# Severe Symptomatic Aortic Stenosis in an Octogenarian with Congenitally Corrected Transposition of the Great Arteries



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## INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA), also known as L-transposition of the great arteries, is rare, accounting for 0.5% to 1.4% of congenital heart disease.[1] Patients are frequently asymptomatic at birth and present in the first few decades of life. This defect is characterized by the presence of discordant connections at the atrioventricular (AV) and ventriculoarterial junctions.[2] This variation of AV discordance is most commonly of the S,L,L type by which there is a ventricular L-loop and L-malposition of the aorta. Morphologically, the left ventricle (LV) lies to the right side and the right ventricle (RV) lies to the left side. The right atrium is connected to the right-sided morphological LV, while the left atrium aligns with the left-sided morphological RV.[3] Anatomically, the morphologic, subpulmonary LV lies anterior and rightward to the morphologic, subaortic RV. The clinical presentation of ccTGA is variable and depends on the presence and severity of associated intracardiac lesions.[4] Congenitally corrected TGA may manifest as exercise intolerance during early childhood or growth failure when associated with ventricular septal defects and valvular abnormalities such as pulmonic stenosis or regurgitation from an Ebstein-like anomaly of the systemic AV valve. The initial presentation may be secondary to complete heart block. Patients with morphologic abnormalities of the tricuspid valve (TV) are at risk of developing severe tricuspid regurgitation and can rapidly deteriorate into RV failure. The survival rate of patients with ccTGA appears to be strongly influenced by tricuspid regurgitation severity.[5] There have been reports of asymptomatic patients with ccTGA found on autopsy, and only a few patients have been described in the literature that are older than 70 years of age and remain asymptomatic.[6] We describe an octogenarian patient with ccTGA and severe stenosis of the aortic valve requiring replacement.

## CASE PRESENTATION

An 84-year-old man without any major medical problems was referred from an outside institution for increasing dyspnea on exertion and intermittent chest pressure. The patient had an oxygen saturation of 96% and blood pressure of 90/56 mm Hg in both arms; the physical examination revealed a 4/6 systolic ejection murmur over the precordium. Electrocardiogram is shown in Figure 1. Investigations performed at the referring institution are presented in Figure 2.

We performed a transthoracic echocardiogram (TTE) that suggested the diagnosis of ccTGA. The subcostal view demonstrated normal visceral and atrial situs (Video 1). The systemic ventricle was dilated with decreased systolic function (LV ejection fraction 30%; Video 2, Figure 3). There was no evidence of an atrial septal defect, ventricular septal defect, or pulmonic stenosis. There was moderate systemic AV valve regurgitation. The aortic valve leaflets were heavily calcified with a peak gradient of 60 mm Hg and a mean gradient of 40 mm Hg (Figure 4, Video 3), and there was mild aortic regurgitation (Video 4). For transcatheter aortic valve implantation (TAVI) planning and diagnosis of the anatomy, cardiac computed tomography was preferred over cardiovascular magnetic resonance imaging and confirmed the diagnosis of ccTGA (Figure 5).

Given that the patient had an excellent prior functional status and that the patient's quality of life had significantly declined after developing symptoms, a shared decision was made to intervene on the aortic valve. A multidisciplinary team discussion was held regarding management options, and TAVI was initially considered. In contrast to the normal LV outflow tract, the team perceived that the muscle bundle separating the systemic outflow tract (morphologic RV outflow tract) from the systemic AV valve (morphologic TV) did not provide an adequate anchor for the subvalvular portion of the transcatheter valve. The risks of TAVI outweighed the benefits; therefore, surgical aortic valve replacement (SAVR) was recommended.

During SAVR, the aorta was observed to be located anterior to the pulmonary artery and arising from the morphologic RV (Figure 6).

The aortic valve was tricuspid and heavily calcified. Excision of the valve leaflets and decalcification of the aortic annulus was performed, making sure to avoid involvement of the subvalvular apparatus. The patient underwent an uncomplicated SAVR using a 23 mm third-generation pericardial aortic bioprosthesis. The postoperative course was

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## VIDEO HIGHLIGHTS

**Video 1:** Two-dimensional TTE, subcostal view, demonstrates cardiac apex pointing to the left, depicting situs solitus with levocardia. A small pericardial effusion is seen.

**Video 2:** Two-dimensional TTE, apical 4-chamber view, demonstrates AV discordance. The systemic trileaflet (tricuspid) valve is determined by the more apical insertion of the left AV valve compared to the right AV valve at the cardiac crux. The systemic RV is dilated and dysfunctional, and the RV wall is hypertrophied and trabeculated.

**Video 3:** Two-dimensional TTE, apical long-axis view, demonstrates the dilated morphologic RV (functional LV) connected to the aorta and the severely calcified, restricted aortic valve.

**Video 4:** Two-dimensional TTE, apical long-axis view with color-flow Doppler, zoomed in at the aortic valve, demonstrates a mild eccentric aortic regurgitation jet.

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complicated by heart block requiring permanent pacemaker placement. The patient was discharged on postoperative day 10.

## DISCUSSION

Sequential segmental analysis is used routinely in the assessment of cardiac morphology in children and adults with congenital heart disease. The de novo diagnosis of complex cardiac morphological abnormalities in adults is uncommon, and therefore, the parasternal view is often a standard starting point. One must consider visualizing the heart from the subcostal and suprasternal views to assess the visceral situs, atrial situs, AV connections, and interrelation of great arteries when undiagnosed adult congenital heart disease is suspected during the TTE.

There are several characteristics that differentiate the morphologic RV from the morphologic LV. The presence of a moderator band, septal attachment of the AV valve with chordal attachment to the interventricular septum (septophilic), and the more apical position of the AV valve are the most reliable characteristics of the morphologic RV. This was missed on the initial TTE of our patient.

The significance of this case is manifold. First, it reinforces the concept that patients with systemic RV can do well clinically for decades in the setting of a competent systemic TV.[7] This patient only became symptomatic in the eighth decade due to the increased afterload of severe aortic stenosis (AS). Interestingly, the severity of AS needed to cause symptoms was similar to what is observed in patients with a systemic LV. A case report of a patient with bicuspid aortic valve who developed AS in the fifth decade of life is consistent with this observation.[8] Second, consideration should be given to epicardial systemic lead placement in all adult patients with ccTGA undergoing open heart surgery. The risk of complete heart block is high, and endovascular systemic lead placement is known to present technical challenges.[9] Unfortunately, our patient did not have this done. Therefore, should the patient meet the criteria for biventricular pacing in the future, we will need to attempt an endovascular lead placement. Third, as adult congenital heart disease patients live longer, we will encounter more congenital heart patients that require treatment for acquired heart disease.[10] Clinical trials and guidelines typically do not include these patients. Optimal care will require collaboration between structural interventional cardiologists, adult cardiac surgeons, congenital cardiac surgeons, imaging cardiologists, and adult

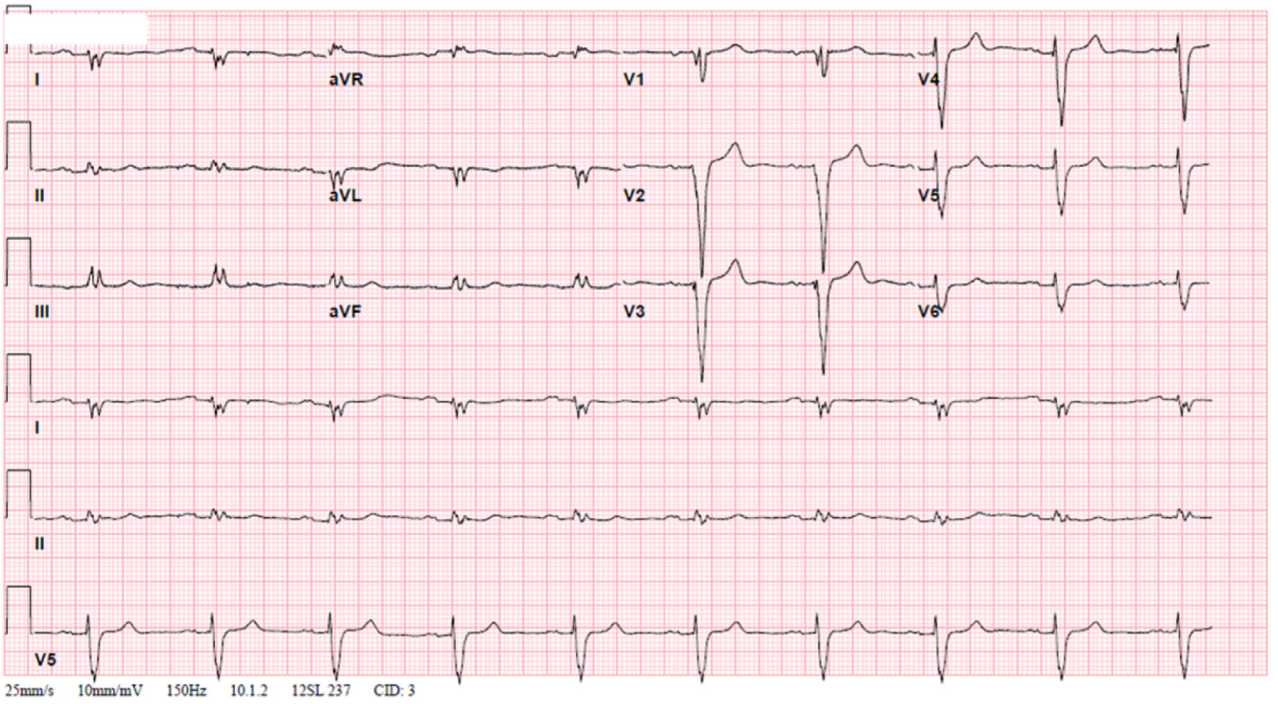


Figure 1 Electrocardiogram demonstrates sinus bradycardia (ventricular rate 58 bpm) with first-degree AV block (PR interval 226 ms), right-axis deviation (135°), and nonspecific intraventricular conduction delay (QRS duration, 162 ms).

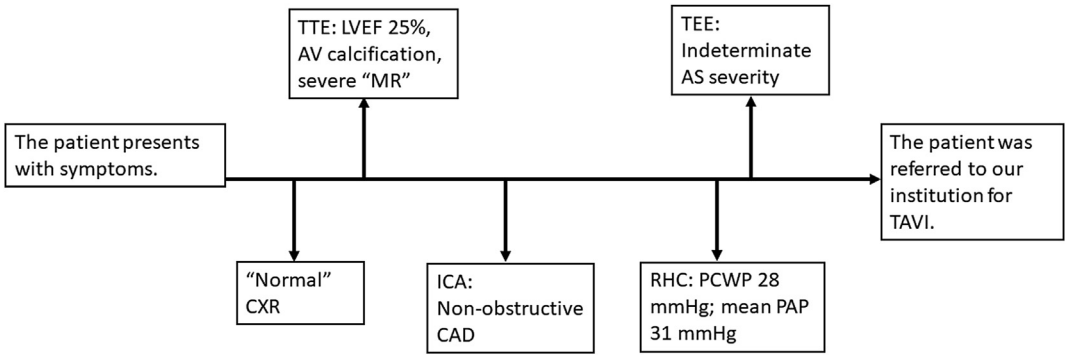


**Figure 2** Investigations reported at the referring institution. *AV*, Aortic valve; *CXR*, chest x-ray; *ICA*, invasive coronary angiography; *MR*, mitral regurgitation; *PA*, pulmonary artery; *PCWP*, pulmonary capillary wedge pressure; *RHC*, right heart catheterization; *TEE*, transesophageal echocardiogram.

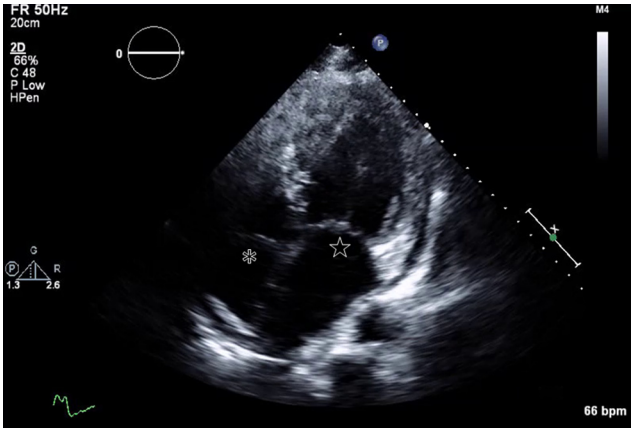


**Figure 3** Two-dimensional TTE, apical 4-chamber view, systolic phase, demonstrates AV discordance, apically displaced left AV valve (star symbol) compared to the right AV valve (asterisk symbol), and small pericardial effusion. Suboptimal image quality is due to poor acoustic transmission.

congenital heart disease experts, as in this case. Fourth, management of AS in patients with ccTGA presents unique challenges. Given that the morphologic RV forms the ventriculoaortic junction, there are inherent risks with aortic valve replacement in these patients. In patients with normal anatomy, the right coronary leaflet rests on the muscular part of the interventricular septum, the noncoronary leaflet is adjacent to the membranous septum and the anterior mitral leaflet, and the left coronary leaflet is continuous with the anterior mitral leaflet (aortomitral curtain) and muscular interventricular septum.[11] In patients with ccTGA, the fibrous continuity for the leaflets of the mitral, aortic, and tricuspid valves is not present. Instead, there is fibrous continuity between the leaflets of the pulmonary and mitral valves in the roof of the right-sided morphologically LV.[2] This makes the anatomy of the outflow tract unsuitable for transcatheter-based approaches. We hypothesize that this is also why if care is not taken to avoid subaortic structures during decalcification of the aortic annulus, it may be associated with worse procedural outcomes. Surgical literature on replacement of aortic valve in patients with ccTGA is limited. As catheter-based approaches improve and indications expand, valvular pathologies in adults with congenital heart disease that are currently strictly managed with surgery will potentially be managed percutaneously. However, further research is warranted before utilizing catheter-based approaches for severe AS in patients with ccTGA.

## CONCLUSION

Adults with ccTGA are frequently diagnosed later in life, and it is not uncommon for this condition to go unrecognized during their initial cardiology evaluation.[12] Congenitally corrected TGA can present with symptomatic AS, which can be challenging to diagnose and treat. Multimodality imaging is essential for the diagnosis. Surgical aortic valve replacement remains the mainstay of treatment given the anatomy of the outflow tract of the morphological RV.

## DATA AVAILABILITY

Data supporting this research article is available from the corresponding author on reasonable request.

## ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

## CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

## FUNDING STATEMENT

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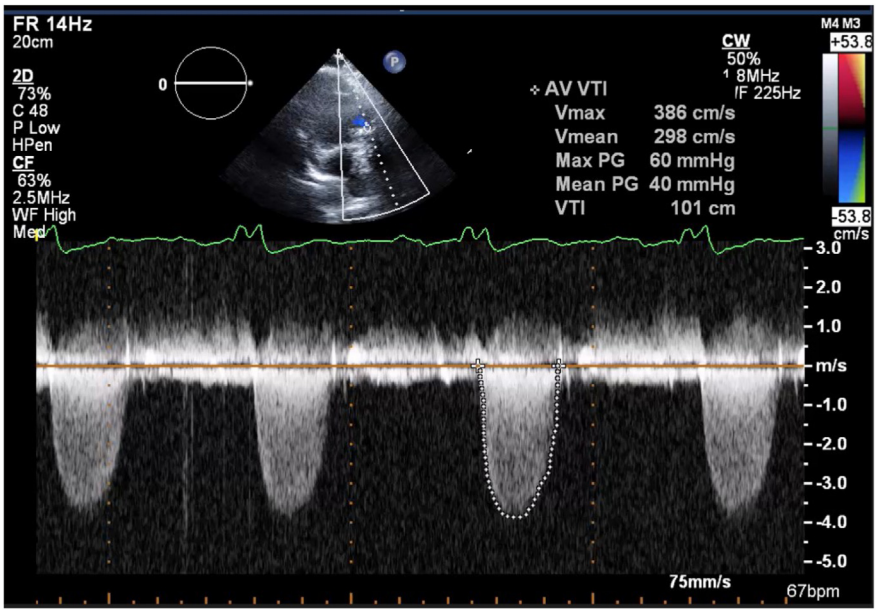


**Figure 4** Two-dimensional TTE, apical 3-chamber view, color-flow Doppler-guided continuous-wave Doppler across the aortic valve, demonstrates elevated aortic valve gradients (peak gradient of 60 mm Hg and a mean gradient of 40 mm Hg).

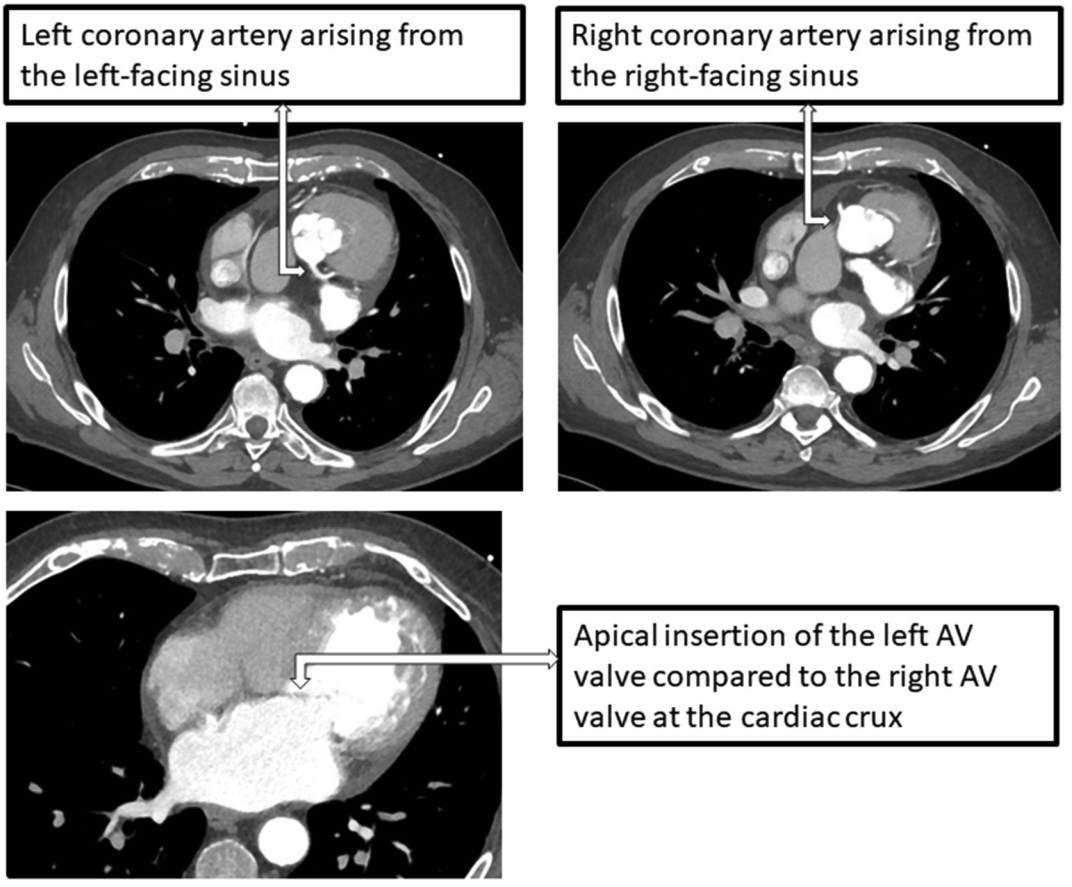


**Figure 5** Cardiac computed tomography, maximal intensity projection, axial displays at the level of the left (*top left*) and right (*top right*) coronary artery ostia, demonstrates the L-malposition of the great arteries, and at the midventricular level demonstrates the apically displaced systemic TV and the dilated and hypertrophied systemic RV.

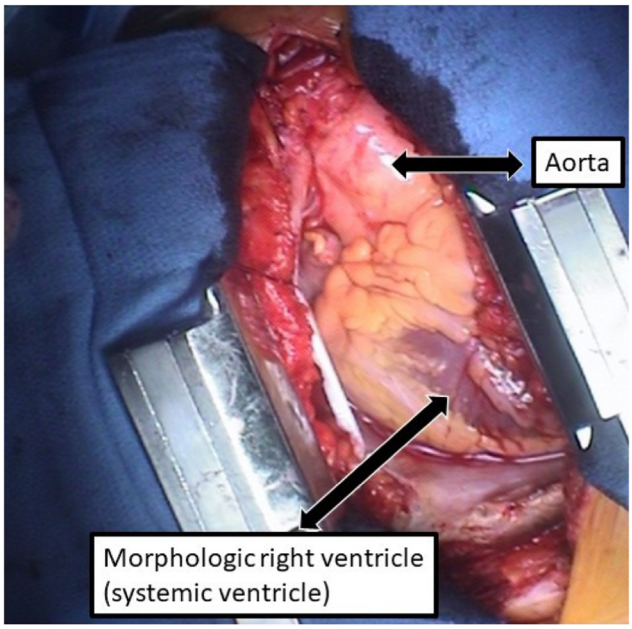


**Figure 6** Aorta arising from the morphologic RV observed during SAVR.

## DISCLOSURE STATEMENT

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2023.12.032.

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