Congenitally corrected transposition: not correct at all

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Purpose of review
Congenitally corrected transposition of the great arteries is a rare congenital defect with several management options. Disagreement continues on strategies, such as anatomic repair, physiologic repair or observation-only. This review discusses recent data that provide further guidance for clinical decision-making.

Recent findings
New data provide greater insights into practice patterns and outcomes. Recent data from high-volume centers show progressively high rates of systemic right ventricle dysfunction over time with lower rates of systemic left ventricle dysfunction following anatomic repair; there is a statistical trend towards better survival of anatomic repair patients. Data comparing anatomic repair to observation showed that anatomic repair patients had a lower hazard of reaching a composite adverse outcome. These complex operations are predominantly performed at a small subset of congenital heart surgery centers.

Summary
Anatomic repair compared with physiologic repair may have better outcomes, although there are relatively high rates of morbidity for both approaches. In the patient without associated lesions, nonsurgical management can have excellent outcomes but is complicated by right ventricular failure over time. Multicenter research will help determine risk factors for bad outcomes; management at high volume, experienced centers will probably be beneficial for this complex patient population.

Keywords
complex congenital heart disease, congenitally corrected transposition of the great arteries, systemic right ventricle

INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare form of congenital heart disease first described by Von Rokitanski in 1875 [1]. It is characterized by atrioventricular (AV) and ventriculoarterial discordance, resulting in the morphologic right ventricle (mRV), supporting the systemic circulation and the morphologic left ventricle (mLV), supporting the pulmonary circulation. ccTGA is rare, with an incidence of 1 of 33 000 live births, accounting for 0.05% of congenital heart disease. It is associated with other abnormalities in approximately 85–90% of cases [2]. Postmortem studies of ccTGA described associated pulmonary obstruction (30–79%), ventricular septal defect (VSD) (70–88%), dextrocardia (20–37%), and systemic AV valve abnormalities (20–38%), such as Ebsteinoid tricuspid valve or straddling tricuspid valve [2–4]. The conduction system varies in ccTGA depending on atrial situs; development of complete heart block is common.

The ideal management of the patient with ccTGA is controversial. Surgical management can be dichotomized into 'physiological repair', in which associated lesions such as VSDs and valve abnormalities are addressed, leaving the mRV as the systemic ventricle, or 'anatomic repair', in which the circulation is rearranged so that the mLV becomes the systemic ventricle. Some patients with no associated lesions are managed expectantly without surgery. Patients can also follow a single ventricle pathway culminating in Fontan surgery,

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Curr Opin Cardiol 2023, 38:000–000

DOI:10.1097/HCO.0000000000001052

KEY POINTS

- Decision making for management of congenitally corrected transposition of the great arteries is complex, with risks of morbidity and mortality associated with all options.
- The anatomic repair, which results in making the left ventricle the systemic ventricle, has lower risk of systemic ventricle dysfunction and trends toward lower long-term mortality compared with the physiological repair.
- Congenitally corrected transposition of the great arteries has a high risk of complete heart block and of pacemaker-induced ventricular dysfunction; newer pacemaking strategies may lead to fewer complications.
- Highly complex congenital heart surgery such as the double switch operation or atrial switch/Rastelli operation is best performed at high-volume congenital heart centers with high levels of expertise in the surgical and postoperative management of patients with congenitally corrected transposition of the great arteries.

which will not be treated at length in this review. In a young child presenting with ccTGA without associated lesions, the decision to operate versus expectant management is difficult. The low incidence of this lesion has limited the availability of outcome-based data. However, recent publications have better defined differential outcomes of these management strategies. Pacemaker management in the population that develops heart block is also complex and complicated by pacemaker-induced ventricular dysfunction, with newer strategies emerging. Surgical and postoperative management of this patient population is highly complex and requires an expert and experienced team of surgeons, cardiologists and anesthesiologists typically found at high-volume congenital heart centers.

NATURAL HISTORY

The natural history of unrepaired ccTGA is largely determined by the function of the systemic mRV, tricuspid valve regurgitation, and presence or absence of associated lesions. Multiple studies have demonstrated that associated defects in combination with poor systemic ventricular function and systemic AV valve regurgitation are associated with poorer outcomes [2–5]. Development of systemic mRV dysfunction is common and often accompanied by tricuspid regurgitation. Cui *et al.* [6▪▪] found a 37% incidence of moderate or severe systemic mRV dysfunction in follow-up of nonoperated patients. An earlier study of 44 patients with unrepaired ccTGA presenting after 18 years of age showed moderate-to severe tricuspid valve regurgitation in 59% of those with depressed mRV function at the time of diagnosis [5]. Graham *et al.* [2] showed that by 45 years of age, 67% of patients with ccTGA and associated lesions had heart failure symptoms, concluding that mRV function was maintained in childhood (<10 years) but mRV dysfunction and subsequent heart failure symptoms became more common after 17 years of age, suggesting inability of the mRV to sustain systemic cardiac output over a normal life span. Heart block is also common. A multicenter, retrospective observational review of 30 fetuses with ccTGA showed the risk of congenital complete heart block to be 5–7%, usually occurring in the third trimester [3]. Progression to de novo AV block is 1.3–2% per year [3,7], with recent data from Barrios *et al.* [8▪▪] describing an incidence of non-iatrogenic complete heart block of 29.8%.

SURGICAL MANAGEMENT

Options for surgical management of ccTGA include physiologic repair procedures, anatomic repair procedures, pulmonary artery banding, or single ventricle palliation. Discussion of single ventricle palliation is outside the scope of this review.

Physiologic repair

Surgical management of ccTGA was historically focused on correction of the physiologic abnormalities occurring from associated cardiac defects, most importantly VSDs, LVOT (subpulmonary) obstruction, and tricuspid valve abnormalities. This approach, first described by Anderson *et al.* [9] and later refined by de Leval *et al.* [10], includes VSD closure through either a right atrial (transmitral), transaortic, or left ventriculotomy approach. The suturing technique for VSD closure in this lesion is unique, owing to the displacement of the atrioventricular bundle to the right side (mLV side) of the ventricular septum along the anterior and superior border of the VSD. In this area, sutures are placed into the mRV side of the septum, working somewhat blindly through the VSD. Placement of an LV to pulmonary artery conduit is often used to address LVOT obstruction, because the presence of the right-sided coronary artery in the AV groove prohibits transannular patch repair.

Anatomic repair

The surgical approach to ccTGA has evolved in recent years towards anatomic repair procedures,

as initially described by Ilbawi in 1990 [11], whereby the mRV is removed from the systemic circulation. These repair procedures consist of either a Senning-type or Mustard-type atrial switch with an arterial switch with or without VSD closure ('double switch'), or in the presence of important LVOT obstruction, an atrial switch with an RV to PA conduit and VSD closure (Senning with Rastelli, Nikaidoh, or REV procedure). As these atrial-baffling procedures are better performed after infancy, many patients planned for anatomic repair undergo initial pulmonary artery banding to maintain or retrain the mLV for systemic workloads [6▪▪,8▪▪].

Pulmonary artery banding

Pulmonary artery banding in ccTGA may be used for mLV maintenance or retraining prior to anatomic repair, or as definitive palliation. Some centers suggest prophylactic pulmonary artery banding at diagnosis, whereas others recommend banding for patients with mLV pressure below a certain threshold (<70–75% systemic). Regardless of the approach, the age at banding is associated with ultimate mLV dysfunction and late mortality, with the greatest preservation of mLV function seen when banding is performed prior to 2 years of age [12,13]. Primarily by affecting ventricular septal interactions, pulmonary artery banding may also help to stabilize mRV, tricuspid valve, and aortic valve function, and as a result, some centers espouse banding as a definitive palliation strategy, although long-term outcomes of this approach have been mixed.

Challenges of surgical care

Ongoing surgical innovation is likely to benefit patients with ccTGA. Three-dimensional modeling is becoming increasingly utilized for preoperative planning in congenital cardiac surgery, especially for complex intracardiac baffling procedures as is often required for ccTGA repair [14]. Intraoperative conduction mapping allows real-time identification of the conduction areas in complex cardiac anatomy, and may help to reduce the risk of iatrogenic heart block [15▪].

OUTCOMES

Optimal surgical management of ccTGA remains controversial and is complicated by the diverse spectrum of associated lesions in an already rare disease state (Table 1). Recent data suggest that if presenting at less than 10 years of age, anatomic repair may be associated with improved survival [8▪▪] or with improved freedom from death, transplant, systemic ventricular dysfunction or AV valve regurgitation [6▪▪]. Although these studies are relatively large, there are still power limitations for detecting statistically significant differences in outcomes between management groups.

A retrospective review of 240 patients diagnosed with ccTGA by Barrios *et al.* [8▪▪] found that patients with physiologic repair had equivalent early and mid-term survival to those having anatomic repair, but the transplant-free survival curves diverged around 12 years after repair (71 vs. 80% at 15 years, $P = 0.09$). This statistical trend towards worse long-term survival with physiologic repair supports previous data that that the mRV is unable to sustain systemic cardiac output in the long-term [2,4,5]. Furthermore, this patient population was found to have significant risk for reoperations, most commonly on the tricuspid valve. Long-term survival in anatomic repair was comparable to those patients with expectant management in these data. However, only 27% of the expectant management cohort remained classified that way at 20 years

Table 1. Long-term outcomes by management strategy

	Expectant management	Physiologic repair	Double switch operation	Atrial switch/ Rastelli
Surgical mortality[a]	N/A	2.3%[b]	3.7%[b]	8.4%[b]
Surgical complexity	N/A	+	++	++
Need for future intervention	+++	++	+	+
Late systemic ventricular dysfunction	++	+++	+	+
Late systemic atrioventricular valve regurgitation	++	++	+	+
Risk of heart block	+	+++	++	+++

[a]Percentages from Chew *et al.* [18▪].
[b]Mortality rate of Atrial Switch/Rastelli significantly higher than physiologic repair or double switch. No statistical significance between physiologic repair and double switch.

follow-up, whereas the majority had progressed to the need for physiological repair.

Another recent study of 128 patients diagnosed with ccTGA with intact ventricular septum over 39 years by Cui *et al.* [6▪▪] found that double switch operation with or without mLV training from pulmonary artery banding was associated with decreased hazard of reaching a composite endpoint including death, requiring a transplant, having severe systemic ventricular dysfunction or significant systemic AV valve regurgitation (hazard ratio 0.29, P value = 0.039). In the subgroup of patients who presented before the age of 5, there was a higher magnitude difference between anatomic repair and no anatomic repair with a hazard ratio of 0.16, P value = 0.008.

Earlier anatomic repair may provide long-term benefits towards preservation of mLV function [6▪▪,8▪▪]. This suggests there is a window of opportunity to perform an anatomic repair in patients who do not have systemic mLV pressure because of LV myocyte remodeling and inability for the mLV to be retrained adequately. Older age at the time of repair has been associated with increased risk of death or transplant [6▪▪,16▪]. However, the absolute age ceiling and optimal age for complete repair are unknown.

These data suggest that if a patient undergoes surgery at a young age, anatomic repair is superior to physiologic repair. Practice patterns at large centers have shifted towards anatomic repair, but this remains controversial in practice because of perceived higher levels of surgical morbidity and mortality associated with this approach. The double switch operation can be performed in experienced centers with low operative mortality and encouraging long-term survival [6▪▪,8▪▪,17]. In a recent analysis of the STS database by Chew *et al.* [18▪], surgical mortality was 3.7% for double switch operation with or without VSD repair, and 8.4% for atrial switch plus Rastelli. The same database showed a 2.3% surgical mortality rate for physiologic repair.

Ventricular dysfunction

Although the data from Cui *et al.* show that mRV dysfunction at presentation is associated with worse outcomes in nonintervention patients, surgical versus expectant management for the patient who presents at a young age with ccTGA without significant associated lesions and without systemic mRV dysfunction remains challenging, without a clear path delineated by available data. Ultimately, systemic ventricular dysfunction is a problem with any of these approaches but is more common with time in the systemic mRV (Fig. 1). With expectant management, systemic mRV dysfunction has a reported incidence of 50% by 30 years of age in patients with

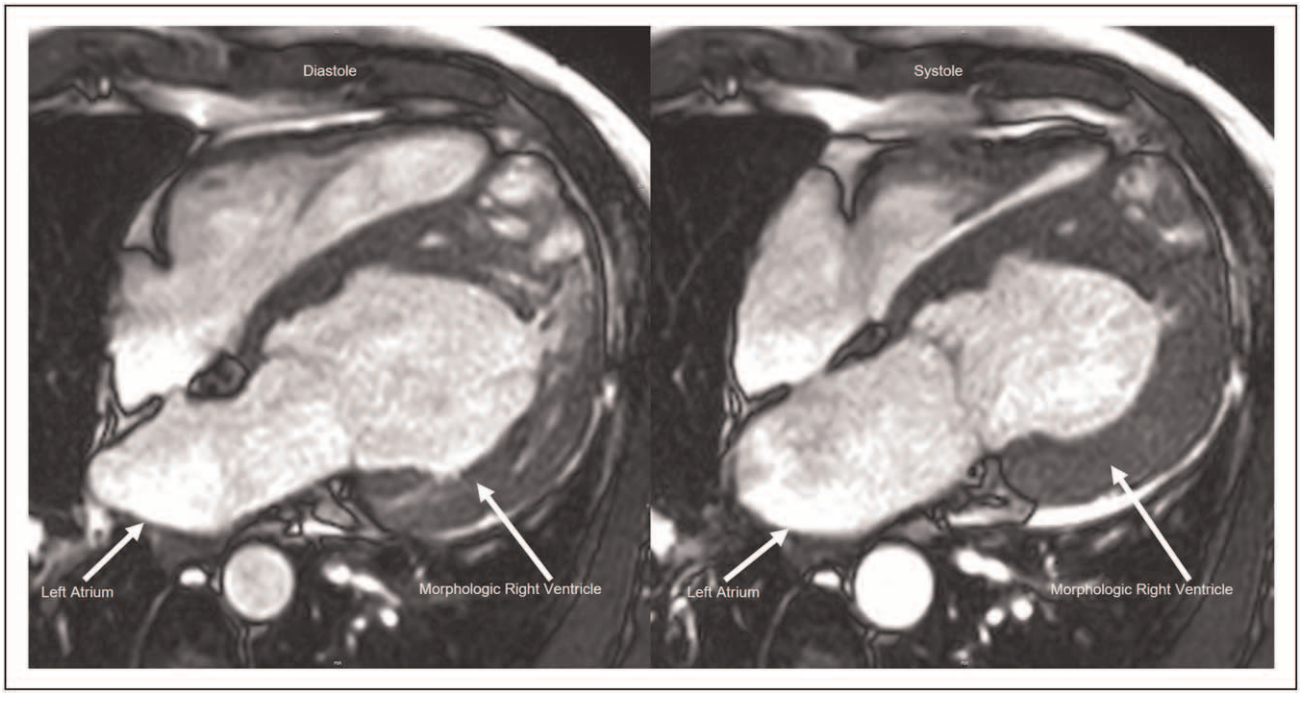

FIGURE 1. Dilated systemic right ventricle with dysfunction. Still images at end diastole and end systole from cardiac MRI steady-state free precession imaging of a 37-year-old with ccTGA with no associated lesions and no cardiac surgical history. Note the severely dilated and hypertrophied systemic, morphologic right ventricle on the left side of the chest, with little area change between diastole and systole compared with the right-sided, morphologic left ventricle.

an intact ventricular septum, whereas systemic left ventricular dysfunction following double switch at last echo was 14% in the same study [6▪▪]. In addition, physiologic repair may result in a higher risk of systemic mRV dysfunction compared with expectant management [19]. In the Barrios study, 10 years post physiological repair, 84% of patients had mRV dysfunction versus 28% of patients with mLV dysfunction 10 years post anatomic repair [8▪▪].

Heart center size

Complex repairs of congenital heart disease can be performed with lower mortality at centers with high surgical volume. This has been shown using the STS database [20] and the European Congenital Heart Surgeons Association database [21]. Although this has not been demonstrated specifically for ccTGA, it is reasonable to infer that anatomic repair fits this pattern. Especially considering the decline in the use of atrial switch procedures for d-looped TGA, many centers and surgeons have unfamiliarity with the complex atrial baffling required to effectively perform an anatomic repair. Postoperative physiology is highly complex, with nearly ubiquitous ventricular systolic and diastolic dysfunction and high risk for complications requiring expert cardiac intensive care management [22,23]. More recent data from the STS database published on outcomes of 985 ccTGA patients at 101 centers showed the six highest volume centers accounted for 41% of all the operations. Only 26 of the centers performed more than 10 operations in this series [18▪]. High-volume, comprehensive congenital heart centers include surgical expertise as well as experienced perfusionists, pediatric cardiac anesthesiologists, cardiac critical care and imaging teams, all of whom are essential in a team approach for achieving the best outcomes.

Heart block and arrhythmia

In addition to the abnormal conduction system and risk of de novo AV block, iatrogenic AV block is common. A single-center evaluation of adults with ccTGA describes high-grade AV block in 8% of patients at presentation and that subsequent pacemaker insertion was necessary in 48% of patients after intervention [24]. The recent analysis of the STS database found the rate of arrhythmia requiring permanent pacemaker in both anatomic and physiologic repairs of 12.7–19%, which was the highest in the setting of VSD closure [18▪].

Need for pacemaker is an important contributor to morbidity in this population, with studies showing that pacing can result in worse systemic ventricular dysfunction [7,16▪,25,26]. Primary cardiac resynchronization therapy (CRT) or biventricular pacing is associated with less systemic ventricular dysfunction when compared with single subpulmonary ventricular lead pacing [7,16▪]. CRT in systemic mRV patients with previous traditional pacing results in improved QRS duration and New York Heart Association functional class [27▪]. However, the response to biventricular pacing is variable, and the benefit of CRT in the setting of a narrow QRS is not yet established. In a European multicenter study, CRT was assessed in 109 pediatric and CHD patients that included 30 patients with ccTGA and found that patients with a systemic mLV were more likely to have improved ventricular function or decreased systemic AV valve regurgitation after CRT than patients with a systemic mRV [25]. Conventional CRT can be technically difficult because of the high incidence of coronary sinus venous drainage abnormalities (~20%). His bundle, Purkinje or proximal left bundle branch pacing is feasible because of the unique location of the conduction system in ccTGA, resulting in a narrowed paced QRS [26]. However, additional studies of this pacing strategy and comparison with conventional CRT are needed.

Late development of atrial arrhythmias has not been widely reported in the ccTGA population, but this may reflect the relatively short follow-up. Late development of both sinus node dysfunction and atrial arrhythmias is a hallmark after atrial baffle surgery in the atrial switch population with up to 48% incidence of sinus node dysfunction and up to 35% of intra-atrial re-entrant tachycardia at 20 year follow-up [28,29]. One can surmise that the cause of atrial arrhythmias secondary to atrial scarring would infer an increased risk following the double switch operation. Further investigation of atrial arrhythmia burden in the ccTGA population is needed.

CONCLUSION

ccTGA is a complex lesion with no risk-free management strategy. As such, the long-term morbidity and mortality of the anatomic repair should be considered in the context of high rates of systemic mRV failure with expectant management and physiologic repair, rather than comparing with a hypothetical ideal that is more realistic with simpler forms of congenital heart disease. New data show that anatomic repair, especially the double switch operation, can be performed with relatively low short-term risks, and compared with physiologic repair has lower rates of systemic ventricular failure and at least trends to better long-term mortality. In infants presenting with ccTGA with no associated lesions, anatomic repair is preferable to expectant management in the context of systemic mRV dysfunction.

In the absence of mRV dysfunction, available data do not point to a clear management decision and further research is needed, preferably from multicenter, prospective databases. Large databases in the United States and Europe show that complex CHD outcomes are best at high-volume surgical centers, and this is likely the case for the challenging surgical and postoperative management necessary for this specific population of patients.

Acknowledgements

None.

Financial support and sponsorship

No relevant sources of funding to this article. A.L.D. is supported in part by the FORCE registry, funded by Additional Ventures through a consortium with Boston Children's Hospital.

Conflicts of interest

There are no conflicts of interest.

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