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ORIGINAL ARTICLE

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A great imitator in adult cardiology practice: congenitally corrected transposition of the great arteries

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Abstract

Introduction: Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital disease that frequently remains undiagnosed until adulthood, especially when there is an absence of other congenital anomalies. Adults with ccTGA may remain asymptomatic and their diagnosis could be missed on initial evaluation, or it could be diagnosed incidentally as an evaluation of murmur. We aim to report the different presentations of ccTGA in eight adult patients and review the key features required to suspect the diagnosis during an initial visit.

Cases: We present some illustrative cases of ccTGA patients who had diverse presentations ranging from being completely asymptomatic to presenting with an acquired heart disease resulting in sudden cardiac arrest. Overall, most of these patients had isolated ccTGA with no other significant associated cardiac anomalies and were either undiagnosed or lost to follow-up until adulthood. These case illustrations represent the challenges confronted in adult practices when patients with unrecognized ccTGA present during an initial visit.

Conclusions: Congenitally corrected transposition of the great arteries poses a challenge in the adult cardiology practice because of its diverse clinical presentation. It is crucial that internists, cardiologists, and sonographers maintain a high degree of suspicion after meticulous physical examination for the early recognition of ccTGA, and thus avoid associated morbidities. Through some case examples, we provide clues to the key diagnostic features that could help them to be vigilant in making a diagnosis.

KEYWORDS
adult congenital heart disease, congenitally corrected transposition of great arteries

1 | INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital anomaly, accounting for less than 1% of all congenital anomalies.[1] Since its initial description by Rokitansky in 1875,[2] many cases have been described in the pediatric and adult populations. It presents with a wide spectrum of morphologic characteristics and clinical features.[3,4] In this anomaly, the right atrium is connected to a right-sided morphological left ventricle (LV), which is connected to the pulmonary artery (PA). The left atrium is connected to a left-sided morphological right ventricle (RV) from which arises the aorta. Thus, atrioventricular (AV) and ventriculoarterial discordance exists, resulting in "double discordance" (Figure 1A,B). The aorta usually is anterior and to the left of the pulmonary artery (levo-transposition), with both great arteries running parallel to each other. A majority of patients with ccTGA have associated cardiac anomalies such as ventricular septal defect (VSD), pulmonary stenosis, abnormalities of the systemic AV valve, ie, tricuspid valve, and the presence or absence of these can alter the natural history.[1]

Patients with ccTGA, especially those without associated defects, may remain undiagnosed until adult life.[3,4] The diagnosis of ccTGA often is overlooked in adult cardiology practice because of its rarity and a failure to recognize the abnormal position of the ventricles and the associated AV valves.[5] Among unoperated adults who present with ccTGA, 66% of patients present initially in adulthood, with 17% older than 60 years of age.[5] The majority of these

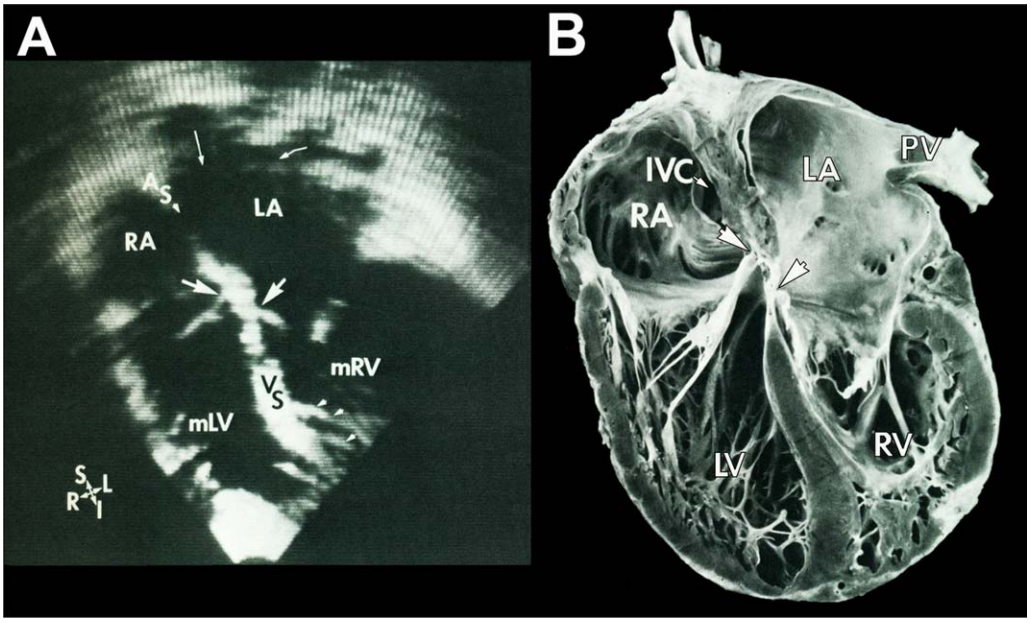

FIGURE 1 Corrected transposition of the great arteries. Atrioventricular discordance (ventricular inversion). Two-dimensional echocardiogram (**A**) and tomographically cut specimen (**B**) from the same patient. The pulmonary veins enter the left-sided atrium (dashed arrows). By posterior scanning from this apical tranducer position, the inferior vena cava was confirmed to be committed to the right-sided atrium. The internal cardiac crux has a mirror-image appearance, with the right-sided atrioventricular valve clearly inserting higher than the left-sided atrioventricular valve (arrows). The higher valve is mitral and the lower is tricuspid. The prominent apical trabeculations (arrowheads) in the left-sided ventricle help in establishing it as a morphologic right ventricle (mRV). AS, atrial septum; IVC, inferior vena cava; LA, left atrium; LV, left ventricle; mLV, morphological left ventricle; PV, pulmonary vein; RA, right atrium; RV, right ventricle; VS, ventricular septum. (Panel A from Seward JB, Tajik AJ, Edwards WD, Hagler DJ. Two-Dimensional Echocardiographic Atlas, Vol 1: Congenital Heart Disease. New York, NY: Springer-Verlag; 1987, with permission of Springer. Panel B from Hagler DJ, Tajik AJ, Seward JB, Edwards WD, Mair DD, Ritter DG. Atrioventricular and ventriculoarterial discordance (corrected transposition of the great arteries). Wide-angle two-dimensional echocardiographic assessment of ventricular morphology. Mayo Clin Proc 1981;56:591-600, with permission of Elsevier.)

patients present with systemic ventricular failure, and thus its early recognition during the initial cardiology visit is crucial to prevent adverse consequences. We report several illustrative cases of ccTGA that emphasize the protean manifestations of this entity in the adult cardiology practice.

2 | ILLUSTRATIVE CASE PRESENTATIONS

2.1 | Patient 1: 56-year-old, healthy, asymptomatic patient presenting with cardiac arrest

A 56-year-old, asymptomatic, white man first presented with out-of-hospital cardiac arrest. He is an executive and was at work when he suddenly collapsed. An automated external defibrillator was available at his workplace, and he was found to be in ventricular fibrillation. He required three shocks prior to return of spontaneous circulation. He was intubated, stabilized and then taken immediately to the cardiac catheterization laboratory. He was found to have anomalous coronary arteries, which involved common ostia, shared with left and right coronary arteries arising from the right coronary cusp. He had severe left circumflex artery stenosis that was treated with percutaneous coronary intervention (PCI). Electrocardiogram (ECG) showed first-degree AV block (Figure 3). Echocardiogram was performed to assess the LV function, and, surprisingly, it revealed a diagnosis of ccTGA with a systemic RV ejection fraction of 45% and mild tricuspid valve regurgitation (TR). He underwent placement of an implantable cardioverter defibrillator (ICD). The patient was doing well at 6-year follow-up and remained asymptomatic. A recent echocardiogram showed a moderately increased systemic RV cavity size, a mildly decreased ejection fraction of 48%, and mild left AV valve regurgitation, ie, TR. The most recent cardiopulmonary stress test showed excellent exercise tolerance with maximum oxygen consumption of 30.6 mL/kg/min (111% predicted).

2.1.1 | Remarks

This case represents the detection of ccTGA in a patient presenting with an acquired cardiovascular disease. These patients might develop other acquired diseases, such as coronary artery disease in this patient, which might be their initial presentation. Cardiac arrest or acute coronary syndrome also may be caused by coronary anomalies,[6] of which the most common is a single coronary arising from the right-facing sinus. Thus, interventional cardiologists should be aware of such varying coronary artery anatomy, and angiography in patients with ccTGA should be performed with vigilance because of the possibility of significant abnormalities that could complicate the procedure.[6] This challenge was particularly faced in this patient; the interventional cardiologist had difficulty engaging the coronaries and performing PCI, especially in the emergent situation.

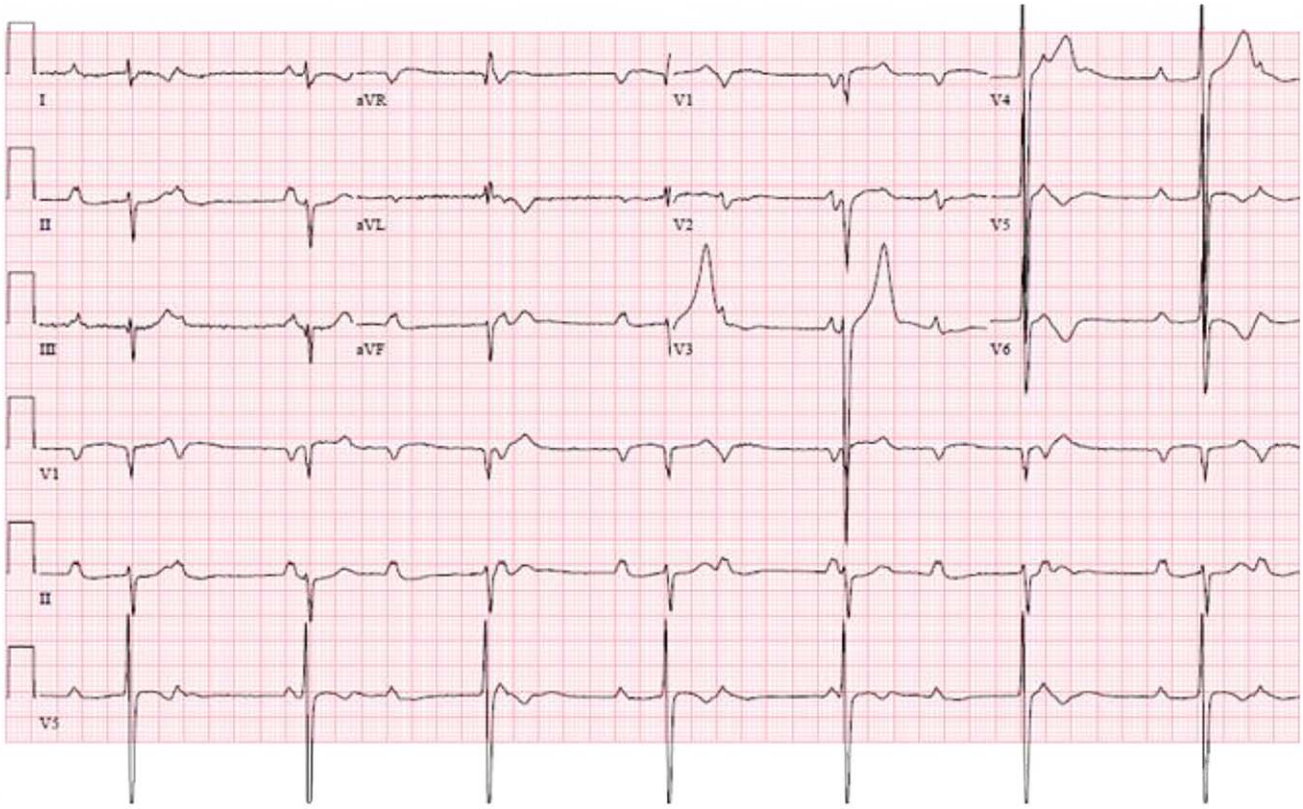

FIGURE 2 Electrocardiogram of Patient 3 during a routine clinic visit showing complete heart block

2.2 | Patient 2: 34-year-old healthy patient presenting with symptomatic complete heart block

A 34-year-old white woman with no other past medical history presented with dizziness and shortness of breath with New York Heart Association (NYHA) class II symptoms. An ECG and 24-hour Holter monitor revealed intermittent complete heart block (CHB). A chest x-ray demonstrated an abnormally straight upper cardiac silhouette (Figure 4). Subsequent echocardiogram demonstrated features of ccTGA with a systemic ventricular ejection fraction of 48% and mild TR. She underwent permanent pacemaker (PPM) placement. At 5-year follow-up, she was clinically doing well and her echocardiographic findings remained stable.

2.2.1 | Remarks

This case represents the evaluation of a young patient presenting with symptomatic CHB. The AV node and His bundle in patients with ccTGA have an unusual position and course, and many patients have dual AV nodes.[7,8] The incidence of CHB among patients with ccTGA ranges from 29% to 45%.[9] Approximately 10% of patients with ccTGA initially present with third-degree block, and 20–30% will have first-degree or second-degree block initially, which tends to progress with age to third-degree.[10] A progressive incidence of complete AV block occurs at 2% per year.[10] Thus, including ccTGA in the differential diagnosis of CHB, especially in young adults, along with other acquired heart disease is the key in making a diagnosis.

2.3 | Patients 3 and 4: 27-year-old and 36-year-old asymptomatic patients evaluated during routine physical examination

2.3.1 | Patient 3

A 32-year-old white man first presented at the age of 27 years with a systolic murmur noticed during routine physical examination. He was completely asymptomatic. The initial echocardiogram revealed a diagnosis of ccTGA (Figure 5) with subpulmonic membrane with a peak gradient of 38 mm Hg (mean 20 mm Hg), a hypertrabeculated RV (Figure 6A,B), moderate TR (systemic AV valve), and a moderately reduced systemic (RV) ventricular ejection fraction of 35%. Also, the rhythm strip on the echocardiogram showed an AV block. Subsequently, ECG showed CHB (Figure 2). He underwent ICD implantation. A cardiopulmonary stress test showed good exercise tolerance with maximum oxygen consumption of 39.3 mL/kg/min (89% predicted). Over the past 5 years, the patient has developed severe regurgitation of his systemic AV valve (tricuspid valve) (Figure 7) and a severely enlarged morphologic RV (Figures 8 and 9). He is currently being evaluated for tricuspid valve surgery. The most recent cardiopulmonary stress test showed good exercise capacity with maximum oxygen consumption of 38.7 mL/kg/min (90% predicted).

2.3.2 | Patient 4

A 36-year-old woman was referred because of an incidental finding of CHB noticed on an ECG performed as part of a preoperative evaluation for hysterectomy. She was completely asymptomatic and was

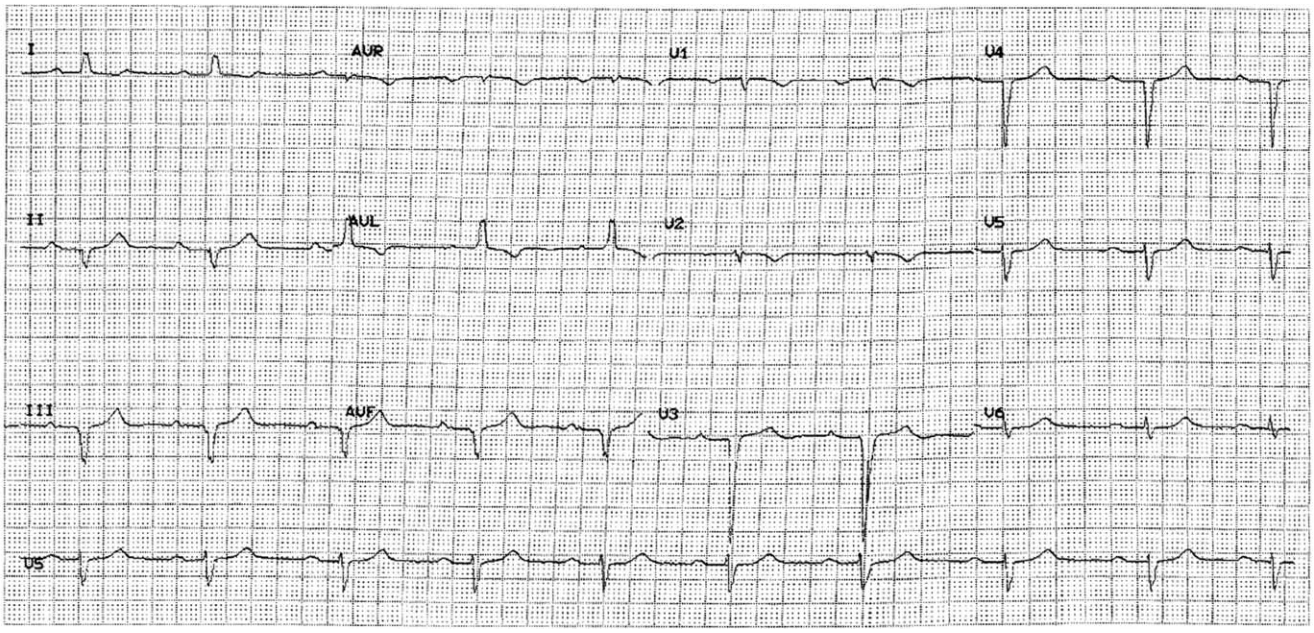

FIGURE 3 Electrocardiogram of Patient 1 showing an inferior and anteroseptal Q wave infarct pattern that is caused by inversion of the bundle branches leading to reverse activation of the septum from right to left

unaware of the presence of any congenital heart disease (CHD). A subsequent echocardiogram for further evaluation revealed a diagnosis of ccTGA with mild tricuspid and mitral valve regurgitation, and a systemic (RV) ventricular ejection fraction of 39%. There were no other associated anomalies. She underwent dual-chamber PPM placement, which was upgraded to a biventricular pacer at age 51 years. She has been followed in our CHD practice for 5 years now. She has occasional fatigue but no other cardiac symptoms. Her most recent echocardiogram showed a systemic ventricular (RV) EF of 37% and mild systemic AV valve regurgitation. Cardiopulmonary stress testing revealed moderate limitation of exercise capacity and a maximum oxygen consumption of 18.7 mL/kg/min (69% predicted). She is being managed medically with up-titration of heart failure medications.

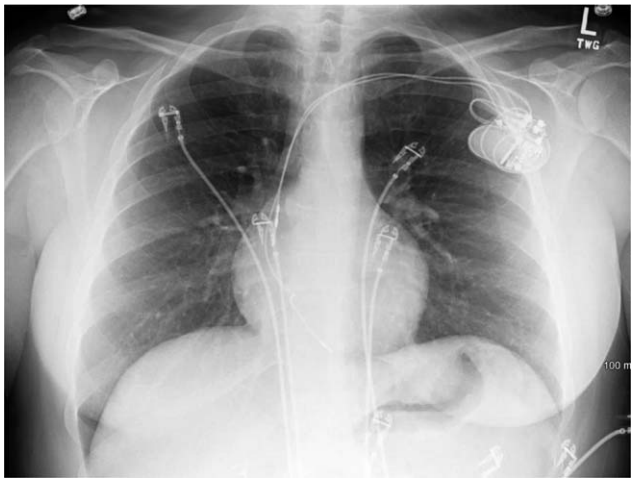

FIGURE 4 Chest x-ray of Patient 2 reveals an abnormally straight upper left heart silhouette due to loss of normal arterial relationships

2.3.3 | Remarks

These cases emphasize that often ccTGA patients with CHB may be completely asymptomatic when they present at their primary care physician's office. In 24% of patients with ccTGA, the diagnosis had been missed at the time of a prior cardiology consultation despite the use of cardiac imaging (echocardiogram, catheterization, or both).[5] This is more prevalent for the 1% of patients with ccTGA who do not have any associated lesions.[11] Patients may present with a variety of

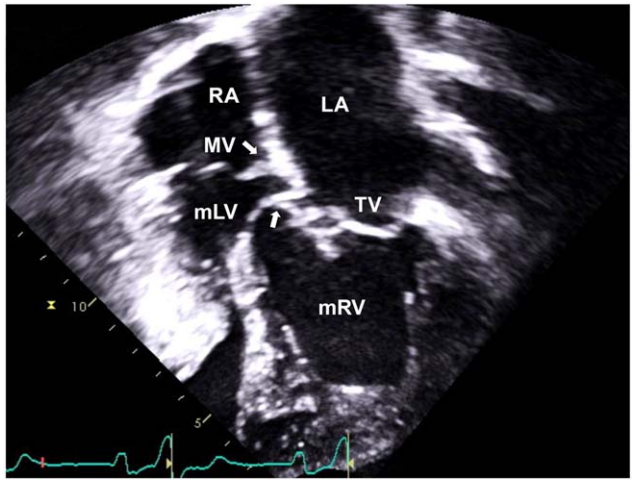

FIGURE 5 Illustrative four-chamber 2-dimensional echocardiographic view of Patient 3 showing the atrioventricular discordance (right atrium [RA] connected to the morphologic left ventricle [mLV] while left atrium [LA] connected to morphologic right ventricle [mRV]). Note, at the crux of the heart where the RV is recognized by the apically displaced tricuspid valve (TV) relative to mitral valve (MV) (arrows)

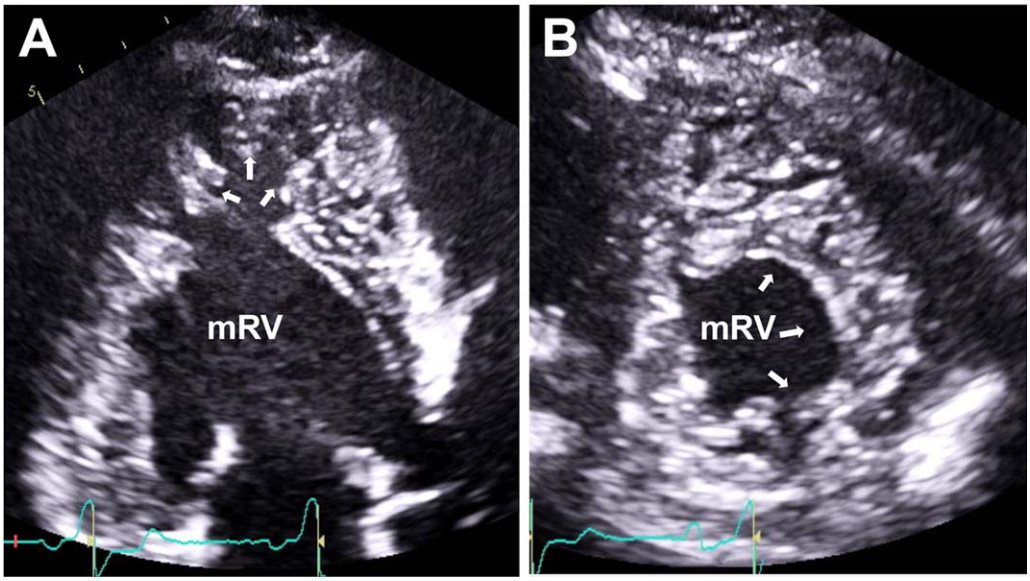

FIGURE 6 (**A**) Apical two-chamber view of Patient 3 showing hypertrabeculated apex (arrows) of morphological right ventricle (mRV). (**B**) Parasternal short-axis view of the same patient showing hypertrabeculated apex of mRV

nonspecific symptoms, and thus a high degree of suspicion about and awareness of this entity is crucial to diagnose ccTGA, especially during initial presentation and before the development of significant morbidity. Timely referral to a CHD center is necessary for their long-term follow-up to prevent significant morbidity from the disease.

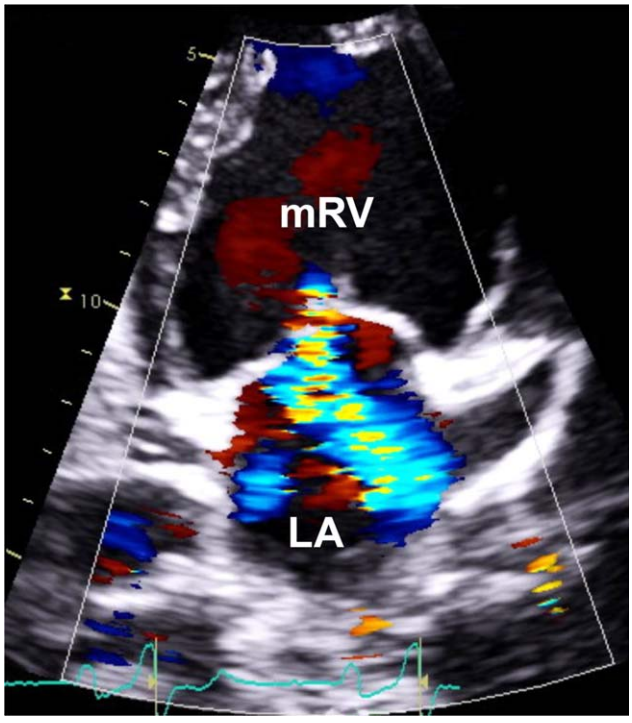

FIGURE 7 Apical two-chamber view of Patient 3 showing systemic atrioventricular valve (tricuspid valve) regurgitation that was quantified as severe regurgitation. LA, left atrium; mRV, morphological right ventricle

2.4 | Patients 5 and 6: 62-year-old and 50-year-old patients presenting with heart failure

2.4.1 | Patient 5

A 62-year-old white woman presented to our adult congenital center for the first time with symptoms of heart failure and NYHA class II symptoms. She was found to have CHB requiring PPM implantation. Further history revealed that at 19 years of age, she was evaluated for shortness of breath with NYHA class II symptoms. She was noticed to have cardiomegaly on chest x-ray. A subsequent echocardiogram revealed a diagnosis of ccTGA with mild tricuspid and mitral valve regurgitation, and mildly reduced systemic (RV) ventricular function. She was then lost to follow-up until presentation to our clinic with heart failure symptoms at age 62 years. An echocardiogram revealed severe TR as well as bileaflet mitral valve prolapse with mild regurgitation and a systemic (RV) ventricular ejection fraction of 37%. She also was noted to have azygous continuation of the inferior vena cava, with the hepatic veins draining directly into the RA.[12] She continued to remain symptomatic despite optimal medical management and subsequently underwent tricuspid valve replacement at 65 years of age. Preoperative coronary angiography revealed an anomalous origin of the right coronary artery from the left cusp and left coronary artery from the right cusp, but no significant atherosclerosis. She was doing well at 3-year follow-up post valve replacement.

2.4.2 | Patient 6

A 50-year-old white man was referred for severe heart failure symptoms resulting in NYHA class III disability. At 18 years of age, his admission into the army was rejected due to a finding of an abnormal murmur on physical examination. He was completely asymptomatic at that time. An ECG was unremarkable, but a subsequent

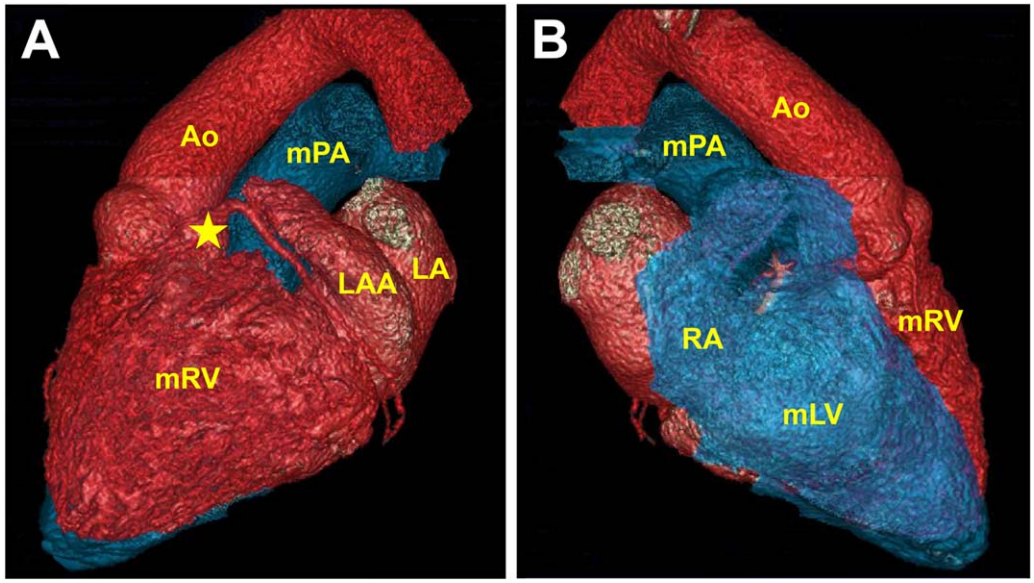

FIGURE 8 (**A**) A 3-dimensional reconstruction by cardiac computed tomography (CT) of Patient 3 showing the atrioventricular discordance (LA connected to mRV), ventriculoarterial discordance (mRV connected to the aorta [Ao]) and transposition of great arteries (anterior location of the Ao compared to the pulmonary artery [PA]). Also noted is the origin of the right coronary artery from the left coronary cusp (*). (**B**) A 3-dimensional reconstruction by CT showing atrioventricular and ventriculoarterial (double) discordance in a patient with congenitally corrected transposition. LAA, left atrial appendage; mLV, morphological left ventricle; mPA, main pulmonary artery; RA, right atrium

echocardiogram led to the diagnosis of ccTGA. He was then lost to follow-up since he remained asymptomatic until 50 years of age, after which he started developing shortness of breath and was found to have severe systemic (tricuspid) AV valve dysfunction requiring tricuspid valve ring #34 placement at age 52. Despite optimal medical management, he deteriorated further with severe systemic (right) ventricular dysfunction and NYHA class IV symptoms requiring an LV assist device as a bridge to transplant at age 60 years. He finally underwent cardiac transplantation at age 61 years. He was doing very well at 5-year follow-up after transplantation, and was asymptomatic.

2.4.3 | Remarks

These two examples are illustrative of the fact that despite the diagnosis having been established earlier in life, these patients are often either given the perception that there is no need for further long-term follow-up or they interpret the physician's recommendation of a stable cardiac condition as one that does not need any follow-up. A minority of patients may be relatively normal from a functional standpoint, and survival to the seventh and eighth decades has been reported when no associated anomalies exist.[3,13] However, failure of the systemic ventricle is much more common earlier in life, usually with concomitant systemic AV valve regurgitation,[14] and, thus, surgery should be considered

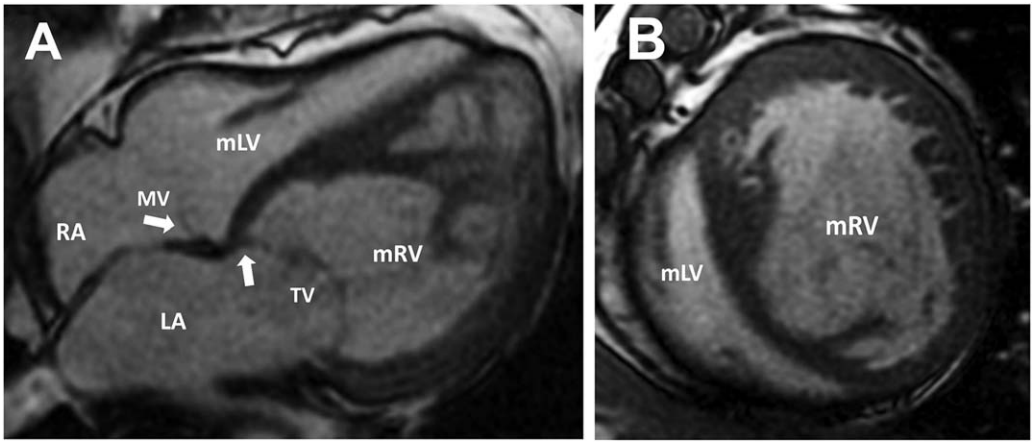

FIGURE 9 (**A**) Cardiac magnetic resonance image (MRI) of Patient 3 showing coarsely trabeculated morphologic right ventricle (mRV) with its associated tricuspid valve (TV) on the left and morphologic left ventricle (mLV) with its associated mitral valve (MV) on the right. (**B**) Cardiac MRI showing an enlarged hypertrabeculated mRV on the left and a small crescent-shaped mLV. LA, left ventricle; RA, right atrium

early, before irreversible ventricular function ensues. Even in patients with ccTGA and no significant associated lesions, more than one-third will have congestive heart failure by the fifth decade.

There are several mechanisms contributing to failure of the systemic RV in patients with ccTGA. In a true morphological left ventricle, the myocytes are arranged in a helical pattern that twists and untwists during the cardiac cycle, producing energy efficient forces to maintain the demands of systemic circulation.[15,16] The RV lacks such myofibril arrangement, and hence in ccTGA patients, when the RV becomes the systemic ventricle it is unable to generate such mechanics and eventually fails.[17–19] Eventually, these patients develop systemic AV valve regurgitation, which leads to volume overload and further deterioration of the systemic RV. Other factors that can lead to worsening RV function include myocardial ischemia and reduced coronary flow.[20,21]

The outcomes of these two patients clearly emphasize the importance of detailed counseling regarding the long-term prognosis of this condition at initial diagnosis and the need for life-long surveillance at an experienced adult CHD center. Recently, it has been clearly shown that referral to specialized adult CHD care is independently associated with a significant mortality reduction in patients with CHD.[22,23]

2.5 | Patients 7 and 8: presentation of a typical patient with ccTGA and other associated anomalies

2.5.1 | Patient 7

A 34-year-old Asian man first presented at age 27 years with palpitations, shortness of breath, and NYHA class II disability. He was noted to have atrial fibrillation on ECG, and a chest x-ray showed dextrocardia. An initial echocardiogram done at an outlying hospital was read as normal except for isolated dextrocardia. A subsequent echocardiogram showed situs solitus, dextrocardia, ccTGA with large infundibular subpulmonic VSD, and pulmonary stenosis. His systemic ventricular (morphologic RV) ejection fraction was 30%, and he had mild atrioventricular valve regurgitation. A cardiopulmonary stress test was performed to further evaluate his functional capacity. His maximum oxygen consumption was 18.9 mL/kg/min (44% predicted). He then underwent complete repair of the ccTGA with patch closure of the membranous VSD, right AV valve (mitral) repair with a 32 mm Cardiomedics annuloplasty ring, pulmonary valve replacement with a Carpentier Edwards 23 mm bovine pericardial bioprosthesis, pulmonary ventricle to pulmonary artery confluence conduit placement with Contegra bovine jugular 22 mm graft, patent foramen ovale closure, and resection of a subpulmonary fibrous obstruction. Postoperatively, he developed CHB requiring PPM placement. He was doing well at 7-year follow-up with mild AV valve regurgitation and a systemic ventricular ejection fraction of 30% and peak oxygen consumption of 18.3 mL/kg/min (44% predicted).

2.5.2 | Patient 8

A 31-year-old Arabic man presented to our adult congenital heart disease clinic with progressively worsening shortness of breath on exertion for the past year. He was diagnosed with dextrocardia, ccTGA, and CHB requiring PPM as an infant. Prior to being evaluated in our clinic, the patient was managed for two years with diuretics for heart failure symptoms. On initial echocardiogram at our center, the patient was found to have a dilated systemic RV with moderately reduced function (ejection fraction of 40%), moderate TR, moderate morphologic LV dysfunction, a large perimembranous VSD, and severe pulmonary hypertension. A right-heart catheterization was performed that showed a PA pressure of 105/57 mm Hg with a mean PA pressure of 75 mm Hg and systemic aortic pressure of 128/94. The patient currently is being managed for his Eisenmenger VSD and severe pulmonary hypertension with riociguat, digoxin, valsartan, and diuretics.

2.5.3 | Remarks

The above 2 cases highlight the presentation of ccTGA with associated cardiac anomalies. Up to 80% of patients with ccTGA have accompanying associated cardiac lesions. VSDs, usually perimembranous in location, can be seen in 60% to 80% of cases and pulmonary stenosis in about 50% of cases.[1,18,19] Patients with large VSDs and consequent left-to-right shunt usually present earlier in life (infancy or childhood) with heart failure symptoms. It is important to recognize and appropriately manage these associated cardiac lesions earlier in life to prevent complications such as Eisenmenger syndrome, pulmonary hypertension, and heart failure.

3 | DISCUSSION

We present the different faces of presentation of ccTGA in our adult congenital heart disease center (Table 1). Most of these patients were not diagnosed with their congenital condition until adulthood when they had varied presentations ranging from being completely asymptomatic to presenting with an acquired disease, ie, cardiac arrest from acute myocardial infarction. Thus, in the current era, when the population of adults with congenital heart disease (CHD) is rapidly growing, constant vigilance in general cardiology and adult medicine practice is key to the early recognition and management of these patients. In the United States, there are now more people over the age of 20 with CHD than under that age.[24]

The population of adult CHD patients is growing at a rate of 5% per year, with 2010 data estimating 1.4 million people in the U.S. having CHD as adults.[25] Using this estimated growth rate, currently there should be approximately 2 million adults with CHD. The medical management of adult patients with CHD will continue to be a challenge because the number of CHD patients increases on a yearly basis while the expertise and available personnel remain limited and focused within very few centers nationally. There are 70 self-proclaimed adult CHD clinics in the U.S. that see a total of about 50,000 patients—only 5% of the 1 million to 1.3 million patients who need care.[24] Thus, most patients continue to be seen by family medicine practitioners, internists, and occasionally by adult cardiologists, who in fact are often the first medical contact these patients might have. Congenital defects like ccTGA, in the absence of other associated anomalies, not only frequently remain unrecognized until adulthood, but also are often

TABLE 1 Clinical characteristics of 8 patients with congenitally corrected transposition of great arteries

Clinical Characteristics	Patient 1	Patient 2	Patient 3	Patient 4	Patient 5	Patient 6	Patient 7	Patient 8
Age (current)	62	39	32	57	68	66	34	34
Age at diagnosis	As a child	34	At birth but lost to follow-up, seen in clinic at 27	36	19	18	27	Infancy
Sex	Male	Female	Male	Female	Female	Male	Male	Male
Race	White	White	White	White	White	White	Asian	Arabic
Clinical presentation at diagnosis in adult practice	Cardiac arrest at age 56	Dizziness	Murmur on physical exam	Pre-op abnormal ECG	Cardiomegaly on CXR	Dyspnea on exertion	Atrial fibrillation	Dyspnea on exertion
NYHA Class	1	2	2	1	2	4	2	3
Symptoms	None	Dizziness, SOB	None	None	SOB	SOB	Palpitation, SOB	SOB
Other anomalies	Anomalous coronaries	None	Subpulmonic membrane with a peak gradient of 38 (mean 20), RV apical noncompaction	None	Azygous continuation of IVC with hepatic veins directly draining into RA, anomalous coronaries	None	Dextrocardia, large infundibular subpulmonic VSD, pulmonary stenosis	Dextrocardia, large perimembranous VSD, Eisenmenger syndrome
ECG	1st degree AV block	Intermittent CHB	CHB	CHB	CHB	LBBB	CHB after surgery	CHB
Systemic ventricle (RV) EF (%)	49	48	39	37	37	20	30	40
Left AV (tricuspid) valve regurgitation	Mild	Mild	Severe	Mild	Severe	Severe	Mild	Moderate
Right AV (mitral) valve regurgitation	Trace	Trace	Trace	Mild	Bileaflet prolapse with mild MR	Trace	Mild	Trace
CP stress test (most recent)	VO2 30.6 mL/kg/min (111% predicted)	VO2 15.8 mL/kg/min (47% predicted)	VO2 38.7 mL/kg/min (90% predicted)	VO_2 18.7 mL/kg/min (69% predicted)	None	None	VO2 18.3 mL/kg/min (44% predicted)	17.5 ml/kg/min (40% of predicted)
ICD	Yes	No	Yes	Yes, at age 51	No	Yes	No	No
PPM	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes, in infancy
Surgery	Circumflex PCI	None	Yes—Planned for systemic AV valve replacement	None	Yes, tricuspid valve replacement	TV ring # 34 at age 52, LVAD as a bridge to transplant at age 60, cardiac transplant at age 61	Complete repair of ccTGA at age 27	No

AV, atrioventricular; CHB, complete heart block; CP, cardiopulmonary; ECG, electrocardiogram; EF, ejection fraction; ICD, implantable cardioverter defibrillator; LBBB, left bundle branch block; NYHA, New York Heart Association; PCI, percutaneous coronary intervention; PPM, permanent pacemaker; TV, tricuspid valve; SOB, shortness of breath; VO_2, maximum oxygen consumption.

overlooked in cardiology practice.[5] This is mainly because of the varied clinical presentation and failure to recognize the abnormal position of ventricles and associated AV valves. The most important and definitive diagnostic test for ccTGA is a meticulous echocardiogram with close attention to the crux of the heart and the position of morphologic right and left ventricles.[1] Imaging should be focused on the parallel or abnormal orientation of the great arteries, with the aorta located anteriorly and to the left of the pulmonary artery.

Among all adult CHD hospital admissions, 20% are admissions for heart failure, at a mean age of 69 years.[26] There is an increasing incidence of systemic ventricular dysfunction and clinical congestive heart failure with advancing age in ccTGA patients,[27] seen in more than one-third of patients by the fifth decade, in the absence of any significant associated lesions. It is crucial to increase awareness of this condition so it can be diagnosed during the initial visit, thus avoiding the associated morbidity and mortality.

4 | CONCLUSIONS

Congenitally corrected transposition of the great arteries is a rare congenital anomaly that poses a challenge in the adult cardiology clinic due to its diverse clinical presentation, which remains relatively asymptomatic, especially in the absence of any associated significant congenital defects. Thus a high-degree of suspicion with an increased awareness about the key diagnostic features among cardiologists and sonographers is crucial for the early recognition of ccTGA and avoidance of associated morbidities.

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AUTHOR CONTRIBUTIONS

Anushree Agarwal: Dr. Agarwal was responsible for the initiation of the study, the study design, data acquisition and collation, interpretation of results, drafting and revising the manuscript, and approval of the final manuscript.

Fatima Samad: Dr. Samad was responsible for interpretation of results, drafting and revising the manuscript, and final approval of the manuscript.

Lindsey Kalvin: Ms. Kalvin was responsible for acquisition of data and images, and final approval of the manuscript.

Michelle Bush: Ms. Bush was responsible for data acquisition and approval of the final manuscript.

A. Jamil Tajik: Dr. Tajik was responsible for the acquisition and collection of data, analysis and interpretation of the results, revision, and approval of the final manuscript.

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The authors have indicated they have no potential conflicts of interest to disclose.

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REFERENCES

[1] Warnes CA. Transposition of the great arteries. *Circulation*. 2006; 114:2699–2709.

[2] Rokitansky C. Die Defecte Der Scheidewände Des Herzens: Pathologische Anatomische Abhandlung. Vienna, Austria: W. Braumüller; 1875.

[3] Presbitero P, Somerville J, Rabajoli F, Stone S, Conte MR. Corrected transposition of the great arteries without associated defects in adult patients: clinical profile and follow up. *Br Heart J*. 1995;74: 57–59.

[4] Bullock-Palmer RP, Rohen A. Congenitally corrected transposition of the great arteries (CCTGA) initially presenting in the sixth decade. *Echocardiography (Mount Kisco, NY)*. 2009;26:1118–1120.

[5] Beauchesne LM, Warnes CA, Connolly HM, Ammash NM, Tajik AJ, Danielson GK. Outcome of the unoperated adult who presents with congenitally corrected transposition of the great arteries. *J Am Coll Cardiol*. 2002;40:285–290.

[6] Lampropoulos KM, Kotsas D, Iliopoulos TA. Acute coronary syndrome in congenitally corrected transposition of the great arteries. *BMJ Case Rep*. 2013;2013:bcr2012008354. Published online 2013 Apr 29. doi: 10.1136/bcr-2012-008354

[7] Wilkinson JL, Smith A, Lincoln C, Anderson RH. Conducting tissues in congenitally corrected transposition with situs inversus. *Br Heart J*. 1978;40:41–48.

[8] Anderson RH, Becker AE, Arnold R, Wilkinson JL. The conducting tissues in congenitally corrected transposition. *Circulation*. 1974;50: 911–923.

[9] Fischbach PSLI, Serwer GS. Congenitally corrected L-transposition of the great arteries: abnormalities of atrioventricular conduction. *Prog Pediatr Cardiol*. 1999;10:37–43.

[10] Juneja R, Rowland E, Ho SY. Atrial morphology in hearts with congenitally corrected transposition of the great arteries: implications for the interventionist. *J Cardiovasc Electrophysiol*. 2002;13: 158–163.

[11] Baumgartner H, Bonhoeffer P, De Groot NM, et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). *Eur Heart J*. 2010;31:2915–2957.

[12] Paterick TE, Schmidt M, Jan MF, et al. Congenitally corrected transposition of the great arteries with anomalous inferior vena cava drainage: multimodality imaging. *Echocardiography (Mount Kisco, NY)*. 2012;29:E16–E19.

[13] van Son JA, Danielson GK, Huhta JC, et al. Late results of systemic atrioventricular valve replacement in corrected transposition. *J Thorac Cardiovasc Surg*. 1995;109:642–652; discussion 652-3.

[14] Prieto LR, Hordof AJ, Secic M, Rosenbaum MS, Gersony WM. Progressive tricuspid valve disease in patients with congenitally corrected transposition of the great arteries. *Circulation*. 1998;98: 997–1005.

[15] Shapiro EP, Rademakers FE. Importance of oblique fiber orientation for left ventricular wall deformation. *Technol Health Care*. 1997;5: 21–28.

[16] Sengupta PP, Tajik AJ, Chandrasekaran K, Khandheria BK. Twist mechanics of the left ventricle: principles and application. *JACC Cardiovasc Imaging*. 2008;1:366–376.

[17] Peterson RJ, Franch RH, Fajman WA, Jones RH. Comparison of cardiac function in surgically corrected and congenitally corrected transposition of the great arteries. *J Thorac Cardiovasc Surg*. 1988; 96:227–236.

[18] Wallis GA, Debich-Spicer D, Anderson RH. Congenitally corrected transposition. *Orphanet J Rare Dis*. 2011;6:22.

[19] Filippov AA, Del Nido PJ, Vasilyev NV. Management of systemic right ventricular failure in patients with congenitally corrected transposition of the great arteries. *Circulation*. 2016;134:1293–1302.

[20] Hornung TS, Bernard EJ, Celermajer DS, et al. Right ventricular dysfunction in congenitally corrected transposition of the great arteries. *Am J Cardiol*. 1999;84(9):1116 a10.

[21] Hornung TS, Bernard EJ, Jaeggi ET, Howman-Giles RB, Celermajer DS, Hawker RE. Myocardial perfusion defects and associated systemic ventricular dysfunction in congenitally corrected transposition of the great arteries. *Heart (British Cardiac Society)*. 1998;80:322–326.

[22] Mylotte D, Pilote L, Ionescu-Ittu R, et al. Specialized adult congenital heart disease care: the impact of policy on mortality. *Circulation*. 2014;129:1804–1812.

[23] Webb G, Landzberg MJ, Daniels CJ. Specialized adult congenital heart care saves lives. *Circulation*. 2014;129:1795–1796.

[24] Moodie D. Adult congenital heart disease: past, present, and future. *Tex Heart Inst J*. 2011;38:705–706.

[25] Gilboa SM, Devine OJ, Kucik JE, et al. Congenital heart defects in the United States: estimating the magnitude of the affected population in. *Circulation*. 2016;134:101−109.

[26] Rodriguez FH, 3rd, Moodie DS, Parekh DR, et al. Outcomes of hospitalization in adults in the United States with atrial septal defect, ventricular septal defect, and atrioventricular septal defect. *Am J Cardiol*. 2011;108:290–293.

[27] Graham TP, Jr., Bernard YD, Mellen BG, et al. Long-term outcome in congenitally corrected transposition of the great arteries: a multiinstitutional study. *J Am Coll Cardiol*. 2000;36:255–261.

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