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Outcomes of congenitally corrected transposition of the great arteries: our five-year single-center experience from Turkey

Büyük damarların doğuştan düzeltilmiş transpozisyonunun sonuçları: Türkiye'den beş yıllık, tek merkezli deneyimimiz

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ABSTRACT

Background: This study aims to report the outcomes of congenitally corrected transposition of the great arteries and to identify possible complication-related factors affecting the prognosis.

Methods: Between August 2009 and September 2014, data of a total of 81 patients (48 males, 33 females; mean age 4 years; range, 15 days to 50 years) with congenitally corrected transposition of the great arteries were retrospectively analyzed. The patients were divided into two groups: congenitally corrected transposition of the great artery patients with (group 1; n=66) and without (group 2; n=15) associated cardiac lesions. Age and gender of the patients, surgical history, atrioventricular block, arrhythmia, and systemic atrioventricular valve regurgitation were noted.

Results: The median follow-up was 15 (range 1 to 52) months. Sixteen patients had clinical signs of congestive heart failure in group 1, while none of the patients had in group 2. Surgery was performed in 31 patients in group 1 and none in group 2. Twelve patients were directed to biventricular correction (nine physiologic and three anatomic corrections). The Glenn shunt was performed in six patients and Fontan circulation in one. Of these patients with the Glenn shunt, one planned to underwent physiologic correction and four to the hemi-Mustard-Rastelli surgery. During follow-up, seven patients died and four patients developed systemic ventricular dysfunction. Cardiac resynchronization therapy was performed in two patients. Previous heart surgery, association of pulmonary atresia or stenosis, the presence of a large ventricular septal defect, tricuspid regurgitation, and atrioventricular block were found to be statistically significant risk factors for clinical congestive heart failure ($p<0.05$). Moderate to severe tricuspid regurgitation, arrhythmias, and the history of an open heart surgery were found to be significant risk factors for the right and left ventricular dysfunction ($p<0.05$).

Conclusion: Timing of clinical presentation and survival of the patients with congenitally corrected transposition of the great arteries mainly depend on the presence of associated cardiac defects. The development of tricuspid regurgitation, atrioventricular block and systemic ventricular dysfunction, shortly after physiologic repair highlights the consideration of anatomic correction methods in children, such as the double-switch at first hand.

Keywords: Children; congenitally corrected transposition; ventricular dysfunction.

ÖZ

Amaç: Bu çalışmada büyük damarların doğuştan düzeltilmiş transpozisyonunun sonuçları bildirildi ve prognozu etkileyen komplikasyon ile ilişkili muhtemel faktörler belirlendi.

Çalışma planı: Ağustos 2009 - Eylül 2014 tarihleri arasında, büyük damarların doğuştan düzeltilmiş transpozisyonu olan toplam 81 hastanın (48 erkek, 33 kadın; ort. yaş 4 yıl; dağılım 15 gün-50 yıl) verileri retrospektif olarak incelendi. Hastalar ilişkili kardiyak lezyonu olan (grup 1; n=66) ve olmayan (grup 2; n=15) büyük damarların doğuştan düzeltilmiş transpozisyonu olarak iki gruba ayrıldı. Hastaların yaş ve cinsiyeti, ameliyat geçmişi, atriyoventriküler blok, aritmi ve sistemik atriyoventriküler kapak yetersizliği kaydedildi.

Bulgular: Medyan takip süresi 15 (dağılım 1-52) ay idi. Grup 1'de 16 hastada klinik olarak konjestif kalp yetersizliği bulguları var iken, grup 2'deki hastaların hiçbirinde yoktu. Grup 1'de 31 hastaya cerrahi müdahale yapılırken, grup 2'de hiçbir hastaya yapılmadı. Hastaların 12'si iki ventrikül tamirine (dokuzu fizyolojik ve üçü anatomik düzeltmeye) yönlendirildi. Altı hastaya Glenn şantı ve bir hastaya Fontan sirkülasyonu yapıldı. Glenn şantı yapılan hastaların birine fizyolojik düzeltme, dördüne hemi-Mustard-Rastelli cerrahisi uygulanması planlandı. Takip sırasında yedi hasta kaybedildi ve dört hastada sistemik ventrikül disfonksiyonu gelişti. İki hastaya kardiyak resenkronizasyon tedavisi uygulandı. Açık kalp ameliyatı öyküsü, pulmoner atrezi veya darlık birlikteliği, geniş ventriküler septal defekt varlığı, triküspid yetersizliği ve atriyoventriküler blok klinik olarak konjestif kalp yetersizliği için istatistiksel olarak anlamlı risk faktörleri olarak saptandı ($p<0.05$). Orta ila şiddetli düzeyde triküspit yetersizliği, aritmiler ve açık kalp cerrahisi öyküsü sağ ve sol ventrikül disfonksiyonu için anlamlı risk faktörleri olarak bulundu ($p<0.05$).

Sonuç: Büyük damarların doğuştan düzeltilmiş transpozisyonu olan hastalarda klinik tablonun ortaya çıkışı ve sağkalım, esas olarak ilave kardiyak defekt varlığına bağlıdır. Fizyolojik düzeltme sonrası kısa sürede triküspit yetersizliği, atrioventriküler blok ve sistemik ventrikül disfonksiyonu gelişmesi çocuklarda double switch gibi anatomik tamir yöntemlerinin ilk olarak düşünülmesi gerektiğine işaret etmektedir.

Anahtar sözcükler: Çocuklar; doğuştan düzeltilmiş transpozisyon; ventrikül disfonksiyonu.

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Congenitally corrected transposition of the great arteries (ccTGA) is a rare cardiac condition, accounting for 0.05 to 1.4% of all congenital heart defects.[1,2] It may present as an isolated defect, but frequently is associated with other cardiac defects, such as ventricular septal defect (VSD), pulmonary stenosis (PS), and Ebstein's anomaly of the tricuspid valve.[1-3] The timing of clinical presentation of a patient with ccTGA mainly depends on the type and severity of the associated lesions.[3]

In this study, we aimed to report the outcomes of ccTGA and to identify possible complication-related factors affecting the prognosis in a single center during a five-year follow-up period.

PATIENTS AND METHODS

Between August 2009 and September 2014, data of a total of 81 patients (48 males, 33 females; median age 4 years; range, 15 days to 50 years) with ccTGA were retrospectively analyzed. The patients were divided into two groups: ccTGA patients with (group 1) and without (group 2) associated cardiac lesions. Data related to the demographic and clinical features [clinical congestive heart failure (CHF)], electrocardiographic and echocardiographic findings [cardiac situs, VSD location and size, Ebsteinoid tricuspid valve, severity and degree of tricuspid regurgitation (TR), degree of PS, aortic regurgitation (AR), right and left ventricular dysfunction], and angiographic findings were noted. Duration of follow-up, mortality, type of surgery, arrhythmias, atrioventricular block (AVB), pacemaker insertion, cardiac resynchronization therapy (CRT), and ablations were also recorded.

The study protocol was approved by the Istanbul Mehmet Akif Ersoy, Thoracic and Cardiovascular Surgery Center Ethics Committee. The study was conducted in accordance with the principles of the Declaration of Helsinki.

Statistical analysis

Statistical analysis was performed using SPSS version 15.0 software (SPSS Inc., Chicago, IL, USA). The Mann-Whitney U test was used to compare median values between the groups, while the chi-square and Fisher's exact tests were performed to compare the rates between the groups. Association of selected variables with clinical CHF, right ventricular (RV) dysfunction, and left ventricular (LV) dysfunction was assessed by calculating odds ratio (OR) estimates and 95% confidence intervals (CI). These estimates were combined over both groups using the Mantel-Haenszel method in the absence of statistically significant differences between the groups. A p value of <0.05 was considered statistically significant.

RESULTS

Group 1 consisted of the patients with significant associated lesions, including large VSD, moderate or severe PS, pulmonary atresia (PA), moderate or severe TR, and Ebsteinoid tricuspid valve. Group 2 consisted of the patients with minor or no significant associated lesions (Table 1). Demographic and clinical characteristics of the patients are shown in Table 2.

Our institutional algorithm depends on systemic ventricle (SV) and systemic atrioventricular valve (SAVV) functions and associated cardiac defects (Figure 1). Prophylactic atrioventricular (AV) pacemaker electrodes were inserted during surgery, based on the preoperative Holter monitoring.

Moderate to severe TR was demonstrated in 24 patients (36%) in group 1. Eleven patients (17%) had varying degrees of an Ebsteinoid tricuspid valve with moderate to severe TR in six of them. Only three patients (20%) in group 2 demonstrated moderate to severe TR during follow-up.

At baseline, no patients demonstrated any systemic ventricular dysfunction (SVD). During follow-up, three patients in group 1 and one patient in group 2 demonstrated SVD.

In group 1, morphological LV dysfunction was present in five patients; three of them had biventricular dysfunction. In group 2, no morphological LV dysfunction was present.

Table 1. Classification of patient groups with congenitally corrected transposition of great arteries

	n	%
Group 1		
ccTGA with associated significant lesions	66	81.5
Ventricular septal defect	52	78.7
Pulmonary stenosis	25	38
Pulmonary atresia	19	28
Tricuspid regurgitation (moderate or severe)	14	21
Ebsteinoid tricuspid valve	11	16.6
Group 2		
ccTGA without associated significant lesions	15	18.5
No associated lesions	2	13.3
Ventricular septal defect (small)	6	40
Pulmonary stenosis (mild)	6	40
Tricuspid regurgitation (mild)	7	46.6

ccTGA: Congenitally corrected transposition of great arteries.

Table 2. Demographic and clinical characteristics of patients

Demographic features	Group 1 (n=66)				Group 2 (n=15)				
	n	%	Median	Range	n	%	Median	Range	*p*
Age (year)			3	15 days to 46 years			4.5	1.16 years to 50 years	0.045
Duration of follow-up (month)			15	1 to 52			28	8 to 50	0.030
Gender									0.574
Male	38				10				
Female	28				5				
Exitus	7	11.3			-	-			
Cardiac situs									0.115
Solitus	53	80.3			15	100			
Inversus	13	19.7			-	-			
Cardiac apex									0.301
Levocardia	34	51.5			11	73.3			
Mesocardia	10	15.2			1	6.7			
Dextrocardia	22	33.3			3	20.0			
Congestive heart failure									0.035
Yes	16				0				
No	50				15				
Right ventricular dysfunction									0.567
Any	63				14				
Moderate or severe	3				1				
Left ventricular dysfunction									0.578
Any	61				15				
Moderate or severe	5				-				
Aortic regurgitation	13	19.7			1	6.7			0.448
Tricuspid regurgitation									0.312
Any	42	63.6			12	80.0			
Moderate or severe	24	36.4			3	20.0			
Arrhythmia	17	25.8			4	26.7			0.981
Pacemaker implantation	8	12.3			3	20.0			0.423
Ablation	2	3			-	-			0.991
Cardiac resynchronization therapy	2	3.1			1	6.7			0.468
Open heart surgery	31	46.9			-	-			0.001
The age at first surgery, (year)			2	10 days to 40 years			-	-	

Clinical CHF was present in 16 patients (24%) in group 1 and none in group 2. The odds ratio (OR) of clinical CHF, RV dysfunction, and LV dysfunction were calculated for all patients for possible risk factors (Table 3). Open heart surgery, PA, moderate to severe PS, and large VSD were found to be significant risk factors for CHF. In addition, moderate to severe TR, arrhythmia, and open heart surgery were found to be significant risk factors for RV and LV dysfunction ($p<0.05$).

Various types of rhythm disorders were present in 19 patients (29%) in group 1 and in five patients (33%) in group 2. Ablation was performed in two patients with the Wolf-Parkinson-White (WPW). Pacemakers were implanted in eight patients (12%) in group 1 and in two patients (13%) in group 2 due to complete AVB on follow-up. Of these 10 patients, six were implanted postoperatively (after surgical VSD closure in 5 of them).

Ductal stenting was performed in five patients and they are still under the follow-up.

Cardiac surgery was performed in 31 of 66 patients (46.9%) in group 1, while no surgeries were performed in group 2 (Table 4). Twelve patients were directed to biventricular correction, of whom nine underwent physiological correction and three underwent anatomical correction. Bidirectional Glenn shunt was performed in six patients (at baseline in two patients, after ductal stenting in two patients, and after aortopulmonary shunting in two patients) and Fontan circulation was completed in one patient with PA and hypoplastic LV.

In addition, pulmonary artery banding (PAB) was performed in seven patients in group 1. Of these patients, four had large VSD, two had moderate to severe TR with Ebstein anomaly, and one had large VSD with an Ebsteinoid valve. Bidirectional Glenn

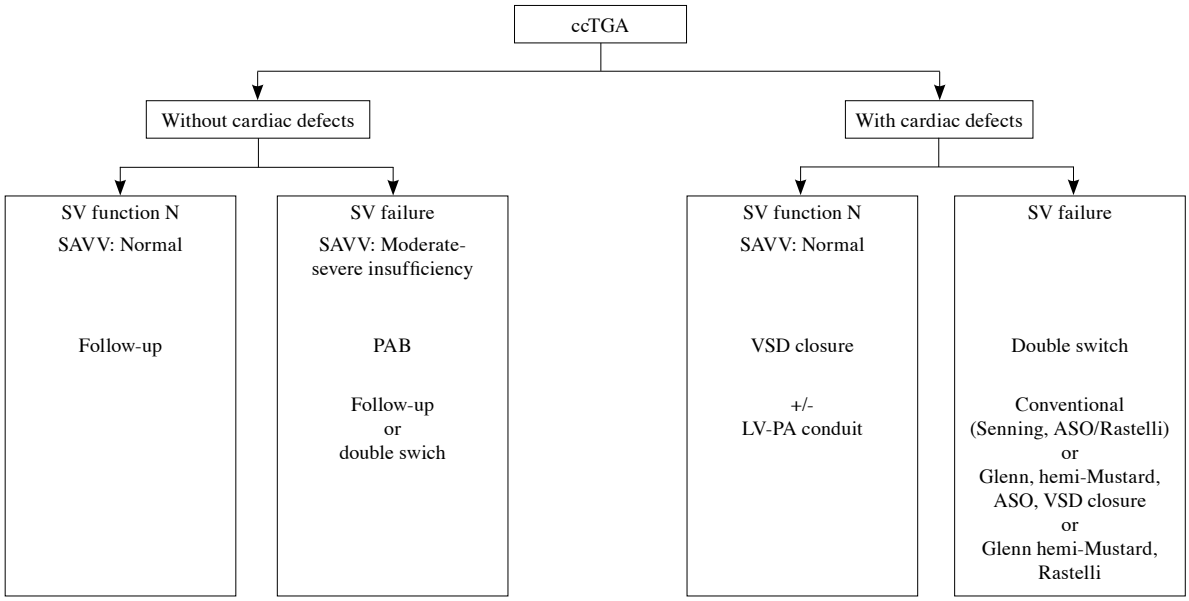

Figure 1. The institutional algorithm of treatment approach in patients with congenitally corrected transposition of great arteries.

*c*TGA: Congenitally corrected transposition of great arteries; SV: Systemic ventricle; SAVV: Systemic atrioventricular valve; PAB: Pulmonary banding procedure; VSD: Ventricular septal defect; LV-PA: Left ventricle-pulmonary artery; ASO: Arterial switch operation.

shunt with arch reconstruction was planned for one patient who had PAB.

Two patients were reoperated; one due to conduit stenosis after initial Senning-Rastelli operation and one due to severe TR. The latter underwent tricuspid valve replacement after the first operation for VSD closure.

Table 3. Surgical data of patients with congenitally corrected transposition of great arteries and significant associated lesions

Surgical procedures	Number
Physiologic correction	
VSD closure and LV-PA conduit	5
VSD closure	4
Anatomic correction	
Senning-Rastelli procedure	2
Double Switch procedure	1
Univentricular correction	
Glenn shunt	6
Fontan procedure	1
Others	
Modified Blalock-Taussig shunt	3
Central shunt	2
Pulmonary banding procedure	7
Total	31

VSD: Ventricular septal defect; LV-PA: left ventricle-pulmonary artery.

The median follow-up was 15 (range 1 to 52) months. Nine patients were lost during follow-up and seven patients died (11%). All deaths were documented in group 1. Of the seven deaths, one patient died after ductal stenting and three died after aortopulmonary shunt procedures during the early postoperative period due to either low cardiac condition or shunt obstruction. A 14-year-old patient with PA who previously underwent the Senning-Rastelli procedure died after the conduit replacement in the early postoperative period due to low cardiac condition. In addition, a two-year-old female patient in whom double-switch with correction of the Ebsteinoid valve was planned died in the preoperative period due to respiratory arrest. A 21-year-old male patient with large VSD and Eisenmenger syndrome was also admitted to the emergency department with cardiac arrest.

Of the six patients who received bidirectional Glenn shunts, one underwent biventricular correction by physiological means, while another was directed to the Fontan circulation, and four were scheduled for the hemi-Mustard-Rastelli procedure.

DISCUSSION

The timing of clinical presentation of the patient with ccTGA mainly depends on the type and severity of the associated lesions.[3-5] Patients with major

Table 4. Odds ratios for clinical congestive heart failure, and right and left ventricular dysfunction

	CHF			RVD			LVD		
	OR	95% CI	*p*	OR	95% CI	*p*	OR	95% CI	*p*
Situs	1.4	0.2-7.1	0.981	1.2	0.2-18.8	0.510	1.3	0.2-12.9	0.990
Tricuspid regurgitation	0.4	0.2-1.6	0.143	0.3	0.2-0.4	0.010	0.3	0.2-0.4	0.003
Ablation	4.2	0.2-7.2	0.358	0.9	0.8-1.0	0.980	0.9	0.8-1.0	0.990
Arrhythmia	1.3	0.4-4.6	0.751	0.2	0.1-1.0	0.050	0.2	0.1-0.7	0.015
Surgery	0.2	0.1-0.5	0.030	0.4	0.3-0.6	0.040	0.4	0.3-0.5	0.020
Pacemaker	0.9	0.2-4.6	0.981	0.2	0.1-1.0	0.080	0.2	0.1-1.4	0.130
Gender	2.1	0.7-6.6	0.171	0.6	0.1-5.0	0.980	1	0.2-6.5	0.980
Pulmonary atresia	8.4	1.0-69.2	0.027	1.2	0.2-14.6	0.990	0.5	0.1-5.7	0.990
Pulmonary stenosis	2	1.5-2.2	0.001	0.8	0.1-9.4	0.990	0.3	0.1-3.6	0.640
VSD	0.2	0.1-1	0.041	1.7	0.1-17	0.990	2	0.-21	0.650

CHF: Congestive heart failure. RVD: Right ventricular dysfunction. LVD: Left ventricular dysfunction; OR: Odds ratio; CI: Confidence interval; VSD: Ventricular septal defect.

associated lesions, such as large VSD, PA, moderate to severe TR, and moderate to severe PS, present earlier than those with minor associated lesions due to the premature appearance and more severe course of clinical symptoms.[3-5] In the present study, a total of 15 patients in group 2, all of whom had minor or no associated defects, were significantly older on admission than the patients in group 1. This suggests that the initial presentation of these patients may be delayed compared to the ccTGA patients with hemodynamically significant associated lesions.

In cases of severe PS or PA of the morphological LV outflow tract, the infant will be cyanotic, often having a ductus-dependent pulmonary circulation or in case of a hemodynamically significant VSD, heart failure will develop earlier in life, evidenced by fatigue, poor weight gain, and feeding intolerance.[6] In the present study, 19 patients with ccTGA and PA were admitted with varying degrees of cyanosis, whereas 16 patients with large VSD and pulmonary hypertension were admitted with clinical CHF.

Furthermore, systemic atrioventricular valve regurgitation is a significant problem in patients with ccTGA. In 90% of these patients, the valve has an abnormal structure; however, TR is present in only about 30% of them.[7] In the present study, consistent with the literature data, there were varying degrees of systemic valve abnormality in 86% of the 81 patients and the valve was Ebsteinoid in 11 (17%) of these patients. There was also moderate to severe TR in 27 patients (33%), 89% (24/27) of whom were in group 1. Of these 24 patients with moderate to severe TR in group 1, the valve was Ebsteinoid in six and SVD was diagnosed in three.

The ability of the SV, namely the RV, to sustain the systemic output over time is the primary concern during the long-term follow-up of ccTGA patients. To date, various potential risk factors for SVD in ccTGA have been implicated, including SAVV regurgitation (tricuspid regurgitation), associated congenital heart defects, AVB, and surgical interventions, particularly open heart surgeries.[5] In a multi-center study including 182 patients, Graham et al.[5] reported RV dysfunction in 39% of patients with ccTGA and associated defects and in 32% of patients without associated defects with a mean follow-up of 32 years. In a study of 40 patients with ccTGA, Prieto et al.[8] reported TR as a major risk factor for SVD. Zias et al.[9] showed that in patients with VSD and PS, the conventional surgical repair (VSD closure and LV-PA conduit placement or correction of stenosis without conduit) has an increased mortality and AVB risk in the early postoperative period and was complicated by conduit stenosis, progressive SAVV regurgitation, and SVD in the long-term follow-up. In the present study, SVD (together with LV dysfunction) developed in three of the 66 patients in group 1 during follow-up. Of these, two were operated for VSD closure and one for VSD closure together with LV-PA conduit placement. Of note, in group 2, one patient without any associated defect developed SVD during follow-up. All four patients with SVD had moderate to severe TR. Consistent with the previous study findings, we observed that moderate to severe TR, arrhythmia, and history of surgical procedure were risk factors for SVD development. We also found TR to be a risk factor for developing SVD (p=0.01, OR: 0.3, 95% CI: 0.2-0.4), as were arrhythmia and history of surgical procedures.

About one-tenth of patients born with ccTGA have complete AVB.[10] In patients born with normal

cardiac conduction, the risk of developing heart block increases by 2% per year, until it reaches a prevalence of 30% in adulthood.[11] In the present study, AVB was not diagnosed initially in any of the patients. Of a total of 81 patients with ccTGA, complete AVB developed in 11 (14%) during follow-up. Complete AVB developed in 12% (8/66) of patients in group 1 and in 20% (3/15) of patients in group 2. Permanent pacemakers were implanted in six patients with associated lesions due to postoperative AVB. It was done after VSD closure in five and after double-switch procedure in one patient. The increase in the number of AVB cases is an expected result, as most of our patients are still in the early childhood.

Despite optimal medical therapy, clinical studies have demonstrated that CRT improves the functional class and echocardiographic variables in severe LV systolic dysfunction, electromechanical delay, and heart failure.[12-15] Dubin et al.[14] in 2005 and Janousek et al.[15] in 2009 conducted large, multi-center, retrospective studies in children and reported that CRT resulted in a significant increase in the ejection fraction (EF) values and a decrease in the QRS duration during a median follow-up of four months and 7.5 months, respectively. The latter study also reported an improved New York Heart Association (NYHA) functional class. In addition, it showed the presence of a systemic LV to be the strongest predictor of improved cardiac functions with CRT.[15] Cardiac resynchronization therapy was performed in two patients with SVD who developed after the Senning-Rastelli procedure in one patient and after VSD closure and LV-PA conduit placement in the other. During follow-up, these patients had improved systemic ventricular functions with an increased functional capacity. These outcomes suggest that many patients with congenital heart disease and progressive heart failure, particularly those with ccTGA, may benefit from CRT.

Moreover, patients with ccTGA are a heterogeneous mixture of anatomical and functional subtypes who also may need various anatomic and physiologic corrective surgeries. Depending on the associated defect and preoperative clinical status, biventricular or univentricular correction can be achieved by different techniques. Along with univentricular approaches, the procedures of choice include biventricular functional correction by VSD closure alone or with PS correction (LV-PA conduit), biventricular anatomical correction by double-switch or atrial switch-Rastelli or by using Glenn at the first stage and, then, completing with the hemi-Mustard-Rastelli. Although the optimal strategy seems to be to restore morphologic LV to the systemic position, recent studies favor different techniques to maintain biventricular correction.[16,17]

Hiramatsu et al.[16] reported the mean age at double-switch operation (DSO) of 18 patients as 4.3 years. Murtuza et al.[17] carried out a retrospective analysis of 113 ccTGA patients and reported the median age at repair as 3.2 years (25 days to 40 years). Bautista-Hernandez et al.[18] reported the median age at surgery of 106 patients who had anatomical correction as 1.2 years (2 months to 43 years). Consistent with these study findings, in the present study, the median age at surgery was two years (10 days to 40 years) in patients with major associated defects.

Zias et al.[9] suggested that atrial switch-Rastelli operations increased early postoperative mortality, particularly in patients with RV hypoplasia. The authors also reported that these patients developed several sequelae of atrial-switch operation, such as atrial arrhythmias and conduit stenosis. In accordance with this study, in the present study, two patients developed LV dysfunction after the Senning-Rastelli procedure during follow-up. In addition, one of them received a pacemaker due to postoperative AVB.

Considering these issues, alternative surgical procedures have been proposed for such patients. The removal of the right ventricular volume load without total decompression of the LV by VSD closure, leaving the RV as systemic ventricle, as is done in conventional treatment, and performing limited correction of PS together with a Glenn shunt have been shown to prevent negative morphological changes in the RV and tricuspid valve.[9] Alternatively, compared to the conventional repair for ccTGA, DSO was expected to improve the long-term prognosis by using the morphological LV in the systemic circulation.[19] In light of these new data, we revised the treatment options for ccTGA patients, taking into consideration the complications of physiologic repair and atrial-switch. The Glenn shunt procedure was performed in four patients and was planned for another, followed by a hemi-Mustard-Rastelli procedure. It has been also reported that compared to the conventional surgical repair, the results of patients who underwent DSO or anatomical repair were much favorable, even in patients with significant TR.[19,20] In accordance with the literature data, in the present study, moderate to severe TR was demonstrated in all (5/5) patients who underwent VSD closure and LV-PA conduit placement, in 50% (2/4) of patients who underwent isolated VSD closure, and in all (2/2) patients who underwent atrial-switch during follow-up.

Furthermore, PAB is an important concern in atrial-arterial switch and has been identified as a risk factor for developing pulmonary root dilatation and subsequent neo-aortic valve regurgitation. In the presence of an associated hemodynamically significant VSD, primary repair can be discussed early in life to avoid potential deterioration of the pulmonary valve after PAB.[21] However, Metton et al.[22] reported that in neonates with isolated ccTGA, PAB was safe with low morbidity rates, which allowed DSO, whenever indicated. In the present study, PAB was performed in seven patients in group 1, five of whom had large VSD and two of whom had severe TR with Ebsteinoid tricuspid valve. After PAB, preoperative severe TR decreased to a mild degree in both patients with an Ebsteinoid tricuspid valve and no TR was identified in the other patients. No pulmonary root dilatation or deterioration of pulmonary arteries was either detected in any patients during follow-up. In the short term, tricuspid valve function was improved and systemic competence of the LV was maintained in these patients without any damage to the pulmonary arteries by PAB.

On the other hand, this study was limited by its retrospective design. Also, the young age of the patients and the short follow-up period may be responsible for the low number of surgical procedures and low incidence of late complications, such as TR or SVD, compared to the adult studies. In addition, as our center is a tertiary center with a wide referral base, asymptomatic patients without associated defects and hemodynamically well-balanced patients might be underestimated. Therefore, this study might not reflect the true population due to the referral of numerous patients with severe associated lesions, such as PA and severe PS in need of an emergent treatment.

In conclusion, the timing of clinical presentation and the survival of the patients with congenitally corrected transposition of the great arteries mainly depend on the presence of associated lesions. In addition, complications, such as systemic ventricular dysfunction and tricuspid regurgitation, develop overtime and are much rarer in childhood. Development of tricuspid regurgitation, atrioventricular block and eventually systemic ventricular dysfunction shortly after physiologic repair leads to the consideration of anatomic correction treatment modalities in children, such as double-switch at first hand.

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