that is, the coronary artery originating from the right aortic sinus supplied the left ventricle and further divided into the anterior interventricular and circumflex branches ([Figure 3a & b](#)) whilst the coronary artery ([Figure 3c & d](#)) arising from the left coronary sinus supplied the right ventricle and gave rise to marginal and infundibular branches. In view of these findings, the patient was implanted with a permanent DDDR pacemaker. The ventricular lead was placed in the morphological left ventricle. The ventricular lead had a peculiar position as it was placed in the morphological left ventricle ([Figure 3e & f](#)). Pacing parameters were checked and found to be adequate. The patient was asymptomatic in the post-operative period. Telemetry done after one month and then after six months revealed normal pacing parameters.

DISCUSSION

Congenitally corrected transposition of the great arteries may present with either mirror-image atrial arrangement or typical atrial arrangement. When the atria are arranged in mirror image (sinus inversus), the sinus node is located in the left atrium and thus maintains normal association with the terminal crest and entrance of the superior caval vein. However, in typical atrial (sinus solitus) arrangement, a ventricular and atrial septa misalignment gap prompts considerable conduction abnormalities. ^[1] Oliver et al. ^[3] in their study of 38 patients documented complete AV block in 17 (57%) patients with situs solitus compared to 1 (12%) patient with situs inversus. In line with these observations, our case also followed the pattern of complete heart block accompanying presentation of situs solitus.

Sinus node dysfunction is frequently associated with senescence of the node and its surrounding tissues. ^[4] Thus, sinus node

disease could have justified irregular, slow heart rate and presyncope observed in our patient. However, Holter recording revealed AV dissociation, making this diagnosis unlikely. Hyperkalemia can lead to AV dissociation. ^[5] However, serum electrolytes were normal in our patient, thus ruling out hyperkalemia. Due to the history and physical examination, complete heart block was suspected in our patient. ECG revealed AV dissociation hence, permanent pacemaker implantation was planned. Echocardiography was subsequently performed and revealed ccTGA.

Hofferberth et al. ^[6] conducted a study on ccTGA patients with AV block. They found 40% patients implanted with univentricular pacing systems required an upgrade to DDDR pacing systems due to an indication of ventricular dysfunction. Furthermore, of those who primarily received DDDR pacing systems, none developed ventricular dysfunction during short term follow-up. We too opted for DDDR pacemaker for our patient.

Permanent pacemaker implantation in contact with the septum in patients with ccTGA may provoke paradoxical motion of the interventricular septum causing functional compromise of the morphological right ventricle. Worsening of the systemic right ventricular function can lead to congestive cardiac failure. ^[7] Vasliu et al. ^[7] placed the lead in contact with the ventricular lateral wall. In our patient the pacemaker was implanted in the morphological left ventricle.

CONCLUSION

Very few cases of ccTGA present as isolated ccTGA remaining asymptomatic throughout life. Conduction disease gradually manifests into complete AV block through the years. Such patients may require conventional pacemaker therapy, which in the presence of ventricular dysfunction may further compromise cardiac performance. Our case highlights the utility and efficacy of DDDR pacing devices in patients with conduction disease.

Conflict of Interest: The authors declare no conflict of interest.

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