Functional outcome of anatomic correction of corrected transposition of the great arteries

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Abstract

Objective: Anatomic correction of corrected transposition of the great arteries, utilizing the morphologic left ventricle as a systemic pumping chamber, is considered the preferred method. The purpose of the study was to analyze the intermediate functional outcome following anatomical correction. Methods: Between 1997 and 6/2010, 23 patients with corrected transposition of the great arteries and associated lesions underwent anatomical correction. Seventeen (74%) and six patients (26%) had situs solitus {S,L,L} and situs inversus {I,D,D}, respectively. Fifteen patients (65%) had undergone 18 palliations before the corrective operation. The median age at palliation was 0.23 years, with a range of 0.016–8.4 years. A corrective, modified Senning–arterial switch procedure was performed in nine patients, 13 patients underwent a modified Senning–Rastelli procedure, and in one patient a combination of modified Senning and aortic translocation (Bex/Nikaidoh) was used. The median age at the corrective operation was 2 years (from 0.3 to 15.7 years). Results: There was no mortality or heart transplant within the mean follow-up of 3.4 years. Freedom from reintervention was 77% at 5 years. There were no signs of obstruction of the systemic and pulmonary venous tunnels. The function of both ventricles was normal in all patients, even in the four patients who required retraining of the left ventricle. Mild aortic regurgitation was noticed in three patients. Preoperatively detected significant tricuspid regurgitation either disappeared or became trivial after the operation in all the six patients. All patients except two are in sinus rhythm; one patient is pacemaker-dependent preoperatively and one is pacemaker-dependent postoperatively. There were no clinically apparent neurological problems. All patients, but one, are in the New York Heart Association (NYHA) class I. Conclusions: Anatomic correction of corrected transposition of the great arteries can be performed in selected patients without mortality and with acceptable morbidity. The mid-term functional outcome is excellent, resulting in normal ventricular function, even in retrained left ventricles, and minimal incidence of complete heart block. The long-term function of the aortic valve, intraventricular tunnels, conduits, and ventricles requires close surveillance.

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Keywords: Corrected transposition of the great arteries; Senning procedure; Rastelli procedure; Arterial switch operation; Aortic root translocation

1. Introduction

The long-term outcome of patients with corrected transposition of the great arteries (ccTGA) and associated lesions after 'classical' surgical repair is uncertain [1,2]. Anatomical correction, utilizing the morphologic left ventricle (LV) as a systemic pumping chamber and the mitral valve (MV) as the systemic atro-ventricular (AV) valve, is considered the preferred method [3–7], especially for patients either with tricuspid valve (TV) regurgitation before surgery, with Ebstein's malformation of the TV, or with right ventricle (RV) dysfunction [2]. Anatomical correction represents a group of procedures in which the atrio-ventricular discordance is 'corrected' by an atrial switch (Senning or Mustard) [8], and ventriculo-arterial discordance is 'corrected' by an arterial switch operation (ASO) [9], by the Rastelli procedure [10], or by translocation of the aortic root (Bex/Nikaidoh (BN) operation) [11], depending on the underlying anatomy of the left ventricular outflow tract (LVOT) and/or morphology of the ventricular septal defect (VSD). Three different types of anatomical correction are therefore recognized: (1) double switch (Senning + ASO (S–ASO)); (2) Senning + Rastelli (S–R); and (3) Senning + Bex/Nikaidoh (S–BN). The purpose of this study was to evaluate our midterm experience with anatomical correction of ccTGA.

2. Materials and methods

2.1. Methods

A retrospective chart review was undertaken to identify all patients with ccTGA in whom an anatomical correction had been performed.
2.2. Patients

Between 1997 and June 2010, 23 consecutive patients with ccTGA and well-balanced ventricles (Table 1) underwent anatomical correction at three institutions: Pediatric Cardiac Centre, Bratislava, Slovakia (four patients); Eppendorf University Hamburg, Germany; German Pediatric Heart Centre (two patients); and Asklepios Clinic Sankt Augustin, Germany (17 patients). One primary surgeon (VH) was involved.

There were 12 (52%) female patients. Seventeen (74%) and six patients (26%) had situs solitus [S,L,L] and situs inversus [I,D,D], respectively. In the [S,L,L] group, 13 patients had levocardia and four had dextrocardia. In the [I,D,D] group, three patients had levocardia, two had mesocardia, and one patient had dextrocardia.

A complex left ventricular outflow tract obstruction (LVOTO) was present in 15 patients. Apart from one patient with only an LVOTO (subpulmonary membrane with accessory MV tissue), the other patients presented with a combination of VSD with pulmonary atresia (five patients) and VSD with subvalvar and/or valvar pulmonary stenosis (nine patients). Seven patients presented with a VSD only; three of them had a restrictive type of VSD. All patients with a VSD had the conoventricular type, but three patients had an inlet type of VSD. Six patients had Ebstein’s anomaly of the TV associated with mild-to-severe regurgitation. One patient had Ebstein’s anomaly of the TV only. One patient had a straddling of the TV; another patient had a straddling of the MV. Two patients had aygous continuation of the inferior vena cava (IVC); one of them had [I,D,D] and levocardia with left isomerism. In this patient, the interatrial septum with restrictive foramen ovale was found ‘shifted’ to the right; therefore, all pulmonary veins were connected with the left-sided right atrium, creating the appearance of total anomalous pulmonary venous drainage. One patient had hypoplasia of the distal transverse arch with coarctation of the aorta.

2.3. Treatment management

Fifteen patients (65%) had undergone a total of 18 palliations before the final operation (Table 2). The median age at palliation was 0.23 years, with a range of 0.016—8.4 years.

2.3.1. 1st palliation

Seven patients underwent pulmonary artery banding (PAB), either to control the pulmonary blood flow (three patients) or to train the LV (four patients). A ‘loose’ PAB was placed to achieve 50% of systemic pressure in the LV and to let the patients ‘grow into’ the PAB.

Eight patients had undergone a shunt type of procedure to increase pulmonary blood flow. Four patients had received a modified Blalock–Taussig (BT) shunt and one had a stented patent ductus arteriosus. In different institutions, two patients had undergone bidirectional Glenn anastomosis + atrial septectomy. One patient was palliated using the Fontan procedure.

2.3.2. 2nd palliation

Three patients underwent additional palliations in different institutions. In one patient, the BT shunt was replaced by a BDG. In another, in whom the pulmonary blood flow was provided by a BDG, an additional central shunt was placed. In the patient with the Fontan operation, the intracardiac tunnel was taken down due to tunnel thrombosis.

2.3.3. Anatomical correction

The indications, timing, and type of anatomical correction of ccTGA varied according to the morphology of the heart, the clinical state of the patient, and the patient’s age [4,12]. There was a trend toward performing the final correction at about 2 years of age.

Primary correction was achieved in eight patients (35%). The median age at final operation was 2.03 years, with a range of 0.3—15.7 years. The median interval between palliation and correction was 1.34 years, with a range of 0.4—11 years. Anatomical correction included nine patients with S–ASO, nine patients with S–R; four patients with Senning–Rastelli (S–R); and one patient with S–BN (Table 3).

2.3.3.1. Senning—arterial switch procedure. Nine patients underwent an S–ASO procedure, two of them as a primary procedure, the rest after PAB. The median interval between PAB, placed for training the LV, and S–ASO was 1.13 years. The median age at correction was 2.7 years, with a range of 0.7—9 years. Apart from S–ASO with VSD closure and debanding of the pulmonary artery where required, pulmonary valvotomy and shaving of the pulmonary valve were performed in one patient and another patient underwent resection of the accessory tissue of the MV and resection of the subpulmonary membrane. One patient required a pacemaker implantation.

2.3.3.2. Senning–Rastelli procedure. Nine patients underwent the S–R procedure, five of them as a primary

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Table 1. Patients characteristics.

<table>
<thead>
<tr>
<th>Situs</th>
<th>Pts</th>
</tr>
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<tbody>
<tr>
<td>Solitus [S,L,L]</td>
<td>17 pts</td>
</tr>
<tr>
<td>Invs [I,D,D]</td>
<td>6 pts</td>
</tr>
<tr>
<td>Anatomic subtype</td>
<td></td>
</tr>
<tr>
<td>ccTGA only</td>
<td>1 pt</td>
</tr>
<tr>
<td>ccTGA + LVOTO</td>
<td>1 pt</td>
</tr>
<tr>
<td>ccTGA + VSD</td>
<td>7 pts</td>
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<tr>
<td>ccTGA + VSD + LVOTO</td>
<td>14 pts</td>
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</tbody>
</table>

Pts – patients; ccTGA – corrected transposition of the great arteries; LVOTO – left ventricular outflow tract obstruction; VSD – ventricular septal defect.

Table 2. Palliative procedures.

<table>
<thead>
<tr>
<th>Type of procedure</th>
<th>N</th>
</tr>
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<tbody>
<tr>
<td>Pulmonary artery banding</td>
<td>7</td>
</tr>
<tr>
<td>Modified Blalock-Taussig shunt</td>
<td>4</td>
</tr>
<tr>
<td>PDA stenting</td>
<td>1</td>
</tr>
<tr>
<td>Other systemic to pulmonary shunt</td>
<td>1</td>
</tr>
<tr>
<td>Bidirectional Glenn anastomosis + atrial septectomy</td>
<td>1</td>
</tr>
<tr>
<td>Bidirectional Glenn anastomosis</td>
<td>2</td>
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<tr>
<td>Fontan</td>
<td>1</td>
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<td>Fontan takedown</td>
<td>1</td>
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</tbody>
</table>

PDA – patent ductus arteriosus.
Sizing maker

Concomitant Type correction full

2.3.3.4. underwent MV procedure, the rest after shunt procedures or duct stenting to increase pulmonary blood flow. The median age at correction was 1.2 years, with a range of 0.3–6.8 years. As a concomitant procedure, one patient required pacemaker implantation. In three patients, the VSD was enlarged, one patient underwent resection of accessory tissue of the MV and in two patients the pulmonary artery was enlarged using a patch. In all patients, the right ventricular outflow tract was reconstructed using a Contegra conduit (Contegra® Medtronic Ltd).

2.3.3.3. Senning–Rastelli procedure. Four patients reported to our institution with the BDG in place. A half-modified Senning procedure was performed in three patients, and in one patient with {I,D,D} and levocardiectomy the IVC was detached from the right ventricle and anastomosed with the left atrium, while the pulmonary veins were committed to the left-sided MV. Subsequently, a Rastelli type of correction was performed. As a concomitant procedure, one patient underwent an aortic valve plasty (sub-commissural downsizing of the annulus) due to more than moderate aortic regurgitation.

2.3.3.4. Senning–Bex/Nikaidoh procedure. One patient underwent a primary S–BN procedure with translocation of the TV attachments at the age of 2 years.

2.3.4. Surgical technique and morphological considerations

The standard technique of cardiopulmonary bypass with full flow and moderate hypothermia (28 °C) was used. Myocardial protection was provided by crystalloid antegrade cardioplegia.

2.3.4.1. Atrial switch — modified Senning. Our technique of a modified Senning procedure differs from the original concept [8], in order to adapt to the specific atrial morphology of the ccTGA and to be ‘situs’ independent. The right atrium is opened in an oblique manner, aiming toward the medial aspect of the IVC—right atrial junction, but close to the AV groove. The idea is to develop and preserve as large a flap of free anterior wall of the right atrium as possible for the systemic venous baffle. Subsequently, the interatrial septum is completely excised. The posterior wall of the systemic baffle is developed with a trapezoid-shaped Goretex patch (Gore®), which is sutured anterior to the left-sided pulmonary veins and posterior to the base of the left atrial appendage. The patch should be seated as low as possible beneath the superior vena cava (SVC)—right atrial junction (area of resected limbus of the fossa ovalis) to prevent obstruction. A large incision is made at the entrance point of the right pulmonary veins. The anterior wall of the systemic venous baffle is developed by suturing the right atrial free wall to the edge of the excised atrial septum between the MV and TV. Preferably, the coronary sinus is kept in the systemic venous atrium so that it is approachable for future electrophysiological studies. However, in {I,D,D} it is better to run the suture line behind the coronary sinus to avoid a ‘normal’ (inferior) position of the AV node and the conduction system [12]. As a part of the double-switch procedure, when the systemic venous baffle is completed, the VSD is approached working through the MV. Interrupted pledged sutures are placed from the left side of the septum and a Dacron patch is seated. Alternatively, one could consider closing the VSD by working through the right ventriculotomy, thus eliminating undue tension on the crux cords. Routinely, for completion of the pulmonary venous atrium, the in situ pericardium is utilized (Shumaker modification) [8,9]. The suture line commences between the pericarial wall and the pericardial reflection, which was left intact around the superior aspect of the right pulmonary artery and vein. The suture line is kept superior and away from the sinus node. Another suture line is commenced at the very inferior end of the anterior lip of the incised right pulmonary veins and pericardium. The suture line swings anterior, reaching the remnant of the right atrial wall. Adequate capacity of the left atrial chamber is created, preserving the optimal position of the MV.

2.3.4.2. Atrial switch — half-Senning procedure. Baffle rerouting of the IVC to the TV was indicated if a BDG was already in place. After resection of the interatrial septum, the IVC entrance was connected to the TV annulus using an appropriately shaped and longitudinally opened Gore-Tex prosthesis (Gore®). The coronary sinus was incorporated into the baffle.

2.3.4.3. Arterial switch procedure. The arterial switch is the procedure of choice if the LV has been exposed to a sufficiently high blood pressure and is therefore able to take over acutely at systemic pressure, and if the LVOT is free, or surgical relief of obstruction on any level is amenable. The technique of ASO for ccTGA is the same as for D-TGA [9,13].

2.3.4.4. Rastelli procedure. The Rastelli operation was chosen in the case of pulmonary atresia or pulmonary stenosis with a non-resectable LVOTO and adequate capacity of the RV. The technique does not differ from a Rastelli correction of D-TGA with LVOTO [10].

2.3.4.5. Bex/Nikaidoh procedure. A BN operation was indicated if creation of a straight intraventricular tunnel was impossible due to an inlet and/or restrictive VSD. A modified

<table>
<thead>
<tr>
<th>Table 3. Corrective procedures.</th>
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<tr>
<td>Type of procedure</td>
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<tr>
<td>S–ASO</td>
</tr>
<tr>
<td>S–R</td>
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<tr>
<td>S–BN</td>
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<tr>
<td>Concomitant procedure</td>
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<tr>
<td>Pulmonary valve plasty</td>
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<tr>
<td>Pulmonary artery plasty</td>
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<tr>
<td>Resection of accessory tissue of MV</td>
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<tr>
<td>Aortic valve plasty</td>
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<tr>
<td>Enlargement of VSD</td>
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<tr>
<td>Translocation of TV chordal attachments</td>
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<tr>
<td>PM implantation</td>
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<tr>
<td>S–ASO — Senning and arterial switch procedure; S–R — Senning and Rastelli; S–R — half Senning and Rastelli; S–BN — Senning and Bex/Nikaidoh procedure; MV — mitral valve; VSD — ventricular septal defect; TV — tricuspid valve; PM — pacemaker.</td>
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technique of posterior translocation of the aortic root with coronary artery detachment was used [11,14].

3. Functional outcome and follow-up

The postoperative result was assessed in all patients at discharge and during follow-up. The patients underwent clinical assessment, two-dimensional echocardiography (ECG), and Doppler assessment of the intracardiac repair. Follow-up data were complete in all but one of the surviving patients (96%). By using a questionnaire, as well as by direct contact with the referring cardiologists, data were collected on morbidity, echocardiography, ECG, New York Heart Association (NYHA) classification, and actual medication.

4. Statistical analysis

Data were analyzed using a statistical program (JMP Statistical Analysis, Cary, NC, USA). Descriptive statistics are expressed as median, or mean and range. The Kaplan—Maier method was used to determine event-free survival curves.

5. Results

There was no early or late death, with a mean follow-up of 3.4 (0.1—15.4) years. In the early postoperative period (>30 days), three patients suffered from low cardiac output and nine patients required treatment for hemodynamically relevant arrhythmias. Due to malignant arrhythmias, one patient underwent successful resuscitation in the early postoperative period. Two patients needed revision due to bleeding. One of these patients underwent an early transcatheter intervention, in which a stent was placed into the kinked left pulmonary artery with subsequent rupture, thus requiring urgent patch plasty of the ruptured pulmonary artery. The chest was left open in six patients. Chylothorax, requiring a diet, was noted in six patients. Infection complications, either catheter-related sepsis or pneumonia, were treated in eight patients. One patient with a low cardiac output, arrhythmias, and sepsis, suffered from pancreatitis.

The event-free survival rate was 94% (0.681—0.992), 57% (0.244—0.947), and 57% (0.130—0.854) at 1, 5, and 10 years, respectively. There were three redos and no reintervention (Fig. 1). All reoperations were associated with S—R repair ($p = 0.03$). Two patients underwent a conduit change due to conduit obstruction, on average 4.5 years after corrective operation. The patient who suffered from pancreatitis during the early postoperative period was readmitted with a pancreatic pseudo-cyst and required operation.

At the last echo examination, there were no signs of obstruction of the systemic or pulmonary venous tunnels. The function of both ventricles was normal in all patients, even in those who required retraining of the LV before anatomical correction. Preoperatively detected significant TV regurgitation either disappeared or became trivial after operation in all six patients. Two patients suffered from mild MV regurgitation. A hemodynamically restrictive re-VSD was found in three patients.

After the S—ASO procedure, trivial/mild neoaortic regurgitation was noticed in 5/2 patients; in one patient the neoaortic root was significantly dilated.

Following S—R and S—R, none of the patients showed a significant (>30 mmHg) LVOTO. A peak gradient of <20 mmHg was measured in the LVOT in two patients. One patient who underwent aortic root plasty suffered from mild neoaortic regurgitation without progression.

After the S—BN procedure, there was no aortic regurgitation and a free, but elongated, LVOT (Fig. 2).

All patients are in sinus rhythm, but three, one preoperatively and two postoperatively, suffered from complete heart block. Two surgical blocks were related to VSD closure; however, one patient is no longer dependent on a pacemaker. One patient has a grade I AV block and another patient has a complete right bundle branch block.

All patients but one are in the NYHA class I. Physical limitations are obvious in one patient, who has suffered from abdominal problems (status post pancreatitis).

Cardiac medication is needed in 13 patients. Apart from diuretics, four patients are receiving antiarrhythmic treatment and three are taking angiotensin-converting enzyme inhibitors. There were no clinically apparent neurological

Fig. 1. Event-free survival in years. Numbers of patients at risk are indicated in the rectangle.

Fig. 2. Status post modified Senning–Bex/Nikaidoh correction in ccTGA (I,D,D) with inlet ventricular septal defect. (A) Angiography shows elongated newly created left ventricular outflow without obstruction. (B) Angiography shows translocated aorta without any regurgitation and with normal coronaries.
problems. The patients are doing well and growth is adequate.

6. Discussion

Results with the traditional approach, preserving the RV as a systemic ventricle and correcting only associated lesions, have clearly demonstrated that TV and RV function is the Achilles heel of the physiology of ccTGA [1,2]. Anatomic correction, utilizing the LV as the systemic pumping chamber and the MV as the systemic AV valve, has therefore been proposed [3] in the hope that it might serve patients better in the long run. At present, the midterm outcomes after anatomical correction are encouraging [4–7,13]; however, long-term outcomes show that anatomical correction has only a slight advantage over other types of surgical treatment. Long-term survival and functional benefits after anatomic correction were particularly demonstrated in patients with preoperative TV regurgitation [2].

Our data show that all types of procedures are safe and provide excellent midterm outcome with an acceptable degree of morbidity for such complex procedures. In this study, there was no death and no need for heart transplant among the 23 patients who underwent anatomical correction, at a mean follow-up 3.4 years. This result corresponds with the early mortality rate, which was between 0% and 15%, and midterm survival benefits between 70% and 100% at 10 years of follow-up, with no difference between S—ASO and S—R [4–6,13]. An event-free survival rate of between 70% and 85% in 10 years has been reported. Reinterventions or reoperations are related to obstructions of the systemic and venous baffles, obstructions of the intraventricular tunnel, to aortic or mitral regurgitation, residual VSDs, a failed conduit, or to a complete heart block with the need for pacemaker implantation [2–7,13].

Deterioration of LV function is the main concern after anatomical correction. The incidence of late LV dysfunction is up to 20% in midterm follow-up, with a constant need for heart transplant. The negative impact of complete heart block, progressive aortic regurgitation, the need for retraining the LV, especially in older patients, and intraventricular tunnel obstruction, on the long-term function of the LV, are well documented [5,6,15]. Our data demonstrate the preservation of normal left ventricular function after correction, even in patients who required LV retraining. The low pacing requirements, the low incidence of neoaortic regurgitation, the gradual retraining of the LV in younger patients, and unobstructed intraventricular tunnels may contribute to the preservation of LV function in our patients. However, the short follow-up precludes any firm conclusions in this regard.

Universal improvement in the function of the RV and TV occurs when they are exposed to the lower pressures in the pulmonary circulation after anatomical correction. Even in the presence of dysplasia of the TV with severe preoperative regurgitation, concomitant TV repair is only rarely needed at the time of the anatomical correction [4,5]. In our series, preoperatively detected significant TV regurgitation either disappeared or became trivial after operation in all six patients.

Patients with ccTGA are at risk for complete heart block, even without surgery, and it has been estimated to affect approximately 2% of patients per year after diagnosis [16]. Surgically acquired heart block is related to resection of the LVOT or to closure and/or enlargement of the VSD. The incidence of surgically acquired heart block is between 5% and 20% [5–7,13], with freedom from pacemaker implantation 72% at 15 years [2]. All our patients are in sinus rhythm, but one patient was pacemaker-dependent preoperatively, and one postoperatively. The high incidence of hemodynamically relevant arrhythmias in the early postoperative period is worrisome and might indicate an ongoing risk for the future.

As the long-term problems are more or less specific to the type of procedure, we believe that modification of the treatment protocol might decrease late morbidity and have a positive impact on late survival.

6.1. Issues related to the Senning procedure

Based on the experience with the Senning procedure for D-TGA, the incidence of baffle-related complications is less than 10%, and the sinus rhythm is preserved in nearly 80% of the patients in the long run [17]. The Senning procedure for ccTGA is technically more challenging because of the misalignment of the interventricular and interatrial septum (in {S,L,L}) and the reduced size of the free right atrial wall, which is associated with the high incidence of mesocardia or dextrocardia in {S,L,L} or levocardia or mesocardia in {I,D,D}. The proposed modified technique is simple, highly reproducible, and applicable, regardless of the situs and position of the apex of the heart. There are several important technical points to be borne in mind:

1. The initial incision of the right atrium should be close to the AV groove to preserve the anterior free wall of the right atrium for the systemic venous baffle.
2. The interatrial septum should be completely resected.
3. The superior aspect of the limbus of the fossa ovalis must be effectively cut off to avoid baffle obstruction from the SVC to the TV. At the same time, the trapezoid-shaped patch, which is used for the posterior wall of the systemic baffle, must be seated low beneath the SVC—right atrial junction.
4. It is important to provide adequate capacity of the pulmonary venous atrium and to prevent distortion and tension on the AV groove, thus keeping the MV in an optimal position.

With the technical aspect of the modified Senning procedure well accomplished, the operation is feasible, regardless of age, and allows an earlier indication for correction. Our youngest patient was 3 months old. The risk of systemic and pulmonary baffle obstructions is minimal, even in {S,L,L} with dextrocardia or on {I,D,D} with levocardia. Furthermore, this technique has the potential to provide adequate capacity of the pulmonary venous atrium, to preserve optimal geometry of the MV, to minimize damage to the sinus node, and to make the coronary sinus accessible for electrophysiological studies or intervention.
The incidence of atrial baffle obstructions, which lead to intervention or reoperations after anatomical correction in patients with ccTGA, is reported to be between 3% and 10%. More frequently, obstruction of the upper part of the systemic venous tunnel has been seen [4–6,13]. In our group of patients, there were no baffle-related obstructions.

6.2. Issues related to ASO

The first problem is the progressive dilatation of the neoaortic root, with the risk of development of neoaortic valve regurgitation. Trivial-to-mild neoaortic regurgitation is frequently seen [5,13]. The incidence of moderate-to-severe aortic regurgitation is up to 15% in midterm follow-up. PAB has a negative impact on the root geometry and postoperative function of the neoaortic valve [5,6]. If technical imperfections of the PAB are ruled out, one can speculate that after too tight a PAB, the sudden increase in wall stress on the pulmonary root may be associated with elastic fiber fragmentation, which leads to significant root dilatation, as is typically seen after a failed Ross procedure [18]. This would support the placement of a 'loose' PAB, thus allowing mechanical adaptation phenomena to gradually take place in the wall of the pulmonary root.

Another technical issue is the preservation of the diameter of the sino-tubular junction during coronary transfer. In our series of nine S–ASO patients, seven had a PAB. In the mean follow-up of 3.4 years, trivial regurgitation was noticed in five patients and mild regurgitation in two — one of them had root dilatation.

The second problem is the management of LV retraining. Perioperative tightening of the PAB is guided by measurement of pressure differences between the systemic RV and LV, following the echocardiographic changes in ventricular size and contractile function and the degree of shift of the interventricular septum toward the RV, as well as the decrease in TV regurgitation [19]. The basic target is to reach 50–75% of systemic pressure in the LV. Excessive PAB tightening leads to a reduced LV function and edema of the myocardium [4]. Sometimes, retraining of the LV can be achieved only by sequential PAB [13]. The results in prepubescent patients are good, but the response of older patients is less predictable and is associated with a higher early and late mortality. The cutoff for retraining the morphologic LV is about 15 years of age [5,9]. A ratio of left-to-right ventricular systolic pressure of >0.8, in the presence of a well-preserved LV function, with the indexed LV mass to LV volume ratio >1.5, is considered to be adequate for anatomical correction [13]. Quinn et al. [15] demonstrated that up to 50% of patients who underwent LV training before anatomic repair had moderate-to-severe LV dysfunction at intermediate follow-up, compared with a prevalence of 20% in patients who did not require training. In our series, four patients underwent PAB for retraining the LV. These patients were young (median age at PAB 1.9 years) and had a normal LV function and no MV regurgitation; however, three of them had moderate-to-severe TV regurgitation. A 'loose' PAB was placed to achieve 50% of systemic pressure in the LV and to allow patients to 'grow into' the PAB. The median interval between PAB and S–ASO was more than a year. This policy simplifies the postoperative course after PAB and training of the LV is gradual. After correction, none of our patients suffered from LV dysfunction during the follow-up.

The incidence of surgically acquired heart block associated with VSD closure in S–ASO is about 10% [5,6,13]. VSD closure is performed with sutures placed on the morphologic RV side of the septum [12]. However, traction on the crux of the heart, particularly in [S,L,L] when the heart is in a mesocardiac or dextrocardiac position, may create permanent heart block even if the sutures are carefully placed [5,9]. In this specific anatomy, one should consider closing the VSD, working through the right ventriculotomy, thus eliminating undue tension on the crux cordis.

6.3. Issues related to the Rastelli procedure

An unfavorable anatomy, such as a non-committed and/or restrictive VSD or significant straddling of the AV valve, may prevent the performance of a Rastelli operation. Under these circumstances, consideration should be given conversion either to the Fontan procedure, or, in suitable patients, to aortic translocation [20].

An obstruction-free and straight intraventricular tunnel is essential for preserving LV function [21]. In ccTGA with pulmonary atresia, the VSD is not restrictive and is usually naturally committed to the aorta, thus allowing the creation of a straight intraventricular tunnel. In theory, this should be the best morphology for intraventricular rerouting. The Ann Arbor group [7] reported results with 30 S–R patients. The outcomes for patients with pulmonary atresia were not significantly different from those of patients with other forms of LVOT obstruction. However, patients with pulmonary atresia tended to have better hospital survival and less morbidity.

The enlargement of a borderline VSD, while achieving better commitments of the VSD toward the aorta, is a significant risk for surgically acquired heart block. The anterior position of the conduction system in [S,L,L] and a posterior position in patients with [I,D,D] is to be generally expected (see below), and enlargement of the VSD is performed accordingly. However, the variability of the conduction system is probably significant [2]. In three of our patients with [I,D,D], enlargement of the VSD was performed without heart block, and the desired functional effect. The position of the AV node is anticipated with respect to the degree of misalignment between the interventricular and the atrial septum. The direction of the enlargement of the VSD is guided accordingly. In [S,L,L], if atrial septum is significantly misaligned, there is most likely to be an anterior position of conduction system. With mesocardia and dextrocardia, misalignment is not as prominent; therefore, a posterior position of the AV node is a possibility, or there are dual AV nodes that connect to a sling of conduction tissue [22]. In [I,D,D] there is no misalignment and the AV node is in a posterior position. In levocardia, misalignment can develop, shifting the AV node to an anterior position.

6.4. Issues related to the Bex/Nikaidoh procedure

Aortic translocation in ccTGA with a complex LVOTO is a challenging procedure that should be considered only if the
anatomy is inadequate for an intraventricular baffle as part of a Rastelli operation. The presence of an inlet and/or restrictive VSD, the association with straddling of the AV valves, and borderline RV volume are considered contraindications for a Rastelli operation [11].

The specific issue in ccTGA is the risk of damage to the conduction system during the division of the outlet septum. In \( I,D,D \) the conduction AV bundle arises from the posterior node to follow the conventional path along the posterior-inferior margin of the VSD, which allows the risk-free transection of the outlet septum. By contrast, in \( S,L,L \) the AV conduction axis runs anterior and cephalad to the pulmonary valve and then descends along the anterior margin of the VSD before diverging into the bundle branches [12]. Division of the muscular outlet septum does not, therefore, result in complete heart block; however, care should be taken when suturing the patch around the septal defect so as not to injure the conduction tissue [11].

Other issues are related to the transfer of the coronary artery, which in ccTGA must be detached and extensively mobilized, and to progressive aortic valve regurgitation. Usually, aortic regurgitation is apparent immediately after surgery, confirming the importance of the technical aspects of aortic root transfer [21].

Experience with aortic translocation combined with a Senning procedure as part of anatomic correction of ccTGA is very limited [11,14,20,23,24]. Morel at al. [11] reported three cases of ccTGA \( S,L,L \) with one early death and one complete heart block. Hu et al. [23] reported four cases of ccTGA \( S,L,L \) with no death; however, one patient required extracorporeal membrane oxygenation (ECMO) support. There was no complete heart block. Interestingly, the whole series of anatomically corrected ccTGA included seven patients. Apart from four survivors after S–BN, three other patients underwent the Senning operation and intraventricular rerouting with two early deaths, suggesting an excellent performance of S–BN, despite the complexity of the procedure. The Melbourne group [24] reported an excellent result in one patient with ccTGA \( S,L,L \) with pulmonary stenosis and a restrictive VSD. Our series includes one patient with ccTGA \( I,D,D \) and an inlet VSD; the patient is in perfect clinical condition with no heart block, a competent aortic valve, and free LVOT (Fig. 2) [14].

6.5. Proposed changes in treatment protocol

Based on our limited experience, and in order to minimize the recognized morbidity associated with the different procedures, one might consider:

1. Earlier indication for correction, while minimizing the number of palliations.
2. Simplification of the required palliation, e.g., avoiding shunting by stent placement.
3. Using a modified Senning, which is applicable regardless of situs and age.
4. Earlier placement of a 'loose' PAB to provide gradual training of the LV and/or to prevent significant dilatation of the pulmonary root.
5. Minimizing the risk of complete heart block by closing the VSD through the right ventriculotomy when undue tension on the crux cordis is expected (e.g., in mesocardia or dextrocardia in \( S,L,L \)).
6. Guiding the direction of the enlargement of the restrictive VSD based on the anticipated position of the AV node with respect to the degree of misalignment between the interventricular and atrial septum.
7. Using the BN procedure if the VSD is uncommitted.

6.6. Is the classical approach still justifiable?

The high survival benefit achieved with the conventional Rastelli operation (VSD closure and LV pulmonary artery conduit) might justify indication of this type of surgery in the patient with a remote and/or restrictive VSD, where S–R treatment is precluded. Shinoka et al. [2] reported on a series of 195 patients who had undergone repair of congenitally corrected transposition. The overall survival in 31 patients after the conventional Rastelli operation was 78.5% at 27 years, and was not different from anatomic types of repair. The authors concluded that the results of conventional repairs were satisfactory, except in patients with significant tricuspid regurgitation.

At our institution, patients unsuitable for LV retraining (older than 15 years and without an LVOTO), who present with progressive TR and/or RV dysfunction, are palliated by loose PAB and biventricular permanent pacing.

6.7. When is a single ventricle pathway indicated?

The modified Fontan procedure might be the preferable option if there is an unfavorable anatomy for anatomical correction, or if there is dysfunction of the LV or complex systemic and/or pulmonary venous anatomy. Whether functionally biventricular hearts treated by conversion to the Fontan circulation may perform better or be more durable than those with functionally single ventricular arrangements is purely a matter of speculation [20]. Thirty-eight patients from the Shinoka’s study [2] had been converted to the Fontan circulation rather than being submitted to complex biventricular repair. The actuarial survival at 22 years was 79.3%; therefore, it was not different from the other operative groups. Furthermore, the actuarial event-free survival was higher for those with the Fontan circulation than for any of the groups undergoing biventricular repair.

6.8. Limitations of the study

The distribution of the different treatment modalities is unequal, which makes comparison among the surgical groups difficult. Follow-up is too short to provide meaningful analysis of the suggested alterations in treatment protocol. The long-term benefits of these approaches need to be evaluated with a larger number of patients and careful follow-up.

In conclusion, anatomic correction of ccTGA can be performed in selected patients with no mortality and acceptable morbidity. In the midterm, an excellent functional outcome can be achieved, which leads to normal ventricular function, even in a retrained LV, and with minimal
incidence of complete heart block. The long-term function of the aortic valve, intraventricular tunnels, conduits, and ventricles requires close surveillance.

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References


Appendix A. Conference discussion

Dr E. Bacha (New York, New York): We are going to have to wait for the long-term results to be definitive about our statements. Although your mortality was zero, your morbidity up front was fairly high, so you are paying a price that is fairly steep up front for a potential benefit down the line.

A certain percentage of these patients, roughly 30%, have no associated malformations and no TR; thus, I expect that they may have been asymptomatic. So you are, in fact, doing a pretty aggressive operation for a subset of patients that may be asymptomatic. You know I like the operation, but this has to be said in terms of the indication.

A quick question about the PA band and then a quick question about those indications.

Do you think an adjustable PA band is something that is useful in this indication?

Dr Hralška: There are technical limitations with the adjustable band. The band is suitable only in babies up to 10 kg in weight. At this point, the company is not willing to manufacture a bigger band. In addition, the adjustable band is pretty bulky and is not easy to place. Equally important, it is extremely expensive.

Dr Bacha: The second quick question. Do you have an age limit as to when you would embark on LV retraining if you have ccTGA with an intact ventricular septum?

Let’s say you have a 5-year-old child, asymptomatic, no tricuspid regurgitation, corrected transposition. What would you do with this patient?

Dr Hralška: You mentioned that we operated on asymptomatic patients. In fact all our patients were significantly symptomatic. Patients who needed LV training had either no VSD or restrictive VSD; however, there was always hemodynamically relevant TR. To answer your question, we prefer not to take an asymptomatic patient to the operating theatre.

Dr Bacha: So for asymptomatic patients who are doing perfectly well, no TR, you would rather sit and do nothing and observe?

Dr Hralška: Yes.

Dr Bacha: How about a patient who has some mild or more TR without many symptoms, where is your age limit as to LV retraining as opposed to deciding to replace the tricuspid valve?

Dr Hralška: I do not know what the age limit is for LV retraining. Most likely the age limit is somewhere between 10 and 15 years. All our patients who underwent LV training were young. The loose banding was placed with the aim of allowing patient growth into the band.

I think that is the strategy which most likely will preserve the function of the left ventricle. Training of the ventricle is smooth, allowing gradual development of the appropriate muscle mass and enlargement of capacity of the left ventricle.