

# Congenitally Corrected Transposition of Great arteries with AV block -cohabitation of structural and electrical cardiac abnormality: a case report

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## Abstract:

**Congenitally Corrected Transposition of Great Arteries (c-TGA) is an anomaly with atrioventricular and ventriculoarterial discordance where left atrium connects to right ventricle and right atrium to left ventricle. However in this case of double inversion, aorta carries saturated blood and pulmonary artery carries venous blood. So, normal physiological circulation is maintained. Our patient, a 38-years-old Bangladeshi male presented with palpitation, dizziness and effort intolerance (NYHAI) during exertion since childhood, which has recently become much worse. However, he**

**had no history of central cyanosis, chest pain or syncope. We diagnosed him as a case of Corrected TGA and second-degree AV block with intermittent complete heart block.**

**A Dual chamber permanent Pacemaker was inserted without any complications. No invasive treatment including corrective surgery was performed because patient's cardiac function was almost normal. Patient became totally asymptomatic after pacemaker implantation**

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## Introduction:

Congenitally corrected transposition of great arteries (c-TGA) is a rare heart disease with an incidence of 1 in 33,000 live births and prevalence of 0.05% of all congenital heart malformations.<sup>1</sup> It occurs most frequently in males with a ratio of 1.5:1.<sup>2</sup> Congenitally corrected transposition of the great arteries (c-GTA) is characterized by transposition of great arteries and inverted ventricles, atrioventricular valves and conduction system but normal atrial situs.<sup>2</sup> The cause of the malformation is not currently

known,<sup>3,4</sup> although a familial association has been found.<sup>5</sup> More than 90% of cases of c-TGA have other associated anomalies like VSD, PS, tricuspid valve or mitral valve anomalies.<sup>6-8</sup> These patients may remain asymptomatic for many years and usually diagnosed in later decades of life due to abnormal ECG, echocardiography, cardiac computed tomography or cardiac MRI. They may also present with severe life threatening complications like systemic ventricular

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dysfunction, heart block and ventricular arrhythmia.<sup>9</sup> No treatment is required for patients with isolated c-TGA (without other defects), which is not commonly found,<sup>6-8</sup> because their life expectancy has been reported to be near normal.<sup>10</sup> But patients with other malformation or heart block i.e either structural or electrical abnormality, require corrective surgery or pacing according to the type of defects.

#### Case Report:

We present a case of non-diabetic, hypertensive, non-smoker, male patient of 38 years who presented with complaints of aggravated effort intolerance, fatigue, palpitation and slow heart rate (between 40 to 50 beats/min) for 1 month. According to the patient he had been suffering from recurrent effort intolerance and fatigue during exertion since his childhood. But he gave no history of generalized bluish discoloration during exertion, fever, chest pain or syncope. His birth history was uneventful and all members of his family are healthy. In 2011 he was first diagnosed as a patient of Congenitally Corrected Transposition of great arteries with intermittent complete heart block. He was advised for permanent pacemaker implantation. But patient did not follow the advice. Recently patient again became symptomatic with same complaints in more aggravated form.

Investigations showed- normal hemogram (Hb-14.5 gm/dl, ESR- 02mm in first hour, normal total and differential count of WBC). Thyroid function was normal (h.TSH-2.12 micro IU/ml), serum creatinine - 0.9mg/dl and serum

electrolyte was normal (Na-141mmol/L, K- 3.5 mmol/L, Cl- 100mmol/L, TCO<sub>2</sub>- 28mmol/L). Patient was mildly dyslipidemic (total cholesterol – 195mg/dl, HDL-38 mg/dl, LDL-137mg/dl, TG- 113mg/dl).

Resting ECG showed bradycardia (HR 45 beats/min) with 2:1 AV block (Fig 1). 24 hours Holter ECG showed occasional first, second and intermittent complete heart block and significant number of extrasystole.

Echocardiography showed hypertrophied & dilated morphological RV on left side, from which aorta originated; small morphological LV on right side from which main pulmonary artery originated; dilated LA due to moderate TR, good biventricular systolic function & levocardia, situs solitus, but no VSD (Fig-2,3,4). Systemic ventricular EF was 62% (morphological RV).

Patient was implanted with a dual chamber permanent pacemaker. There was some difficulty during temporary pacemaker (TPM) insertion done before PPM implantation. As the right atrium connects to morphological LV through mitral valve, it was quite difficult to insert the TPM lead. The positioning of ventricular lead of the permanent pacemaker was also difficult, because it enters the morphological LV instead of the usual RV. However, a good result with stable efficient pacing was achieved by using a screw-in ventricular lead. (Fig-5) After implantation, patient quickly achieved relief of his symptoms. At follow up, he was completely asymptomatic and happy.



Fig.-1: Resting ECG showed 2:1 AV block



Fig-2: A4CH view- showed AV discordance



Fig-3: PSAX view- showed AO anterior to PA

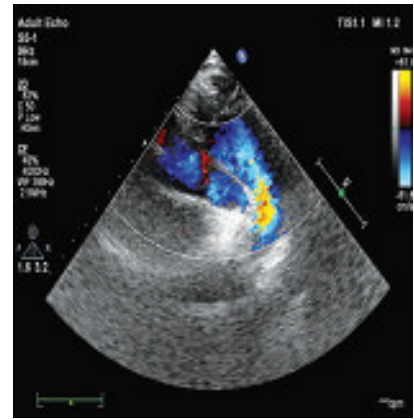


Fig-4: CMF shows two parallel flow indicating TGA.

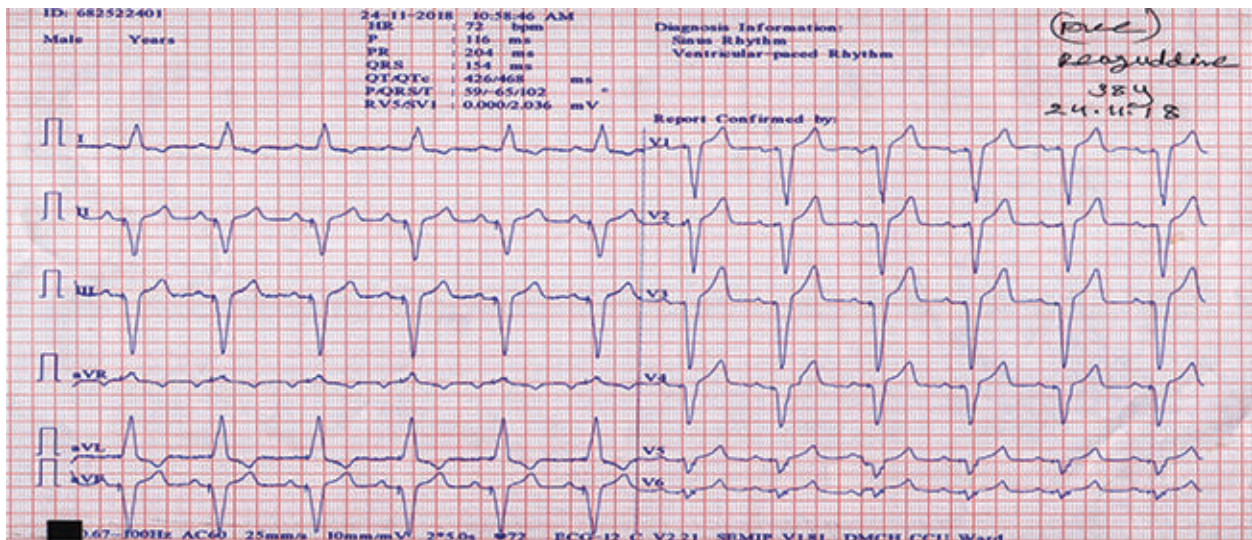


Fig.-5: Resting 12 lead ECG after permanent pacemaker implantation showing atrial tracking & ventricular pacing.

**Discussion:**

Corrected TGA is associated in approximately 70-80% cases with VSD and in 30-50% cases with pulmonary stenosis. Tricuspid valve abnormality is present in almost 90% cases.<sup>4,8,11-13</sup> Ebstein like anomaly of the tricuspid valve occur in 20-53% cases.<sup>12,13</sup> Conduction and coronary anomalies are also found. Prognosis depends on AV conduction, arrhythmia, structural anomalies, and degree of hemodynamic dysfunction. A multicenter study of congenitally corrected transposition of the great arteries demonstrated that 25% of patients without associated cardiac lesions and 67% of patients with other cardiac abnormalities developed congestive heart failure by the age of 45.<sup>7</sup> During embryological development, left handed looping of the heart tube results

in AV discordance and aorto-pulmonary septum fails to rotate 180° resulting in ventriculoarterial discordance. Blood however flows in effective sequence due to double inversion; hence it is called corrected TGA.

In c-TGA, usually the sinoatrial (SA) node is located in its normal position. The AV node is typically located along the anterosuperior margin of the VSD and is usually accompanied by an elongated His bundle and a second subsidiary AV node may exist in some cases.<sup>4,11,12</sup> With progression of age, complete heart block may ensue with progressive incidence of 2 percent per year.<sup>4</sup> With ventricular inversion, conduction bundles are also inverted and this makes electrical activation from right to left. This causes characteristic ECG findings of Q waves in the inferior leads and an absence of Q waves in the

left-sided precordial leads,<sup>13</sup> which may be misinterpreted as an inferior myocardial infarction.<sup>4</sup> Also reentrant tachyarrhythmias and Wolff-Parkinson-White syndrome may be noted.<sup>4</sup>

Without serious conduction defect there are two treatment options, one is periodic follow-up with echocardiography for worsening ventricular function or aggravation of TR, and other is corrective surgery.<sup>14</sup> With progressive conduction defect, patient would require permanent pacemaker implantation. During this procedure difficulty may occur as one has to pass the lead from RA to LV (instead of RV) through a bicuspid mitral valve (instead of tricuspid valve). We also faced this problem during pacing of the patient. ACE inhibitor or beta blocker has not been well studied in corrected TGA population. Therefore most of the patients received conventional systemic LV protection strategies.<sup>12</sup> This patient was prescribed ARB for the hypertension and ventricular dysfunction protection. In such cases, for periodic follow up, only echocardiography is not adequate as RV (which is acting as LV) has multiple coarse trabeculations. Therefore, cardiac magnetic resonance imaging is considered as an additional tool for measurement of ejection fraction in patients with corrected TGA.

#### Conclusion:

In congenitally corrected TGA, double discordance of AV and ventriculoarterial connections maintain normal circulation unless presence of other malformations complicates the condition. This case presented with effort intolerance despite normal systemic ventricular function (morphologic RV, EF-62%) due to second degree and intermittent complete AV block and those symptoms improved after implantation of permanent pacemaker. The prognosis of corrected TGA without associated cardiac anomalies depends on the appearance of anatomical tricuspid regurgitation and subsequent development of systemic (morphological right) ventricular dysfunction and advanced heart block. We plan to follow up this patient to detect any future systemic ventricular dysfunction by echocardiography and, if necessary, by cardiac MRI.

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