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Case Report

# Congenitally Corrected Transposition of the Great Arteries at Age 88 Years

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## ABSTRACT

**Most patients with congenital heart disease are diagnosed shortly after birth; however, some patients are overlooked and diagnosed in adulthood. We present the case of a man in whom congenitally corrected transposition of the great arteries (CCTGA) was first diagnosed at the age of 88 years. CCTGA is a rare disease; patients without frequently associated cardiac anomalies are considered to have long life expectancies; however, their prognosis remains unclear. This case is the oldest patient at the time of CCTGA diagnosis in the literature, which may provide new insights for CCTGA without frequently associated cardiac anomalies.**

## RÉSUMÉ

**La plupart des cardiopathies congénitales sont diagnostiquées peu après la naissance; certains cas passent toutefois inaperçus et sont diagnostiqués une fois le patient arrivé à l'âge adulte. Nous présentons le cas d'un homme chez qui une transposition congénitalement corrigée des gros vaisseaux (TCCGV) a été diagnostiquée alors qu'il avait 88 ans. La TCCGV est une maladie rare; les patients qui ne présentent pas aussi l'une ou l'autre des anomalies cardiaques qui y sont fréquemment associées ont généralement une bonne espérance de vie, bien que le pronostic demeure obscur. De tous les cas rapportés dans la littérature médicale, le patient qui est présenté ici est le plus âgé qui ait jamais reçu un diagnostic de TCCGV, ce qui pourrait éclairer sous un jour nouveau la TCCGV non accompagnée des autres anomalies cardiaques qui y sont fréquemment associées.**

Most patients with congenital heart disease are diagnosed shortly after birth. Even if overlooked during the perinatal period, patients usually develop a range of symptoms at younger ages. However, a small number of patients remain free of symptoms and undiagnosed for years or even decades. We describe a case of congenitally corrected transposition of the great arteries (CCTGA) that was first diagnosed at a very advanced age.

## Case

An 88-year-old man presented with dyspnea on exertion. His blood pressure was 176/64 mm Hg, pulse rate 53 beats per minute, and oxygen saturation value 90% on room air. Physical examination revealed coarse crackles in bilateral lung fields and pitting edema in both legs. Electrocardiography showed bradycardia due to third-degree atrioventricular block, inversion of all complexes in lead I, marked right axis deviation, reverse R-wave progression in precordial leads, and Q-wave in leads I, aVL, and V6 (Fig. 1). Transthoracic echocardiography showed systemic ventricular dysfunction (ejection fraction of 27%) with mild systemic atrioventricular valvular regurgitation. He was known to have dextrocardia, which was diagnosed on the basis of chest radiography when he visited a clinic for ill health in his thirties. Additionally, he had a past history of appendicitis surgery, which led to the detection of heterotaxy. He had not been hospitalized previously for any cardiovascular disease.

After transvenous temporary pacing catheter placement and the administration of diuretics, his condition improved; however, his bradycardia persisted, and we decided transvenous permanent pacemaker implantation was required. We performed chest computed tomography to make an operative plan. The 3 major findings were as follows: First, the aorta arose from the anatomic right ventricle with the moderator band and the subaortic infundibulum; second, the anatomic right ventricle was connected to the left atrium; and third, the

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See page 727 for disclosure information.

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**Novel Teaching Points**

- This case suggests that the prognosis for CCTGA patients without frequently associated cardiac anomalies is potentially as good as that for healthy individuals.
- There is a possibility of undiagnosed congenital heart disease even in elderly patients without a history of adult congenital heart disease.

right atrium, which was connected to the superior and inferior vena cava, was located on the left side with the presence of situs inversus (Fig. 2, A-F). These findings indicated atrioventricular and ventriculoarterial discordance with atrial situs inversus. Other than a small atrial septal defect, we did not find any other cardiac anomalies, including ventricular septum defects and pulmonary stenosis. He was therefore diagnosed with CCTGA. He underwent transvenous permanent pacemaker implantation (Fig. 2G) and has remained free of symptoms since the operation.

## Discussion

CCTGA is a rare disease that accounts for less than 1% of congenital heart diseases. CCTGA without other frequently associated cardiac anomalies sometimes remains undiagnosed for decades because symptoms are often minimal and nonspecific, and many asymptomatic patients are first given a diagnosis of CCTGA in adulthood.[1] CCTGA patients often present with anatomic tricuspid regurgitation and advanced atrioventricular block at a younger age, which lead to the development of chronic heart failure and morphologic right ventricular dysfunction with increasing age. Graham et al. reported that by age 45 years, chronic heart failure was present in 67% of CCTGA patients with frequently associated cardiac anomalies, but in only 25% of those without.[2] Although the prognosis of CCTGA without other frequently associated cardiac anomalies remains unclear, some case reports suggest that the potential life expectancy is as long as that of healthy individuals.[3-5] In our case, the patient remained asymptomatic until age 88 years, and he seems to be the oldest patient at the time of CCTGA diagnosis in the literature. CCTGA patients with situs inversus are considered to have a better prognosis than those with situs solitus,[6] and this might be another reason why his manifestations of CCTGA occurred very late in life.

## Conclusions

This case report describes the case of a man with CCTGA that went undiagnosed until he was 88 years of age. This case may provide new insights for the natural history of CCTGA without frequently associated cardiac anomalies.

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## Disclosures

The authors have no conflicts of interest to disclose.

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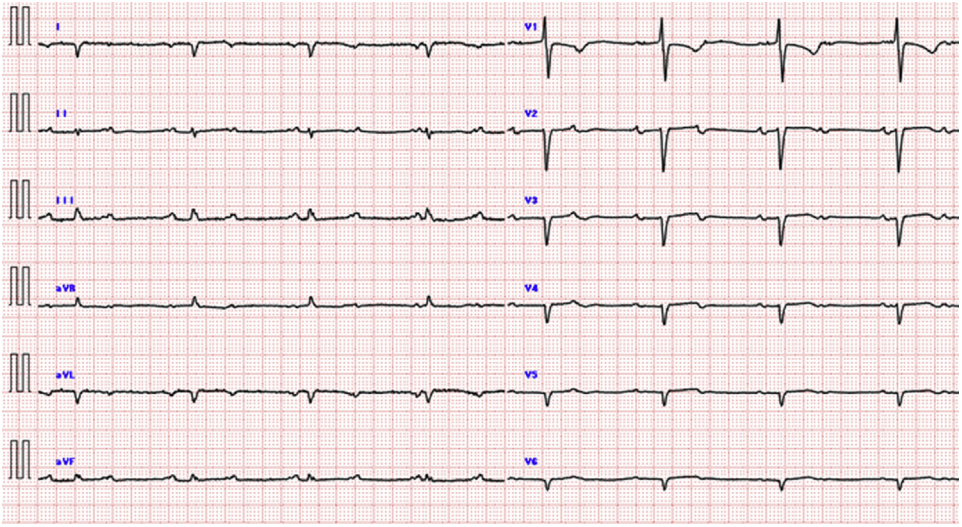

**Figure 1.** An electrocardiogram showed bradycardia of 48 beats per minute due to third-degree atrioventricular block, inversion of all complexes in lead I, marked right axis deviation with 152 degree, reverse R-wave progression in precordial leads, and Q-wave in leads I, aVL, and V6.

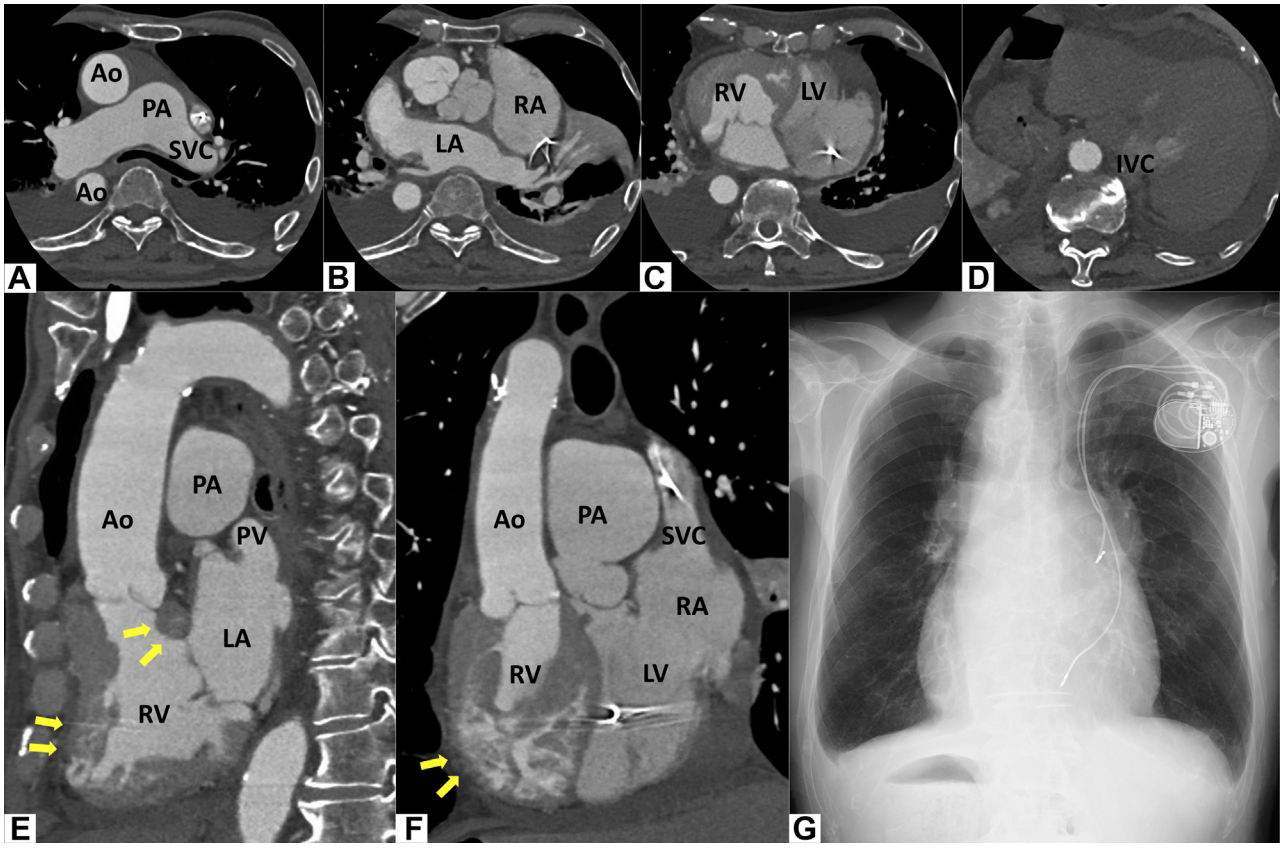


**Figure 2.** (**A-F**) Chest computed tomography images. The aorta arose from the anatomic right ventricle with the moderator band and the subaortic infundibulum (**arrows**). The anatomic right ventricle was connected to the left atrium. The right atrium, which was connected to the superior and inferior vena cava, was located on the left side with the presence of situs inversus. (**G**) A chest radiograph image after transvenous permanent pacemaker implantation. Ao, aorta; IVC, inferior vena cava; LA, left atrium; LV, anatomic left ventricle; PA, pulmonary artery; PV, pulmonary vein; RA, right atrium; RV, anatomic right ventricle; SVC, superior vena cava,.