

Complete atrioventricular block in an adult with congenitally corrected transposition of the great arteries with interrupted inferior vena cava

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A 56-year-old man got admitted as he was suffering from dizziness for 3 days. Electrocardiogram (ECG) showed complete atrioventricular (AV) block with ventricular rhythm of 35/min. We found that he had no inferior vena cava (IVC) which drained into right atrium in the middle of temporary pacing lead insertion. Venous drainage into superior vena cava from dilated azygos vein was identified after venogram. Echocardiogram revealed a congenitally corrected transposition of the great arteries (CCTGA). Chest computed tomography (CT) angiogram revealed AV and ventriculoarterial discordance with reversed ventricles and interrupted IVC with azygos continuation. DDD pacemaker was implanted via left axillary vein without any problem.

Key words: Complete atrioventricular block, congenitally corrected transposition of the great arteries, interrupted inferior vena cava

INTRODUCTION

As the survival of patients with congenital heart diseases continues to improve, adult cardiologists will increasingly become involved in the management of these patients. Also, in the cardiac rhythm management era, the situation is the same. Infrequently, we face difficulties during device implantation. Congenitally corrected transposition of the great arteries (CCTGA) is one of them.

We describe a middle-aged patient with CCTGA who presented with complete atrioventricular (AV) block. Interestingly, he did not have inferior vena cava (IVC). Thus, we inserted a temporary pacing lead via an azygos vein. To the best of our knowledge, there has been only one case^[1] with CCTGA, interrupted IVC, and combined complete AV block in an adult reported in the literature until 2012. This is the second report.

CASE REPORT

A 56-year-old man presented to Chonnam National University Hospital, Gwangju, Korea, in 2009 with dizziness and chest tightness for 3 days. He was a

30 pack-year current smoker. His blood pressure was 140/90 mmHg and heart rate was 35/min. Grade 1 systolic murmur was audible at the apex. The electrocardiogram (ECG) showed complete AV block with a ventricular beat of 35/min [Figure 1]. Sinus rate was 54/min. Mild cardiomegaly was seen on chest X-ray. Temporary pacemaker was tried via transfemoral approach at first. However, electrode catheter was not advanced into the right ventricle despite several trials. We found that he had no IVC which drained into right atrium. Venous drainage into superior vena cava from dilated azygos vein was identified after venogram [Figure 2]. Thus, an electrode was positioned in the right side ventricle via azygos vein and superior vena cava. Echocardiogram revealed that the position of the two ventricles was reversed resulting in the following: the right atrium connecting to the left ventricle, the left atrium connecting to the right ventricle, also the aorta arising from the right ventricle and supplying systemic circulation, and the pulmonary artery arising from the left ventricle and supplying pulmonary circulation [Figure 3]. Chest computed tomography (CT) angiogram revealed AV and ventriculoarterial discordance with morphologically left ventricle in the right side and morphologically right ventricle in the left side and interrupted IVC with azygos continuation [Figure 4]. Abdomen CT angiogram revealed situs ambiguous with polysplenia. Anyway, he needed permanent pacemaker definitely. Thus, DDD pacemaker was implanted via left axillary vein [Figure 5]. The pacemaker parameter showed ordinary feature. We have followed him up without any problem.

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Received: 28-02-2012; **Revised:** 13-05-2012; **Accepted:** 24-05-2012

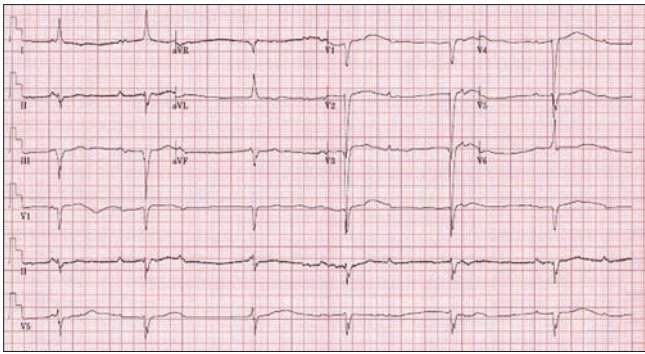


Figure 1: ECG showed complete AV block with junctional escape rhythm with a rate of 35/min. Sinus rate was 54/min. Q waves in the inferior and right chest leads and absence of q waves in V5–6 are the features of typical QRS pattern of CCTGA

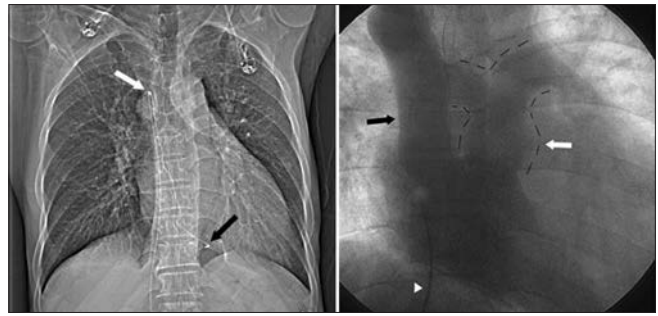


Figure 2: Left panel: Pacing electrode (black arrow) was positioned in right side ventricle via azygos vein and superior vena cava (white arrow). Right panel: Venous drainage into superior vena cava from dilated azygos vein (black arrow) was identified after venogram (white arrow and dashed line, pulmonary artery; white arrowhead, interrupted inferior vena cava)

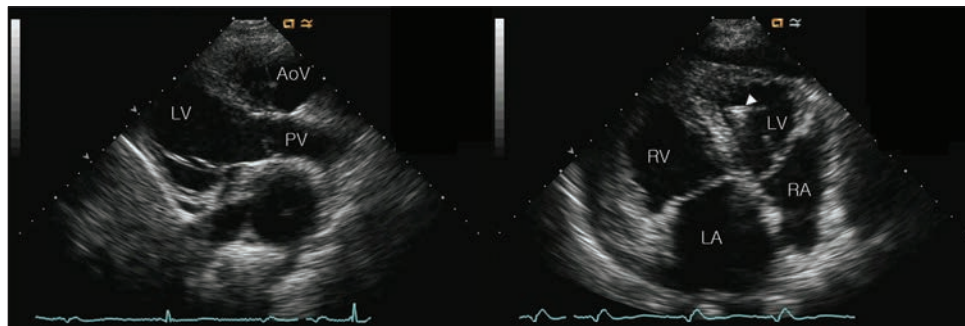


Figure 3: Echocardiogram revealed that the position of the two ventricles was reversed so that the right atrium (RA) connected to the left ventricle (LV). In normal heart, we display an LV in the opposite side to echo marker. In this image, the LV is in the same side as that of echo marker. So, transposition (being corrected) of both ventricles is seen. The left atrium (LA) connects to the right ventricle (RV); also, the aorta arises from the right ventricle and supplies the systemic circulation; the pulmonary artery arises from the left ventricle and supplies the pulmonary circulation. Arrowhead is the pacing electrode in the left ventricle (AoV, aortic valve; PV, pulmonic valve)

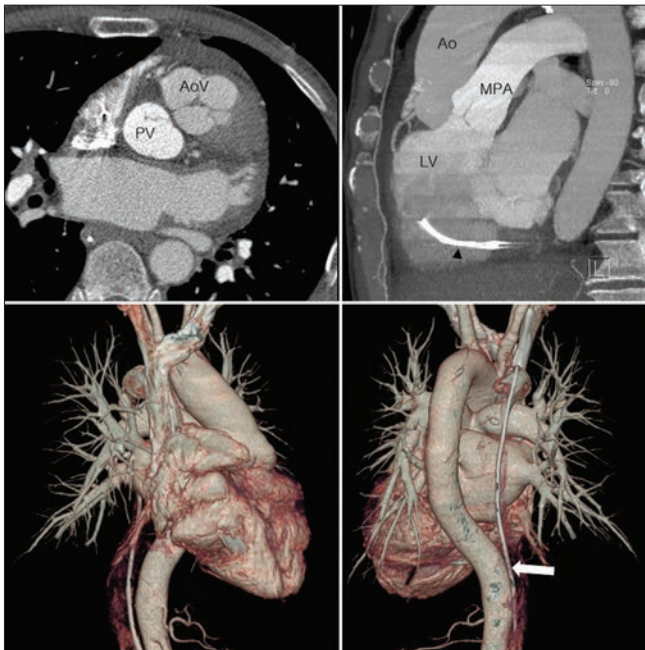


Figure 4: Chest CT angiogram revealed atrioventricular and ventriculoarterial discordance with morphologically left ventricle (LV) in the right side and morphologically right ventricle in the left side and interrupted inferior vena cava with azygos continuation (black arrowhead and white arrow is a pacing electrode catheter) (AoV, aortic valve; PV, pulmonic valve; MPA, main pulmonary artery)

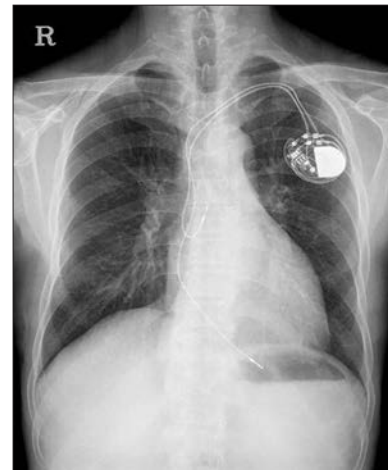


Figure 5: DDD pacemaker was implanted via left axillary vein

DISCUSSION

CCTGA has a prevalence of 0.4–0.6% of all congenital heart disease cases.^[1] It is usually accompanied by additional cardiac malformations, but may occur without associated defects. 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.^[2] 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.^[3] 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.^[1] When complete heart block is present, the QRS complex is narrow because conduction block occurs in the proximal site. The ventricular rate is usually greater than 50/min and rarely dominates the clinical picture, which is determined by the additional anatomical abnormalities.^[2] In this case, the escape rhythm was ventricular rhythm, QRS complex was wide, and heart rate was about 35/min. 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.^[4] Some authors reported that conduction system fibrosis was recognized to contribute to a complete AV block.^[5] Some patients present early, while others present late. The combination of the congenitally malpositioned conducting tissues and acquired changes such as a fibrosis that develops throughout the years seem to be the cause of conduction block.

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.^[6] Pacing may also be associated with septal shift, which may exacerbate SV dilatation and cause worsening of systemic AV valve regurgitation. It is probably prudent, therefore, to have more frequent clinical and echocardiographic monitoring in an unoperated patient after pacemaker implantation.^[2]

A complicating factor in this case was the interrupted IVC. Interrupted IVC with azygos continuation is rare in adults.^[7] Its prevalence is 0.6–2.0% in patients with congenital heart disease. Usually, we insert a temporary pacing lead via femoral vein and IVC. Thus, we could find abnormal drainage of venous system and abnormal orientation of cardiac chambers and arteries. There is no problem in venous access for permanent pacemaker implantation because the connection between a brachiocephalic vein, a superior vena

cava, and a right atrium is ordinary. However, lead fixation in left ventricle is another complicating factor. In contrast to right ventricle, the trabeculation of left ventricle is relatively smooth. So, probability of dislodgment of pacing lead is high using passive fixation lead. In fact, epicardial lead used to be chosen for providing long-term stable lodgment of electrode. Screw fixation lead can overcome this instability. So, a screw fixation lead was chosen in a morphologic left ventricle in this patient. Stable lodgment of lead has been confirmed through pacemaker follow-up. Only 1% of patients with CCTGA are uncomplicated without associated lesions such as ventricular septal defect, pulmonary stenosis, and Ebstein's anomaly of the systemic AV valve.^[2] In a study cohort, 66% of patients with CCTGA presented initially in adulthood and 17% of these patients were older than 60 years of age. Most of these patients had significant regurgitation of the systemic AV valve.^[2] Interestingly, our patient had only mild regurgitation of the systemic AV valve. Perhaps, that is another cause of his late presentation.

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How to cite this article: Yoon N, Kim KH, Park HW, Cho JG. Complete atrioventricular block in an adult with congenitally corrected transposition of the great arteries with interrupted inferior vena cava. *J Res Med Sci* 2012;17:808-10.

Source of Support: Nil, **Conflict of Interest:** None declared.