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# Asymptomatic Isolated Congenitally Corrected Transposition of the Great Arteries in a 25-Year-Old Male: A Case Report

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**Abstract**

Congenitally corrected transposition of the great arteries (ccTGA) is a complex cardiac abnormality that represents less than 1% of all congenital heart defects. It is characterized by a unique pathophysiology involving both atrioventricular and ventriculoarterial discordance and may occur with or without cardiac abnormalities such as ventricular septal defects, pulmonary stenosis, or tricuspid valve anomalies. A man in his 20s presented with a 3-week history of mild dyspnea during strenuous activities. The patient was diagnosed with isolated ccTGA based on electrocardiogram and echocardiogram findings. However, as a long-term complication of ccTGA, the patient exhibited mild tricuspid regurgitation, mild mitral regurgitation, and right ventricular hypertrophy. The dyspnea was explained by the failing systemic right ventricle. Despite his cardiac anomaly, the patient leads a normal lifestyle, with ongoing monitoring to ensure optimal management of his condition. ccTGA is even rarer in the absence of additional cardiac abnormalities, and its diagnosis could be delayed due to being asymptomatic. Patients must avoid risk factors and that could potentially aggravate their condition. Regular surveillance is imperative for the early detection of potential complications.

**Keywords**

congenitally corrected transposition of the great arteries, isolated ccTGA, levo-transposition

## Introduction

Congenitally Corrected Transposition of the Great Arteries (ccTGA), first described by Von Rokitansky in 1875, is a complex cardiac abnormality that represents less than 1% of all congenital heart defects.[1,2] It is characterized by a unique pathophysiology involving both atrioventricular and ventriculoarterial discordance.[2] The deoxygenated blood flows through the superior/inferior vena cavae into the right atrium (RA). It then passes through the tricuspid valve into the morphological left ventricle (LV) before being pumped into the pulmonary artery. From there, it travels to the lungs where it undergoes oxygenation before returning via the pulmonary veins to the left atrium (LA) and moves through the mitral valve into the morphological right ventricle (RV), which pumps it to the aorta and then circulates back to the body as oxygenated blood.[1,3] Other names of ccTGA include L-looped transposition of the great arteries (L-TGA), double discordance, or ventricular inversion.[4] (Table 1) showcases a summary of ccTGA morphology.

Individuals with ccTGA may go undetected well into their adulthood, especially if they do not have additional cardiac anomalies.[1] The timely identification and effective management of ccTGA, along with its related structural heart issues, are essential for determining the outcomes and life expectancy of those affected.[5] Here, we discuss a case of a male patient in his mid-20s diagnosed with ccTGA, who enjoys a good, asymptomatic life without having undergone surgical or medical treatment, only under regular follow-ups.

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**Table 1.** Summary of ccTGA Morphology.

| Structure | Morphology in ccTGA |
|---|---|
| Atria | Normal position and morphology |
| AV connection | Discordant:<br>RA is connected to morphological LV<br>LA is connected to morphological RV |
| Ventricles | Morphological RV (systemic ventricle) is on the left<br>Morphological LV (sub pulmonary ventricle) is on the right |
| Great arteries | Discordant:<br>Aorta arises from the morphological RV<br>Pulmonary artery arises from the morphological LV |
| Coronary arteries | Inverted:<br>Right coronary supplies morphological LV<br>Light coronary supplies morphological RV |
| Conduction system | Abnormal:<br>AV node is often displaced and hypoplastic<br>Bundle of His may have an abnormal course |

Abbreviations: ccTGA, congenitally corrected transposition of the great arteries; RA, right atrium; LV, left ventricle; LA, left atrium; RV, right ventricle; AV, atrioventricular.

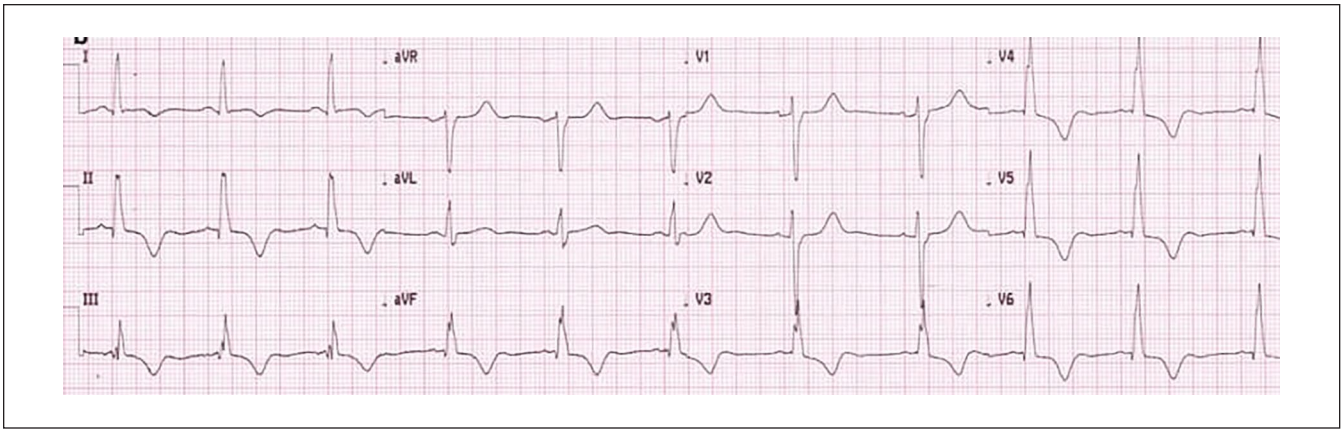


**Figure 1.** The ECG demonstrates an absence of Q waves in leads V5 and V6. ECG, electrocardiogram.

## Case Presentation

A 20-year-old Arab male with no significant medical history presented with a 3-week history of dyspnea when preforming strenuous activities. Clinical examination showed mild dyspnea, classified as New York Heart Association Class II; his vital signs were normal. He had not undergone any annual health check-up and was not on any medication. Hematologic, serological, and biochemical investigations were within normal limits. The electrocardiogram (ECG) displayed a sinus rhythm featuring Q waves in leads V1 and V2, and notably, Q waves were absent in leads V5 and V6 (Figure 1). There was no evidence of heart block or arrhythmias on the ECG. Further investigation with an echocardiogram (Figure 2) revealed normal situs solitus, the findings revealed that the RA was connected to a right-sided morphological LV, which was connected to the pulmonary artery. The LA was connected to a left-sided morphological RA, from which arose the aorta. This configuration resulted in atrioventricular and ventriculoarterial discordance, leading to a "double discordance." The aorta was anterior and to the left of the pulmonary artery (levo-transposition), with both great arteries running parallel to each other. No other heart anomalies were noted, and the patient was diagnosed with congenitally corrected transposition of the great arteries (ccTGA). Mild Tricuspid Regurgitation (TR), alongside mild mitral regurgitation and RV hypertrophy, were also identified on the echocardiogram (Figure 2). The dyspnea was explained by the failing systemic RV. The patient received counseling on adhering to a healthful lifestyle and engaging in mild physical activities. Regular echocardiographic monitoring was instituted at biennial intervals. The patient, upon follow-up over 5 years, has been leading a normal lifestyle, with ongoing monitoring in place to ensure optimal management of their condition.

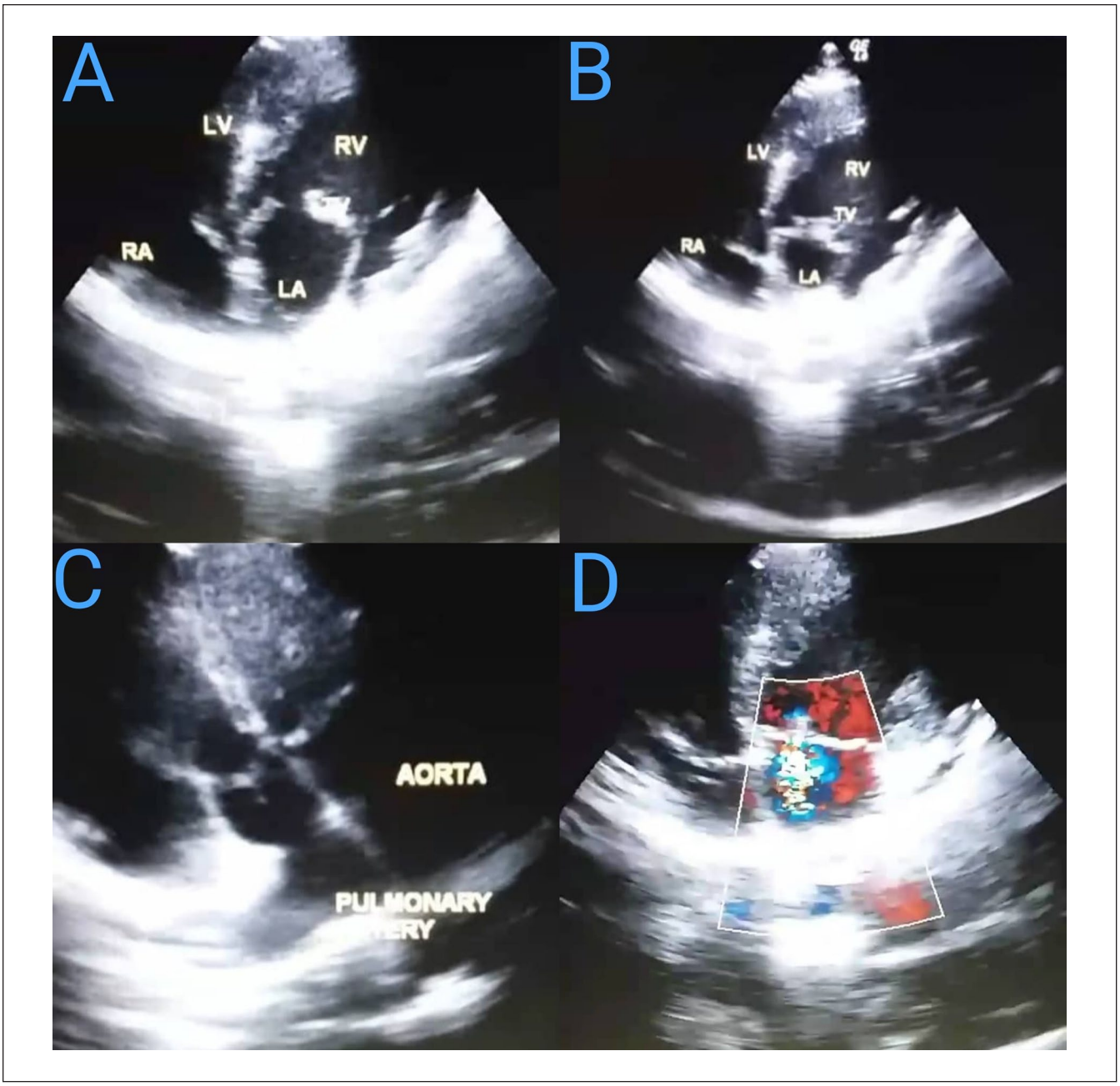


**Figure 2.** The echocardiogram (A and B) of the apical 4-chamber view reveals atrioventricular discordance with the tricuspid valve being slightly closer to the apex. (C) The parasternal long-axis view (Off-axis) reveals the great arteries in parallel orientation, with the aorta positioned anteriorly and the pulmonary arteries posteriorly. (D) The apical 4-chamber view reveals moderate tricuspid regurgitation. The echocardiogram reveals atrioventricular and ventriculoarterial discordances, along with moderate tricuspid regurgitation, with no other anomalies detected.

## Discussion

The anatomy of ccTGA can be considered "corrected" because there is a double switch in the heart's structure. The ventriculoarterial discordance, where the great arteries are connected to the wrong ventricles, is accompanied by atrioventricular discordance, where the atria and ventricles are also switched.[1,3] Despite the "corrected" anomaly, over 90% individuals with ccTGA have associated cardiac abnormalities such as ventricular septal defects, pulmonary stenosis, or tricuspid valve anomalies.[1] Normally, the RV is adapted for the lower pressures of the pulmonary circuit. However, when it functions as the systemic ventricle in ccTGA, it may become strained under the higher systemic pressures over time.[5] Symptoms related to the systemic right ventricular

dysfunction (sRVd) in ccTGA can vary widely among individuals. While some may remain asymptomatic for many years, others may exhibit signs of cyanosis, dyspnea, heart failure, arrhythmias, or heart murmurs caused by structural abnormalities or valve issues.[1,3]

Patients are usually diagnosed early in life; however, in the absence of other anomalies, the diagnosis may be delayed.[2] In adults, the symptoms are likely to be associated with sRVd,[1] as asymptomatic mild TR is commonly detected during echocardiography for other concerns.[6] Like in this case, the patient went undiagnosed until the age of 20, when he presented with mild dyspnea. His diagnosis was ultimately established based on characteristic findings observed in ECG and echocardiogram. Although cardiac MRI can offer more detailed imaging, echocardiogram continues to be the primary method for diagnosis.[1] Signs of heart block can be detected on ECG in certain patients,[2] but this was not observed in this patient. In specific cases, chest X-ray and cardiac catheterization may be useful, although they are not typically employed.[2]

Surgical intervention decisions are generally based on the physiological impacts of the accompanying defects,[7] and the type of abnormality requiring repair does not lead to significant differences in survival rates.[1] While medical therapy with beta-blockers, angiotensin-converting enzyme inhibitors, angiotensin receptor blockers, or aldosterone antagonists lacks robust evidence of efficacy in patients with isolated sRVD and heart failure,[5] reduction of afterload may provide benefit in patients with left ventricular dysfunction.[2] In the present case, neither surgical intervention nor medical therapy was considered necessary. Surgical intervention in asymptomatic patients with preserved RV function and tricuspid valve function is controversial; however, a failing systemic RV or increasing TR typically warrant surgical consideration.[2] However, the most effective surgical strategy for these patients remains largely undefined.[7]

Individuals with isolated ccTGA and associated lesions of minimal clinical significance may experience near-normal longevity due to the remarkable adaptability of the RV to systemic pressure.[2] Notably, there is a documented case of a 92-year-old patient with ccTGA, highlighting this adaptability.[8] These patients should receive ongoing care from a pediatric or adult cardiologist specializing in congenital heart disease, typically with biennial visits, and should be counseled on maintaining a healthy lifestyle, avoiding smoking, and encouraged to engage in regular moderate exercise, while strenuous or endurance exercise may be advised against.[1] Given the 2% annual risk of developing TR, patients should undergo ECG and echocardiogram at each visit and regular assessment of functional capacity through cardiopulmonary exercise testing.[1,9]

## Conclusion

In managing this patient's condition, the decision against surgical intervention or medical therapy underscores the significance of careful, individualized assessment in cases of asymptomatic patients with preserved right ventricular and tricuspid valve function. This case highlights the necessity for ongoing assessment and a good understanding of patient-specific factors in guiding treatment decisions and ensuring optimal patient outcomes.

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### Ethics Approval

Our institution does not require ethical approval for reporting individual cases or case series.

### Informed Consent

Written informed consent was obtained from the patient for their anonymized information to be published in this article.

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