Congenitally Corrected Transposition of the Great Arteries

11

Kiyozo Morita

Abstract

Congenitally corrected transposition (ccTGA) is a rare cardiac malformation, characterized by the combination of discordant atrioventricular and ventriculo-arterial connections, accompanied by other cardiovascular malformations in the majority of cases. The clinical presentation and the age of onset of ccTGA largely depend on the presence and severity of associated malformations. Currently, most patients who underwent a surgical repair in childhood survive to reach adulthood with or without postoperative residual lesions or late complications to be repaired, while there is also a considerable population of unoperated patients in adulthood. The correct diagnosis is often made for the first time in adulthood, because of a symptom of heart failure or incidentally when an ECG, chest X-ray, or echocardiogram is performed for other reasons. With advancing age, ccTGA even in the setting of no associated malformations can often lead to progressive systemic atrioventricular (AV) valvar regurgitation and the systemic right ventricular dysfunction late in adulthood, which might be the most important and common manifestation for adult ccTGA. The physiological repair, the anatomical repair, or the tricuspid valve replacement can be indicated to adult unoperated patients, as well as the redo operation for the late postoperative complication after the initial intracardiac repair. This chapter deals with the specific morphological features, which relates to the natural and unnatural history of this disease, and the indication and procedures of the interventional approaches for both unoperated and postoperative patients.

K. Morita, M.D., Ph.D.
Division of Pediatric Cardiovascular Surgery, Department of Cardiovascular Surgery,
Jikei University School of Medicine, 3-25-8 Nishi-Shinbashi, Minato-Ku,
Tokyo 105-8461, Japan
e-mail: kiyozo@jikei.ac.jp

M. Masuda, K. Niwa (eds.), *Adult Congenital Heart Disease*,
DOI 10.1007/978-981-10-4542-4_11

Keywords
Physiological repair • Anatomical repair • Tricuspid valve replacement • Double switch operation • Systemic right ventricular dysfunction

11.1 Introduction: Overview of ccTGA in Adulthood

Congenitally corrected transposition (ccTGA) is a rare cardiac malformation accounting for approximately 0.5–1.0% of congenital heart disease, characterized by the combination of discordant atrioventricular (AV) and ventriculo-arterial (VA) connections, accompanied by other cardiovascular malformations in more than 90% of cases [1]. The clinical presentation and the age of onset of ccTGA largely depend on the presence and severity of associated malformations.

Adults with ccTGA categorize to the unoperated and operated patients. Currently, most patients who underwent a surgical repair in childhood survive to reach adulthood with or without postoperative residual lesions or late complications to be repaired, while there is also a considerable population of unoperated patients in adulthood.

Some patients may be diagnosed in childhood but not require operation. In a majority of adult ccTGA, however, the correct diagnosis is made for the first time in adulthood, because of a symptom of heart failure or incidentally when an ECG, chest X-ray, or echocardiogram is performed for other reasons. With advancing age, ccTGA even in the setting of no associated malformations can often lead to progressive atrioventricular (AV) valvar regurgitation and the systemic right ventricular dysfunction late in adulthood, which might be the most important and common manifestation for adult ccTGA. Therefore, ccTGA is a unique entity among a variety of complex congenital heart malformations, in which the initial surgical intervention (i.e., intracardiac repair for associated anomaly, tricuspid valve replacement, and the cardiac resynchronization therapy: CRT) can be definitely indicated to adult unoperated patients, as well as the redo operation for the late postoperative complication.

This chapter deals with the specific morphological features, which relates to the natural and unnatural history of this disease, and the indication and procedures of the interventional approaches for both unoperated and postoperative adult ccTGA patients.

11.2 Anatomical Type and Associated Lesions

11.2.1 Definition and Anatomical Type

ccTGA is a complex congenital anomaly with a wide spectrum of morphological features and clinical profiles. The underlying malformation is the combination of discordant atrioventricular and ventriculo-arterial connections (“duplicated discordance”) and results in normal physiology despite the presence of the transposition of the great arteries.

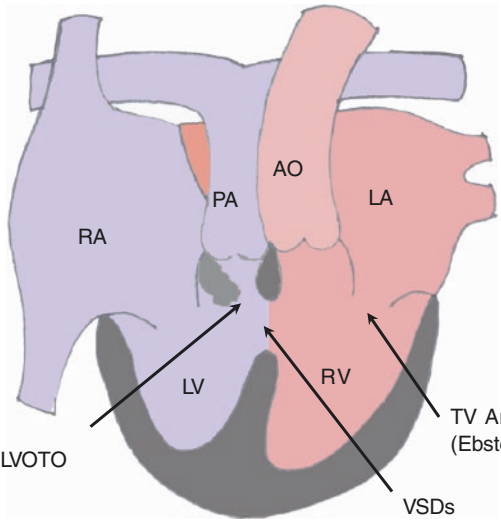

Fig. 11.1 Congenitally corrected transposition of the great arteries (ccTGA). Definition and associated malformations: the combination of discordant atrioventricular and ventriculo-arterial connections ("duplicated discordance") results in normal physiology despite the presence of the transposition of the great arteries

Thus, the morphologically right atrium is connected to a morphologically left ventricle across the mitral valve, which gives rise to the pulmonary artery. The morphologically left atrium is connected to the morphologically right ventricle across the tricuspid valve, which gives rise to the aorta (Fig. 11.1).

The most common anatomical arrangement is situs solitus with L-looping of the ventricles and the aorta anterior and leftward of the pulmonary artery (S,L,L) in 95 percent of cases [2], while situs inversus with D-looping of the ventricles and the aorta anterior and rightward (I,D,D) is also seen in the other 5% of ccTGA cases.

The majority of cases of ccTGA have any associated anatomical abnormalities. The most common lesions include ventricular septal defect (VSD) in 60–80% of patients, the left ventricular outflow tract obstruction (LVOTO) or pulmonary stenosis/atresia (PS/PA) in 40–50% of patients, and abnormalities of the morphologically tricuspid valve in up to 90% of patients with Ebstein's anomaly in 50% [3–5]. The isolated ccTGA without associated malformation is seen only 1% of cases (Fig. 11.1).

11.2.2 Associated Malformations

11.2.2.1 Ventricular Septal Defect (VSD)

The majority of VSDs are perimembranous, located below the pulmonary valve, with the posterosuperior margin of the defect formed by an extensive area of fibrous continuity between the leaflets of the pulmonary, mitral, and tricuspid valves, most characteristically in this anomaly. Often the perimembranous defects become at least partially obstructed by aneurysmal membranous atrioventricular (AV) septum tissue or the straddling tricuspid valve tissue. Other types of defects are also described including doubly committed subarterial (conal) defects, muscular defects, and AV canal type defects.

11.2.2.2 Left Ventricular Outflow Track Obstruction (LVOTO)

The LVOT obstruction may comprise subpulmonary stenosis and/or pulmonary valve stenosis. Pulmonary atresia (PA) can also be seen. The subpulmonary stenosis is often related to the presence of a fibromuscular shelf on the septum, muscular hypertrophy, tunnellike hypoplasia, or an aneurysmal dilation of fibrous tissue derived from the interventricular component of the membranous septum or an accessory mitral or tricuspid valve tissue.

11.2.2.3 Lesions of the Morphologically Tricuspid Valve

The tricuspid valve has been reported to be abnormal in the great majority of ccTGA patients. There is a wide spectrum of severity. Ebstein's anomaly of the systemic AV valve also occurs but is different from the typical right-sided Ebstein's anomaly, only characterized by the septal and posterior leaflets displaced inferiorly toward the cardiac apex [6]. It is rare to find the large "sail-like" deformity of the anterosuperior leaflet, as seen in the setting of concordant atrioventricular connections. And the atrialized portion of the RV inflow is also relatively small.

Another morphological variation is straddling of the tricuspid valve, with overriding the ventricular septum in the presence of VSD. The morphologically mitral valve also straddles and overrides, often in combination with double outlet right ventricle [7].

11.2.3 The Atrioventricular Conduction System in ccTGA: Clinical and Interventional Considerations [1, 8, 9]

With the arrangement of AV discordance, there is reversed offsetting of the attachments of the leaflets of the atrioventricular (the morphological mitral and tricuspid) valves to the septum, with the mitral valve on the right side attached appreciably higher than the tricuspid valve on the left side at the crux of the heart. Almost always there is fibrous continuity between the leaflets of the pulmonary and mitral valves in the roof of the right-sided morphologically left ventricle. The pulmonary outflow tract is deeply wedged between the atrial septum and the mitral valve, thus deviating the atrial septum away from the ventricular inlet septum. Consequently, this gap of septal malalignment is occupied by an extensive membranous septum [1] (Fig. 11.2).

Due to this malalignment of the atrial and inlet ventricular septum, the regular atrioventricular node, which is located at the apex of the triangle of Koch in the base of the atrial septum, is anatomically impossible to connect to the atrioventricular bundle in ccTGA with situs solitus {SLL} arrangement. Instead, the anterior AV node is located beneath the ostium of the right atrial appendages at its junction with the anterior atrial wall and is positioned above the lateral margin of the area of pulmonary to mitral fibrous continuity. The anterior AV node gives rise to a penetrating bundle (PB) through fibrous trigone, and the non-branching bundle (NBB) or the His bundle encircles pulmonary valve anteriorly and descends onto the anterior part of the trabecular septum on the right side (morphologic LV) surface of the septum. (with intimately close relationship to the anterosuperior edge of pm VSD, if present). And then the NBB bifurcates into a cord-like right bundle branch (RBB), which extends leftward to reach the morphologically right ventricle. The fanlike left

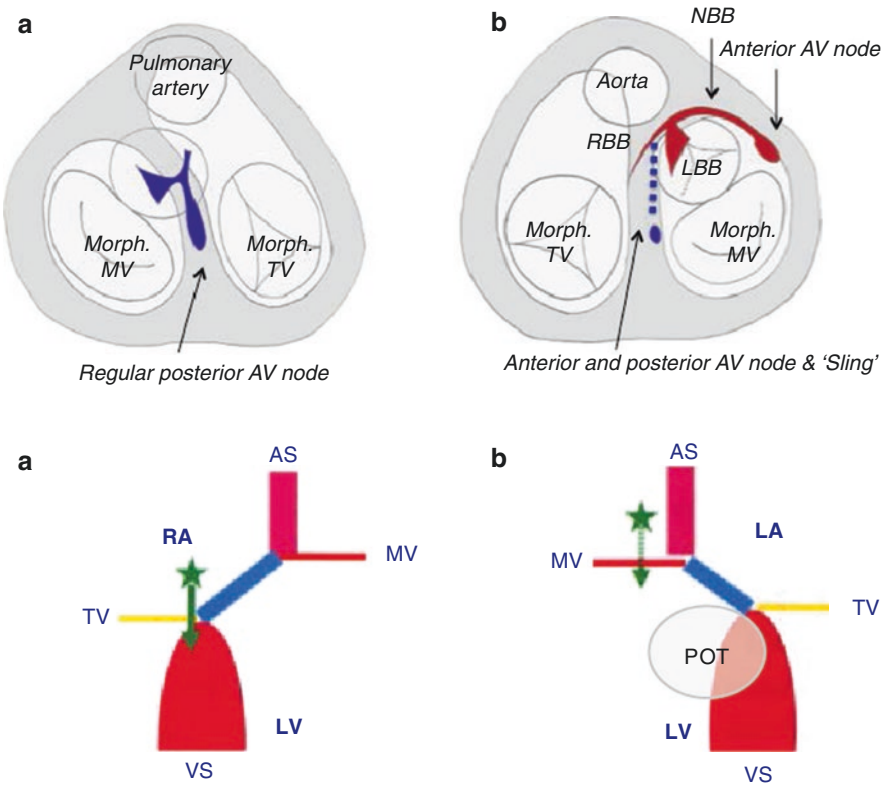

Fig. 11.2 The atrioventricular conduction system in ccTGA. (**a**) Normal heart and (**b**) ccTGA in situs solitus. *AS* the atrial septum, *VS* the ventricular septum, *TV* tricuspid valve, *MV* mitral valve, *AVS* the atrioventricular septum, *POT* the pulmonary outflow tract

bundle branch (LBB) cascades down the smooth surface of the morphologically left ventricle (Fig. 11.2). This abnormal course of the conduction system is of crucial significance to the surgeons in the presence of a ventricular septal defect or subpulmonary obstruction (Fig. 11.3).

In contrast, the normal AV conduction bundle from regular posterior AV node is present in situs inversus, which arrangement of ccTGA is associated with better alignment of the atrial and inlet septum with less "wedging" of the pulmonary outflow tract. Furthermore ccTGA in situs inversus and the other variations may have the dual AV node with possible sling, resulting in the occurrence of reentrant supraventricular tachycardia [10, 11].

11.2.3.1 Clinical Implications of Abnormal Conduction System

The long penetrating and non-branching bundle in ccTGA are vulnerable to fibrosis with advancing age. This makes the conduction system somewhat tenuous, with a progressive incidence of complete AV block.

Other conduction disturbances described include sick sinus syndrome, atrial flutter, reentrant atrioventricular tachycardia due to a dual AV node, and an accessory pathway along the atrioventricular junctions.

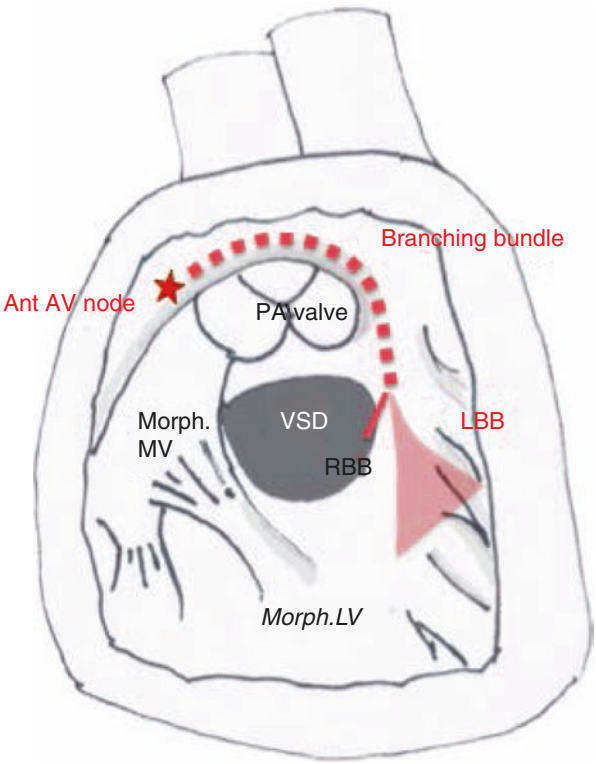

Fig. 11.3 The relationship of the abnormal course of the AV conduction bundle to the pulmonary valve and a ventricular septal. From the left ventricular view

11.3 Overview of Surgical Intervention

11.3.1 Surgical Management Strategies

11.3.1.1 Physiological Repair vs Anatomical Repair

Figure 11.4 depicts the classification and modifications of surgical procedure for ccTGA: *Physiological repair* vs *anatomical repair*. Until recently, patients with associated abnormalities underwent conventional physiological repair. The goal of the operation is to repair the cardiac defects by closing VSD, repairing the tricuspid valve, and reliving LVOTO, while leaving the morphologically right ventricle as the systemic ventricle. The physiologic approach is straightforward but has the shortcomings over time. After this type of repair, the RV remains the systemic ventricle, and long-term results of this type of surgical repair are disappointing with the late occurrence of the right ventricular dysfunction, tricuspid valve regurgitation, and complete heart block. Because of this poor prognosis over the long term, there is an increasing trend toward achieving anatomical correction.

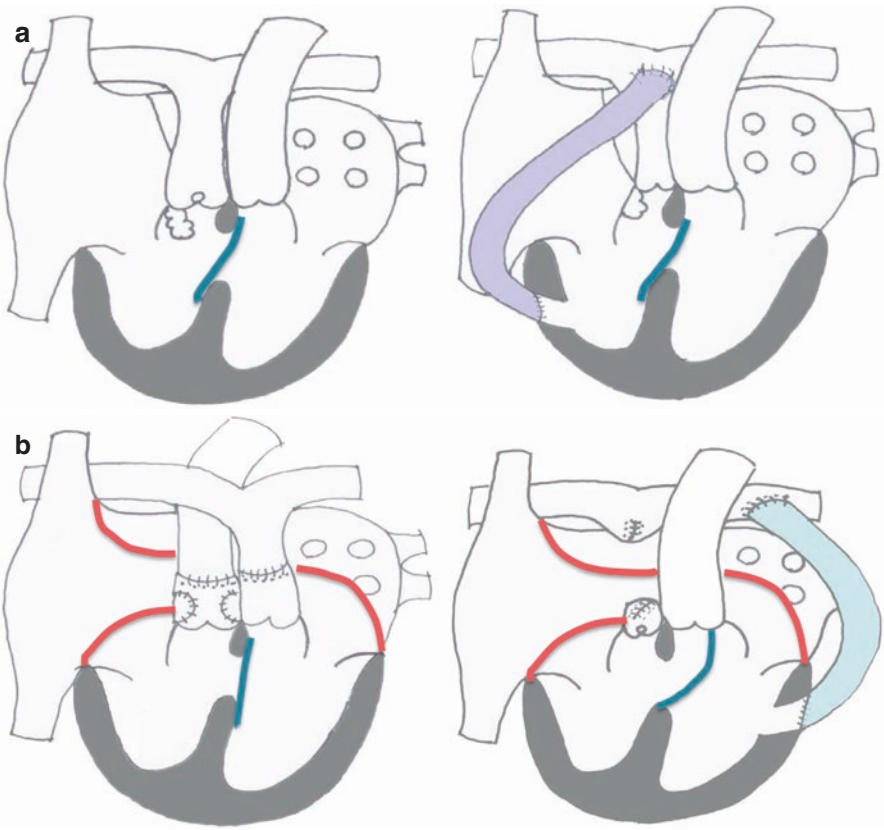

Fig. 11.4 The classification of surgical procedure for ccTGA: *Physiological repair* (**a**) vs *anatomical repair* (**b**). (**a**) (1) VSD closure (2) LVOTO relief using LV-PA conduit (conventional Rastelli) with VSD closure (**b**) (1) The double switch operation (DSO) (2) The Rastelli–Senning/Rastelli–Mustard Operation

The concept of the anatomical correction is the rerouting of the pulmonary venous return to the morphologically left ventricle and aorta and the systemic venous return to the morphologically right ventricle and pulmonary arteries, thus restoring a normal anatomic pattern of circulation. This can be achieved by the combination of atrial switch operation and either arterial or intraventricular switch operation. Anatomic correction for ccTGA is tailored to the patients' specific anatomies: (1) the arterial–atrial switch procedure or the double switch operation (DSO) for patients with normal LVOTO regardless of the presence of VSD and (2) the Rastelli–atrial switch procedure [the Rastelli/Senning or the Rastelli/Mustard] for patients with an unrestrictive VSD and LVOTO (pulmonary atresia or severe subpulmonary stenosis). Current surgical strategies for ccTGA in relation to the anatomical types are shown in Fig. 11.5.

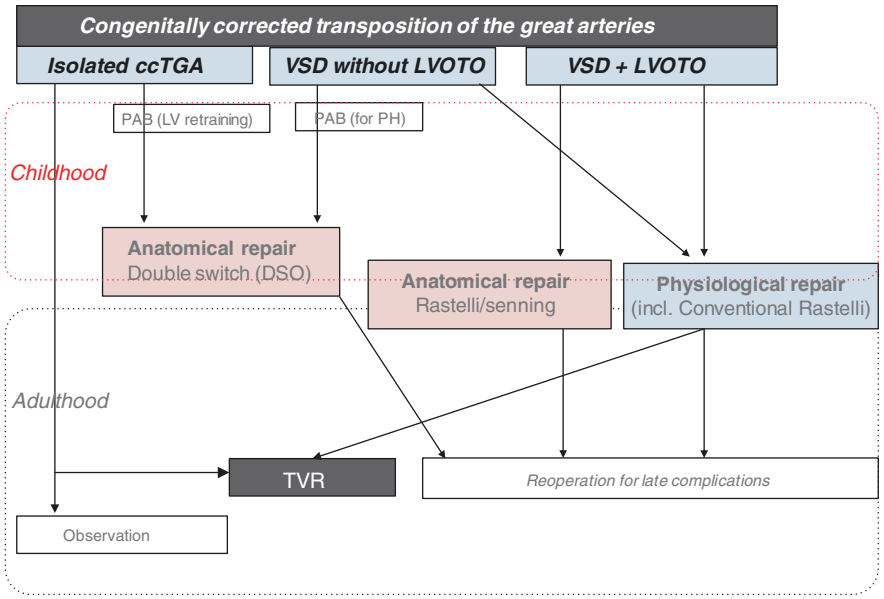

Fig. 11.5 Current surgical strategies for ccTGA in relation to the anatomical types

Despite anatomical repair may improve long-term prognosis, this operation represents major and technically challenging surgical procedures. The physiological repair may be still indicated to a certain group of patients with associated lesion who are contraindicated to anatomical repair.

11.3.1.2 Operative Indication for ccTGA

In general the diagnosis of ccTGA is not in itself an indication for surgical correction. The operative indication should be based on the hemodynamic impact of the associated lesions and the potential of the systemic RV to fail. Definitive indication includes the patients with heart failure due to a large VSD or a VSD with significant LVOTO/PS. When a VSD is present, the indications for operation will be determined by its hemodynamic impact (i.e., Qp/Qs, pulmonary arterial pressure, and pulmonary resistance), as well as the specific morphology of the defect itself. In the presence of pulmonary stenosis the indication for surgery is dictated by the severity of the obstruction (i.e., transpulmonary pressure gradient >50 mmHg) and the degree of cyanosis, hypoxemia, and polycythemia. Other major indications for surgery include the severity and progression of tricuspid valve regurgitation (TR). Replacement of the morphologically tricuspid valve is usually recommended, if the regurgitation is more than grade 2/4 at the time of intracardiac repair of other lesions [12]. Systemic AV valve replacement is relatively common in the adult age.

If the decision is made to achieve the anatomic correction in order to retain anatomically normal physiology to avoid the future deterioration of the systemic RV and TR, the timing and strategies of surgery are always difficult decision, especially in asymptomatic patients with the isolated ccTGA or with hemodynamically

well-balanced lesions. Most centers would not recommend a prophylactic double switch operation for patients without associated abnormalities in whom the RV and tricuspid valve function are normal. Instead, pulmonary arterial banding (PAB) is indicated to the isolated ccTGA to retrain the LV for the subsequent anatomical correction in childhood, especially in patients with the presence of significant TR or RV dysfunction. Some centers consider the anatomical repair is their institutional first choice for the patients who have a balanced systemic and pulmonary blood flow due to VSD and pulmonary stenosis without clinically significant cyanosis to treat [13].

11.3.1.3 PAB for Retraining the Left Ventricle in Isolated ccTGA for DSO [14–16]

In the isolated ccTGA without associated abnormalities, LV pressure will be at pulmonary artery pressure; thereby the LV should be retrained in younger children by the placement of a pulmonary artery band (PAB) to increase LV pressure for DSO. This strategy is thought to be particularly required in pediatric patients with significant tricuspid regurgitation. Some report proposed the early prophylactic pulmonary artery banding in neonate with isolated ccTGA considering the future application of DSO.

In childhood the LV can be successfully retrained using a strategy of progressive pulmonary artery banding in order to provide an increased pressure load on the LV and promote LV hypertrophy. The possible predictors for a successful LV retraining include an LV systolic pressure of at least 70–80% of systemic pressure in childhood or 100% in adolescence and an LV wall thickness or LV mass index that is equivalent to that in a systemic LV. One of the best predictors of failure of retraining is age, with most successfully retrained patients being <10 years of age [17].

11.3.1.4 Pulmonary Artery Banding as a Definitive Palliation

Pulmonary artery banding intended to train the LV has been shown simultaneously to reduce the tricuspid regurgitation and subsequently to improve the RV dysfunction [18]. As regards the mechanism of this phenomenon, it has been speculated that an increase in the LV to RV pressure ratio also may reduce the degree of TR by the shift of the interventricular septum toward the RV [18, 19]. Recently Cools et al. [20] proposed PAB as "open-end" palliation for systemic RV dysfunction and progressive tricuspid regurgitation in 14 patients aged 0.9–14.9 years. Whether this strategy will be applied to adults with isolated ccTGA remains to be seen.

11.4 Clinical Manifestation in Adulthood

11.4.1 Clinical Manifestation of Adult Unoperated ccTGA and the Timing of Diagnosis

The adult with ccTGA presents in various ways, and the clinical course is quite variable depending on the presence and severity of associated lesions. If there are no associated defects, or well-balanced circulation due to the combination of the lesions, the patient will typically be asymptomatic early in life. Diagnosis can be

made later in adulthood, when patients present with complete heart block or congestive heart failure, or incidentally when an ECG, chest X-ray, or echocardiogram is performed for other reasons.

According to the report from the Mayo Clinic adult congenital heart disease database from 1986 to 2000 of the unoperated adult (≧18 years old) with ccTGA, the correct diagnosis was first made at the age older than 18 years old in 66% of patients, 17% of whom were more than 60 years old at the time of diagnosis [21]. And more strikingly, adult patients with unoperated ccTGA are often misdiagnosed in adulthood and are referred late with the presence of symptomatic systemic AV valve (SAVV) regurgitation, significant right ventricular dysfunction, and AV block. Beauchesne et al. reported that 53% of adult unoperated patients had advanced systemic ventricular dysfunction at the time of referral, and, often, they have had severe SAVV regurgitation ≧3/4 in 59% of them [21] and therefore were deemed as "referred late."

11.4.2 Natural History

The natural history of patients with ccTGA is largely dictated by the function of the systemic RV and by the presence or absence of associated abnormalities.

In general, ccTGA is known to have an unfavorable natural history. Most patients with lesions will develop progressive RV dysfunction, relatively early in life, and may not, in great majority, live over 50 years of age. A single center database review from the Toronto Congenital Cardiac Centre for Adults of 131 patients with ccTGA showed a mortality rate of 21% in patients >18 years of age, with a mean age at death of 33 years. Several studies documented an increasing incidence of cardiac failure with advancing age; this late consequence is extremely common by the fourth and fifth decades. A multicenter study conducted by the International Society for Adult Congenital Cardiac Disease of 182 patients with adult ccTGA (≧18 years old) has demonstrated that by the age of 45 years, 67% of patients with associated lesions and 25% of patients without significant associated lesions had congestive heart failure [22].

In contrast, in the setting of ccTGA without any other associated anomalies, the ventricular function is adequate to maintain a "normal" physical activity into adulthood [23, 24]. Some patients, but very rare, may survive to the seventh and eighth decades when no associated anomalies exist being discovered as a chance finding at autopsy [25, 26].

11.4.2.1 Mechanism of Systemic Right Ventricular Failure

The exact mechanism of SV failure is unknown but may relate to microscopic structural features and fiber orientation of the RV myocardium. The morphologically RV lacks the helical myocytic arrangement, essential to the twisting or torsion of the ventricle, and thus being unable to sustain the demands of a systemic ventricle, unlike LV [27].

Other possibilities include coronary perfusion mismatch, because the cardiac hypertrophy caused by the pressure overload on the morphological right ventricle

may surpass the coronary artery oxygen supply, which comes mainly from the right coronary artery. A high incidence of myocardial perfusion defects with regional wall-motion abnormalities and impaired ventricular contractility has been reported [28]. Systemic ventricular dysfunction is also caused by volume overload due to AV valve regurgitation or complete AV block.

11.4.2.2 The Left-Sided Morphologically Tricuspid Valve

The natural history of the left-sided morphologically tricuspid valve is variable. The valve tends to remain competent during the first decade of life but slowly becomes progressively incompetent during the second to fifth decades of life. If there is an Ebsteinoid malformation of the valve, the regurgitation can be seen at birth. In the majority of patients, the SAVV is morphologically abnormal, and with time, there is increasing regurgitation. Beauchesne et al. [21] reported that adult unoperated ccTGA obtained prevalence of systemic valvar regurgitation of 59%, and 68% of them underwent systemic AV valve replacement.

11.4.2.3 Rhythm Abnormality: Complete Atrioventricular Block and Atrial Tachyarrhythmias

Approximately one-tenth of infants born with congenitally corrected transposition have complete heart block [29, 30]. In patients born with normal cardiac conduction, the risk of developing heart block over time increases by 2% per year until it reaches a prevalence of 10–15% by adolescence and 30% in adulthood [31].

Around two-fifths of adult patients, nonetheless, will have normal cardiac conduction throughout their lives. As time passes, the PR interval prolongs, until complete heart block becomes manifest [2].

11.4.2.4 Impact of Atrial Situs on Natural History

Oliver et al. [32] reviewed the long-term outcome of 38 adult ccTGA patients (≧18 years old) to compare the natural history of two anatomic arrangements, situs inversus vs situs solitus, and concluded that the natural prognosis was significantly better in situs inversus than that in situs solitus. Ebstein-like anomaly or spontaneous complete atrioventricular block was rare, and late complications are uncommon in patients with ccTGA with situs inversus. It has been speculated that the good septal alignment in ccTGA with situs inversus might preclude of AV conduction disorders and tricuspid valve abnormalities.

11.4.3 Unnatural History of Operated Patients

11.4.3.1 Long-Term Outcome and Complications After the Physiological Repair

Long-term results of this type of surgical repair are disappointing [33, 34]. Hraska et al. [33] reported the outcomes of 123 patients with ccTGA who underwent a variety of surgical procedures. The 10-year survival after physiological repair was only 67%, and systemic RV dysfunction occurred in 44% of patients. Freedom from RV dysfunction was approximately 40% at 15 years with factors predicting RV

dysfunction being Ebstein's anomaly, tricuspid valve replacement, and postoperative complete heart block.

As regards the impact of the individual procedures, Bogers et al. [35] reviewed the long-term outcome of Rastelli-type and non-Rastelli-type physiological repairs for ccTGA. Actuarial survival at 20 years was 62% for the non-Rastelli group and 67% for the Rastelli group. Freedom from reoperation was 47% at 20 years in the non-Rastelli group and 21% at 19 years for the Rastelli group. Tricuspid valve regurgitation was more often seen in the non-Rastelli group, whereas redo operations were performed predominantly in the Rastelli group mainly for conduit obstruction.

The impact of preoperative TR is crucial in the physiological repair, and it has been documented that when no tricuspid valve regurgitation is present preoperatively, a survival rate of 72% at 30 years can be reached with the physiological repair [36].

11.4.3.2 Long-Term Outcome and Complications After the Anatomical Repair: The DSO and the Rastelli/Senning Procedure

Series of the DSO and the Rastelli/Senning procedure have been published by several groups in large centers with early mortality currently of 0–10% and long-term survival at 10 years of 80–95% [14, 36–41]. Another factor to be considered for interpreting the outcome of anatomical repair is the learning curve for such complex operations and the recent progress in technical modification.

As regards the type of anatomical repair (the DSO vs Rastelli/Senning), more recently it has been reported that the long-term outcome of the DSO was more favorable than the other with an excellent reoperation-free survival, and this procedure should be considered as the optimal procedure of choice [38, 39]. Hiramatsu et al. [38] reviewed 90 patients with ccTGA from 1983 to 2010 underwent the Rastelli–Senning/Mustard procedure (group I) and the DSO (group II). Although survival, including hospital and late mortality at 20 years, was similar (75.7% in group I vs. 83.3% in group II), freedom from reoperation was 77.6% in group I vs. 94.1% in group II. Similarly, Gaies et al. [39] demonstrated that the 10-year survival was superior in the patients who underwent the DSO compared to the Rastelli–Senning.

In contrast, the outcome of the Rastelli/Senning operation for ccTGA with LVOTO and VSD has been improved with the technical modification and the improvement in the materials for extracardiac conduit. For patients with pulmonary stenosis and restrictive VSD, additional DKS anastomosis seems to be an effective approach to avoid postoperative systemic ventricular outflow tract obstruction and surgical heart block, as reported by Hoashi et al. [13].

The Complication of the Anatomical Repair

There is a definite incidence of late occurrence of various complications relating to anatomical repair. The complications after the atrial switch include sinus nodal dysfunction, supraventricular arrhythmias, and problems with the atrial baffle obstruction. If the ventricular switch is used, the left ventricular outflow obstruction due to the stenosis of VSD or intracardiac rerouting baffle, aortic regurgitation, or RV-PA

conduit obstruction and regurgitation can occur. After the DSO, coronary arterial obstruction or stenosis, aortic valvar regurgitation, and pulmonary arterial stenosis may be evidenced.

Outcome of Retraining the Morphological Left Ventricle for Isolated ccTGA

In the isolated ccTGA, during childhood, the subpulmonary LV has been successfully retrained using a strategy of multi-staged PAB in order to provide an increased pressure load on the LV and promote LV hypertrophy. Recent studies documented that the failure rate of LV training before anatomical repair from 0 to 20% is mainly due to LV dysfunction. One of the best predictors of failure of retraining is age, with most successfully retrained patients being <10 years of age [14, 16, 17]. Devaney et al. [16] reported that the two oldest patients, aged 12 and 14 years, had LV failure necessitating removal of the band PAB among 15 patients who underwent PAB for the LV retraining. Poirier et al. demonstrated that the success rate was only 20% in patients older than 12 years old, whereas 62% of the patients less than 12 years old successfully completed LV retraining [17]. Therefore, surgical retraining of the LV is an option in children but not in adults due to unfavorable outcomes. LV retraining is even a challenging task in older children.

LV Dysfunction After DSO

Systolic dysfunction of the LV is relatively common after the double switch procedure [15, 40, 41]. In a retrospective, single-institution study of a total of 113 ccTGA patients, Murtuza et al. [41] demonstrated that freedom from death, transplantation, or poor LV function at 10–14 years was significantly lower in the DSO compared to the Rastelli–Senning group, despite the better actuarial survivals at 10 years in the former group. Quinn et al. [15] reported that patients requiring LV retraining prior to the DSO may be more at risk of developing significant LV dysfunction (6 of 11 patients: 55 %) than patients whose the left ventricle remains at systemic pressure at birth (6 of 33 patients 16 %).

LV dysfunction has been also noticed by several groups, regardless of the proceeded PAB to retrain LV, and is attributed to various factors including complete AV block, aortic regurgitation, and mitral regurgitation [41, 42]. LV function in patients who did not have a large left to right shunt preoperatively demonstrated to be better than in the group who had a large VSD or patent ductus arteriosus as a cause of LV preparedness [40].

The occurrence of LV dysfunction indicates that the DSO is still fraught with imperfections. The Rastelli group apparently more often required conduit revisions but has otherwise performed well.

11.4.3.3 Anatomical vs Physiological Repair

As regards the benefit of the anatomical repair in the actuarial survival rate, Shin'oka et al. [36] reported that there were no statistical differences in the long-term survival rates between the physiological and anatomic surgical repairs. Nevertheless, the outcome of the physiological repair was much worse in patients with significant preoperative TR (the survival at 30 years: 71.9% without TR vs 19.1% with TR). In

contrast, results of the anatomical repair were favorable even for patients with significant TR (71.8% 30 years), and, therefore, the anatomical repair should be a procedure of choice for those patients with significant TR. Conversely, the physiological repair still remains a viable option with an acceptable long-term survival when patients do not have significant TR before their operation.

In 2010 Lim et al. [43] analyzed 167 patients with ccTGA who underwent biventricular repairs and demonstrated the superiority of anatomical repair compared to the physiological repair in the long-term outcome for systemic AV valve and ventricular function.

Freedom from TR and RV dysfunction was significantly higher in the anatomical repair than in the physiological repair (74.5% at 18 year vs 7.4% at 22 year for freedom from TR and 86.3% at 18 years vs 19% at 23 years for freedom from RV dysfunction, respectively). The reoperation-free ratio was 10.1% at 22 years after the physiological repair and 46.2% at 15 years after the anatomical repair. Meta-analysis of individual patient data reported by Alghamdi et al. [44] including 11 studies revealed superiority of the anatomical repair for in-hospital mortality.

11.4.3.4 Long-Term Outcome and Fontan Operation for ccTGA

The Fontan operation has been shown to have a better late survival over the physiological repair and comparable with anatomical repair in terms of freedom from reoperation and the late survival other than in the presence of preoperative significant TR [33, 36, 45–47].

In 2005, Hraska et al. [33] demonstrated that among 123 cases there was a much better 5-year survival in the Fontan patient group than the physiological repair, despite the Fontan patients having lower cardiac output. In a recently published retrospective review of 56 ccTGA patients by Hsu et al. [48], early and late mortality were lower after the Fontan operation compared with the anatomical and conventional repair after 6 and 25 years of follow-up. The 30-day in-hospital mortality rate was 13.3% after the physiological repair (group I), 22.2% after the anatomical repair (group II), and 0% after the Fontan (group III). The overall survival rate was 80% at 16 years in group I, 53% at 13 years in group II, and 100% at 13 years in group III. Freedom from reintervention was 74% in group I, 53% in group II, and 84% in group III. Despite these excellent clinical outcomes of Fontan for ccTGA, however, the advantages of the Fontan must be weighed against its potential disadvantages and long-term complications related to progressive cyanosis, liver cirrhosis, protein-losing enteropathy, and the development of plastic bronchitis.

11.5 Practice of Therapeutic Intervention and Indication

11.5.1 Medical Management

Medical treatment involves management of the failing systemic RV [25, 26] with diuretics, inhibitors of angiotensin-converting enzyme, and digoxin. Afterload reduction with ACE inhibitors or angiotensin II receptor blockers may be less successful for systemic morphological right ventricle than when those used for a

morphological left ventricle [49]. Data are lacking to support the use of beta-blockers to improve ventricular function in ccTGA. For medical therapy of arrhythmias, it is prudent to start antiarrhythmic therapy relatively slowly because of the potential for complete AV block and the possible need for pacemaker implantation.

11.5.2 Intervention for Arrhythmia

Spontaneous complete heart block may be present from birth in approximately 4% of cases. In addition, progressive deterioration in AV conduction can occur throughout life, with an estimated risk of spontaneous heart block of 2% per year [31]. If there are conduction abnormalities, the patient might require implantation of a pacemaker. The status of AV conduction must be monitored regularly with ECG and periodic Holter monitor in adults with ccTGA. Catheter ablation may be indicated to reentrant supraventricular tachycardia due to the casual presence of the accessory pathways or the dual AV node in a variety type of ccTGA, particularly when there is Ebstein malformation of the left-sided tricuspid valve or a straddling AV valve [50].

11.5.3 Cardiac Resynchronization Therapy (CRT)

A substantial number of ccTGA patients will eventually require permanent pacing for complete heart block, with the ventricular lead being placed in the morphologic LV. This may negatively impact on the systemic RV due to the resulting interventricular dyssynchrony. Resynchronization approaches to the systemic RV appear to be an attractive option for ccTGA patients, showing ventricular dyssynchrony. Although numbers are small, this approach appears to have produced a symptomatic improvement in some patients with class III and IV symptoms [51]. A coronary sinus lead is used to pace the RV in ccTGA, since the CS is situated adjacent to the systemic RV and drains into the systemic right atrium in ccTGA patients, which makes transvenous CRT feasible in this group of patients. Because of the heterogeneous anatomy of coronary sinus, visualization of the CS anatomy before attempting CRT implantation is strongly recommended, using different modalities such as CS venography, cardiac CT, and magnetic resonance imaging.

11.5.4 Surgical Management

The indication, procedure, pitfall, and complication of the intracardiac repair of ccTGA in adulthood.

11.5.4.1 Special Consideration for Indication in Adulthood

Initial operation for associated lesions or late consequences of ccTGA is not uncommon even in the elderly patients (i.e., in the fortieth–fiftieth decade). The indications for surgery in adult patients are usually the onset of symptoms due to associated systemic AV valve regurgitation and rarely due to pulmonary overcirculation (a large

VSD) or a significant cyanosis (a VSD with pulmonary stenosis). Nevertheless, even nowadays, the late presentation in presence of cyanosis is particularly common in developing countries due to delay in diagnosis and/or referral. Surgical intervention in the adult, therefore, often consists of systemic AV valve replacement (tricuspid valve replacement: TVR) alone or with a concomitant procedure (i.e., VSD closure) and ideally should be performed before the SV ejection fraction has deteriorated (may be below 45%) [52].

In adult ccTGA, the physiological repair is predominantly adopted, since the morphological and physiological conditions are rarely appropriate to the anatomical repair, and the anatomical repair in adults is associated with a higher mortality. Despite the limitations of significant risk for the systemic right ventricle dysfunction, the physiologic operative approach may be useful in the appropriately selected cases with good right ventricular function and tricuspid valve function.

11.5.4.2 Tricuspid Valve Replacement (TVR) for Tricuspid Regurgitation

Indication procedure and outcome: Tricuspid valve replacement (TVR) for ccTGA is often indicated in adulthood in conjunction with intracardiac repair or the other procedures in unoperated patients or after the physiological repairs. Recently Mongeon et al. [53] reported 46 patients of TVR for ccTGA, 5–72 years of age with a mean age of 40.8 + 14.8 years, with concomitant procedures in approximately 60% of the cases.

In patients with situs solitus, TVR is usually accomplished through a standard left atriotomy anterior to the right pulmonary veins and rarely through a right atriotomy and transseptal approach. In the patient with situs inversus, right-sided tricuspid valve is approached transseptally through the left-sided morphologically right atrium. The selection of prosthetic valves may depend on the patient age. The modification of chordal preservation to retain the RV function is utilized in the high-risk patients [53].

Van Son et al. [52] reported the outcome of 40 patients who underwent TVR at the Mayo Clinic between 1964 and 1993, as the early mortality was 10% and late survival was 68% at 10 years. The survival in patients with RVEF < 44% was 20% at 10 years, whereas in patients with RVEF > 44% the survival was 100%. The authors concluded that surgery for significant TR should be considered at the earliest sign of progressive RV dysfunction before EF more than 44%. Scherptong et al. [54] recently published a multicenter Dutch experience of tricuspid valve surgery in 16 patients with a systemic RV. Eight patients underwent the tricuspid repair, and eight underwent TVR. This study showed a decreased survival after valve repair compared to TVR, which supports the advantage in the approach of primary tricuspid valve replacement in ccTGA. Recent report from Mayo Clinic [53] has demonstrated the 10-year survival or freedom from cardiac transplantation after TVR in 46 patients was still 64.1%, despite the excellent surgical mortality of 0%. Therefore TVR may be only moderately successful at improving survival because it was performed too late. In this report, preoperative variables associated with late mortality were an RVEF < 40%, an RV systolic pressure ≧50 mm Hg, atrial fibrillation, and New York Heart Association functional class III to IV. And also preoperative RVEF

was the only independent predictor of postoperative systemic RV function (RVEF% at >1 year postoperatively), as the postoperative EF was preserved in 63% of patients who underwent surgery with an SVEF ≧ 40% compared with 10.5% of patients who underwent surgery with an SVEF < 40%.

Lundstrom et al. [12] proposed operation for asymptomatic or barely symptomatic patients with ccTGA and cardiac enlargement due to systemic ventricular volume overload.

11.5.4.3 Conventional Repair

In adult ccTGA, a conventional physiological repair consists of closure of the VSD, relief of LVOTO if present, and replacement of the systemic tricuspid valve (Fig. 11.4).

VSD Closure: de Leval Operation (Fig. 11.6)

The VSD is usually well visualized and closed through the mitral valve. In this circumstance, the surgical injury to the long non-branching AV bundle (NBB) should be avoided since it runs immediately adjacent to the anterosuperior edge of the perimembranous VSD on the LV side of the septum. By the DeLaval method in order to avoid damage to the NBB, the interrupted horizontal mattress sutures, reinforced with pledgets, are placed through the septal defect and anchored approximately 4 mm from the edge of the defect within the left-sided anatomical RV surface, particularly along its superior and anterior rims. The remainder of the patch is sutured to the right-sided surface of the septum [55] (Fig. 11.6).

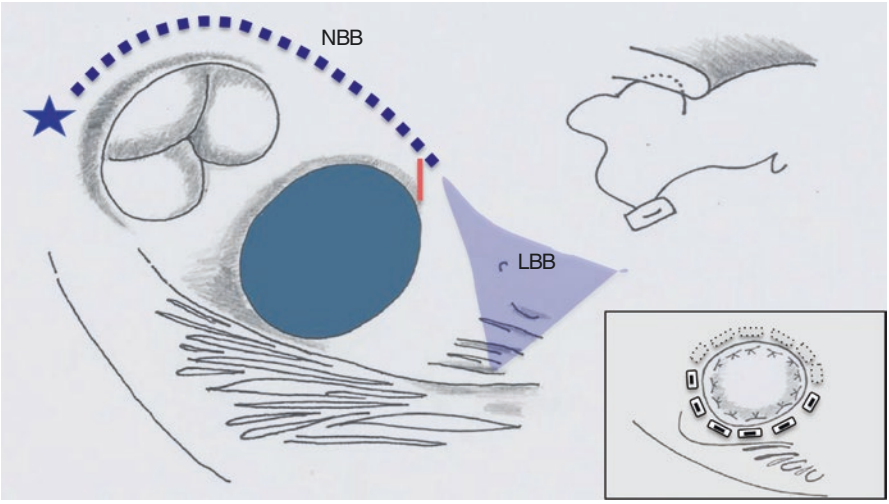

Fig. 11.6 VSD closure (The DeLaval method): in order to avoid damage to the His bundle (NBB), the interrupted horizontal mattress sutures, reinforced with pledgets, are placed through the septal defect within the left-sided anatomical RV surface, particularly along its superior and anterior rims. The remainder of the patch is sutured to the right-sided surface of the septum

In older children or in adult, the VSD may be also approached from the aorta through the aortic valve by placing a patch on left-sided RV surface without a special concern about the conduction disturbance. If the tricuspid valve is concomitantly replaced, access from the LA gives excellent exposure both to the left AVV and to the VSD. Placement of stiches on the morphological RV side of the septum is very easy.

Left Ventricle–Pulmonary Artery Conduit (LV-PA Conduit): Conventional Rastelli (Fig. 11.7)

The placement of the left ventricle–pulmonary artery conduit is needed for subpulmonary LVOTO stenosis, unless there is a resectable obstructing mass of endocardial cushion tissue. Direct relief of muscular subpulmonary stenosis is contraindicated because of the risk of causing permanent AV block and inadequate relief of the obstruction. The placement of the LV-PA conduit is at risk of compression by the sternum, heart block, or injury to the mitral anterior papillary muscle.

The left ventriculotomy is made lower in the midportion to avoid the conduction system, and therefore possibility of injury to the papillary muscles must be considered. Explore the left ventricular free wall digitally through the mitral valve to allow a direct determination of location of the papillary muscles and selection of a safe area for ventriculotomy (Fig. 11.7). Apical placement of the left ventriculotomy for the inflow conduit rather than in the midportion or base placement may avoid these complications [56], although this results in a long and winding extracardiac conduit that may be short-lived because of the proliferation of pseudointima.

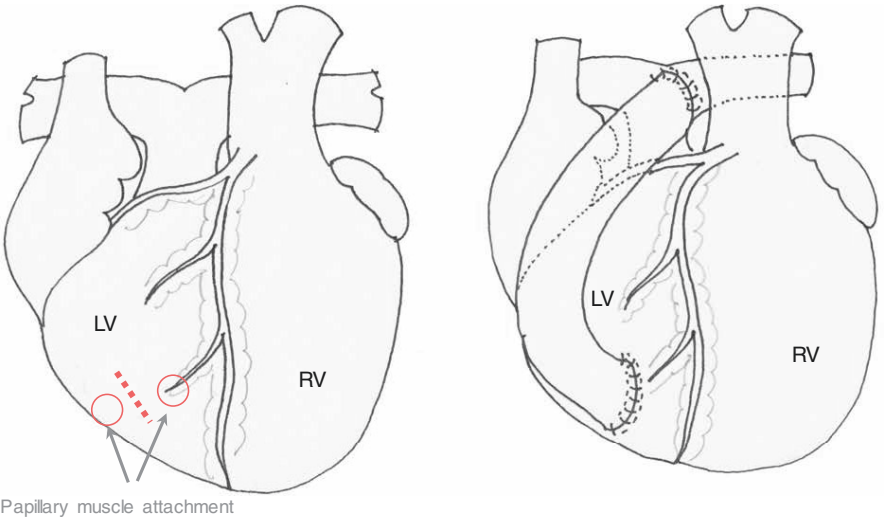

Fig. 11.7 Conventional Rastelli operation: left ventricular to pulmonary artery (LV-PA) conduit

Pitfall for Conventional Repair: Progression of TR After Relieving LVOTO

The physiological repair has been often demonstrated to worsen systemic AV valve regurgitation to significant extent [19, 57]. This may be attributed to the changes of the position of the ventricular septum after relief of pulmonary stenosis, leading to the remodeling of the tricuspid annular configuration and dilatation [12, 18].

Kollars et al. [18] demonstrated that the worsening of TR was presented in 16 patients who underwent either the conventional Rastelli-type repair involving VSD closure or the replacement of a stenotic LV to the pulmonary artery conduit, both of which procedures would decrease the LV pressure postoperatively compared to prior to operation.

Recent studies have demonstrated that the positions of the ventricular septum and septal attachments of the tricuspid valve depend on the pressure gradient between the RV and LV in patients with a ccTGA. Thereby, LV to right ventricle (RV) pressure ratio also may affect the degree of TR [18, 19]. When the LV is exposed to pulmonary pressure, the ventricular septum protrudes into the LV and the septal attachments are pulled away from their annulus, causing coaptation zone deficiency and progressive TR [18, 19]. The study by Koh et al. [19] on 15 patients with discordant atrioventricular connection and pulmonary outflow tract obstruction who underwent the physiological repair using the LV to the pulmonary artery conduit indicated that the left ventricular pressure of <60% of the systemic right ventricular pressure at the systolic phase could be among the deleterious factors in terms of TR and dilatation of the right ventricular cavity. They suggested that the surgeon can choose a smaller conduit than usually used so as to provide some pressure gradient between the LV and the pulmonary artery remained.

Erek et al. [58] experienced an acute severe TR during conduit re-replacement, in a 17-year-old male after the physiological repair, and the degree of tricuspid regurgitation successfully decreased by the pulmonary conduit banding under transesophageal echocardiography guidance, to increase the left ventricular to right ventricular pressure ratio increased from 0.33 to 0.60.

Modification: The Physiological Repair with the One and a Half Ventricular Repair Strategy

Complete relief of LVOTO by pulmonary valvotomy in ccTGA is usually difficult without the use of an extracardiac conduit, due to the outflow tract anatomy and conduction system location. In this regard, the 1.5 ventricle physiological repair in conjunction with the bidirectional Glenn appears to be an effective solution for selected cases and does avoid the need for a valved conduit.

11.5.4.4 Anatomical Repair

Poor right ventricular function or tricuspid valve function would be compelling reasons to choose the anatomic over physiological repair. In some adult ccTGA circumstances, consideration may be given to restoring the left ventricle to the SV, but careful evaluation of its function must be made. If the reroutable unrestrictive VSD

is present, then a Rastelli type of reconstruction for the LV outflow can be done by baffling the VSD to the aorta. Right ventricle-to-pulmonary artery continuity is achieved with a conduit. The DSO has not been considered in adulthood unless preceded PAB for retraining LV or as a palliation for a large VSD has been appropriately done sufficiently early in childhood. Surgical retraining of the LV is an option in children but not in adults due to unfavorable outcomes [59].

Atrial Switch Operation: Senning vs Mustard

The Senning operation utilizes the patients' own tissue to create the venous baffles, while the Mustard operation excises the interatrial septum and uses a pantaloon-shaped baffle usually of heterogeneous materials to complete the inflow switch. In the past, Senning or Mustard procedures were selected as atrial switch procedures, depending on the size of the right atrium, and a large-sized right atrium was thought to be essential for Senning procedure. However, it has been shown that Senning atrial switch can be performed without postoperative caval obstruction for all types of anatomic features including the apicocaval juxtaposition [38]. Furthermore an augmented patch in Mustard has been demonstrated to be easily calcified without growth potential and result in late caval obstruction. Currently, the Senning procedure has become the most widely used procedure as a part of anatomical repair for ccTGA, because of the lower incidence of pathway obstruction, baffle leak, and significant late arrhythmias.

The Senning Procedures: Pitfalls in ccTGA

The Senning procedure is feasible in all atrial switch procedures. When the free wall of the right atrium is relatively narrow, as in ccTGA with dextrocardia or mesocardia, a large pericardial patch to augment a new functional left atrium is a useful option.

The suture line of the free wall of the RA for creating the systemic venous pathway needs the care for the coronary sinus (CS) and conduction system. In the classical Senning, the inferior suture line would run along the tendon of Todaro, leaving the coronary sinus and the triangle of Koch anterior to the suture line, avoiding damaging the normally positioned posterior AV node. The modification of the cutback of orifice of CS to LA is useful. In contrast, in ccTGA, the suture line can be allowed to deviate anteriorly, incorporating the coronary sinus within the IVC pathway. However, the normal topology of the AV node in the triangle of Koch might be seen in the setting of situs inversus and the other varieties of the anatomical type.

Hemi-Mustard and the Bidirectional Glenn Operation

Malhotra et al. [60] propose a modified atrial switch procedure, consisting of a hemi-Mustard procedure to baffle inferior vena caval return to the tricuspid valve in conjunction with a bidirectional Glenn operation in order to avoid complications associated with the complete atrial switch and to reduce inflow volume load into RV.

The hemi-Mustard modification obviates the need for the superior caval suture lines and, as a result, might result in a reduced incidence of these late complications of traditional atrial baffle procedures. Use of the BDG operation also decreases the systemic venous return into the failing RV and reduces volume load to both a dysplastic or Ebsteinoid tricuspid valve and the RV-PA conduit.

The implicated benefits include (1) reduction of atrial baffle related- complications (i.e., sinus node dysfunction and SVC obstruction), (2) prolonged extracardiac RV-PA conduit life, and (3) promoting better coaptation of tricuspid valve and (4) technical simplicity.

The septum primum is resected completely, and, if necessary, the limbus is cut in a superior direction to further enlarge the atrial septal opening. The polytetrafluoroethylene patch is always circular in shape.

Arterial Switch Operation (ASO)

Double switch operation [Senning and arterial switch operation (ASO)] relies on successful coronary translocation. The arterial switch operation was performed using the techniques generally utilized for D-transposition of the great arteries with trapdoor flaps method for coronary implantation and the LeCompte maneuver for neopulmonary reconstruction. The LeCompte maneuver is generally performed if the vessels were in an anteroposterior relationship but not if the vessels were side by side.

In ccTGA, coronary anatomy other than the most frequently encountered 1LCx 2R (according to the Leiden classification) may be associated with incremental operative risk [61].

During DSO, the ventricular septal defect is usually closed through the morphological RV or through the aorta without concern of damaging the conduction system unlike transmitral approach.

Anatomical Rastelli Operation: Intraventricular Rerouting and RVOTR with RV-PA Conduit

Through a right ventriculotomy, a piece of 0.6-mm Gore-Tex patch is placed to direct the left ventricular flow through the ventricular septal defect into the aorta using multiple interrupted sutures inferiorly and posteriorly and continuous sutures around the aortic annulus. The right ventricle and pulmonary artery were established using a pulmonary homograft or other valved conduit (RV-PA conduit).

Previous study revealed that RV volume reduced approximately 20% after the Rastelli operation, because a part of the morphologic RV cavity is occupied by the intraventricular route. Thus, at least 120% of normal size seemed to be essential for better RV function after the anatomical repair [13, 38].

The degree of leftward rotation of the aorta and its relationship to the pulmonary artery dictate the position of the right ventriculo-pulmonary artery conduit. Placement of a conduit to the right side of the aorta results in compression of the conduit by the sternum, which may accelerate conduit dysfunction [62]. It can also result in compression of the right coronary artery by the conduit and myocardial

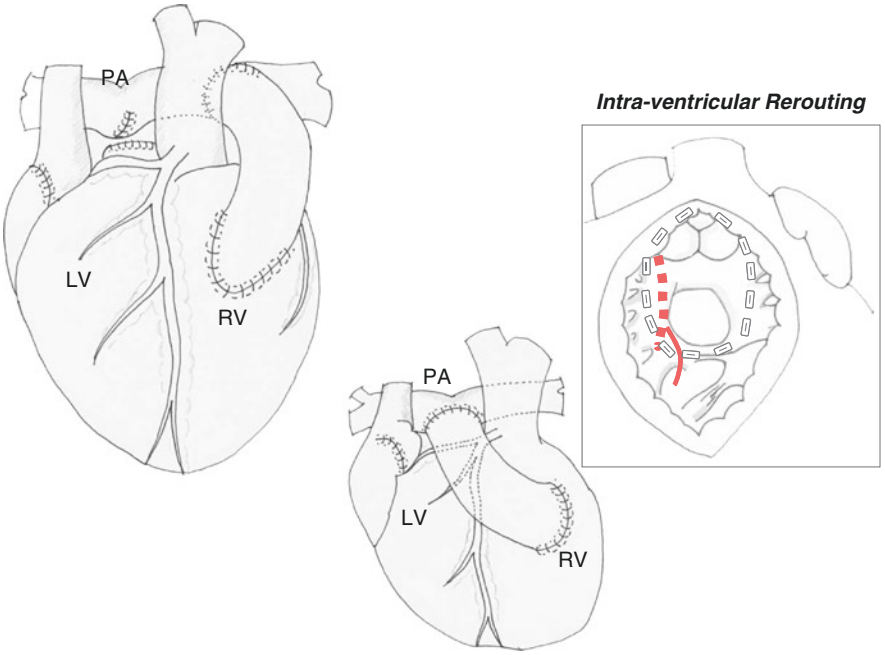

Fig. 11.8 Anatomical Rastelli operation: intraventricular rerouting and the RVOT reconstruction with the right ventricular to pulmonary artery conduit (RV-PA conduit)

ischemia. Therefore, it is ideal to place the conduit on the left side of the aorta whenever possible. Positioning of the conduit to the right of the aorta might be necessary in patients with an aorta to the extreme left (Fig. 11.8).

Additional DKS Anastomosis

Modification of Rastelli procedure for patients with pulmonary stenosis and restrictive VSD [13].

For patients with pulmonary atresia (PA), VSD size was theoretically large enough to create a nonobstructive intraventricular route. Nevertheless, for patients with pulmonary stenosis and restrictive VSD, it is required to create a nonobstructive systemic ventricular outflow tract usually by the VSD enlargement. Although the safe application of VSD enlargement needs the exact location of the conduction system, which may vary depending on the anatomical type. In this situation, additional DKS anastomosis without VSD enlargement has been demonstrated to be an effective approach to avoid postoperative systemic ventricular outflow tract obstruction and surgical heart block (Fig. 11.9).

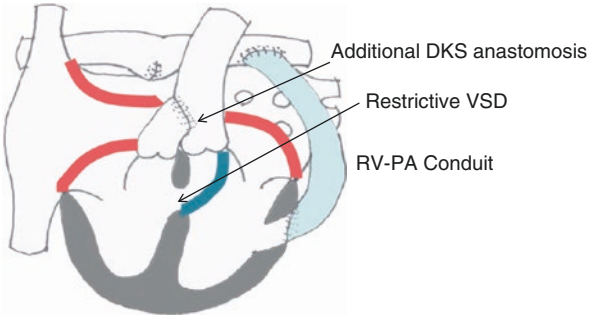

Fig. 11.9 Additional DKS anastomosis: modification of Rastelli procedure for patients with pulmonary stenosis and restrictive VSD

Aortic Root Translocation with Atrial Switch in ccTGA [63, 64]

The ccTGA with isolated pulmonary stenosis is contraindication to the anatomical repair. In some patients, the aortic root translocation (Nikaidoh operation) along with the Senning operation may be applied to achieve the anatomical repair.

The aortic root is harvested with the isolation of coronary bottom of both the right and left or left coronary artery. The harvested aortic root was turned 180, translocated posteriorly, and anastomosed to the original pulmonary root site. The detached coronary arteries were reimplanted in the aortic root. The VSD is closed using a 0.6-mm-thick expanded polytetrafluoroethylene (ePTFE) patch. The right ventricular outflow tract reconstruction was performed using extracardiac conduit.

11.5.4.5 Reparative Procedures and Indication for Late Complications for ccTGA

Indications for redo surgery in patients who have undergone the physiological repair include the initial or repeated TVR and the replacement of obstructive LV to PA conduit. As regards the anatomical repair, there is a definite incidence of late occurrence of various complications relating to each individual procedure in the anatomical repair. The indication of redo operation after the atrial switch includes the atrial baffle obstruction either systemic or pulmonary venous pathway. If the ventricular switch (Rastelli) is used, the left ventricular outflow obstruction due to the stenosis of VSD or intraventricular baffle obstruction, aortic regurgitation, or RV-PA conduit obstruction can be the reasons for reoperation. The indication of redo operation after the ASO as a part of the double switch may include coronary arterial obstruction or stenosis, aortic regurgitation, and pulmonary stenosis. Aortic valve regurgitation is seen more commonly in patients who underwent pulmonary artery banding before ASO as part of staged anatomical repair.

References

1. Wallis GA, Debich-Spicer D, Anderson RH. Congenitally corrected transposition. Orphanet J Rare Dis. 2011;6:22.
2. Schiebler GL, Edwards JE, Burchell HB, Dushane JW, Ongley PA, Wood EH. Congenitally corrected transposition of the great vessels: a study of 33 patients. Pediatrics. 1961;27:II849.
3. Carole AW. Transposition of the great arteries. Circulation. 2006;114:2699–709.
4. Allwork SP, Bentall HH, Becker AE, et al. Congenitally corrected transposition of the great arteries: morphologic study of 32 cases. Am J Cardiol. 1976;38:908e23.
5. Van Praagh R, Papagiannis J, Grunenfelder J, et al. Pathologic anatomy of corrected transposition of the great arteries: medical and surgical implications. Am Heart J. 1998;135:772e85.
6. Anderson KR, Danielson GK, McGoon DC, Lie JT. Ebstein's anomaly of the left-sided tricuspid valve: pathological anatomy of the valvular malformation. Circulation. 1978;58(3 Pt 2):I87–91.
7. Becker AE, Anderson RH, Smith A. Straddling right atrioventricular valves in atrioventricular discordance. Circulation. 1980;61:1133–41.
8. Anderson RH, Becker AE, Arnold R, Wilkinson JL. The conducting tissues in congenitally corrected transposition. Circulation. 1974;50:911–23.
9. Anderson RH. The conduction tissues in congenitally corrected transposition. Ann Thorac Surg. 2004;77:1881–2.
10. Wilkinson JL, Smith A, Lincoln C, Anderson RH. Conducting tissues in congenitally corrected transposition with situs inversus. Br Heart J. 1978;40:41–8.
11. Losekoot TG, Anderson RH, Becker AE, Danielson GK, Soto B. Anatomy and embryology. In: Modern pediatric cardiology; congenitally corrected transposition. New York: Churchill Livingstone; 1983. p. 21.
12. Lundstrom U, Bull C, Wyse RK, Somerville J. The natural and "unnatural" history of congenitally corrected transposition. Am J Cardiol. 1990;65:1222–9.
13. Hoashi T, Kagisaki K, Miyazaki A, Kurosaki K, Shiraishi I, Yagihara T, Ichikawa H. Anatomic repair for corrected transposition with left ventricular outflow tract obstruction. Ann Thorac Surg. 2013;96(2):611–20.
14. Langley SM, Winlaw DS, Stumper O, Dhillon R, De Giovanni JV, Wright JG, Miller P, Sethia B, Barron DJ, Brawn WJ. Midterm results after restoration of the morphologically left ventricle to the systemic circulation in patients with congenitally corrected transposition of the great arteries. J Thorac Cardiovasc Surg. 2003;125:1229–41.
15. Quinn DW, McGuirk SP, Metha C, et al. The morphologic left ventricle that requires training by means of pulmonary artery banding before the double-switch procedure for congenitally corrected transposition of the great arteries is at risk of late dysfunction. J Thorac Cardiovasc Surg. 2008;135:1137e44.
16. Devaney EJ, Charpie JR, Ohye RG, et al. Combined arterial switch and Senning operation for congenitally corrected transposition of the great arteries: patient selection and intermediate results. J Thorac Cardiovasc Surg. 2003;125:500e7.
17. Poirier NC, Yu JH, Brizard CP, Mee RB. Long-term results of left ventricular reconditioning and anatomic correction for systemic right ventricular dysfunction after atrial switch procedures. J Thorac Cardiovasc Surg. 2004;127:975–81.
18. Kollars CAK, Gelehrter S, Bove EL, Ensing G. Effects of morphologic left ventricular pressure on right ventricular geometry and tricuspid valve regurgitation in patients with congenitally corrected transposition of the great arteries. Am J Cardiol. 2010;105:735–9.
19. Koh M, Yagihara T, Uemura H, Kagisaki K, Kitamura S. Functional biventricular repair using left ventricle-pulmonary artery conduit in patients with discordant atrioventricular connections and pulmon- ary outflow tract obstruction—does conduit obstruction maintain tricuspid valve function? Eur J Cardiothorac Surg. 2004;26:767–72.

20. Cools B, Brown SC, Louw J, Heying R, Meyns B, Gewillig M. Pulmonary artery banding as open end palliation of systemic right ventricles: an interim analysis. Eur J Cardiothorac Surg. 2012;41:913–8.
21. Beauchesne LM, Warnes CA, Connolly HM, Ammash NM, Tajik AJ, Danielson GK. Outcome of the unoperated adult who presents with congenitally corrected transposition of the great arteries. J Am Coll Cardiol. 2002;40:285–90.
22. Graham TP, Bernard YD, Mellen BG, Celermajer D, Baumgartner H, Cetta F, Connolly HM, Davidson WR, Dellborg M, Foster E, Gersony WM, Gessner IH, Hurwitz RA, Kaemmerer H, Kugler JD, Murphy DJ, Noonan JA, Morris C, Perloff JK, Sanders SP, Sutherland JL. Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. J Am Coll Cardiol. 2000;36:225.
23. Dimas AP, Moodie DS, Sterba R, Gill CC. Long-term function of the morphological right ventricle in adult patients with congenitally corrected transposition of the great arteries. Am Heart J. 1989;118:896.
24. Kishon Y, Shem-Tov AA, Schneeweiss A, Neufeld HN. Congenitally corrected transposition of the great arteries without associated defects—a study of 10 patients. Int J Cardiol. 1983;3:112.
25. Pezard P, Banus Y, Laporte J, Geslin P, Garnier H, Tadei A. Corrected transposition of the great vessels in aged adults: apropos of 2 patients aged 72 and 80 [in French]. Arch Mal Coeur Vaiss. 1986;79:1637–42.
26. Presbitero P, Somerville J, Rabajoli F, Stone S, Conte MR. Corrected transposition of the great arteries without associated defects in adult patients: clinical profile and follow up. Br Heart J. 1995;74:57–9.
27. Shapiro EP, Rademakers FE. Importance of oblique fiber orientation for left ventricular wall deformation. Technol Health Care. 1997;5:21–8.
28. Hornung TS, Bernard EJ, Jaeggi ET, Howman-Giles RB, Celermajer DS, Hawker RE. Myocardial perfusion defects and associated systemic ventricular dysfunction in congenitally corrected transposition of the great arteries. Heart. 1998;80:322.
29. Cardell LS. Corrected transposition of the great vessels. Br Heart J. 1956;18:186–92.
30. Friedberg DZ, Nadas AS. Clinical profile of patients with congenitally corrected transposition of the great arteries. A study of 16 cases. N Engl J Med. 1970;282:1053.
31. Huhta JC, Maloney JD, Ritter DG, Ilstrup DM, Feldt RH. Complete atrioventricular block in patients with atrioventricular discordance. Circulation. 1983;67:1374–7.
32. Oliver JM, et al. Comparison of outcomes in adults with congenitally corrected transposition with situs inversus versus situs solitus. Am J Cardiol. 2012;110:1687–91.
33. Hraska V, Duncan BW, Mayer JE Jr, et al. Long-term outcome of surgically treated patients with corrected transposition of the great arteries. J Thorac Cardiovasc Surg. 2005;129:182e91.
34. Yeh T Jr, Connelly MS, Coles JG, et al. Atrioventricular discordance: results of repair in 127 patients. J Thorac Cardiovasc Surg. 1999;117:1190e203.
35. Bogers AJ, Head SJ, de Jong PL, et al. Long term follow up after surgery in congenitally corrected transposition of the great arteries with a right ventricle in the systemic circulation. J Cardiothorac Surg. 2010;5:74.
36. Shin'oka T, Kurosawa H, Imai Y, et al. Outcomes of definitive surgical repair for congenitally corrected transposition of the great arteries or double outlet right ventricle with discordant atrioventricular connections: risk analyses in 189 patients. J Thorac Cardiovasc Surg. 2007;133:1318e28.
37. Duncan BW, Mee RB, Mesia CI, et al. Results of the double switch operation for congenitally corrected transposition of the great arteries. Eur J Cardiothorac Surg. 2003;24:11e19.
38. Hiramatsu T, Matsumura G, Konuma T, Yamazaki K, Kurosawa H, Imai Y. Long-term prognosis of double-switch operation for congenitally corrected transposition of the great arteries. Eur J Cardiothorac Surg. 2012;42(6):1004–8.

39. Gaies MG, Goldberg CS, Ohye RG, Devaney EJ, Hirsch JC, Bove EL. Early and intermediate outcome after anatomic repair of congenitally corrected transposition of the great arteries. Ann Thorac Surg. 2009;88:1952–60.
40. Sharma R, Talwar S, Marwah A, Shah S, Maheshwari S, Suresh P, Garg R, Bali BS, Juneja R, Saxena A, Kothari SS. Anatomic repair for congenitally corrected transposition of the great arteries. J Thorac Cardiovasc Surg. 2009;137(2):404–12.e4.
41. Murtuza B, Barron DJ, Stumper O, Stickley J, Eaton D, Jones TJ, Brawn WJ. Anatomic repair for congenitally corrected transposition of the great arteries: a single-institution 19-year experience. J Thorac Cardiovasc Surg. 2011;142(6):1348–57.
42. Bautista-Hernandez V, Marx GR, Gauvreau K, Mayer JE Jr, Cecchin F, del Nido PJ. Determinants of left ventricular dysfunction after anatomic repair of congenitally corrected transposition of the great arteries. Ann Thorac Surg. 2006;82:2059–65.
43. Lim HG, Lee JR, Kim YJ, Park YH, Jun TG, Kim WH, Lee CH, Park HK, Yang JH, Park CS, Kwak JG. Outcomes of biventricular repair for congenitally corrected transposition of the great arteries. Ann Thorac Surg. 2010;89:159–67.
44. Alghamdi AA, McCrindle BW, Van Arsdell GS. Physiologic versus anatomic repair of congenitally corrected transposition of the great arteries: meta-analysis of individual patient data. Ann Thorac Surg. 2006;81:1529–35.
45. Horer J, Schreiber C, Krane S, et al. Outcome after surgical repair/palliation of congenitally corrected transposition of the great arteries. Thorac Cardiovasc Surg. 2008;56:391–7.
46. Talwar S, Ahmed T, Saxena A, et al. Morphology, surgical techniques, and outcomes in patients above 15 years undergoing surgery for congenitally corrected transposition of great arteries. World J Pediatr Congenit Heart Surg. 2013;4:271–7.
47. Talwar S, Bansal A, Choudhary SK, et al. Results of Fontan operation in patients with congenitally corrected transposition of great arteries. Interact Cardiovasc Thorac Surg. 2016; 22:188–93.
48. Hsu KH, Chang CI, Huang SC, Chen YS, Chiu IS. 17-year experience in surgical management of congenitally corrected transposition of the great arteries: a single-centre's experience. Eur J Cardiothorac Surg. 2016;49(2):522–7.
49. Dore A, Houde C, Chan KL, et al. Angiotensin receptor blockade and exercise capacity in adults with systemic right ventricles: a multicenter, randomized, placebo-controlled clinical trial. Circulation. 2005;112:2411–6.
50. Chetaille P, Walsh EP, Triedman JK. Outcomes of radiofrequency catheter ablation of atrioventricular reciprocating tachycardia in patients with congenital heart disease. Heart Rhythm. 2004;1:168–73.
51. Al Fagih A, Al Najashi K, Dagriri K, Al Otay A, Al Ghamdi SA. Feasibility of cardiac resynchronization therapy in a patient with complex congenital heart disease and dextrocardia, facilitated by cardiac computed tomography and coronary sinus venography. Hell J Cardiol. 2010;51(2):178–82.
52. van Son JA, Danielson GK, Huhta JC, et al. Late results of systemic atrioventricular valve replacement in corrected transposition. J Thorac Cardiovasc Surg. 1995;109:642–52.
53. Mongeon FP, Connolly HM, Dearani JA, et al. Congenitally corrected transposition of the great arteries: ventricular function at the time of systemic atrioventricular valve replacement predicts long-term ventricular function. J Am Coll Cardiol. 2011;57(20):2008–17.
54. Scherptong RW, Vliegen HW, Winter MM, et al. Tricuspid valve surgery in adults with a dysfunctional systemic right ventricle: repair or replace? Circulation. 2009;119(11):1467–72.
55. De Leval MR, Bastos P, Stark J, et al. Surgical technique to reduce the risks od heart block following closure of ventricular septal defects in atrioventricular discordance. J Thorac Cardiovasc Surg. 1979;78:515.
56. Aeba R, Katogi T, Koizumi K, et al. Apico-pulmonary artery conduit repair of congenitally corrected transposition of the great arteries with ventricular septal defect and pulmonary outflow tract obstruction: a 10-year follow-up. Ann Thorac Surg. 2003;76:1383–8.

57. Sano T, Riesenfeld T, Karl TR, Wilkinson JL. Intermediate-term outcome after intracardiac repair of associated cardiac defects in patients with atrioventricular and ventriculoarterial discordance. Circulation. 1995;92(9 Suppl):II272–8.
58. Erek E, Abud B, Oz K, Güzeltas A. Preservation of systemic tricuspid valve function by pulmonary conduit banding in a patient with corrected transposition of the great arteries. Interact Cardiovasc Thorac Surg. 2012;15:332–4.
59. Duncan BW, Mee RB. Management of the failing systemic right ventricle. Semin Thorac Cardiovasc Surg. 2005;17:160–9.
60. Malhotra SP, Reddy VM, Qiu M, Pirolli TJ, Barboza L, Reinhartz O, Hanley FL. The hemi-Mustard/bidirectional Glenn atrial switch procedure in the double-switch operation for congenitally corrected transposition of the great arteries: rationale and midterm results. J Thorac Cardiovasc Surg. 2011;141(1):162–70.
61. Ismat FA, Baldwin HS, Karl TR, Weinberg PM. Coronary anatomy in congenitally corrected transposition of the great arteries. Int J Cardiol. 2002;86:207–16.
62. Ilbawi MN, Ocampo CB, Allen BS, Barth MJ, Roberson DA, Chiemmongkoltip P, Arcilla RA. Intermediate results of the anatomic repair for congenitally corrected transposition. Ann Thorac Surg. 2002;73(2):594–9.
63. Kwak JG, Lee CH, Lee C, Park CS. Aortic root translocation with atrial switch: another surgical option for congenitally corrected transposition of the great arteries with isolated pulmonary stenosis. J Thorac Cardiovasc Surg. 2010;139(6):1652–3.
64. Ikai A, Koizumi J, Kin H, Mukaida M, Takahashi S, Oyama K, Okabayashi H. Aortic translocation using the hemi-mustard procedure for corrected transposition. Ann Thorac Surg. 2012;94(3):996–8.