The results of a two-stage double switch operation for congenital corrected transposition of the great arteries with a deconditioned morphologically left ventricle

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Abstract

OBJECTIVES: The purpose of this retrospective study was to evaluate a two-stage double switch operation, morphological left ventricular (mLV) retraining followed by an atrial–arterial switch operation, in the management of patients with congenitally corrected transposition of the great arteries (CCTGA) and a deconditioned mLV.

METHODS: Between May 2005 and May 2011, 14 patients with CCTGA and a deconditioned mLV anomaly underwent the two-stage double switch operation. There were eight males and six females aged between 2.5 and 72 months (mean: 34.4 ± 24.0 months) old and weighing from 5 to 23 kg (mean: 12.7 ± 4.9 kg). The major associated malformations included: tricuspid regurgitation (TR, n = 13); restrictive ventricular septal defect (n = 10); atrial septal defect or patent foramen ovale (n = 7); mild pulmonary stenosis (n = 5) and patent ductus arteriosus (n = 4). These patients underwent morphological left ventricular retraining by means of pulmonary artery banding under general anaesthesia, which was then followed by a double switch operation under general anaesthesia and cardiopulmonary bypass.

RESULTS: There were no deaths or complications during the hospital stay or follow-up for the mLV retraining. In comparison with pre-operative conditions, the mLV end-diastolic diameter (mLVEDd), the posterior wall thickness of the mLV and the mLV/mRV pressure ratio were all increased; the interventricular septum had moved partially to the midline position and TR had decreased. After the atrial–arterial switch procedure, 2 patients died during the perioperative period. The causes of death included serious cardiac arrhythmia with circulatory collapse and sudden death. The others were followed up for 2–8 years: 1 patient died from serious cardiac arrhythmias with circulatory collapse in the follow-up period. With regard to the others, 8 were evaluated as New York Heart Association Functional Class I, and the other 3 as Class II. Moderate aortic valve regurgitation was noted in 3 patients and moderate mitral regurgitation in 1 patient.

CONCLUSIONS: For CCTGA children with degraded mLV, the two-stage double switch procedure can be performed with low mortality and morbidity and may be an appealing alternative to conventional repair. mLV retraining should be performed as early as possible. The second-stage atrial–arterial switch procedure showed satisfactory early and mid-term results. More attention should be paid to the long-term function of the mLV and the aortic valve.

Keywords: Left ventricular retraining • Double switch operation • Congenitally corrected transposition of the great arteries • Congenital heart disease

INTRODUCTION

Congenitally corrected transposition of the great arteries (CCTGA) is a complex congenital heart defect characterized by atrioventricular and ventriculo-arterial discordance. The morphological left ventricular (mLV) and the morphological right ventricular (mRV) support the pulmonary and systemic circulations, respectively. Over time, progressive deterioration of the mLV and/or the systemic atrioventricular valve would, if untreated, lead to significant morbidity and mortality. The optimal management approach for these patients remains controversial. Conventional repair, which leaves the mRV supplying the systemic circulation, is still associated with progressive mRV dysfunction and tricuspid regurgitation. Therefore, anatomical correction might be superior. Nevertheless, an mLV should have adequate pressure, volume load and ventricular mass to sustain a systemic circulation. So, for certain patients with a deconditioned mLV anomaly, the two-stage double switch (DS) operation, mLV retraining followed by the atrial–arterial switch, may be an appealing alternative [1, 2]. Between May 2005 and May 2011, 14 consecutive patients with CCTGA and a deconditioned mLV anomaly received morphological left ventricular (mLV)
retraining followed by the atrial–arterial switch in Beijing Fu Wai hospital. In the current study, we evaluate the clinical outcomes of this two-stage operation on these patients.

**MATERIALS AND METHODS**

**Patient population**

Between May 2005 and May 2011, 14 consecutive patients with CCTGA and a deconditioned mLV anomaly received mLV retraining followed by the atrial–arterial switch. The mean interval between the two stages of the operation was 10.2 ± 9.5 months (0.7–34 months). There were eight males and six females. At the palliation stage, the patients’ ages were from 2.5 to 72 months (mean: 34.4 ± 24.0 months) with a body weight range of 5.1–23 kg (mean: 12.7 ± 4.9 kg), and at the DS operation, the ages were from 11 to 121 months (mean: 47.5 ± 26.0 months) with a body weight range of 8–27 kg (mean: 15.1 ± 5.9 kg).

These patients were examined preoperatively by echocardiography, and nine of them underwent angiography or cardiac catheterization. The major associated malformations included: tricuspid regurgitation (TR, n = 13); restrictive ventricular septal defect (n = 10); atrial septal defect or patent foramen ovale (n = 7); mild pulmonary stenosis (n = 5) and patent ductus arteriosus (n = 4). The 13 patients with TR were divided into three groups: mild (n = 8), moderate (n = 3) and severe TR (n = 2).

**Surgical management**

The patients underwent mLV retraining by means of pulmonary artery banding (PAB) under general anaesthesia. The main pulmonary artery was exposed through an upper midline sternotomy approach. The methods used for PAB were similar to those described by Mee [3]. The PA band comprised a polytetrafluoroethylene strip, with an initial length of 24 mm plus 1 mm in kg in weight, to reduce the circumference of the PA. The banding was placed near the middle of the PA to avoid pulmonary valve damage. A pressure-measuring needle was inserted into the proximal PA to monitor mLV systolic pressure directly. Transoesophageal echocardiography was used to assess ventricular function, tricuspid regurgitation and septal commitment. The banding was applied to increase the mLV pressure to 65–80% of the systemic value while maintaining adequate mLV function. After a period of haemodynamic stabilization and observation of the mLV/mRV pressure ratio, the band was tightened further. Later, the band was secured to the main PA with a fine 5/0 polypropylene suture line to prevent migration. Subsequent assessment of suitability for the DS procedure was by means of transthoracic echocardiography and cardiac catheterization. If the primary PAB was not sufficient to perform the anatomical repair, early rebanding was considered. For patients whose systemic trained LV was suitable for anatomical repair, the arterial switch was accomplished through the LeCompte manoeuvre and standard transplantation of the coronary arteries; meanwhile, the PAB was removed. A modified Senning procedure was performed in all cases under general anaesthesia and cardiopulmonary bypass (CPB) with moderate systemic temperature and low-volume blood flow. The pulmonary venous pathway was routinely enlarged with autologous in situ pericardium and the capacity of the left atrial chamber was optimized. Myocardial protection was afforded by intermittent cold crystalloid cardioplegia during aortic cross-clamping.

Concomitant operative procedures performed in 14 patients included: ventricular septal defect repair (n = 10); atrial septal defect repair (n = 7); patent ductus arteriosus closure (n = 4); tricuspid valve plasty (n = 3); resection of additional discrete membranous subaortic stenosis (n = 1) and aortic sinus angioplasty (n = 1).

**Data analysis**

Demographic and other patient-related data were obtained from clinical records. The values, unless stated otherwise, are expressed as mean ± standard deviation (SD). Student’s paired t-test was chosen for the differences between two groups. The level of statistical significance was set at P <0.05. Early operative mortality was defined as death occurring during hospitalization, or within 30 days postoperatively.

**RESULTS**

The mean operation time of PAB was 2.76 ± 0.35 h (2.3–3.5 h), the mean endotracheal intubation time was 14.22 ± 13.62 h (2–53 h), the mean intensive care unit (ICU) stay was 44.40 ± 23.86 h (21–96 h) and the mean hospital length of stay was 10.07 ± 7.50 days (6–35 days). There was no mortality or complications during the hospital stay or follow-up. Two patients required rebanding at 13 and 35 months, respectively, after the initial PAB. The principal methods of assessing ventricular and valve function were echocardiography, angiography and/or cardiac catheterization at every time point (preband/early post-band: at discharge after mLV retraining/later post-band: at DS procedure). After PAB, mLV end-diastolic diameter (mLVEDd), posterior wall thickness of mLV and the mLV/mRV pressure ratio were increased. PAB was associated with a sustained reduction in TR, with a sustained leftward change in the interventricular septum commitment (Table 1 and Figs 1–3).

All patients proceeded to the second-stage operation by means of an atrial–arterial switch, with a mean interval of 10.23 ± 9.47 months (0.67–34 months). The mean CPB and the aortic blocking time were 291.36 ± 83.71 (180–516) and 190.35 ± 22.64 (135–218) min, respectively. The mean endotracheal intubation time was 153.18 ± 149.19 h (21–360 h). The ICU length of stay was 13.23 ± 11.51 days (3.5–40 days), and the mean hospitalization stay was 23.09 ± 11.48 days (11–52 days). One patient required a brief period of extracorporeal membrane oxygenation (ECMO) support after repair, finally weaned off successfully on postoperative day 5. During the perioperative period, 1 patient required diaphragmatic paralysis for respiratory failure due to diaphragm paralysis; another required a brief period of peritoneal dialysis and 2 patients died with serious cardiac arrhythmias and circulatory collapse (n = 1) or sudden death (n = 1). Major early complications included: pneumonia (n = 6); arrhythmia (n = 2); pleural effusion (n = 2); pneumothorax (n = 1); respiratory tract haemorrhage (n = 1) and mild aortic insufficiency (n = 1). During the 2–8 years of follow-up (clinical visits n = 9 and telephone interviews n = 3): 1 patient died from serious cardiac arrhythmias and circulatory collapse; and 8 patients were evaluated as New York Heart Association Functional Class I; and another 3 as Class II. Moderate aortic valve regurgitation was noted in 3 patients and moderate mitral regurgitation was noted in 1 patient; all 4 of these patients are still in follow-up; none of the surviving patients from the original cohort required a further operation.
DISCUSSION

CCTGA is a complex congenital heart defect characterized by atioventricular and ventriculo-arterial discordance, and in which the mRV cannot support the systemic circulation because it becomes overloaded. Patients need early intervention to avoid progressive dilatation of the mRV and aggravation of TR, which could lead to a vicious circle and eventually result in RV failure or severe TR. The optimal management for these patients remains controversial. Conventional repair, still leaving the mRV in the systemic circulation, comprises VSD repair, tricuspid valve replacement or plasty and pulmonary valvoplasty. Conservative management such as these is still associated with a high incidence of late progressive mRV dysfunction and TR. Anatomical correction may be superior to conventional repair in terms of higher survival rate and better living quality: these rely on the mLV providing an adequate pressure and ventricular mass to support the systemic circulation. In certain patients, especially older children, there is a restricted VSD or intact ventricular septum: in such children, the mLV anomaly is associated to varying degrees with inadequate support for the pulmonary circulation and with a low pressure and volume loading; the ventricular wall of the mLV is thinner; the ventricular septum can shift towards the mLV as a result of the pressure from the mRV, which is overloaded with pressure and volume; and the mLV cannot support the systemic circulation following the DS procedure. These patients require preliminary mLV training to recondition the mLV; then they might be eligible for anatomical repair [4–6]. During the past two decades, the anatomical repair of DS operation for CCTGA has been applied with good clinical results. However, the two-stage DS operation is a complex procedure and according to the literature, the mortality rate is still high and more attention should be paid to mLV function after surgery.

Table 1: Changes in haemodynamic parameters in response to morphological left ventricular (mLV) retraining

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Preband</th>
<th>Early post-band</th>
<th>Later post-band</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>mLVED (mm)</td>
<td>21.20 ± 7.01</td>
<td>24.64 ± 6.26</td>
<td>28.00 ± 7.10</td>
<td>0.0086*</td>
</tr>
<tr>
<td>Posterior wall thickness of the mLV (mm)</td>
<td>4.64 ± 1.65</td>
<td>5.08 ± 1.62</td>
<td>5.38 ± 1.50</td>
<td>0.023*</td>
</tr>
<tr>
<td>PmLV/mRV</td>
<td>0.37 ± 0.10</td>
<td>0.78 ± 0.10</td>
<td>0.72 ± 0.167</td>
<td>0.00012**</td>
</tr>
<tr>
<td>Band velocity (m/s)</td>
<td>-</td>
<td>3.60 ± 0.36</td>
<td>4.35 ± 0.37</td>
<td>0.0000003***</td>
</tr>
<tr>
<td>Ejection fraction (%)</td>
<td>63.00 ± 6.12</td>
<td>65.00 ± 6.32</td>
<td>66.00 ± 4.24</td>
<td>0.072**</td>
</tr>
</tbody>
</table>

mLV: morphological left ventricle; mRV: morphological right ventricle; mLVED: mLV end-diastolic diameter.

*Significant: early post-band versus preband.
**Significant: later post-band versus preband.
The age of banding might be the main determinant of the efficacy of the mLV retraining and the later function of the myocardium. It is generally accepted that, in younger children, especially infants, who have a mainly immature myocardium, the response to the retraining is accompanied by cardiac cell proliferation and regional vascularization and, thus, further myocardial hyperplasia. However, in older patients, with generally mature myocardium, the response to banding is hypertrophy rather than hyperplasia, which might account for the findings of diastolic dysfunction of the mLV and less effective durability of the mLV retraining. Therefore, some studies suggest that, once mLV degradation occurs in CCTGA children, mLV training should be performed as early as possible. Older patients, especially in the later teenage years, may miss the optimum timing for the DS procedure, even though mLV training has been performed [7–11]. Winlaw et al. performed mLV training in 39 CCTGA children with an average follow-up period of 4.3 years (25 days to 12.6 years). The result showed that, in older patients (especially those aged over 16 years), mLV retraining might delay the progressive degradation of the mLV and chronic mRV failure, but the likelihood of eventual success with anatomical repair was small [12]. According to Poirier et al.’s study, there was a greater probability of mLV failure and a higher operative mortality at the DS procedure if the patients were aged over 12 years. They also pointed out that, as patients grew up, the rates of postoperative mLV dysfunction and mortality after the DS procedure became significantly higher [13]. In our series, among 14 patients who underwent the two-stage DS operation, the oldest of the 14 subjects underwent two episodes of mLV retraining, at the ages of 5 and 10 years, respectively, and proceeded to undergo the DS procedure 16 days after his second mLV retraining. He required ECMO support after repair for 5 days and his remaining recovery appeared uneventful, until unfortunately he died suddenly on the day of discharge: the possible cause being serious cardiac arrhythmia.

Furthermore, suitable banding of the pulmonary artery and effective evaluation of the efficacy of LV retraining are of great importance. Banding that is too loose may not produce the expected effect. By contrast, banding that is too tight, too long or repeated may cause myocardial fibrosis, subendocardial oedema or ischaemia, which would lead to myocardial necrosis or hypertrophy and a reduced ventricular work index. These patients could have their mLV pressure increased to systemic levels, but severe mLV diastolic dysfunction may occur after the DS procedure. Overbanding of the pulmonary artery will cause a sudden increase in wall stress on the PA root, which could result in damage or rupture of the elastic fibres in the vascular wall and lead to significant dilatation of the neoaortic sinuses and regurgitation of the neoaortic valve: all these would severely affect postoperative LV function [14–17]. In our opinion, according to the patient’s age, underlying diagnosis and preoperative baseline mLV volume and pressure, we suggest that an ideal mLV/mRV pressure ratio after PAB should be achieved between 0.65 and 0.80. Intraoperative oesophageal and postoperative echocardiography were repeatedly applied to monitor the efficacy of mLV retraining. We considered criteria for effective retraining to include: increase in mLVEDd; increase in the thickness of the mLV posterior wall; partial shift of the ventricular septum towards the mRV; TR improvement or stability; and the progressive increase of the velocity of blood flowing through the pulmonary artery band over time. Given that the retraining effect was less satisfactory at follow-up than immediately afterwards, the second mLV retraining should be performed as soon as possible after the first. However, unsatisfactory results of a DS procedure after multiple mLV retraining episodes have been reported [18, 19]. In our study, 2 patients underwent the DS operation after two episodes of mLV retraining. One patient is described above. The other child suffered from repeated supraventricular tachycardia after the operation, with progressive heart failure, and died 1 year postoperatively due to severe repeated supraventricular tachycardia in the context of mLV dysfunction. Some studies have suggested that, for older children or those with severely degraded mLV, it is better to use looser banding with a lower initial mLV/mRV pressure ratio, of the order of 0.50–0.60. Adjustable PA bands may be even more helpful: the mLV/mRV pressure ratio and the band velocity can gradually be increased over time, to keep up with the relative growth of the PA as the child grows. This can avoid excessive pressure overload, and effectively minimize the subsequent risk of ventricular damage caused by overbanding [20, 21].

Development of new aortic valve (neo-AR) regurgitation and aortic root dilatation can be more frequently seen in patients after the DS procedure, especially for the two-stage DS procedure [22–24]. In our studies, moderate aortic regurgitation was detected in three patients during follow-up. This could be due to: the PA being distorted by PA banding; damage or rupture of the elastic fibres in the vascular wall by the excessive PAB; or damage to the vascular integrity of the neoaortic sinuses, and their ability to expand, during coronary artery transplantation. The banding strip should be placed and secured nearly at the middle of the PA. A banding position that is too low can twist the pulmonary valve, whereas a banding position that is too high can cause distortion of the pulmonary artery bifurcation, especially the left pulmonary artery. Preservation of the diameter of the sinotubular junction during coronary artery transplantation should also be considered.

CONCLUSIONS

Our study shows that, for CCTGA children with a degraded mLV, anatomical correction of the CCTGA through mLV retraining, followed by the DS procedure, can be performed with low mortality and morbidity, and may be an appealing alternative to the conventional approach. mLV retraining should be performed as early as possible. Evaluation of the DS procedure as a second-stage operation indicated satisfactory early and mid-term results in 12 of 14 subjects. The long-term results require further follow-up. During follow-up, attention should be paid to the long-term function of the mLV and the neoaortic valve.

Conflict of interest: none declared.

REFERENCES


