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Original Research Paper

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CASE REPORT : COMPLETE HEART BLOCK IN CCTGA REQUIRING PACEMAKER IMPLANTATION

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ABSTRACT One of the rare congenital heart diseases is congenitally corrected transposition of great arteries. A pacemaker implantation in congenitally corrected transposition of great arteries is quiet challenging due to altered anatomy. we report a case of 14yr / girl presented with bradycardia, with a history of syncope to emergency department, incidentally CCTGA was discovered. Electrocardiogram shown complete heart block with junction escape rhythm of 55bpm and features suggesting of ventricular inversion. Chest X ray showed cardiomegaly. CCTGA was revealed on two-dimensional echocardiography. The implantation of permanent DDDR pacemaker was uneventful, subsequently she recovered and was discharged.

KEYWORDS :

INTRODUCTION

Congenitally corrected transposition of great arteries is characterized by transposition of great arteries and inverted ventricles, atrioventricular valves but normal atrial situs. It accounts for about 1% of patients born with CHD. Of these 1 % have uncomplicated CCTGA. Because of unusual position of atrioventricular node and abnormal conduction system, cardiac conduction disorders are frequent in CCTGA.

Isolated CCTGA remain asymptomatic till later decades of life. They often present with life threatening complications like tricuspid regurgitation, heart block, ventricular arrhythmias, systemic ventricular dysfunction.

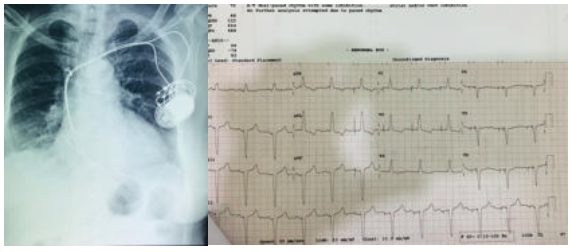
Electrocardiography, echocardiography, cardiac computed tomography (CT), cardiac magnetic resonance imaging (MRI) are used to establish the diagnosis of CCTGA.

We report a case of complete heart block in a 14 year old girl with CCTGA. she was successfully managed with permanent pacemaker and was discharged after she was stabilized.

CASEREPORT

A 14 year young girl with chief complaint of syncopal attack since one day presented to emergency department. Physical examination findings were pulse 55bpm , blood pressures were 100/70 mmhg in sitting position in the right arm , her laboratory investigation included complete blood count , renal function tests , liver function tests , lipid profile and serum electrolytes . All tests were normal except for raised SGPT (ALT) enzyme (70IU/l)

Electrocardiogram shown CHB with AV dissociation with narrow QRS escape rhythm and ventricular inversion features .Echocardiography confirmed congenitally corrected transposition of great arteries (CCTGA), there were two atria and two ventricles with atrioventricular and ventriculoarterial discordance, mild left atrioventricular valve regurgitation. CCTGA with complete heart block and bradycardia indicated permanent pacemaker implantation into the patient. She was implanted with a permanent DDDR pacemaker. The ventricular lead was placed in the morphological left ventricle. The leads position of pacemaker are in right atrium and morphological left ventricle. Pacing parameters were acceptable. The procedure was uneventful, and she was discharged.



A) chest Xray showing dual chamber pacemaker B) ECG after pacemaker implantation

DISCUSSION:

Congenitally corrected transposition of great arteries is a condition in which there is atrioventricular and ventriculo-arterial discordance. The morphologically right atrium is connected to a morphologically left ventricle across the mitral valve, with the left ventricle then connected to the pulmonary trunk. The morphologically left atrium is connected to the morphologically right ventricle across the tricuspid valve, with the morphologically right ventricle connected to the aorta. The systemic venous return is pumped to lungs, while pulmonary venous return is directed to the body resulting in normal physiological circulation.

Isolated CCTGA are asymptomatic and are not associated with anomalies, but complications may occur in the later years of life. Our case presented with syncope and bradycardia and on evaluation was found to have bradycardia, third degree heart block. Hyperkalemia can lead to AV block, but our patient has normal electrolytes [1]. Further investigations revealed the presence of CCTGA, mild valvular regurgitation. Cardiac conduction defects are frequent occurrence in congenitally corrected transposition of great arteries as the location of AV node and course of bundle of his is anomalous though sinus node is normal[2] Implantation of Permanent pacemaker was planned.

The study conducted on CCTGA patients with AV block, by Hofferberth et al. , 40% patients implanted with univentricular pacing systems required an upgrade to DDDR pacing due to

manifestation of ventricular dysfunction. Additionally, patients who received DDDR pacing, none developed ventricular dysfunction during the follow-up[3].

Cardiac transplantation is required in patients with complicated CCTGA, while some patients can be managed with pacemaker implantation. More often, pacemaker implantation in such situation is challenging concerning to complex anatomy [4]. Besides pacemaker implantation in such patients might lead to systemic ventricular function worsening and atrioventricular valve regurgitation, this is probably might be due to modification in ventricular septum position that causes a septal shift and failure of tricuspid valve coaptation[5]. In spite of these complications, pacemaker was successfully implanted in our patient and she is being followed up in favorable condition.

CONCLUSION:

Very few cases of CCTGA present as isolated CCTGA and remain asymptomatic. Conduction disease gradually manifests into complete AV block through the years, this is a rare case presentation of complete heart block, in an incidental diagnosis of CCTGA. We managed the patient with a dual chamber pacemaker. These patients need follow up as they may develop complications related to CCTGA.

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