Outcomes of the anatomical repair in patients with congenitally corrected transposition of the great arteries: lessons learned in a high-volume centre†

Marien Lenoir¹, Ismail Bouhout², Regis Gaudin¹, Olivier Raisky¹ and Pascal Vouhé¹a

¹Division of Pediatric Cardiac Surgery, University Paris Descartes and Necker Sick Children Hospital, Paris, France
²Division of Cardiac Surgery, Montreal Heart Institute, University of Montreal School of Medicine, Montreal, QC, Canada

* Corresponding author. Division of Pediatric Cardiac Surgery, University Paris Descartes and Necker Sick Children Hospital, 149 rue de Sévres, 75015 Paris, France.

Keywords: Congenitally corrected transposition of the great arteries — Senning procedure — Réparation à l'étage ventriculaire — Anatomical repair

Abstract

OBJECTIVES: The physiological repair of the congenitally corrected transposition of the great arteries (ccTGA) has been associated with a long-term risk of the right ventricular dysfunction and tricuspid valve regurgitation. On the other side, the anatomical repair with the restoration of the left ventricle in a systemic position has been hypothesized to improve long-term outcomes. The aim of this study was to determine the results of the anatomical repair.

METHODS: Between 1990 and 2015, 39 patients underwent an anatomical repair of a ccTGA. The median age was 7 years (range from 8 days to 42.8 years). Eighteen (46%) patients had a double switch. Of these, 21 (54%) patients had an atrial switch with a Rastelli technique (n = 16, 41%), with a réparation à l'étage ventriculaire (REV) procedure (n = 4, 10%) or a truncal rotation (n = 1, 3%). The follow-up was 100% complete at 2 years with a median follow-up of 3.17 years (range 0–26 years).

RESULTS: The early mortality was 10% (n = 4) and the late mortality was 20% (n = 8). The long-term survival was 77 ± 10% at 10 years for the double switch group and 62 ± 11% at 10 years for the Rastelli group (P = 0.25). Eight (20%) patients required reintervention. Freedom from reoperation at 10 years was 82 ± 11% in the double switch group and 88 ± 11% in the Rastelli group. At the last follow-up, 23 (82%) patients were in New York Heart Association (NYHA) functional class I and 2 (7%) patients in NYHA class II.

CONCLUSIONS: The anatomical repair of the ccTGA is associated with significant early mortality and morbidity. However, the long-term functional status is satisfactory. The anatomical repair of the ccTGA should be reserved for a carefully selected group of patients.

Keywords: Congenitally corrected transposition of the great arteries — Senning procedure — Réparation à l'étage ventriculaire — Anatomical repair

INTRODUCTION

The congenitally corrected transposition of the great arteries (ccTGA) is an uncommon congenital malformation, representing <0.5% of all congenital heart diseases [1]. In addition to the right ventricular (RV) and left ventricular (LV) inversion, there is a significant disruption of the heart's internal architecture. Indeed, the atrioventricular discordance induces malalignment of the interauricular and interventricular septum, which frequently leads to other associated malformations. Therefore, a ventricular septal defect (VSD) is common with a ccTGA, while other RV anomalies are less frequent but not exceptional. Pulmonary stenosis or atresia is also reported in up to 50% of cases.

Traditionally, the surgical management of these patients is based on ‘physiological’ repair consisting of the correction of associated lesions, including VSD closure, tricuspid valve repair and RV outflow obstruction relief. However, the right ventricle evolving in a systemic position fails in the long term with the development of tricuspid valve regurgitation. Indeed, Kral Kollars et al. [3] reported that the reduction of the LV to RV ratio after physiological repair increases the spherical shape of the right ventricle, which alters the tricuspid valve coaptation surface. More recently developed, anatomical repairs aim for an atrial stage switch (Senning or Mustard procedures) in addition to an arterial switch or a Rastelli procedure [RV to the main pulmonary artery (PA) (RV-PA) conduit]. Therefore, the right ventricle is restored in the pulmonary position, reducing the RV overload and the tricuspid valve regurgitation, thus improving long-term patient outcomes [6]. Nevertheless, the anatomical repair is associated with a prolonged aortic clamping time and a high early mortality. Only a few studies have reported early and long-term results following an anatomical correction of a ccTGA [4–8]. The aim of this study was to report...
the early and late outcome after anatomical repairs for ccTGA in a single-centre cohort.

**MATERIALS AND METHODS**

**Patients**

We conducted a retrospective, single-centre study of patients undergoing anatomical repair of a ccTGA. From February 1990 to December 2015, 207 consecutive patients diagnosed with ccTGA were identified. Of these, patients undergoing palliative procedures alone (n = 77), physiological repairs (n = 63), Fontan palliation (n = 25) and heart transplantation (n = 3) were excluded. A total of 39 patients with anatomical repairs were included in the present study. Permission to undertake this study was obtained from the Paris V University Ethics Committee. The need for individual consent was waived.

**Data collection and outcomes**

Baseline characteristics, surgical details and early and long-term results were gathered from the French Public Health Care database. Additional data were collected from medical records. The median follow-up was 3.17 years (range 0 day–26 years) and was 100% complete at 2 years after study closure (1 January 2017). The primary end point was survival. Secondary outcomes were in-hospital mortality, major early morbidity and reintervention in the long term. In addition, cumulative sum (CUSUM) analyses were used to determine a hinge point for improvement in early mortality.

**Surgical management algorithm**

When possible, the anatomical repair was the preferred approach in our centre. Eligibility criteria were a well-prepared LV and the feasibility of the tunnelization between the left ventricle and the aorta. In cases of the RV dysfunction and tricuspid valve regurgitation, the anatomical repair was performed by a resection of the conal septum in patients with restrictive VSD.

All ccTGA patients with pulmonary artery atresia had a modified Blalock-Taussig (mBT) shunt. In addition, 14 of the 21 patients with pulmonary artery stenosis underwent mBT shunt placement (saturation <70% and clinical cyanosis). Pulmonary artery banding (PAB) was undertaken, if the VSD was large or in order to train the left ventricle for a subsequent anatomical repair (Fig. 1).

The type of anatomical repair varied according to the double discordance anatomy, associated anomalies and the operative period. In the absence of a severe pulmonary valve stenosis, we performed a double switch (DS) procedure. In this case, the atrial switch was always done using the Senning technique. In the presence of severe pulmonary artery stenosis/atresia, a Rastelli procedure [9], a réparation à l'étage ventriculaire (REV) repair [10] or a truncal rotation was used. The Rastelli procedure was carried out in combination with a Mustard procedure [11] before 1996 and with a Senning procedure after 1996. The REV procedure or the truncal rotation was always performed in combination with a...
Senning procedure. When the right ventricle was considered too small, we performed a hemi-Mustard at the atrial level (Fig. 1).

Surgical technique

Briefly, patients were operated on through a median sternotomy and under normothermic cardiopulmonary bypass (CPB) using 2 venous cannulas. The heart was arrested with anterograde warm blood cardioplegia (repeated every 10 min) or more recently with Custodiol® cardioplegia (repeated every 2 h). The patent arterial duct was systematically divided. The pulmonary arteries and their branches were extensively mobilized. Based on anatomical configuration and surgeon preferences, different strategies were used and are briefly described below. As a general rule, we preferred to use the Senning procedure [12] to achieve the atrial switch so as to minimize the risk of the intra-atrial baffle stenosis. All Senning procedures were performed with the Shumaker modification, no matter the position of the heart. Eight patients had dextrocardia and mesocardia.

Statistical analysis

Continuous variables were expressed as mean ± standard deviation or median (range) when skewed. Means were compared using the Student’s t-test or non-parametric Mann–Whitney test as appropriate. Categorical variables were expressed as frequencies (%) and compared using the χ2 or Fisher’s exact test as appropriate. The survival after anatomical repair was analysed using the Kaplan–Meier method and compared with the log-rank test. CUSUM analyses were performed for early mortality using the method described by Rogers et al. [13]. A cumulative observed minus expected failure chart was used. The expected failure rate (early mortality) was set at 10% based on previously published reports [8, 14–16]. Data were analysed using IBM SPSS statistics 21 (IBM Corporation, Armonk, NY, USA) and STATA 13.0 (StataCorp, College Station, TX, USA).

RESULTS

Patient characteristics and anatomy

A total of 39 patients underwent an anatomical repair for ccTGA. The median age at the repair was 7 years (8 days–42.8 years). The median weight was 19 kg (3–69 kg). Morphological characteristics and associated lesions are presented in Table 1. Eighteen (46%) patients had a double switch (DS). Twenty-one (54%) patients with a pulmonary stenosis/ataresia had an atrial switch associated with a Rastelli technique (n = 16, 41%), an REV procedure (n = 4, 10%) or a truncal rotation (n = 1, 3%) (Fig. 1). Concomitant procedures were performed during the anatomical repair in 11 (28%) patients, including 4 tricuspid valve repair, 1 mitral valve repair, 1 coarctation repair, 2 multiple VSD closures and 2 anastomoses between the right atrium and left superior vena cava. Five patients in the Rastelli group underwent a bidirectional Glenn procedure (4 Rastelli/hemi-Mustard and 1 REV/hemi-Mustard).

Eleven (61%) patients had a VSD in the DS group. The pulmonary stenosis was present in 11 (28%) patients and pulmonary atresia in 10 (26%) patients. Seven patients had a RV failure (57% of patients treated with DS) and 10 patients had a severe tricuspid regurgitation rather than 3 (60% of patients treated with DS).

The operative indications in the pulmonary atresia/stenosis group were symptomatic cyanosis (80%) and RV dysfunction in the systemic position (20%). In the DS group, the indications were symptomatic cyanosis (77%) and pulmonary hypertension (23%) in patients with VSD. In those without VSD, the indications were RV dysfunction (40%), dyspnoea (30%) and complete atrioventricular blocks (30%).

Palliation

Twenty-seven (71%) patients had a surgical palliation before definitive anatomical repair. Of these, 13 (33%) patients underwent

<p>| Table 1: Patient demographics, morphologies and palliation characteristics |
|-----------------|-----------------|-----------------|-----------------|-----------------|</p>
<table>
<thead>
<tr>
<th>Outcomes</th>
<th>DS (n = 18)</th>
<th>Pulmonary stenosis/ataresia (n = 21)</th>
<th>Total (n = 39)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Demographics</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Median age at repair (years), median (range)</td>
<td>6 (0–37)</td>
<td>8 (1–43)</td>
<td>7 (0–43)</td>
<td>0.86</td>
</tr>
<tr>
<td>Male, n (%)</td>
<td>11 (61)</td>
<td>11 (52)</td>
<td>22 (56)</td>
<td>0.58</td>
</tr>
<tr>
<td>Morphology, n (%)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Situs inversus</td>
<td>1 (6)</td>
<td>7 (41)</td>
<td>8 (24)</td>
<td>0.02</td>
</tr>
<tr>
<td>VSD</td>
<td>11 (61)</td>
<td>21 (100)</td>
<td>32 (82)</td>
<td>0.002</td>
</tr>
<tr>
<td>Ebstein’s anomaly of TV</td>
<td>5 (29)</td>
<td>0</td>
<td>5 (15)</td>
<td>0.02</td>
</tr>
<tr>
<td>Severe TR</td>
<td>6 (37)</td>
<td>4 (26)</td>
<td>10 (32)</td>
<td>0.52</td>
</tr>
<tr>
<td>Coarctation</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>LVOTO</td>
<td>0</td>
<td>21 (100)</td>
<td>21 (54)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>PA</td>
<td>0</td>
<td>11 (52)</td>
<td>11 (28)</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Palliation, n (%)</td>
<td></td>
<td></td>
<td></td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Banding, n (%)</td>
<td>13 (72)</td>
<td>14 (70)</td>
<td>27 (71)</td>
<td>0.88</td>
</tr>
<tr>
<td>mBT shunt, n (%)</td>
<td></td>
<td></td>
<td></td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Median age at palliation (months), median (range)</td>
<td>1.58 (0.13–9)</td>
<td>0.43 (0.06–334)</td>
<td>1.1 (0.06–334)</td>
<td>0.69</td>
</tr>
<tr>
<td>Median duration of palliation (years), median (range)</td>
<td>7.52 (0.05–36)</td>
<td>4.47 (0.7–15)</td>
<td>6.61 (0.05–36)</td>
<td>0.23</td>
</tr>
</tbody>
</table>

DS: double switch; LVOTO: Left ventricular outflow tract obstruction; mBT: modified Blalock–Taussing; PA: pulmonary atresia; PS: pulmonary stenosis; TR: tricuspid regurgitation; TV: tricuspid valve; VSD: ventricular septal defect.
ECMO. support duration was 5 days (5–9 days), and no patient died on
osis/atresia group (in the DS group versus 9 (53%) patients in the pulmonary sten-
 maker insertions in the DS group, all occurred in patients with a
VSD. A delayed sternal closure was performed in 2 (12%) patients
postoperatively (<30 days) and underwent a permanent pace-
(PPM) insertion [4 (20%) in the DS group and 1 (5%) in the pulmon-
Hinge point at the 15–20 patient mark (Fig. 2).
In the CUSUM, there was a reduction in early mortality with a
early mortality occurred in the first 14 patients (before 2003).
the early mortality in the DS group and 5 patients in the pulmonary stenosis/atresia group.
The causes of death are detailed in Table 3. The overall sur-
in the pulmonary stenosis/atresia group (77 ± 10% vs 62 ± 11% at
10 years, respectively). The freedom from >mild LV dysfunction
was 72 ± 7%, 69 ± 8% and 60 ± 11% at 1, 10 and 20 years, re-
respectively. The long-term survival was similar in the DS group and
in the pulmonary stenosis/atresia group (77 ± 10% vs 62 ± 11% at
10 years, P = 0.54) (Fig. 3). The freedom from death, heart trans-
and LV dysfunction was 70 ± 8%, 66 ± 8% and 61 ± 9% at 1, 5
and 10 years, respectively. The freedom from >mild LV dysfunction
was 92 ± 5%, 80 ± 9% and 80 ± 9% at 1, 5 and 10 years, respectively.
The freedom from LV dysfunction was 100% at 5 years in the PAB
and 78 ± 10% in PAB for VSD (P = 0.47).

**Intraoperative data and early outcome**

The median CPB and cross-clamp times were similar between the
and the pulmonary stenosis/atresia groups [258 min (222–501 min) and 156 min (108–247 min) vs 272 min (204–894 min) and
158 min (113–308 min), P = 0.46 and P = 0.85 respectively (Table 2).
Four (10%) patients died in the early period (<30 days), 2 (10%)
patients in the DS group and 2 (10%) in the pulmonary stenosis/
stenosis/atresia group. The causes of deaths included 2 pulmonary hyper-
tension crises, 1 multiple organ failure and 1 sudden death. All
the early mortality occurred in the first 14 patients (before 2003).
In the CUSUM, there was a reduction in early mortality with a
hinge point at the 15–20 patient mark (Fig. 2).
Reintervention for bleeding occurred in 5 (16%) patients exclu-
sively in patients of the pulmonary stenosis/atresia group
(P = 0.02). Five (14%) patients developed an atrioventricular block
postoperatively (<30 days) and underwent a permanent pacemaker
(PPM) insertion [4 (20%) in the DS group and 1 (5%) in the pulmonary stenosis/atresia group, P = 0.01]. Among the 4 pace-
markers insertions in the DS group, all occurred in patients with a
VSD. A delayed sternal closure was performed in 2 (12%) patients
in the DS group versus 9 (53%) patients in the pulmonary sten-
sis/atresia group (P = 0.014). Three (14%) patients required an
extracorporeal membrane oxygenation (ECMO) in postoperative
period in the pulmonary stenosis/atresia, while no patient was
put on circulatory support in the DS group (P = 0.07). The median
support duration was 5 days (5–9 days), and no patient died on
ECMO.

**Late outcomes**

The long-term follow-up was available for 39 (100%) patients. Of
these, 8 (20%) patients died during the follow-up period: 3 patients
in the DS group and 5 patients in the pulmonary stenosis/atresia
group. The causes of death are detailed in Table 3. The overall sur-
vival was 72 ± 7%, 69 ± 8% and 60 ± 11% at 1, 10 and 20 years, re-
spectively. The long-term survival was similar in the DS group and
in the pulmonary stenosis/atresia group (77 ± 10% vs 62 ± 11% at
10 years, P = 0.54) (Fig. 3). The freedom from death, heart trans-
plant and LV dysfunction was 70 ± 8%, 66 ± 8% and 61 ± 9% at 1, 5
and 10 years, respectively. The freedom from >mild LV dysfunction
was 92 ± 5%, 80 ± 9% and 80 ± 9% at 1, 5 and 10 years, respectively.
The freedom from LV dysfunction was 100% at 5 years in the PAB
group and 78 ± 10%, P = 0.47.
Among the 7 patients with ccTGA intact septum, 1 patient
died of pulmonary hypertension crisis and 1 patient had a rein-
tervention 4.6 months after surgery for atrial baffle enlargement.
Eight of the 39 (20%) patients underwent 14 reinterventions
during the follow-up. The median age at reintervention was
3.6 years (range from 4.7 months to 20.9 years). No differences
were observed between the DS group [4 (22%) patients] and the
pulmonary stenosis/atresia group [4 (19%) patients]. These
procedures included 5 pulmonary valve replacements [3 RV outflow
truct redo in 16 (18%) Rastelli and 1 single redo for 4 (25%) REV].

**Table 2: Intraoperative data and early outcomes**

<table>
<thead>
<tr>
<th>Outcomes</th>
<th>DS (n = 18)</th>
<th>Pulmonary stenosis/atresia (n = 21)</th>
<th>Total (n = 39)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Early deaths, n (%)</td>
<td>2</td>
<td>2</td>
<td>4 (10%)</td>
<td>0.46</td>
</tr>
<tr>
<td>CPB time (min), median (range)</td>
<td>258 (222–501)</td>
<td>272 (204–894)</td>
<td>260 (204–894)</td>
<td>0.85</td>
</tr>
<tr>
<td>Cross-clamp time (min), median (range)</td>
<td>156 (108–247)</td>
<td>158 (113–308)</td>
<td>157 (108–308)</td>
<td>0.07</td>
</tr>
<tr>
<td>ECMO, n (%)</td>
<td>0</td>
<td>3 (14)</td>
<td>3 (8)</td>
<td>0.01</td>
</tr>
<tr>
<td>Mean ECMO time (days), median (range)</td>
<td>2 (12)</td>
<td>9 (53)</td>
<td>11 (33)</td>
<td>0.014</td>
</tr>
<tr>
<td>Delayed sternal closure, n (%)</td>
<td>0</td>
<td>5 (5–9)</td>
<td>5 (5–9)</td>
<td>0.014</td>
</tr>
<tr>
<td>Extubation time (days), median (range)</td>
<td>1 (0–16)</td>
<td>5 (1–29)</td>
<td>2 (0–29)</td>
<td>0.089</td>
</tr>
<tr>
<td>Median ICU length of stay (days), median (range)</td>
<td>7 (0–30)</td>
<td>14 (5–66)</td>
<td>10 (0–66)</td>
<td>0.06</td>
</tr>
<tr>
<td>Redo for bleeding, n (%)</td>
<td>4 (22)</td>
<td>1 (5)</td>
<td>5 (12)</td>
<td>0.1</td>
</tr>
<tr>
<td>Operative (&lt;30 days), n (%)</td>
<td>2 (11)</td>
<td>1 (5)</td>
<td>3 (6)</td>
<td>0.45</td>
</tr>
</tbody>
</table>

CPB: cardiopulmonary bypass; DS: double switch; ECMO: extracorporeal membrane oxygenation; ICU: intensive care unit; PPM: permanent pacemaker.
2 intra-atrial baffle enlargements, 3 PPM implantation, 2 tricuspid valve repairs, 1 mitral valve repair and 1 ECMO. Most of the pulmonary valve replacements occurred in the pulmonary stenosis/atresia group (n = 4 patients). One patient in the DS group needed a new RV-PA conduit for a pulmonary valve endocarditis 3.6 years after surgery. Reinterventions on the atrial level were required in 2 patients (1 in each group) secondary to pulmonary venous obstruction. The freedom from reintervention was 94 ± 6%, 82 ± 11% and 55 ± 23% in the DS group vs 100%, 88 ± 11% (IC), 59 ± 20% in the pulmonary stenosis/atresia at 1, 10 and 20 years, respectively (P = 0.46) (Fig. 4).

The freedom from postoperative pacemaker implantation was 80% at 10 years.

At the last follow-up, 23 patients were in New York Heart Association (NYHA) functional class I and 2 patients were in NYHA class II.

DISCUSSION

Several studies have shown the reduced life expectancy in patients with ccTGA [11, 12]. The surgical approaches for ccTGA include palliation procedures, physiological correction and anatomical repair. The latter was introduced to restore the morphological left ventricle and mitral valve to the systemic circulation. Recent reports [3] have shown a considerable improvement in the incidence of in-hospital mortality and long-term outcomes. Nonetheless, in our experience as well as in other studies [8, 14–16], early mortality and morbidity remain significant. The results of these different studies are summarized in Table 4.

Effect of high-volume centres in improving early outcomes

As highlighted in Fig. 2, the early results of the anatomical repair of the ccTGA improved with experience with a hinge point at 15–20 patients. This improvement could be explained by many factors, mainly surgical experience, enhancement in perioperative management (more aggressive utilization of nitric oxide) and patient selection (avoiding surgery in patients <1-year-old). As the ccTGA is a rare disease, accumulating such a number of cases to overcome the learning curve of the anatomical repair procedure is arduous for a small volume centre. In parallel, Hsu et al. [15] demonstrated a similar learning curve and a high early mortality rate that correlated initial results with a long clamping time. Therefore, we believe that these patients should be offered a physiological repair or referred for an anatomical repair in an expert and high-volume centre. Adachi et al. [19] showed that long-term survival is acceptable with physiological repair.

<table>
<thead>
<tr>
<th>Table 3: Causes of death</th>
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<tbody>
<tr>
<td>n</td>
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<tr>
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<td>11</td>
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<td>12</td>
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</tbody>
</table>

DS: double switch; LV: left ventricle; REV: reparation à l’étage ventriculaire; RSV: respiratory syncytia virus; RV: right ventricle.
Palliation strategy and training of the left ventricle

In patients with left ventricular outflow tract obstruction (LVOTO) without clinical cyanosis, an anatomical repair could be achieved later in life. In patients with clinical manifestations, mBT shunt palliation should be undertaken before the anatomical repair. Indeed, we believe that the LVOTO allows prophylactic banding that prepares the left ventricle to the systemic circulation. In patients without LVOTO and a VSD, we perform a PAB to avoid a right–left shunt and subsequently, a pulmonary arterial hypertension. In patients without LVOTO and without VSD, we prefer resection to carry out a PAB early in life to allow stabilization or even a regression of the tricuspid regurgitation by avoiding a shift in the ventricular septum, in addition to the preservation of the LV function. Similarly, Myers et al. [20] aimed to perform the PAB before 2 years of age, followed by an anatomical repair before 3 years of age.

Conduction disorders and arrhythmia

In our study, the rate of postoperative PPM was higher in the DS group, especially in those with a concomitant VSD closure. Indeed, during the VSD closure in a DS, the exposure is suboptimal, and the patch is applied directly on the interventricular septum. This is in contrast with the Rastelli or the REV procedure, where the patch sutures are distant from the crest and the conduction system. This risk could be mitigated by placing the suture line on the right side of the interventricular septum without opening the systemic ventricle [21]. In addition, as the history of the ccTGA was associated with a high risk of complete heart block [23], we think that PPM wires should be placed in all patients during the procedure. In patients with a pacemaker, the loss of synchronization has been identified as a potential contributing factor in predicting for LV dysfunction. Therefore, cardiac resynchronization therapy should be aimed in all patients in order to avoid heart failure in the long term [17, 18].

Furthermore, there is a substantial risk of atrial tachyarrhythmia following a Mustard or a Senning procedure. Indeed, tachyarrhythmias can contribute to the LV dysfunction through the loss of the atrial contraction, diastolic dysfunction and the negative inotropic effects of antiarrhythmic drugs. To achieve clinical improvements in these patients, the use of the isthmus cryoablation is an interesting avenue, which warrants further validation in the long term [25–27].

Different strategies during the anatomical repair

In our initial experience, the atrial switch was achieved using the Mustard procedure. However, in the 1990s, due to growing evidence showing the detrimental effects of this procedure, we turned to the Senning technique. Indeed, the latter was associated with a lower rate of arrhythmia and baffle obstruction in the long term. In patients with ccTGA, restrictive VSD and pulmonary stenosis, the combination of a Senning and a Rastelli procedure has a high risk of a complete heart block and LVOTO. Therefore, some authors have hypothesized that an additional Damus–Kaye–Stansel anastomosis may be effective in avoiding postoperative systemic ventricular outflow tract obstruction and postoperative complete heart block [16]. Furthermore, these patients could also benefit from a truncal rotation or a Bex-Nikaidoh procedure in order to avoid postoperative LVOTO [27]. Nonetheless, these techniques should be avoided when an abnormal coronary anatomy is suspected.

In our study, the most common reoperations following a Senning–Rastelli procedure were RV-PA conduit replacements. Therefore, the REV was introduced in our centre to avoid the use of an extracardiac conduit and mitigate the risk for reintervention. In addition, the conal septum was systematically resected during a REV procedure, which widened the tunnelization area, reducing the risk of a subaortic stenosis and preserving LV function.

CONCLUSION

While anatomical repairs have emerged as the best option in patients with ccTGA in the long term, early mortality and morbidity remain significant. Therefore, anatomical repair should be reserved to a well-selected patient population. In addition, these early results improve with time which may enhance the potential benefit of these procedures in specialized centres. Finally, some technical modifications that have been introduced recently may improve long-term outcomes in patients with ccTGA and LVOTO and need to be further studied.

Conflict of interest: none declared.

Table 4: Published cohorts of patients undergoing anatomical repairs

<table>
<thead>
<tr>
<th>Study</th>
<th>Study period</th>
<th>Population (DS/RS)</th>
<th>Patients (n)</th>
<th>Early deaths (%)</th>
<th>Survival at 10 years (DS/RS; %)</th>
<th>Survival at 20 years (%)</th>
<th>Reintervention at 10 years (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lim et al. [28]</td>
<td>1983–2009</td>
<td>10/34</td>
<td>44</td>
<td>NA</td>
<td>100 (excluding hospital mortality)</td>
<td>NA</td>
<td>46.2</td>
</tr>
<tr>
<td>Ly et al. [29]</td>
<td>1995–2007</td>
<td>20/0</td>
<td>20</td>
<td>0 (0 of 20)</td>
<td>100</td>
<td>NA</td>
<td>77.4</td>
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<td>Shinoka et al. [8]</td>
<td>1980–2003</td>
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<td>Duncan et al. [30]</td>
<td>1993–2002</td>
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<td>Hoashi et al. [16]</td>
<td>1987–2011</td>
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