Review Article

Diagnosis, management, and outcome of heart failure in congenitally corrected transposition of the great arteries (ccTGA)—a narrative review

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Background and Objective: Congenitally corrected transposition of the great arteries (ccTGA) remains a rare congenital disorder with a wide range of manifestations. The management of heart failure (HF) of the systemic right ventricle (RV), arrhythmias, heart block, and acquired cardiac conditions require a complex and multi-faceted approach. The objective of this manuscript is to present the current evidence regarding diagnostic, treatment, and management strategies for HF in ccTGA, including ventricular assist device (VAD) therapy and heart transplantation.

Methods: A systematic review of the literature was conducted using PubMed, covering the period between 2010 and 2024. The search terms included "heart failure", "ccTGA", "VAD", "heart transplantation", and "systemic RV failure". Two clinical cases were included for illustrative purposes.

Key Content and Findings: HF is a common occurrence in the context of ccTGA, primarily driven by progressive pressure and volume overload of the systemic RV, regurgitation of the systemic atrio-ventricular (AV) valve, and the development of arrhythmias, including complete heart block and (supra-)ventricular tachycardia. The use of HF medication is indicated for symptomatic patients, however, data on the efficacy of standardized HF medication remains limited. Timing of AV-valve replacement is essential to prevent further progression of HF.

Conclusions: In ccTGA, the timing of surgery and interventional treatment approaches, the effect of pharmacological treatment in the context of HF, as well as the timing of initiation of a mechanical circulatory support, VAD and heart transplantation, are based on individualised consensus-level decisions. Optimal management remains a topic of debate due to the scarcity of outcome data. Future investigations should focus on identifying surrogate parameters for guiding treatment.

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Keywords: Adults with congenital heart disease (ACHD); heart failure (HF); congenitally corrected transposition of the great arteries (ccTGA); heart transplantation; ventricular assist device (VAD)

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Introduction

The number of adult patients with congenital heart disease (ACHD) is increasing steadily as a result of advances in both medical and surgical treatment options, which are improving outcomes for children with complex congenital heart disease (CHD) (1,2). In cases of complex or hemodynamically relevant CHD, sequelae often persist or emerge over time, even in the context of primary surgical success or interventions. Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital heart disease, accounting for 0.5% of all ACHD cases (3). The condition presents a significant challenge due to its complex anatomical variations and associated lesions, which give rise to a range of surgical treatment options. Defects accompanying cyanotic shunting frequently manifest as early symptoms, necessitating surgical intervention. In the absence of associated lesions, ccTGA may remain asymptomatic into adulthood. A previous study reported that 66% of ccTGA patients were correctly diagnosed for the first time at age 18 years, while 24% of these patients had a prior cardiology consultation in which the diagnosis was missed, despite the use of cardiac imaging (4). The objective of this review is to describe the complications and management of ccTGA in adult life.

Acquired lesions or arrhythmias manifesting throughout disease progression [e.g., atrial fibrillation, atrio-ventricular-nodal reentrant tachycardia (AVNRT), coronary artery disease, left ventricle (LV)/right ventricle (RV) dilation, thromboembolic events, total heart block, ventricular arrhythmias, and sudden cardiac death] result in a challenging patient cohort with a high burden of morbidity and mortality. New surgical and interventional treatments have transformed the management of this complex condition over the past few decades. However, timing and efficacy of surgical and interventional approaches, the use of heart failure (HF) medication and when to initiate mechanical circulatory support and ventricular assist devices (VADs) or heart transplantation remain topics of debate due to a paucity of data. The objective of this manuscript is to present the current evidence regarding diagnostic, treatment, and management strategies of HF within ccTGA, including VAD therapy and heart transplantation. We present this article in accordance with the Narrative Review reporting checklist (available at https://cdt.amegroups.com/article/view/10.21037/cdt-24-334/rc).

Anatomy

ccTGA is defined by atrio-ventricular (AV) and ventriculo-arterial discordance and was initially delineated by Von Rokitansky in 1875 (5). The condition may be apparent in *situs solitus* or *inversus*, which refers to the visceroatrial constellation. The position of the heart is typically left-sided (laevocardia), though it may also be situated in the midline (mesocardia) or, in 20% of cases, on the right side (dextrocardia) (6). In most cases, an inversion of the ventricular arrangement is evident, with the RV situated to the left and the aorta positioned anteriorly and ascending from the systemic RV (7). The term "congenitally corrected" is therefore a misnomer, as it does not refer to a surgical form of repair, but rather to the redirected blood flow due to intrauterine adaptations. It is not uncommon for associated lesions to be present, although ccTGA can also manifest as an isolated defect. Lesions associated with ccTGA are most commonly ventricular septal defects (VSDs), (sub-) pulmonary stenosis or atresia, tricuspid valvular lesions or atrial septal defects, which can be grouped into up to 13 different anatomic subtypes of ccTGA (8).

Methods

A systematic review of the literature was conducted using PubMed, encompassing the period between 1st April 2010 and 31st March 2024 (*Table 1*). The search terms employed included "heart failure", "ccTGA", "VAD", "heart transplantation", and "systemic RV failure". Two clinical cases were included for illustrative purposes.

Table 1 The search strategy summary

Items	Specification
Date of search	20th May 2024
Database searched	PubMed
Search terms used	"heart failure", "ccTGA", "VAD", "heart transplantation", and "systemic RV failure"
Timeframe	Between 1st April 2010 and 31st March 2024
Inclusion criteria	Literature that represents the current guidelines or multicentre trials (however most of the literature was derived from observational studies and guidelines for patients with ccTGA have the evidence level of expert opinion), consideration of qualitative and quantitative studies, restriction to original research, humans, and papers published in English at any date within the timeframe
Selection process	Review of all abstracts meeting the inclusion criteria. Manual search of the reference lists of all eligible articles. Two authors (i.e., M.K. and M.L.B.V. with support of S.I. and C.S.) assessed the importance of the studies independently prior to their inclusion in the review. A relevant manuscript was defined based on the impact factor of the journal, being a guideline manuscript, a society position paper, or a comprehensive review of the current literature

ccTGA, congenitally corrected transposition of the great arteries.

The search terms had to be identified in the text of the articles, regardless of where they were located. Moreover, the authors selected literature that represents the current guidelines or multicentre trials. However, the majority of the literature is derived from observational studies, and the evidence level for guidelines for patients with ccTGA is that of expert opinion. Both qualitative and quantitative studies were considered in order to elucidate the use of the aforementioned aspects regarding ccTGA, advanced HF, VAD therapy or device therapy in general in ccTGA. The search was limited to original research, human subjects, and papers published in English within the searched time frame. The abstracts were then subjected to a review in order to ascertain whether the articles in question met the requisite inclusion criteria. Subsequently, a manual search of the reference lists of all eligible articles was conducted. Prior to their inclusion in the review, the studies were assessed by two authors (M.K. and M.L.B.V., with support from S.I. and C.S.) for their relative importance. A relevant manuscript was defined as one that met the following criteria: impact factor of the journal, guideline, society position paper, or comprehensive review of the current literature.

Presentation and management of ccTGA

Patients with ccTGA form part of a heterogeneous patient collective, mainly arising from associated anatomical variations and comorbidities. Therefore, most studies examining patient characteristics and outcomes differentiate between those with ccTGA who exhibit early symptom onset and are diagnosed at birth or infancy with prompt surgical intervention and those with less specific, slowly progressing symptoms who do not require immediate intervention or surgery and are therefore likely diagnosed later in life (9).

The natural course of ccTGA

The natural course of the disease is dependent on the presence of concomitant anomalies in the majority of cases, as well as the progression of systemic ventricular dysfunction and conduction disturbances (10). A recent study reported a mean age at diagnosis of 12.4±11.4 years, while the mean age at the last review was 34.3±11.3 years. A total of 56% of the subjects included in the study had undergone surgical intervention. A total of 19% of patients had undergone pacemaker implantation, while two patients had received a defibrillator for the treatment of non-sustained ventricular tachycardia (NSVT). The mortality rate was reported as 1 death per 109 patient-years. Two of the patients had a documented history of supraventricular tachycardia or NSVT. However, all patients were classified as New York Heart Association (NYHA) class I or II, and the systemic (right) ventricular function was recorded as normal, mildly impaired, or mildly to moderately impaired at the most recent follow-up (11).

Presentation and management of ccTGA at birth and infancy

Currently, the diagnosis of ccTGA is primarily based on fetal echocardiography or prenatal karyotyping of the fetus. Consequently, counselling for this condition commences during the fetal stage of development. Patients with ccTGA and an early presentation frequently present with associated cardiac or extracardiac anomalies. The principal manifestations include HF symptoms (e.g., dyspnoea, oedema, restlessness, weakness, failure to thrive, breathing and drinking difficulties or heart murmur) resulting from pulmonary volume overload or cyanosis arising from concomitant VSD or pulmonary artery stenosis (12). A significant proportion of these cases necessitate prompt surgical intervention. The diversity of anatomic characteristics gives rise to a range of physiological manifestations in patients with ccTGA, which in turn result in a spectrum of symptoms.

Consequently, the surgical approach is determined by the physiological behaviour of patients with ccTGA.

The surgical approach can be divided into two main categories: biventricular repair and staged Fontan procedures. It is difficult to make a comparison between the merits of functional and anatomical repair for biventricular repair due to differences in the historical background and the target cases involved. In cases of functional repair, the Rastelli procedure is performed to address pulmonary stenosis, while patch closure is undertaken to address a VSD.

In the context of anatomical repair, intra-atrial blood flow is redirected through the utilisation of the Senning or Mustard technique. Subsequently, the procedure is classified as either an arterial switch or Rastelli procedure, contingent on the presence of pulmonary valve stenosis.

Presentation and management of ccTGA in adulthood

Adult patients suffering from ccTGA often present with rather unspecific symptoms, including palpitations and decreased exercise capacity. Diagnosis is based on the identification of abnormal findings in multimodality imaging, consisting of transthoracic/transoesophageal echocardiography (TOE), chest X-ray, magnetic resonance imaging (MRI), or computed tomography (CT) scans. Patients with late manifestation of ccTGA present with mild or no associated cardiac lesions. Still, VSD and pulmonary valve stenosis can be observed in adult patients with ccTGA and balanced haemodynamics. The tricuspid configuration of the systemic AV-valve renders it susceptible to central maladaptation in the context of a progressive dilation of the systemic RV due to chronic volume overload. Disease management in adulthood is therefore guided by evaluation for systemic AV-valve repair or replacement, primary or secondary preventative device implantation and/or cardiac resynchronization therapy (CRT) as well as end-stage HF management.

Diagnosis, management, and outcome of HF in ccTGA

Patients with ccTGA are at an increased risk of developing HF. Approximately 32% of adults with ccTGA present with signs and symptoms of HF. Current clinical guidelines for acquired heart disease may not be directly applicable to patients with failure of the systemic RV due to a lack of evidence. In patients with ccTGA who develop HF, it is of utmost importance to ascertain whether there are any residual lesions requiring treatment.

Double switch surgery carries an inherent risk of low output syndrome in the immediate postoperative period. It is imperative to monitor for the occurrence of supraventricular tachycardia and venous stenosis in patients who have undergone the Senning procedure. In the late postoperative period, there is an elevated risk of deterioration in LV function due to neo-aortic regurgitation.

In the event that the Rastelli procedure is selected for repair, the extracardiac conduit must be replaced in succession at the subsequent operation. Conversely, in the future, transcatheter pulmonary valve implantation (TPVI) may be a potential alternative to the surgical procedure. However, TPVI has not yet been globally approved for use with artificial conduits.

Complications and anatomical variations causing HF

Systemic RV

The morphologic RV of those who have not undergone any surgical repair for ccTGA constitutes a component of systemic circulation by a thin wall with only a two-layered muscular structure, in comparison to a three-layered muscular in the morphologic LV. Such patients frequently develop valvular regurgitation, which serves to exacerbate the symptoms of HF, particularly in instances of Ebstein-

like tricuspid dysplasia, the most common cardiac anomaly associated with ccTGA.

Pulmonary vascular disease

Pulmonary circulation evaluation is paramount in patients presenting with sub-pulmonary RV failure or sub-pulmonary failure of the anatomic LV in ccTGA. Pulmonary arterial hypertension (PAH) can be associated with ACHD due to anatomical specificity. Then, severe characterization would be necessary to focus on specific therapies for PAH. Patients with ccTGA may have elements of both precapillary and postcapillary pulmonary hypertension, most of which are associated with WHO class 2 or 5 (13-15).

HF management and details for ccTGA

The number of ccTGA patients who visit emergently due to HF symptoms is increasing according to their age and survival. The therapeutic plans would be influenced by whether the patient's anatomical feature is associated with a morphologic left or RV in the systemic circulation, and the extent of the impairment of the sub-pulmonary ventricle. HF of ccTGA is often complex by complications due to previous surgeries.

Diagnosis of HF in patients with ccTGA

A comprehensive understanding of the underlying CHD, along with a detailed account of previous surgical procedures and interventions, is of paramount importance in the diagnosis of HF in ccTGA. The onset of arrhythmias is a common initial symptom in spite of a little alteration. There is a necessity for periodic examinations including electrocardiograms (ECG), CT, cardiac magnetic resonance imaging (CMR). In addition, the fundamental elements of diagnosis are the transition of N-terminal pro-B-type natriuretic peptide (NT-proBNP), and cardiopulmonary exercise testing (CPET). The diagnostic problem lies in two cases: symptomatic patients with normal systemic ventricular function and asymptomatic patients with impaired systemic RV function. When confronted with these two situations, a 25% decrease in peak oxygen consumption on CPET or a doubling of NT-proBNP over a 6-month follow-up period indicates progressive systemic RV systolic dysfunction. As such, they prompt the need for prompt treatment initiation, as evidenced by the following references (16-20).

Medical treatment of HF in ccTGA

Although there is no evidence (21), we often use beta-blockers in asymptomatic patients with systemic RV dysfunction. While the management of symptomatic patients is similar to that of patients with morphological LV dysfunction. A notable distinction from existing HF guidelines is the threshold for initiating medical treatment, which is set at an ejection fraction (EF) of less than 40%. The most common approach involves the use of renin-angiotensin-aldosterone system (RAAS) blockers in conjunction with beta-blockers, mineralocorticoid receptor antagonists (MRAs), diuretics, and/or digoxin. In patients with subpulmonary morphological LV dysfunction, the concomitant diuretics are used only if they show any symptoms.

Treatment of acute heart failure (AHF) with ACHD

Today, we have no therapeutic standard for those with ACHD and AHF. Therefore, they are treated according to the general HF, giving weight to the presence of complications and anatomical variants.

Implantable cardioverter-defibrillator (ICD) implantation for ccTGA

Most data resulted from observational studies of limited experts due to the lack of evidence for the implantation of ICD for ACHD (22,23). Thus, novel studies are focusing on prospectively assessing preventative ICD implantation in ACHD (24). Generally, ACHD patients can undergo ICD implantation as primary prevention based on the same indications for ICD in the latest European Society of Cardiology (ESC) guidelines (25). It is already reported that those who have not undergone any surgical repair for ccTGA tend to suffer from ventricular arrhythmias. So, they often undergo the implantation of ICD as secondary prevention.

There is a consensus in guidelines that ACHD patients who meet the latest ICD criteria of left ventricular ejection fraction (LVEF) <35%, biventricular physiology, HF treated properly, and NYHA symptom class II or III are candidates for ICD implantation as primary prevention. But, when focused on the benefits of transvenous pacing

lead, it still has been difficult to select patients with systemic RV appropriately due to the risks of infection and venous occlusion under the anatomical complex.

CRT implantation for ccTGA

The validity of CRT implantation for ACHD depends on the basic disease itself, anatomical features, and whether the patient is under bi-ventricle or single-ventricle hemodynamic. Although most of the data reported in ACHD resulted from observational studies, CRT implantation is indicated as a class I therapy for those with a systemic morphologic LV when they meet the same criteria as non-ACHD patients (25,26). But, even for those who have systemic morphologic RV, such as ccTGA, CRT could be indicated when they meet the criteria, including EF <35%, dilated ventricles, NYHA class II to IV, and complete right bundle branch block with a QRS complex ≥150 msec.

VADs: special considerations in patients with ccTGA

Considering the rising number of patients with ACHD and the increase in HF in this population, there is a need to evaluate additional treatment options. Among the novel therapies developed for HF, durable mechanical circulatory support in the form of VADs can be a bridge to transplantation. These devices have been shown to decrease mortality in end-stage HF among non-transplant candidates and appear to offer a similar benefit to transplant candidates who are deteriorating while awaiting transplantation (27).

Transcatheter tricuspid valve repair (TTVR)

Percutaneous tricuspid interventions are gradually providing alternative options to conventional surgical procedures for high-risk patients. Among these, edge-to-edge TTVR represents the strategy with the highest global penetration. The compassionate use of MitraClip devices (Abbott Medical, USA) in a tricuspid position was initially proposed, and the results were promising. Nevertheless, the difficulties in achieving the correct catheter height or optimal coaxiality represented significant limitations for the conventional mitral system. The TRILUMINATE trial was the first to assess the performance of a dedicated edge-to-edge TTVR system called TriClip (Abbott Medical). Notwithstanding the utilisation of solely NT devices (of abbreviated length), TriClip exhibited a markedly elevated success rate for the procedure, accompanied by an exceptionally low incidence of complications. The novel TriClip system was made commercially available in Europe in June 2020. The devices are available in two sizes, XT and NT. This article presents the initial experience of the system in Spain. It is conceivable that the TriClip system could be employed in conjunction with ccTGA and, moreover, even following VAD implantation (28,29).

Heart transplantation in patients with ccTGA

The number of heart transplantations for patients with ACHD is gradually increasing. However, outcomes after heart transplantation are significantly worse than those of heart transplantations for patients without CHD, especially in the context of multi-system disorder.

Patients with ACHD are frequently excluded from trials assessing HF therapies. Consequently, there is an urgent need for more data on HF trajectories in ACHD to inform the management of those with ACHD and HF and post-transplantation trajectories with regards to CHD subgroup (such as ccTGA) and disease complexity.

Clinical cases to illustrate the complexity in diagnosis as well as advanced HF treatment in ccTGA

Clinical case 1: incidental diagnosis of ccTGA due to a suspicion of torrential "mitral" regurgitation

A 68-year-old female was admitted to the hospital with worsening palpitations, shortness of breath and the suspicion of severe mitral regurgitation for evaluation of a surgical valve repair or replacement. Transthoracic echocardiography (TTE) showed suggestive findings of ccTGA, with severe regurgitation of the systemic tricuspid AV-valve and a systemic RV with moderately reduced function (*Figure 1A*,*1B*). TOE (*Figure 1C*,*1D*) and MRI confirmed the diagnosis of ccTGA with the aorta in the left anterior position, worsening systemic RV function and significant regurgitation of the systemic tricuspid AV-valve. After interdisciplinary discussion, the indication for surgical valve repair was prompted.

Preoperative CT scans showed signs of coronary artery atherosclerosis. Coronary angiography revealed a stenosis of the proximal circumflex artery. Coronary artery bypass surgery (CABG) with venous graft from the ascending

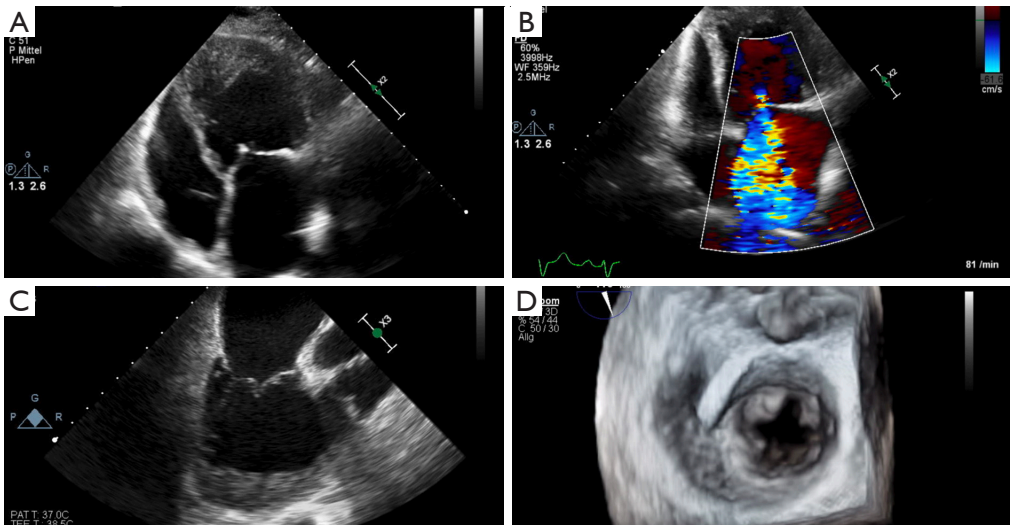

Figure 1 Echocardiographic findings in Case Report 1. (A) Apical four-chamber view showing the hyper trabeculated systemic RV. (B) Transesophageal 3-chamber view depicting the hyper trabeculated systemic RV in subaortic position. (C) Apical four-chamber view with colour Doppler revealing severe regurgitation of the systemic tricuspid AV-valve. (D) Transoesophageal echocardiography with a 3-dimensional view on the systemic tricuspid AV-valve. AV, atrio-ventricular; RV, right ventricle.

aorta to the third major RV branch of the right coronary artery (RCA) and tricuspid valve replacement with a 32 mm Hancock biological valve prosthesis were performed. In the five-year follow-up, the patient's condition showed improvement, with echocardiography revealing stable findings with mild regurgitation of the systemic tricuspid AV-valve after replacement and a mildly reduced systemic RV function. However, palpitations remained a persistent issue, and no further atrial fibrillation could be identified.

Clinic case 2: end-stage HF management in ccTGA—considerations in VAD implantation and heart transplantation

A 35-year-old male patient was admitted with recurrent ventricular tachycardia (VT) and protein losing enteropathy (PLE) attributed to a worsening systemic RV function. The underlying defect was diagnosed as a complex ccTGA necessitating seven previous cardiac surgeries. The initial surgery was a tricuspid valve repair (TVR) at two years due to moderate tricuspid regurgitation, followed by a pacemaker implantation for complete atrioventricular block three months later. Fourteen years after the initial procedure secondary TVR in the setting of recurrent regurgitation was performed.

In the following years, the patient remained stable in NYHA functional class (FC) II. At 35 years of age, the patient presented with recurrent VT and PLE. Right heart catheterization (RHC) revealed severe RV failure with a central venous pressure up to 21 mmHg.

HF and arrhythmia drug treatment was reinforced with amiodarone, additional diuretics, milrinone, and dobutamine. The cardiac function remained unimproved, and he was listed for cardiac transplantation. He underwent HeartMate3™ for anatomical RV failure. Three years later he underwent heart transplantation at the age of 38 years. Previous mediastinal surgeries resulted in major scarring and a prolonged operation time. Weaning trials in the postoperative period led to a tracheotomy.

The patient's persistent PLE and low renal functional reserve resulted in weaning challenges from continuous hemofiltration postoperatively. He fell into sepsis due to multiple infections, including gallbladder perforation and cellulitis of the right upper limb. The patient succumbed to septic shock seven months after heart transplantation.

Conclusions

Due to advancements in diagnostic techniques and an increased recognition of the complexities associated with CHD, coupled with the availability of comprehensive prenatal assessments, the timing of diagnosis in patients

with ccTGA has witnessed a notable improvement over time. A considerable number of ccTGA patients, particularly those with balanced hemodynamics, remain undetected and present therapeutic challenges when diagnosed at a later stage of life. Optimal timing of surgery and interventional treatment approaches, the effect of pharmacological treatment in the context of HF, as well as the timing of initiation of a mechanical circulatory support, VAD and heart transplantation, are based on individualised consensus-level decisions. Optimal management remains a topic of debate due to the scarcity of outcome data. Future investigations should focus on identifying surrogate parameters for guiding treatment.

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Footnote

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