Received: 18 October 2017 | Revised: 12 January 2018 | Accepted: 23 January 2018

DOI: 10.1111/chd.12595

ORIGINAL ARTICLE

WILEY Congenital Heart Disease

Rhythm disturbances and treatment strategies in children with congenitally corrected transposition of the great arteries

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Abstract

Background: We aimed to evaluate rhythm abnormalities in cases of congenitally corrected transposition of the great arteries (ccTGA) and associated treatment strategies.

Patients and Methods: This retrospective cohort study included 65 pediatric patients with ccTGA who were admitted to the clinic between 2009 and 2017. The patients were divided into two groups, and surgical data, Holter electrocardiographic (ECG) recordings, ECG recordings, electrophysiological data, and device implantation data on the two groups were compared.

Results: Group I (n = 53, 82%) consisted of patients with significant associated lesions, and Group II (n = 12, 18%) consisted of those with minor or no associated lesions (isolated ccTGA). Rhythm abnormalities were diagnosed in 22 (34%) of the patients based on initial ECG findings and Holter ECG recordings. Eleven (17%) of these patients had atrioventricular (AV) block of different degrees, and the other 11 (17%) had supraventricular arrhythmia (SVA). The median follow-up was 49 months (range, 9–89 months), and the rhythm remained normal in 26 (42%) of the patients. Three patients died on follow-up. Of 40 patients with normal initial findings, nine required pacemaker implantation due to complete heart block, and SVA developed in seven patients on follow-up. No ventricular tachycardia was seen initially or on follow-up. Ablation was performed in four patients. During the follow-up period, pacemakers were implanted in 12 (23%) of patients in Group I and 4 (33%) of patients in Group II due to complete heart block. Cardiac resynchronization therapy (CRT) was performed in four patients due to systemic ventricular dysfunction. Notably, all four of these patients had a pacemaker implanted postoperatively.

KEYWORDS
arrhythmia, children, congenitally corrected transposition of the great arteries, rhythm disturbances

1 | INTRODUCTION

Congenitally corrected transposition of the great arteries (ccTGA) is a rare pathology, which accounts for less than 1–1.4% of all congenital heart defects.[1–4] Although frequently associated with other cardiac defects, such as ventricular septal defects (VSDs), pulmonary stenosis (PS), and anomalies of the tricuspid valve, it can occur in isolation,[5] Previous studies revealed that spontaneous complete heart block was present from birth in around 4% of patients and that progressive deterioration of the condition was common.[6,7] The presence and severity of associated lesions in ccTGA and conduction disturbances alter the timing of the clinical presentation of the disease.[7]

There are a limited number of studies of rhythm disturbances and their management in children with ccTGA.[6–8] The aim of the present study was to investigate rhythm and conduction disturbances in a relatively large group of pediatric ccTGA patients from a single center and associated treatment strategies.

2 | PATIENTS AND METHODS

Since arrhythmia may develop over time, all ccTGA patients who attended our arrhythmia clinic between November 2009 and February 2017 were routinely followed up, irrespective of whether they had arrhythmia at the time of admission. In this retrospective cohort study, data on 65 pediatric ccTGA patients were recorded.

The diagnosis of ccTGA was done by transthoracic echocardiography. Regarding anatomical arrangements, ccTGA was characterized by situs solitus, with L-looped ventricles and an anterior leftward aorta (S,L,L) or situs inversus, with D-looped ventricles and an anterior rightward aorta (I,D,D). There were 58 patients in the S,L,L group (89% of patients) and 7 patients in the I,D,D group (11% of patients). The patients were divided into two groups. Group I was composed of patients with significant associated lesions and included patients with any of the following lesions: a large ventricular septal defect (VSD), moderate or severe pulmonary stenosis (PS), pulmonary atresia (PA), moderate or severe tricuspid regurgitation, or an Ebstein-like anomaly of the tricuspid valve. Group II (isolated ccTGA) consisted of patients with minor or no associated lesions. There were multiple pathologies in some patients. Ten patients had mild systemic AV valve regurgitation, two patients had mild pulmonary stenosis, and three patients had small VSD.

The demographic data, 12-lead electrocardiography (ECG) data, and Holter ECG recordings were examined. The ECG and Holter ECG recordings of nonoperated patients who were followed up every 6 months were evaluated, as well as readings obtained in the postoperative 1st week, 1st month, 3rd month, 6th month, and every 6 months of follow-up in the operated patients. The recordings were classified as normal sinus rhythm, conduction abnormalities (first-degree atrioventricular block [AVB], 2:1 AVB, or complete AVB), supraventricular arrhythmias (Wolff–Parkinson–White [WPW] syndrome, ectopic atrial rhythm [EAR], premature atrial beats, or focal atrial tachycardia [FAT]), and ventricular arrhythmias. All surgical-related information, ablations, pacemaker insertions, cardiac resynchronization therapy (CRT) data, and mortality during follow-up were recorded.

2.1 | Statistical analysis

Statistical analyses were performed using SPSS version 15.0 for Windows (SPSS Inc., Chicago, Illinois). The Mann–Whitney *U* test was used to compare the mean values of the two groups, and a chi-square and Fisher's exact test were used to compare the findings between the groups. A *P* value of $< .05$ was considered statistically significant.

3 | RESULTS

3.1 | Patient characteristics

In total, 65 patients who were followed up at regular intervals at our arrhythmia clinic and had initial Holter ECG recordings and 12-lead ECG recordings were included in the study. Fifty-three of the 65 (81.5%) patients had significant associated lesions (Group I), and the remaining 12 (18.5%) patients had isolated ccTGA (Group II). Of the 65 patients, 40 (61%) were males. The median age at admission to the arrhythmia clinic was 30 months (range, 3 days to 16.5 years). The median body weight was 13 kg (range, 3–78 kg). Six (9%) of the patients had a univentricular physiology. The atrial situs was solitus in 58 (89%) patients and inversus in 7 (11%) patients (Table 1).

3.2 | Findings of initial ECG and Holter ECG recordings

In the initial ECG and Holter ECG recordings, the cardiac rhythms were normal sinus rhythm in 43 (66%) patients. Thirty-seven (72%) patients in Group I and 6 (50%) patients in Group II had a normal sinus rhythm, with no findings of arrhythmia. The number of patients without arrhythmia was significantly higher in Group I ($P < .05$).

Rhythm and conduction abnormalities were detected in 22 of the 65 (34%) patients in the initial ECG evaluation and/or Holter ECG recordings. Eleven of these 22 patients had conduction anomalies: first-degree AVB (n = 1), second-degree AVB (Mobitz type I AV block, n = 1), and complete AVB (n = 9). The incidence of conduction anomalies was significantly higher in Group I (n = 6) than Group II (n = 5) ($P < .05$).

The initial ECG and Holter ECG recordings revealed supraventricular arrhythmias in 10 (19%) patients in Group I and 1 (8%) patient in Group II. Of these 11 patients, nine had a normal sinus rhythm on the initial ECGs but supraventricular arrhythmias according to the Holter ECG recordings. Among these 11 patients, the diagnoses were as follows: WPW syndrome (n = 2), EAR (n = 6), FAT (n = 1), and premature atrial beats (n = 2). Although the incidence of supraventricular arrhythmias was higher in Group I than Group II, the difference did not reach statistical significance ($P > .05$). No ventricular arrhythmia was detected in the initial evaluation in either group.

3.3 | Angiographic data and surgical procedures

Thirty of the 65 patients underwent diagnostic or interventional cardiac catheterization. Complete AVB developed in one patient after cardiac catheterization during the preoperative period before bidirectional cavopulmonary anastomosis surgery was performed.

Surgery was performed in 29 (44%) of the 65 patients, all of whom belonged to Group I (55% of Group I patients). Physiologic repair involves correction of only the associated cardiac defects, leaving the morphologic right ventricle as the systemic ventricle. Anatomic repair consists of redirection of blood flow at the atrial level with a Senning or Mustard procedure, combined with either an arterial switch or a Rastelli procedure in order to place the morphologic left ventricle in the systemic position. Twenty-two of the 29 had surgery for biventricular circulation. Of these patients, physiological correction was performed in nine patients, anatomical correction was performed in three patients, and pulmonary artery banding was performed in nine patients. In two out of the three patients undergoing anatomic repair, the Senning procedure was combined with Rastelli; in the remaining patient, the Senning procedure was combined with an arterial switch

TABLE 1 The demographic variables and echocardiographic findings of the patients with ccTGA

	n = 65
Age, median (range)	
Group I	12 months (3 days–16 years)
Group II	5.5 years (3 months–16.5 years)
Body weight, kg, median (range)	
Group I	12 (3–78)
Group II	25 (6–60)
Gender	
Male	40 (61%)
Female	25 (39%)
Situs	
Solitus	58 (89%)
Inversus	7 (11%)
Cardiax apex	
Levocardia	49 (65%)
Mezocardia	6 (9%)
Dextrocardia	10 (15%)
Type of cardiac physiology	
Single	6 (9%)
Biventricular	59 (91%)
Significant associated lesions	
Yes	53 (82%)
No	12 (18%)
Type of associated lesions	
Ventricular septal defect (large)	42 (64%)
Pulmonary stenosis (moderate or severe)	28 (43%)
Pulmonary atresia	3 (5%)
Ebstein-like anomaly of the tricuspid valve	10 (15%)
Tricuspid regurgitation (moderate or severe)	17 (26%)
Initial ECG findings	
Normal	52 (80%)
Rhythm abnormalities	13 (20%)
Initial Holter findings	
Normal	43 (66%)
Rhythm abnormalities	22 (34%)

procedure. A double switch operation was performed in one patient with a former Blalock–Taussig shunt. In the remaining seven patients, univentricular surgery was performed using the bidirectional cavopulmonary anastomosis procedure. In one patient, univentricular physiology was planned to be corrected using the hemi-Mustard–Rastelli procedure, whereas the others were planned to be directed to the Fontan procedure. A permanent pacemaker was implanted in 8 (28%) of the patients due to postoperative complete AVB. In all patients with permanent pacemaker implantation, complete AVB continued to persist in the last follow-up. In these eight patients, the surgical procedures were Senning–Rastelli in one patient, Senning–Jatene in one patient, VSD closure in four patients, and VSD closure and left ventricle-pulmonary artery conduit placement in two patients.

3.4 | Follow-up

The median follow-up was 49 months (range, 9–89 months). The duration of follow-up was similar in both groups. Three (5%) patients died, all of whom belonged to Group I: a 14-year-old patient with PA who had previously undergone a Senning–Rastelli procedure died after conduit replacement in the early postoperative period due to the low cardiac output; a 5-month-old patient who had undergone VSD closure and arch reconstruction and developed complete postoperative AVB died in the early postoperative period; and a 6-month-old patient with a large VSD and severe pulmonary stenosis died unexpectedly preoperatively. Rhythm and conduction were normal in 26 (42%) of the 62 surviving patients throughout their follow-up.

Among 22 patients with initial pathological Holter ECG and/or ECG evaluations, 11 had conduction abnormalities. Two of these patients had first-degree and second-degree AVB on the initial Holter ECG recordings and did not show any progression during follow-up evaluations. Seven had complete AVB and had a pacemaker implanted. Another two patients with complete AVB are still being followed up without pacemaker implantation.

Apart from the three patients who died during the follow-up period, of 40 patients with normal initial evaluations, nine patients developed complete AVB and had a pacemaker implanted on follow-up. Pacemaker implantations were performed in eight patients postoperatively and in one patient after a cardiac catheterization procedure. Among 9 patients with pacemaker implantation the median follow-up duration was 30 months (range, 9–72 months)

The following changes were detected in patients who had initial rhythm abnormalities: Of six patients with EAR, second-degree AVB developed in one patient, and no change was seen in the remaining five patients. Ablations were performed in two patients with WPW syndrome. The ablation procedure was successful in one patient, whereas medical treatment was required in the other patient. Medical treatment was continued in the patient with FAT after an unsuccessful ablation attempt. Two patients with a premature atrial beat did not show any change during the follow-up.

Supraventricular arrhythmia developed in seven patients whose initial ECG and Holter ECG recordings were normal. EAR was diagnosed in four patients, and supraventricular tachycardia (SVT) was diagnosed in two patients. Asymptomatic WPW syndrome was diagnosed and clinically followed up in another patient. One of the patients with SVT had two unsuccessful ablation procedures and followed up with medication treatment.

No cases of ventricular tachycardia were diagnosed in the initial or follow-up evaluations. The rhythm findings of the patients on follow-up are summarized in Figure 1.

In total, four patients (WPW syndrome, n = 2; FAT, n = 1; and SVT, n = 1) had seven ablation procedures. The ablation procedures were successful in the two patients with WPW syndrome, whereas the remaining patients were assigned to drug treatment.

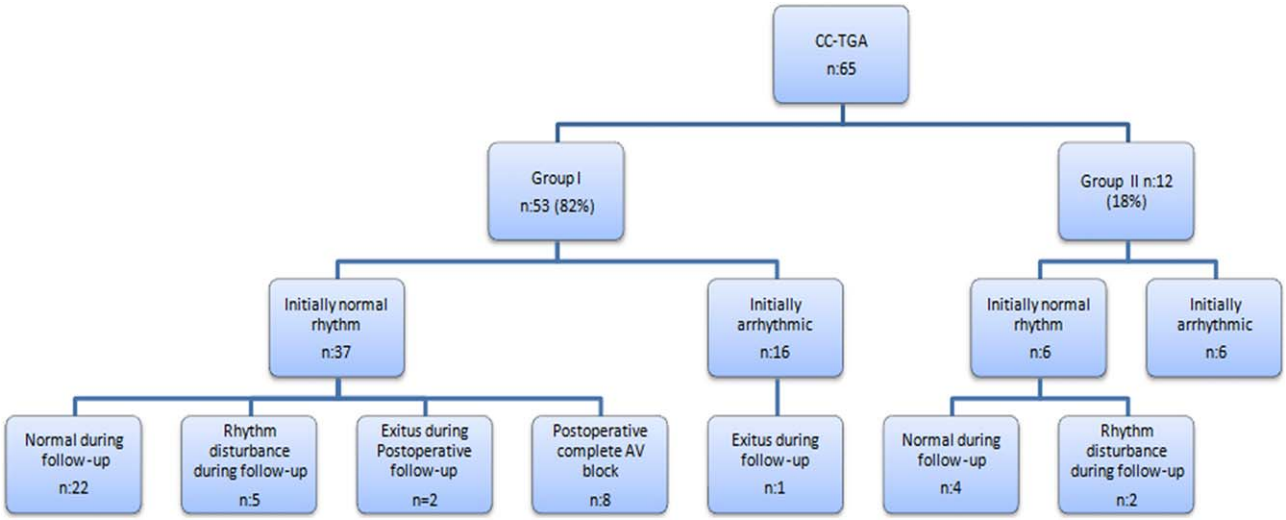

FIGURE 1 The rhythm and conduction features of the study patients

One of the patients with posteroseptal accessory pathway had two recurrences and three ablation procedures, followed up without medication. The other patient had a recurrence followed by a second ablation procedure which was partially successful and he is still on drug treatment. In the patient with FAT, the ablation procedure was accepted as unsuccessful, since we could not stimulate sustained SVT. In the patient with SVT the first ablation attempt was unsuccessful. He died postoperatively while his SVT was under control with drug treatment.

Our success rate for ablation procedure is quite low. It is known that successful posteroseptal ablation rate is quite low, whereas recurrence is quite common in WPW patients. Especially in ccTGA patients with left sided Ebstenoid valve (as in our study), stabilization of catheter and determination of true annulus is very difficult and these factors lower the success rate of the procedure, while increasing recurrence rates.[9,10]

3.5 | Device implantation

Pacemakers were implanted in 12 (23%) patients in Group I and 4 (33%) patients in Group II due to complete AVB. Eight of these pacemakers were implanted in the postoperative period. In all eight cases, VSD closure was part of the surgical procedure (Figure 2).

CRT was performed in four patients who were admitted to the clinic postoperatively due to systemic ventricular dysfunction. One patient had systemic left ventricular failure after a Senning–Rastelli procedure, and the other three patients had systemic right ventricular failure after surgeries for physiological correction. All four cases of CRT involved patients who developed systemic ventricle dysfunction after pacemaker implantation. The preprocedural ejection fractions (EF) of three patients who had benefit after CRT upgrade were 25%, 22%, and 18%, respectively, and the QRS durations on ECG were 192, 144, and 170 msec, respectively. The preprocedural functional classes of the patients were evaluated as New York Heart Association (NYHA) class IV. After the CRT implantation the EF of the patients were 35%, 45%, and 42%, respectively, and the QRS durations on ECG were 140, 128, and 138 ms, respectively. The functional classes were improved to NYHA class II. The preprocedural EF, QRS duration, and functional class of the unresponsive patient were 26%, 155 ms, and NYHA class IV; whereas the postprocedural values were 24%, 158 ms, and class IV, respectively. After CRT implantation, two patients' clinical conditions improved, and they were no longer heart transplant candidates.

4 | DISCUSSION

ccTGA is a rare congenital cardiac pathology, with heterogeneous clinical findings and numerous treatment options. Studies in the literature emphasize the importance of rhythm and conduction pathologies, treatment strategies, and regular follow-up of patients with ccTGA.[6–8] In this study, we evaluated ECG and Holter ECG recordings of ccTGA patients in a pediatric cardiac center.

The results of the study can be summarized as follows:

1. The evaluation of the ECG and Holter ECG recordings revealed that conduction disturbances were much more frequent than reported previously.[6–8]
2. Nearly 20% of the patients with normal initial ECG and Holter ECG recordings developed significant rhythm abnormalities on follow-up.
3. In the present study, we detected almost 10% more conduction abnormality or arrhythmia in Holter ECG recordings compared with ECG. This finding clearly depicts the benefit of Holter ECG recording, even for patients with normal 12 lead ECG.
4. The requirement for a pacemaker was more frequent in isolated ccTGA cases. In addition, the need for a pacemaker increased due to surgery in ccTGA patients with associated lesions. No postoperative AVB occurred after univentricular surgery.
5. Notably, all cases of CRT involved patients who had postoperative pacemaker implantations.

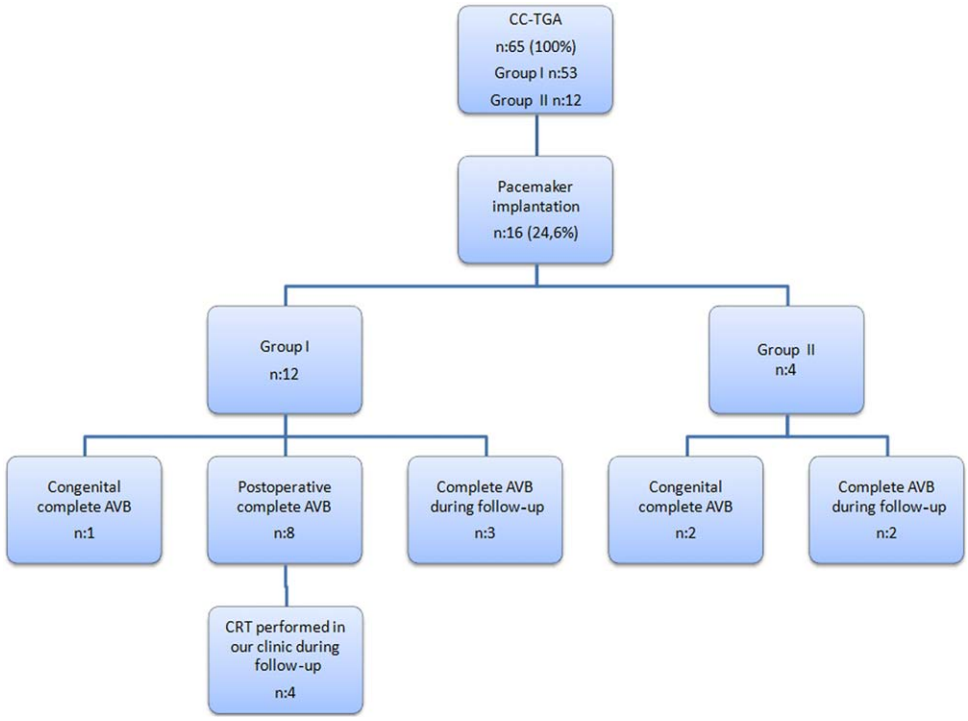

FIGURE 2 The features of the patients with pacemakers

Previous studies reported an estimated 2%-per-year risk of spontaneous heart block in ccTGA patients due to progressive deterioration of conduction,[6] reaching a prevalence of nearly 30% in adulthood.[11] A previous study reported increased sensitivity of the conduction system and a tendency toward the development of fibrosis in ccTGA patients.[12] Due to these factors, spontaneous heart block can develop at any age in ccTGA patients. Pathological studies revealed that not only the looping of ventricles but also the location and course of the AV node and bundle of His are abnormal in the presence of ccTGA. Patients usually have dual AV nodes, with the anomalous second node positioned anteriorly and leading to a long, penetrating bundle, in addition to a sparse conduction system, in which the conduction tissues are frequently vulnerable to trauma.[7,8] Although the probability of an associated anomaly is higher in patients with atrial situs inversus, the frequency of spontaneous heart block is lower due to the normal localization of the conduction system. The percentage of complete AVB is higher in patients without VSDs compared to patients with VSDs.

In the present study, spontaneous complete AVB developed in nine patients during a follow-up period of 49 months. The situs was solitus in all the patients, and six had no VSDs, in accordance with findings in the literature. One patient who developed complete AVB after cardiac catheterization received a pacemaker implant, proving the vulnerability of the conduction system in ccTGA patients.

In ccTGA patients, age has consistently been shown to be a risk factor for developing AVB. Although the patients in Group 2 have a higher incidence of AV block and were considerably older than the patients in Group 1, this difference did not reach statistical significance. We suggested that the most important factor for this finding is probably the younger age of patient groups and the short follow-up period.

A previous study reported that tricuspid valve or VSD surgery can precipitate heart block.[7] A number of studies demonstrated that abnormal ventricular conduction from an aberrant node located at the margin of the right AV valve due to an accessory His bundle coursing anterior to the VSD predisposed ccTGA patients to postoperative AVB.[13–16] In the present study, of the 29 patients who underwent surgery, closure of the ventricular septal defect was performed in 12 patients. Ten of these patients developed postoperative AVB, and two of them died in the early postoperative period before permanent pacemaker implantation. Permanent pacemakers were implanted in the remaining eight patients.

As reported previously, depending on the alterations in the conduction system, there is an equal risk for development of spontaneous complete AVB in both single- and two-ventricle patients.[17] In the present study, none of the single-ventricle patients developed perioperative complete AVB. We believe that surgical interventions for the single-ventricle pathway pose less of a risk to the AV node. The absence of spontaneous AVBs in the single-ventricle patients in the present study might be due to the limited number of patients with this pathology and the relative short duration of the follow-up.

Corrected TGA may be associated with a variety of arrhythmias, including AV reentrant tachycardia, atrial tachyarrhythmia, and ventricular tachycardia.[18] Based on a long period of follow-up, Connelly et al. reported that 36% of patients with ccTGA experienced atrial arrhythmias.[18] In contrast, Kayrak et al. reported that atrial fibrillation or

supraventricular tachycardia was rare in a follow-up of patients with ccTGA.[19] They concluded that were only a few documented cases of ventricular tachycardia in patients with ccTGA. Ventricular arrhythmia might appear on follow-up in ccTGA patients, as ischemia and fibrosis develop after a period of pressure overload on the right ventricle. Previous studies asserted that younger age, together with low incidence of ischemia and fibrosis when compared with that in adults, might explain the absence of ventricular arrhythmia in children with ccTGA.[20,21]

Accessory pathways are present in 2%–5% of patients with ccTGA, especially those with an Ebsteinoid malformation of the systemic tricuspid valve.[22] In a previous study, a sling-like bundle over the anterior margin of the VSD connecting both AV bundles mediated AV reentrant twin AV node tachycardia in some ccTGA patients.[23] Previous studies noted that the ablation of accessory pathways in Ebstein's anomaly or ccTGA can be challenging due to the displacement of AV valves in relation to actual AV grooves.[24–26] They reported an acute success rate of 80% and a recurrence rate of 25%–40%.[24–26] They attributed these success and recurrence rates to several factors, all of which likely affect the success rate of ablation procedures. In the present study, seven ablation procedures were performed in two patients diagnosed with WPW syndrome and two patients with FAT. These were partially successful, and the patients remain under follow-up while receiving medical treatment.

A number of studies showed that CRT improved the functional class in heart failure patients unresponsive to optimal medical therapy.[27–29] In large multicenter studies, an increase in the ejection fraction and an improvement in electromechanical delay were demonstrated after CRT in pediatric patients.[27,28] Janousek and Cecchin et al. evaluated postoperative CRT results of patients with congenital heart disease, including ccTGA patients, and reported that the majority of patients responded positively to CRT therapy.[28,29] In the present study, CRT was performed postoperatively in four ccTGA patients with systemic ventricular failure. One patient with systemic left ventricular failure and two patients with systemic right ventricular failure showed improvements in functional capacity after CRT. The other patient with systemic right ventricular failure did not show any improvement at the end of the 6-month follow-up period. These findings indicate that CRT may be beneficial in patients with ccTGA and systemic ventricular failure.

Although the indications, patient selection, and specifications of responder and nonresponder patients for CRT are known in the adult population, little is known about the effectiveness of CRT in children. Data about pediatric patient selection and clinical outcomes usually relies on retrospective multicenter studies. One of the largest series, published by Janousek et al., included 109 patients from 17 European centers.[28] Over a median follow-up of 7.5 months, they reported an overall improvement in EF of 11.5%, a decrease in QRSd by 30 millisecond, a decrease in systemic ventricular end-diastolic dimension by a median of 1.1. Z scores, and an improvement in NYHA functional class. Interestingly, the majority of these patients (77%) had previous conventional pacemakers before upgrading to CRT. Patients with a systemic LV and prior conventional pacing-induced dyssynchrony showed major clinical improvement and reverse LV remodeling. These findings are consistent with our results confirming that most of the responders were patients with previous pacemaker implantation, and it might be an important predictor for successful CRT implantation.

On the other hand, data about CRT applications in ccTGA patients with systemic RV is even more limited and controversial. Technical feasibility and benefits of CRT in this population were evaluated by Janousek et al. in 2004.[30] Six of the 8 patients (75%) had conventional pacing systems with LV pacing-induced conduction delay and prolonged QRS duration. CRT was achieved by atrial synchronous simultaneous biventricular pacing, resulting in decreased QRSd, interventricular mechanical delay, and improvement in RV function. However, results in subsequent studies demonstrated mixed conflict results for patients with systemic RVs. The multicenter study by Dubin et al.[27] included 17 patients with systemic RVs. Twelve patients had a significant improvement in systemic RV EF, with a decrease in QRSd and clinical improvement. In contrast, Cecchin et al. [27] reported a poor response in patients with systemic RVs, with only 2 of their 9 patients demonstrating improvement with CRT. In conclusion, CRT response is unclear in patients with systemic RV. Even though we had a limited number of patients, we suggested that in patients with systemic ventricle failure due to the postoperative complete AVB and single sided pacing related dyssynchrony, CRT upgrade might be an alternative treatment option for clinical improvement and bridging to heart transplantation.

5 | STUDY LIMITATIONS

This study was limited by its retrospective nature. The young age of the patients and short follow-up periods were other limitations. Another limitation might be related to patient population; since our arrhythmia clinic is one of the largest in Turkey, this may cause referral bias from other pediatric cardiologists. However, we attempted to reduce this referral bias by including all patients with ccTGA irrespective of arrhythmia.

6 | CONCLUSION

In conclusion, conduction and rhythm abnormalities should be routinely investigated at regular intervals using Holter ECG recordings, together with ECG, to ensure a comprehensive assessment of ccTGA patients. Based on in-depth evaluations, the present study revealed that rhythm disturbances were much more frequent than reported previously. Although pacemaker implantation was more frequent in patients with isolated ccTGA, the risk of pacemaker implantation was directly related to surgery in ccTGA patients with associated lesions. Regular follow-up visits are critically important in patients with postoperative pacemaker implantations to detect systemic ventricular dysfunction and identify the need for CRT.

CONFLICT OF INTEREST

The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

AUTHOR CONTRIBUTIONS

Concept/design: Kasar Taner, Ergul Yakup

Data analysis/interpretation, drafting article: Kasar Taner, Ozturk Erkut

Critical revision of article: Gökalp Selman, Ergul Yakup, Guzeltas Alper, Haydin Sertac

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REFERENCES

[1] Samánek M, Voríšková M. Congenital heart disease among 815,569 children born between 1980 and 1990 and their 15-year survival: a prospective Bohemia survival study. *Pediatr Cardiol.* 1999;20(6):411–417.

[2] Kuehl KS, Loffredo CA. Population-based study of l-transposition of the great arteries: possible associations with environmental factors. *Birth Defects Res A Clin Mol Teratol.* 2003;67(3):162–167.

[3] Van Praagh R, Papagiannis J, Grünenfelder J, Bartram U, Martanovic P. Pathologic anatomy of corrected transposition of the great arteries: medical and surgical implications. *Am Heart J.* 1998;135(5 Pt 1):772–785.

[4] 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(2):285–290.

[5] Rutledge JM, Nihill MR, Fraser CD, Smith OE, McMahon CJ, Bezold LI. Outcome of 121 patients with congenitally corrected transposition of the great arteries. *Pediatr Cardiol.* 2002;23(2):137–145.

[6] Warnes CA, Williams RG, Bashore TM, et al. American College of Cardiology.; American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease); American Society of Echocardiography; Heart Rhythm Society; International Society for Adult Congenital Heart Disease.; Society for Cardiovascular Angiography and Interventions.; Society of Thoracic Surgeons. ACC/AHA 2008 guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Develop Guidelines on the Management of Adults With Congenital Heart Disease). Developed in Collaboration With the American Society of Echocardiography, Heart Rhythm Society, International Society for Adult Congenital Heart Disease, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons. *J Am Coll Cardiol.* 2008;52:143–263.

[7] Warnes CA. Transposition of the great arteries. *Circulation.* 2006;114(24):2699–2709.

[8] Daliento L, Corrado D, Buja G, John N, Nava A, Thiene G. Rhythm and conduction disturbances in isolated, congenitally corrected transposition of the great arteries. *Am J Cardiol.* 1986;58(3):314–318.

[9] Kanter RJ. Pearls for ablation in congenital heart disease. *J Cardiovasc Electrophysiol.* 2010;21(2):223–230.

[10] Hebe J, Hansen P, Ouyang F, Volkmer M, Kuck KH. Radiofrequency catheter ablation of tachycardia in patients with congenital heart disease. *Pediatr Cardiol.* 2000;21(6):557–575.

[11] Huhta JC, Maloney JD, Ritter DG, Ilstrup DM, Feldt RH. Complete atrioventricular block in patients with atrioventricular discordance. *Circulation.* 1983;67(6):1374–1377.

[12] Graham TP, 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(1):255–261.

[13] Anderson RH, Becker AE, Arnold R, Wilkinson JL. The conducting tissues in congenitally corrected transposition. *Circulation.* 1974;50(5):911–923.

[14] Allen HD, Driscoll DJ, Shaddy RE, Feltes TF, eds. Moss and Adams' Heart Disease in Infants, Children, and Adolescents Including the Fetus and Young Adult. Philadelphia: Lippincott Williams & Wilkins; 2008.

[15] Cook AC, Anderson RH. The functionally univentricular circulation: anatomic substrates as related to function. *Cardiol Young.* 2005;15(S3):7–16.

[16] Davies MJ, Anderson RH, Becker AE, eds. Atrioventricular conduction tissues in congenital heart disease. In: The Conduction System of the Heart. Stoneham, MA: Butterworth-Heinemann 1983:135–153.

[17] Simmons MA, Rollinson N, Fishberger S, Qin L, Fahey J, Elder RW. Modern incidence of complete heart block in patients with L-looped ventricles: does univentricular status matter? *Congenit Heart Dis.* 2015;10(5):E237–E242.

[18] Connelly MS, Liu PP, Williams WG, Webb GD, Robertson P, McLaughlin PR. Congenitally corrected transposition of the great arteries in the adult: functional status and complications. *J Am Coll Cardiol.* 1996;27(5):1238–1243.

[19] Kayrak M, Kaya Z, Gul EE, et al. Congenitally corrected transposition of great arteries with severe rhythm disturbances. *Indian Pacing Electrophysiol J.* 2010;10:179–183.

[20] Caldarella C, Leccisotti L, Bruno I, Collarino A, Maggi F, Giordano A. Giordano AMyocardial perfusion single-photon emission tomography (SPET) and positronemission tomography-computed tomography (PET-CT) imaging for congenitallycorrected transposition of great arteries. *Pediatr Cardiol.* 2012;33(8):1435–1439.

[21] Hornung TS, Bernard EJ, Jaeggi ET, Howman-Giles RB, Celermajer DS, Hawker RE. Myocardial perfusion defects and associated systemic ventricular dysfunction incongenitally corrected transposition of the great arteries. *Heart.* 1998;80(4):322–326.

[22] Lukac P, Pedersen AK, Mortensen PT, Jensen HK, Hjortdal V, Hansen PS. Ablation of atrial tachycardia after surgery for congenital and acquired heart disease using an electroanatomic mapping system: which circuits to expect in which substrate? *Heart Rhythm.* 2005;2(1):64–72.

[23] Mönckeberg JG. Zur Entwicklungsgeschichte des Atrioventrikularsystems. *Verhandl Deutsch Gesellsch.* 1913;16:228.

[24] Lobo RG, Griffith M, De Bono J. Ablation of arrhythmias in patients with adult congenital heart disease. *Arrhythm Electrophysiol Rev.* 2011;3(1):36–39.

[25] Dave AS, Aboulhosn J, Child JS, Shivkumar K. Transconduit puncture for catheter ablation of atrial tachycardia in a patient with extracardiac Fontan palliation. *Heart Rhythm.* 2010;7(3):413–416.

[26] Attenhofer Jost CH, Connolly HM, Edwards WD, Hayes D, Warnes CA, Danielson GK. Ebstein's anomaly - review of a multifaceted congenital cardiac condition. *Swiss Med Wkly.* 2005;135(19–20):269–281.

[27] Dubin AM, Janousek J, Rhee E, et al. Resynchronization therapy in pediatric and congenital heart disease patients: an

international multicenter study. *J Am Coll Cardiol*. 2005;46(12): 2277–2283.

[28] Janousek J, Gebauer RA, Abdul-Khaliq H, et al. Cardiac resynchronisation therapy in paediatric and congenital heart disease: differential effects in various anatomical and functional substrates. *Heart*. 2009; 95(14):1165–1171.

[29] Cecchin F, Frangini PA, Brown DW, et al. Cardiac resynchronization therapy (and multisite pacing) in pediatrics and congenital heart disease: five years experience in a single institution. *J Cardiovasc Electrophysiol*. 2009;20(1):58–65.

[30] JanoušEk J, Tomek V, Chaloupecký V, et al. Cardiac resynchronization therapy: a novel adjunct to the treatment and prevention of systemic right ventricular failure. *J Am Coll Cardiol*. 2004;44(9): 1927–1931.

How to cite this article: Kasar T, Ayyildiz P, Tunca Sahin G, et al. Rhythm disturbances and treatment strategies in children with congenitally corrected transposition of the great arteries. *Congenital Heart Disease*. 2018;13:450–457. https://doi.org/10.1111/chd.12595