

Classic Case

Congenital corrected transposition of the great arteries manifesting in adult: A classic case

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Abstract

Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital anomaly characterized by atrioventricular and ventriculoarterial discordance, in which the morphologic right ventricle supports systemic circulation, often leading to progressive dysfunction and heart failure in adulthood. We report the case of a 33-year-old man presenting with progressively worsening dyspnea. Chest radiography demonstrated cardiomegaly with dextrocardia and features of pulmonary interstitial edema. Multimodality imaging—including transthoracic echocardiography, coronary CT angiography, and cardiac MRI—was essential in confirming CCTGA and assessing secondary right ventricular failure. This case highlights the critical role of early recognition and comprehensive imaging in guiding management and optimizing outcomes in adult patients with this rare congenital condition.

Keywords: Congenitally Corrected Transposition of the Great Arteries (CCTGA), Dextrocardia, Heart Failure, Multimodality Cardiac Imaging, Systemic Right Ventricle.

Introduction

Transposition of the great arteries (TGA) is a congenital cardiac anomaly characterized by ventriculoarterial discordance, in which the aorta arises from the right ventricle (RV) and the pulmonary artery (PA) arises from the left ventricle (LV) [1].

TGA is classified into two types: complete or dextro-transposition (D-TGA) and congenitally corrected transposition (CCTGA) or levo-transposition (L-TGA). CCTGA is rare, accounting for less than 1% of all congenital heart defects [1]. It differs from D-TGA by the presence of both atrioventricular and ventriculoarterial discordance (Figure 1). Atrioventricular (AV) discordance refers to the connection of the right atrium to the left ventricle and the left atrium to the right ventricle. Despite physiologically normal circulation, the morphologic right ventricle sustains systemic output and is chronically exposed to systemic pressures, predisposing it to progressive dysfunction and eventual failure [1].

Here, we report a classic case of CCTGA diagnosed in adulthood, presenting with acute dyspnea and heart failure. Detailed evaluation included chest radiography, coronary CT angiography, and cardiac MRI.

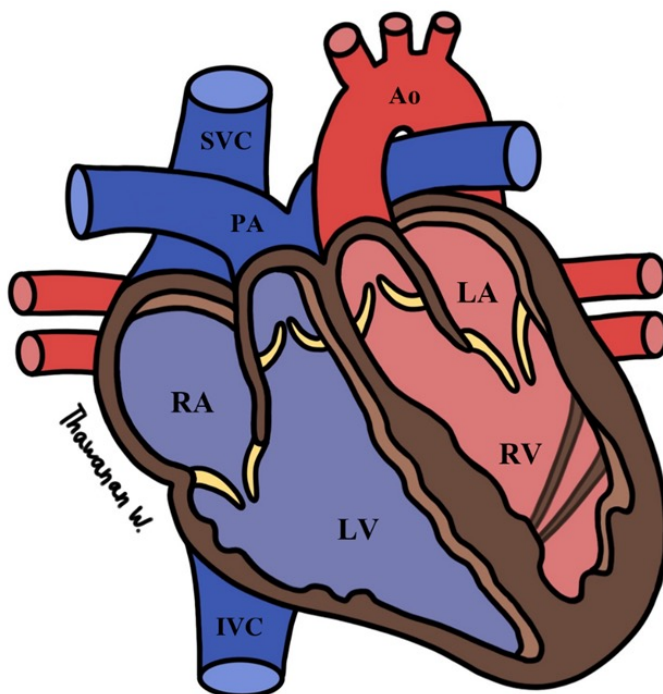


Figure 1. The illustration shows the anatomy of the heart and great arteries in congenitally corrected transposition of the great arteries (CCTGA). The pulmonary artery (PA) originates from the left ventricle (LV) and the aorta (Ao) originates from the right ventricle (RV). The right atrium (RA) connects to the LV and the left atrium (LA) connects to the RV. IVC = inferior vena cava.

Case summary

A 33-year-old man with no known underlying disease presented with progressive dyspnea and a change in functional class over the past month. Physical examination revealed fine crepitations in both lungs without cardiac murmurs. ECG showed sinus bradycardia with absent septal Q waves in the lateral leads, reversed R-wave progression, and widespread T-wave inversions (I, II, III, aVF, aVL, V2–6). Laboratory testing revealed markedly elevated NT-proBNP (9,267 pg/mL). Chest radiography demonstrated cardiomegaly with dextrocardia, bulging of the right cardiac border, prominent hila, and mild blunting of both costophrenic angles (Figure 2). Although these findings are nonspecific, the combination of dextrocardia and cardiomegaly in an adult with heart failure should raise suspicion for an underlying congenital cardiac anomaly and prompt further cardiac evaluation.

Transthoracic echocardiography demonstrated situs solitus with dextrocardia, atrio-ventricular (AV) and ventriculoarterial (VA) discordance, and a dilated systemic right ventricle (RV) with an ejection fraction of 30%, confirming acute heart failure secondary to CCTGA. The patient was admitted for intravenous diuretics and further imaging evaluation.

Coronary CT angiography and cardiac MRI confirmed dextrocardia with L-transposition of the great arteries (L-TGA). The aorta originated from the morphologic RV and the pulmonary artery from the morphologic LV (VA discordance). The right atrium drained into the morphologic LV and the left atrium into the morphologic RV (AV discordance). There was no left main coronary artery; the LAD and LCX arose separately from the right-facing aortic cusp, while the RCA originated from the left-facing cusp (Figure 3). Cardiac MRI further demonstrated severe RV dilatation with moderate systolic dysfunction and concentric RV hypertrophy, consistent with RV cardiomyopathy secondary to congenital heart disease (Figure 4). Following intravenous diuretics, the patient's symptoms improved, and he was discharged on oral diuretics. On follow-up, his clinical status improved, but cardiac function showed no recovery. He is currently on the waiting list for cardiac transplantation.

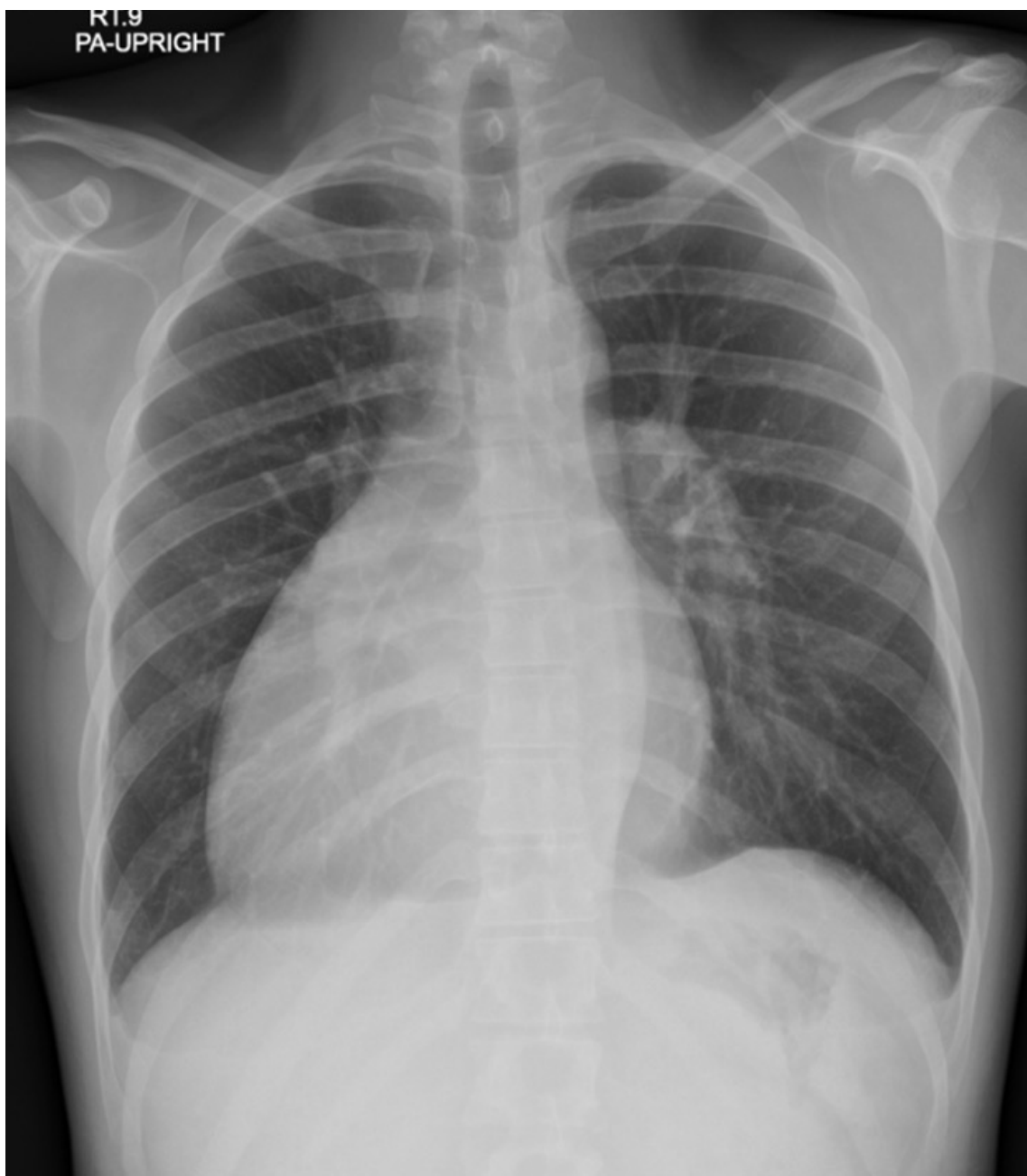


Figure 2. *PA upright chest radiograph demonstrates cardiomegaly with dextrocardia, bulging of the right cardiac border, prominent hila, and mild blunting of both costophrenic angles.*

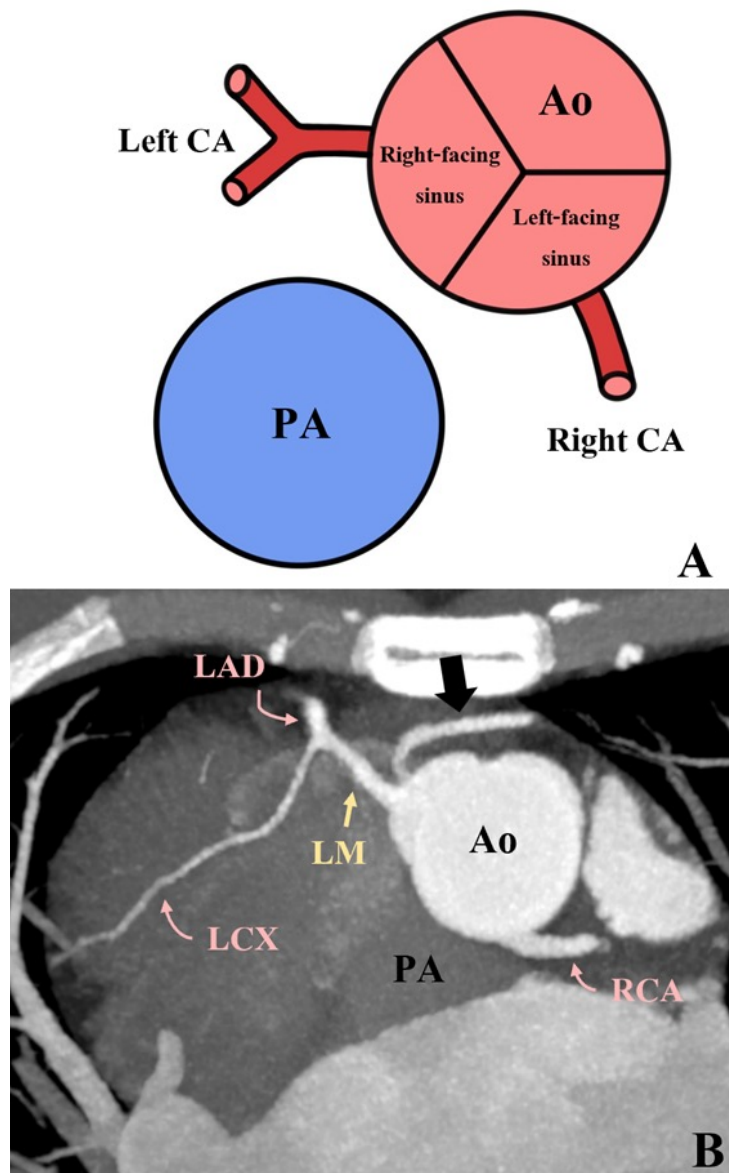


Figure 3. Coronary anatomy of the patient with CCTGA. (A) Illustration of the anatomy of the coronary arteries in CCTGA. CA = coronary artery. (B) MIP cardiac CT image at the level of the aortic sinus of Valsalva, the left anterior descending artery (LAD) and the left circumflex artery (LCX) originate from the left main coronary artery (LM) which arises from the right-facing sinus. The LAD courses anteriorly in the interventricular groove and the LCX courses in the right atrioventricular groove. The right coronary artery (RCA) originates from the left-facing sinus and travels along the left atrioventricular groove. In this patient, another coronary artery branch (black straight arrow) arises directly from the right-facing sinus and supplies the free wall of the right ventricle.

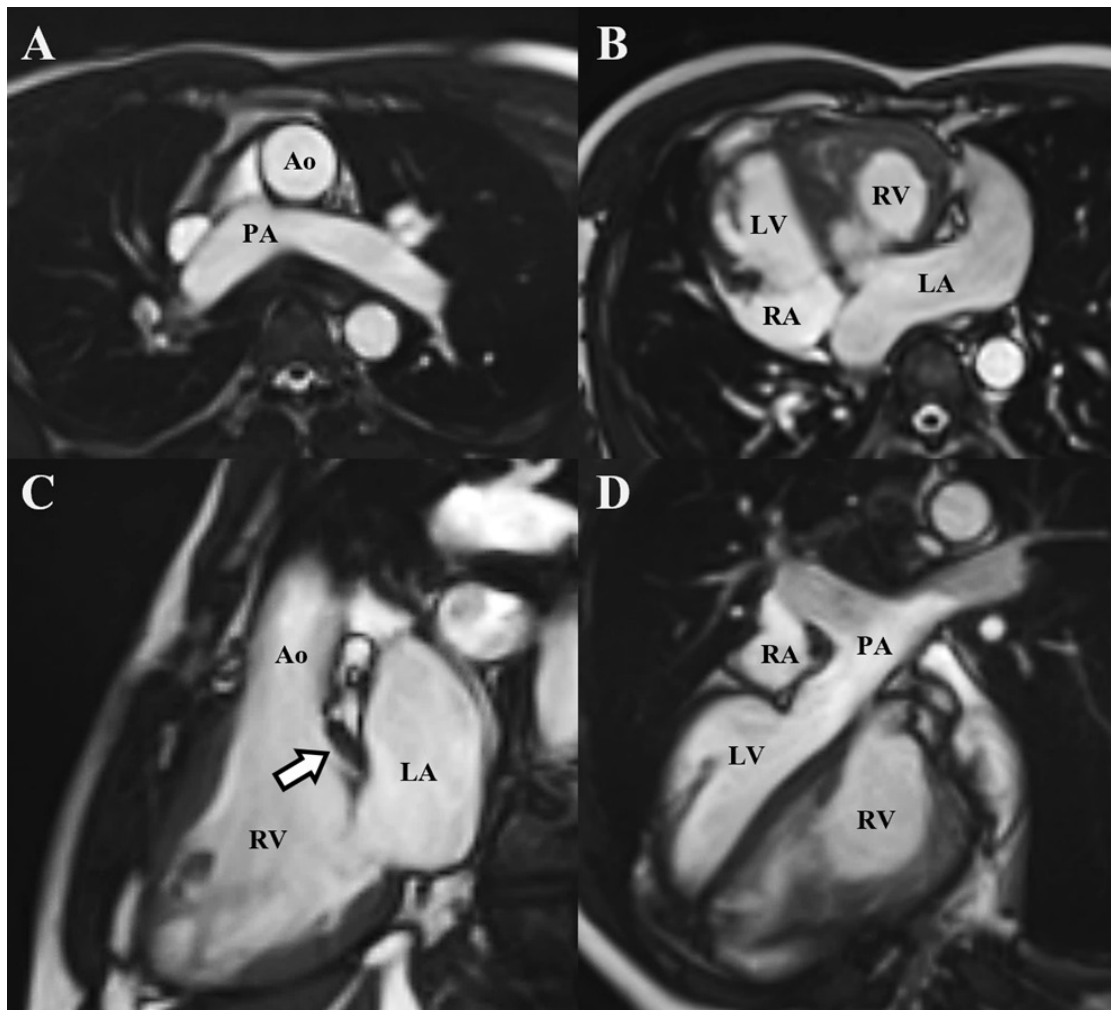


Figure 4. Cardiac MRI of the patient with CCTGA. (A) Axial SSFP MR image, anterior and leftward positioning of the aorta (Ao) relative to the main pulmonary artery (PA) is observed. (B) Axial SSFP MR image illustrates the right atrium (RA) connected to the left ventricle (LV) and the left atrium (LA) connected to the right ventricle (RV). The morphologic LA demonstrates finger-like appearance of the LA appendage. (C) Sagittal SSFP MR image illustrates a muscular band (arrow) between the atrio-ventricular and semilunar valves of the left-sided ventricle, representing the morphologic RV (D) SSFP MRI in the four-chamber view illustrates that the morphologic LV connects to the PA. Coarse inter-ventricular septum of the morphologic RV is also noted.

Discussion

Congenitally corrected transposition of the great arteries (CCTGA) was first described by the Australian pathologist Von Rokitsansky in 1875 [2]. The condition results from abnormal embryonic looping of the primary heart tube which undergoes a left-sided loop (L-loop) instead of the normal right-sided loop (D-loop). This abnormal looping produces atrioventricular discordance, in which each atrium connects to a morphologically mismatched ventricle. In addition, abnormal torsion of the aortopulmonary septum leads to ventriculoarterial discordance (transposition of the great vessels). As a result, the right atrium, mitral valve, and morphologic left ventricle receive deoxygenated systemic venous blood and connect to the posteriorly located pulmonary artery. Conversely, the left atrium, tricuspid valve, and morphologic right ventricle receive oxygenated pulmonary venous blood and connect to the aorta, which is positioned anterior and leftward to the pulmonary artery.

CCTGA can occur in either situs solitus or situs inversus, depending on the viscerotrial arrangement [3]. Cardiac position is usually levocardia or mesocardia, while dextrocardia is seen in approximately 20% of cases, including our patient. Coronary anatomy may also follow a mirror-image pattern [1]. In our patient, the left coronary artery arose from the right-facing sinus and coursed through the right atrioventricular groove posterior to the mitral valve. The right coronary artery originated from the left-facing sinus, ran between the left atrium and right ventricle, and supplied only the systemic right ventricle.

Most patients with CCTGA have associated congenital heart defects that influence their clinical presentation and long-term prognosis. Ventricular septal defect is present in up to 80% of cases, while other common anomalies include pulmonary stenosis, left ventricular outflow tract obstruction (LVOTO), Ebstein anomaly, situs inversus, and dextrocardia [4]. Isolated CCTGA, which accounts for only 1–2% of cases, is usually asymptomatic in childhood due to physiologically corrected circulation without mixing or shunting of blood [5].

Despite this apparent compensation, the morphologic right ventricle is not structurally suited to support the systemic circulation in the long run. Chronic exposure to high afterload and systemic pressures induces myocardial remodeling, initially causing right ventricular hypertrophy and progressing to dilatation. Pathological hypertrophy, combined with microcirculatory impairment, results in inadequate perfusion of the hypertrophied systemic right ventricle—particularly because it is supplied solely by the right coronary artery. Over time, this leads to progressive right ventricular dysfunction and

eventual heart failure. In adults with CCTGA, heart failure occurs in approximately 37% of cases, typically presenting with dyspnea, orthopnea, paroxysmal nocturnal dyspnea, peripheral edema, and jugular venous distension [2]. Elevated N-terminal pro-B-type natriuretic peptide (NT-proBNP) levels may aid diagnosis, as observed in our patient.

Imaging plays an important role in both diagnosis and management. The choice of modality depends on clinical context and resource availability. Transthoracic echocardiography is usually the first-line investigation, providing information on cardiac morphology, ventricular and valvular function, and hemodynamics. For more detailed evaluation, coronary CT angiography (CTA) and cardiac MRI play complementary roles in pre- and postoperative assessment, offering high-resolution visualization of cardiovascular anatomy, coronary artery origins, ventricular function, and myocardial morphology [6].

In our patient, coronary CTA and cardiac MRI confirmed both atrioventricular and ventriculoarterial discordance, establishing the diagnosis of CCTGA. Additional findings included severe right ventricular dilatation, moderately impaired systolic function, and severe concentric hypertrophy, consistent with right ventricular cardiomyopathy secondary to systemic pressure loading. Recognition of morphologic ventricular characteristics was crucial for diagnosis: the morphologic right ventricle was identified by the presence of a moderator band, coarse trabeculation, a muscular infundibulum between the atrioventricular and arterial valves, and papillary muscle attachment to both the septum and free wall [1]. In contrast, the morphologic left ventricle showed fine trabeculation, smooth septal contour, fibrous continuity between the atrioventricular and arterial valves, and absence of a moderator band. The aorta was located anterior and leftward of the pulmonary artery, arising from the morphologic right ventricle, confirming the classic anatomy of CCTGA.

Management strategies depend on associated lesions and clinical presentation. In symptomatic patients with heart failure due to right ventricular dysfunction, initial treatment follows the approach used for morphologic left ventricular failure, with medical therapy initiated when EF is less than 40% [2]. In our patient, diuretics were prescribed as first-line therapy. Surgical approaches include either conventional repair or anatomical correction. The conventional approach addresses associated defects without correcting the discordance, leaving the right ventricle as the systemic ventricle. Anatomical repair, by contrast, can be achieved using a double switch operation or a Senning–Rastelli procedure [1]. Ultimately, cardiac transplantation may be required, particularly in terminal heart failure, as in our patient.

Conclusion

Congenitally corrected transposition of the great arteries (CCTGA) is a rare congenital cardiac anomaly that may remain undetected until adulthood, when patients often present with systemic right ventricular failure. Multimodality imaging, including echocardiography, coronary CT angiography, and cardiac MRI, plays a crucial role in diagnosis, delineation of anatomic relationships, and assessment of ventricular function. Our case highlights the value of advanced imaging in confirming CCTGA in adults and in guiding clinical management, including the need for timely referral for transplantation in cases of progressive systemic RV dysfunction.

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