



## A Rare Case of Congenitally Corrected Transposition of Great Arteries (CCTGA)

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### ABSTRACT

*Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital heart defect associated with multiple cardiac morphological abnormalities and conduction defects. Data from the Baltimore-Washington Infant showed 40 infants per 1,00,000 live births are affected by CCTGA<sup>(1)</sup>; this is less than 1% of all congenital heart defects. The literature reports fewer than 1000 cases, however, the true prevalence of the malformation is not known. We report a case of a 37 year old female diagnosed with CCTGA.*

**Keywords:** Congenitally corrected transposition of the great arteries (CCTGA), congenital heart disease (CHD).

### INTRODUCTION

CCTGA is a rare congenital heart disease (CHD) with a prevalence of less than 1% of all CHD <sup>[1]</sup>. It is characterized by atrioventricular (AV) and ventriculoatrial (VA) discordance. The progressive risk of spontaneous complete AV block throughout life in patients with CCTGA is 2% per year <sup>[2]</sup>. The prognosis rests on clinical presentation, progression of disease and the effect of systemic pressure on the functional Ventricle. Patients undiagnosed until adulthood usually have no associated anomalies and present due to an abnormal chest radiograph or ECG. These patients are asymptomatic until right ventricular dysfunction, tricuspid regurgitation or complete heart block develops. No treatment is required for

patients with corrected transposition who have no other defects because their life expectancy has been reported to be near normal <sup>[3]</sup>.

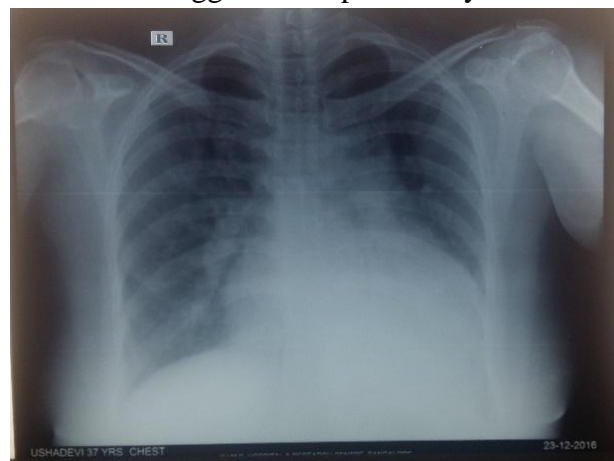
### CASE REPORT

A 37 year old female presented to us with complaints of easy fatigability and breathlessness on exertion since 15 days. Patient had no other comorbidities and was otherwise normal. She was pursuing her daily activity without any complaints. No significant past and family history. Obstetric history- P5L5, all deliveries were normal vaginal deliveries and were uneventful. On examination, vitals were unremarkable except for high blood pressure of 190/140 mmHg. General physical examination was normal with no

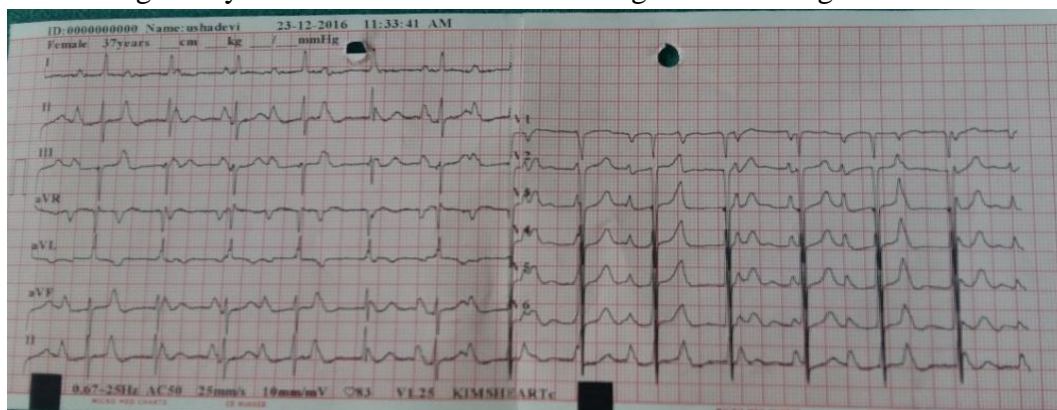
peripheral signs of congenital heart disease or no signs of infective endocarditis. Cardiovascular system examination revealed normal position of the heart and S1 and S2 heard in all areas and Pan-systolic murmur in Mitral Area (Grade 4, Radiating to the back) with ejection systolic murmur in pulmonary area with a loud P2 with Grade 2 left parasternal heave with palpable P2. Respiratory system and abdominal examination were unremarkable. Chest X-ray showed cardiomegaly with features suggestive of pulmonary oedema. ECG showed isorhythmic AV dissociation with Right atrial enlargement. Transthoracic echocardiography was done which showed CHD-Situs Solitus, Corrected Transposition of great vessels with morphological LV to right and morphological RV to the left and LVOT arising from morphological RV and RVOT arising from morphological LV with moderate left AV regurgitation, mild Right AV valve

regurgitation, Pulmonary hypertension (PASP-50 mmhg) with reduced left sided ventricular function (EF-40%). Patient was treated with anti-hypertensive and anti-failure measures. At discharge, patient's vitals were stable.

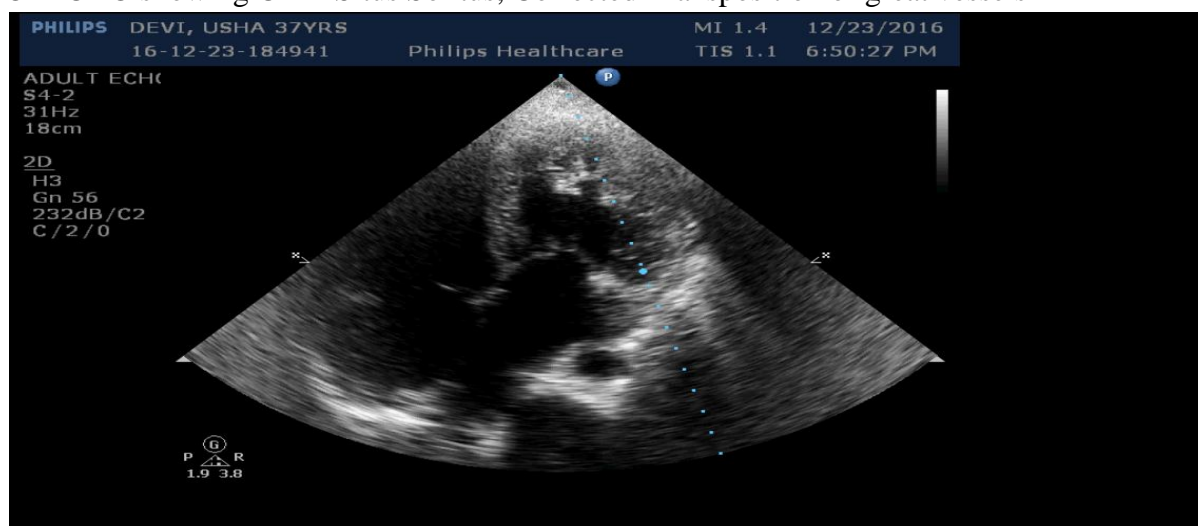
**Figure 1-** CHEST X-RAY showing cardiomegaly with features suggestive of pulmonary oedema

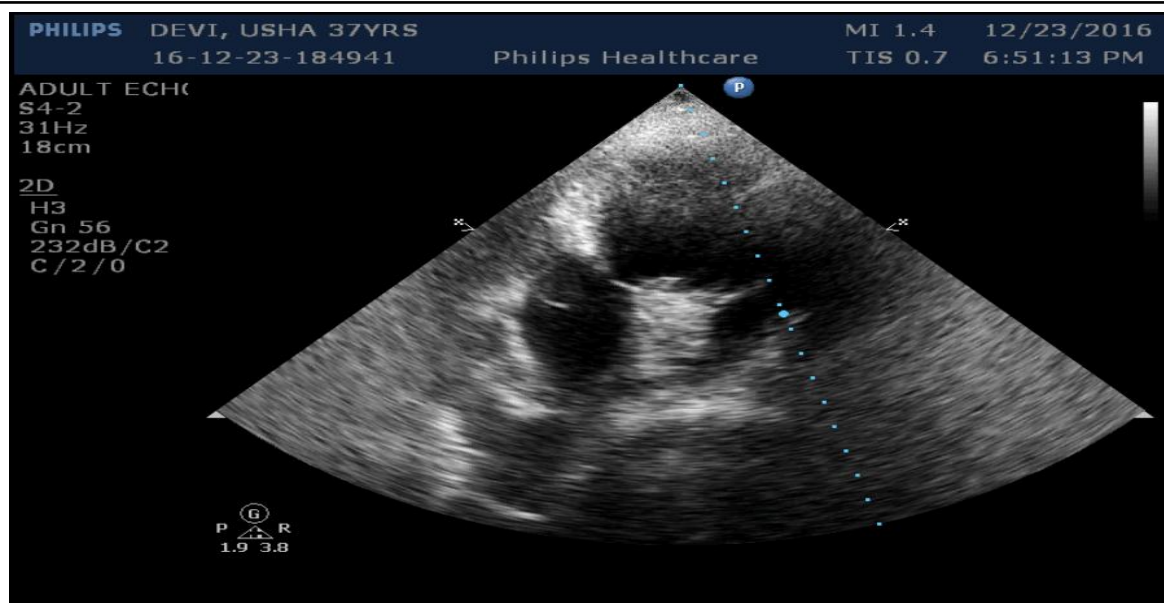


**Figure 2-** ECG showing isorhythmic AV dissociation with Right atrial enlargement.



**Figure 3-** ECHO showing CHD-Situs Solitus, Corrected Transposition of great vessels





## DISCUSSION

CCTGA, is a rare (less than 1% of all CHD) and complex heart defect<sup>[5]</sup>. It is characterized by AV and ventriculoatrial discordance. The aorta is located closer to the anterior and more to the left than the pulmonary artery. The AV valves follow their respective ventricles. Because of the displacement of the AV node and the abnormal course of conduction tissue, there is an increased risk of spontaneous complete AV block. CCTGA is commonly associated with other cardiac defects and its isolated occurrence is rare. Sudden Cardiac Death is still the leading cause of death in patients with CHD<sup>(4,6)</sup>

In conclusion, the highlights of this unique case are: CCTGA is a rare cardiac anomaly where many patients will remain asymptomatic for much of their lives. CCTGA patients have a reduced tolerance for exercise and have reported reduced quality of life compared to a control population. Prompt diagnosis and early recognition of signs of cardiac failure are essential in preventing mortality and morbidity in such cases.

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