Long-term prognosis of double-switch operation for congenitally corrected transposition of the great arteries†

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Abstract

OBJECTIVES: Recently, the double-switch operation for congenitally corrected transposition of the great arteries has become the procedure of choice in our institute; however, the long-term follow-up is uncertain.

METHODS: From 1983 to 2010, 90 patients with congenitally corrected transposition of the great arteries underwent the double-switch operation, which comprised of an atrial switch plus intraventricular rerouting (with or without extracardiac conduits) in 72 patients (group I), and an atrial switch plus arterial switch in 18 patients (group II). The mean age at operation was 7.4 years old in group I vs. 4.3 years old in group II. The mean follow-up period was 12.9 years in group I vs. 10.9 years in group II. Hospital and late mortality, reoperation, arrhythmia and NYHA status were analysed retrospectively.

RESULTS: The Kaplan–Meier survival, including hospital and late mortality at 20 years, was similar (75.7% in group I vs. 83.3% in group II). The freedom from reoperation was 77.6% in group I (redo-Rastelli in five patients, subaortic stenosis resection in three, tricuspid valve replacement in one and mitral valve plasty in one) vs. 94.1% in group II (P < 0.05 vs. group I; aortic valve replacement in one). The freedom from arrhythmia was 57.1% in group I vs. 78.6% in group II (P < 0.05 vs. group I). The ratio of NYHA class I to II at outpatient clinic was similar (86% in group I vs. 86% in group II).

CONCLUSIONS: The long-term prognosis of the double-switch operation for congenitally corrected transposition of the great arteries was acceptable. In particular, an atrial switch plus arterial switch could be performed with low morbidity, and it should be considered as the optimal procedure.

Keywords: Double-switch operation • Long-term prognosis • Corrected transposition of great arteries

INTRODUCTION

The double-switch operation (DSO) is expected to improve long-term prognosis by using the morphological left ventricle in the systemic circulation compared with conventional repair for congenitally corrected transposition of the great arteries (cc-TGA). We have already shown that in spite of a lack of statistical differences between long-term survival rates of patients who underwent conventional surgical repair vs. those of patients who underwent DSO or anatomical surgical repair, the results of anatomical repair were satisfactory even for patients with significant tricuspid regurgitation [1–3], and DSO for cc-TGA has become the procedure of choice in our institute.

In this study, we observed the medical records of patients who underwent DSO to investigate the early and late outcome after DSO and review the long-term prognosis of DSO for cc-TGA.

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employed in 49 of 72 patients, a direct pulmonary artery–right ventricular anastomosis in 22 of 72 patients and an intraventricular conduit in one of 72 patients in group I.

At a prior operation, a Blalock Taussig shunt was performed in all patients, and subclavian flap aortoplasty was performed in one patient in group I. Pulmonary artery (PA) banding (PAB) was performed in 14 patients in group II [PAB for left ventricular (LV) training in eight patients and protective PAB for congestive heart failure in six patients] and subclavian flap aortoplasty was performed in two patients. As a concomitant procedure, aortic valve replacement (AVR) was performed in one patient and mitral valve replacement in one patient in group I.

RESULTS

There were nine hospital deaths (eight of 72 patients in group I and one of 18 in group II) and eight late deaths (six of 64 patients in group I and two of 17 in group II). The causes of the late deaths were sudden death in three patients, multiple organ failure in one, congestive heart failure in one and infective endocarditis in one in group I and sudden death in one and congestive heart failure in one in group II. The Kaplan–Meier actuarial survival rate, including hospital and late mortality at 20 years, was similar (75.7% in group I vs. 83.3% in group II; Fig. 1).

There were 11 reoperations (10 in group I and one in group II). The freedom from reoperation was 77.6% in group I (conduit exchange in five patients, resection of subaortic stenosis in three, tricuspid valve replacement in one and mitral valve plasty in one) vs. 94.1% in group II (AVR in one).

There were 17 reinterventions in group I (conduit stenosis in 12 patients and superior vena cava stenosis in five). Regarding superior vena cava stenosis, the Mustard procedure was used in four patients and the Senning procedure in one. There were no reinterventions in group II.

In the long-term follow up of the survivors, the details of arrhythmia were atrial tachycardia in 14, ventricular tachycardia in three (an implantable cardioverter–defibrillator was implanted in one), atrial fibrillation in three, atrioventricular block in eight (pacemaker was implanted in eight) and sick sinus syndrome in six in group I and atrioventricular block in two (a pacemaker was implanted in two) in group II. The freedom from arrhythmia was 57.1% in group I vs. 78.6% in group II.

Total freedom from all cardiac events, including hospital death, late death, reintervention, reoperation and arrhythmia, were significantly higher in group II (40% in group I vs. 60% in group II; Fig. 2).

Regarding the patient’s quality of life from the NYHA classification, 70% of patients (45 of 64) were in NYHA class I and 16% (10 of 64) in NYHA class II in group I, while 72% of patients (12 of 17) were in NYHA class I and 6% of patients (one 17) in NYHA class II in group II (Fig. 3). The ratio of NYHA I to II at outpatient clinic was similar (86% in group I vs. 78% in group II).

The average plasma brain natriuretic peptide levels at the recent outpatient clinic were 59.1 pg/ml in group I and 48.9 pg/ml in group II, and there were no statistically significant differences.

Cardiac catheterization was performed in 37 patients (51% of the survivors). The average left ventricular ejection fraction was good in both groups (50.9% in group I and 56.5% in group II), and the average cardiac index was likewise also good in both groups of patients, but significantly higher in group II (2.8 l/min/m² in group I vs. 3.8 l/min/m² in group II).

DISCUSSION

A prevalence of systemic right ventricular dysfunction and an abnormal response of the ejection fraction with exercise have been reported in patients with cc-TGA or with an atrial switch...
Figure 3: Clinical status of the survivors. NYHA: New York Heart Association; LD: late death. The ratio of NYHA I to II at outpatient clinic was similar.

repair for dextro-transposition of the great arteries [4–7]. These patients are at risk of right ventricular and tricuspid valve dysfunction with increasing age [8]. The DSO is expected to improve the long-term prognosis by using the morphological left ventricle in the systemic circulation, compared with conventional repair for cc-TGA. Some advantages are particularly notable in terms of improving right ventricle and tricuspid valve function combined with low morbidity and mortality, and the fact that the left ventricle becomes the systemic ventricle [9]. Based upon the theoretical advantages of DSO, DSO for cc-TGA has become the procedure of choice in our institute. We have already compared the results of DSO with those of the conventional surgical repair [1–3], but no study has compared the long-term results between atrial switch plus intraventricular rerouting and atrial switch plus arterial switch for cc-TGA.

Despite its complexity, anatomical repair or DSO nowadays represents a good option in the management of cc-TGA surgery. Ly et al. showed excellent surgical results for the atrial switch plus arterial switch operation [10]. From 1995 to 2007, 20 patients with cc-TGA without right ventricular outflow tract obstruction had the DSO. A median follow-up of 60 months was achieved in all and there were no deaths. However, PAB is one of the important concerns in the atrial switch plus arterial switch operation. PAB was identified as a risk factor for the development of pulmonary root dilatation and subsequent neo-aortic valve regurgitation [11], and in the presence of an associated haemodynamically significant ventricular septal defect (VSD), primary repair can be discussed early in life. It avoids potential deterioration of the pulmonary valve after PAB. However, in the series, patients who presented with cardiac failure during the neonatal period and even later were more likely to undergo a two-stage management, with initial PAB. This was obviously due to the limited experience with the DSO as well as the Senning procedure early in life [12]. Another group mentioned that in neonates with isolated cc-TGA, prophylactic PAB is safe and carries a low morbidity. At mid-term evaluation, tricuspid valve function is stabilized or improved and systemic competence of the left ventricle is maintained, thus allowing the double switch if indicated [13]. Although only one patient presented more than moderate aortic valve regurgitation and needed AVR in our series, close observation is still necessary in future.

To avoid complications associated with the complete Senning and Mustard procedures and to assist right-heart haemodynamics, Malhotra et al. favour 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 [12]. The risks of the hemi-Mustard and bidirectional Glenn operation are minimal, and the benefits of this procedure include prolonged conduit life, reduced baffle- and sinus node-related complications, and technical simplicity, and they described favourable mid-term results. However, the occurrence of pulmonary arteriovenous fistulae (PAVF) remains a serious concern in this procedure, and careful observation for PAVF must be made in future.

Conduction abnormalities are common in discordant atroventricular connections, predisposing the patients to the development of complete heart block [14]. The incidence of heart block or ventricular tachycardia was higher in the atrial-Rastelli group, which was related to the fact that VSD enlargement was aggressively performed to relieve a potential subaortic stenosis in our series. VSD enlargement was a risk factor for complete heart block/pacemaker implantation, although it was not a risk factor for hospital and late death.

The potential risk of supraventricular tachyarrhythmia may exist due to the atrial switch procedure, especially the Mustard procedure. In our series, 41 Senning and 49 Mustard procedures were selected as atrial switch procedures, depending on the size of the right atrium, and the Mustard procedure was more frequently employed in group I. This might be the reason why supraventricular arrhythmia was more frequent in group I in our series. Recently, we suggested that the Senning procedure was feasible in all atrial switch procedures, applying a large pericardial patch to augment a new functional left atrium, as others have described [15, 16]. The Senning procedure has become the most widely used variant because of the lower incidence of pathway obstruction, baffle leak and significant late arrhythmias. Obstruction to the superior vena cava pathway has been recorded in less than 3% of cases and can usually be managed by interventional catheterization. In our series, reintervention due to superior vena cava stenosis was necessary in five patients in group I, of whom four patients were treated with the Mustard procedure. This might be the reason why reintervention was more frequent in group I. Late problems with atrial arrhythmias have not been widely reported, but this may reflect the relatively short follow up for these patient cohorts compared with older series in d-transposition of the great arteries.

The atrial switch plus Rastelli-type group will require change on a regular basis of their ventricular to pulmonary artery valved conduits. In our series, the external conduit was employed in 49 of 72 patients in group I, and only five patients underwent
conduit exchange during the mean follow-up period of 12.9 years. However, the number of reoperations for conduit exchange should increase in the future.

Barron et al. mentioned that in the atrial switch plus arterial switch group after training of the morphological left ventricular by prior pulmonary artery banding, there is an incidence of morphological left ventricular dysfunction, sometimes associated with neo-aortic valve regurgitation, and a minority of patients need aortic valve replacement [17]. Curiously, such LV dysfunction has never been observed in our series, and we speculate that the reason for this difference may be that their LV dysfunction may be due to relatively older age at operation. The LV training was done in 11 of 44 patients, and the median age at DSO of those who required LV training was 61.9 months in the Barron et al. series. In contrast, LV training was done in eight of 18 patients and the median age at DSO was 42.5 months in our series. The young age at operation might be an important factor, as mentioned by Serraf [10]. In any case, careful surveillance of the long-term function of the morphological left ventricle and the aortic valve are needed in the future.

According to the results of this study, the present indications for the double-switch operation for cc-TGA in our institute are thought to be as follows: (i) an age limit of 15 years old; and (ii) estimated right ventricular volume > 70% of normal in the atrial and arterial switch and > 150% of normal in the atrial switch and intraventricular rerouting.

In conclusion, the long-term prognosis of DSO for cc-TGA was acceptable. In particular, an atrial switch plus arterial switch could be performed with low morbidity, and it should be considered as the optimal procedure.

Conflict of interest: none declared.

REFERENCES


APPENDIX. CONFERENCE DISCUSSION

Dr W. Brawn (Birmingham, UK): We are at an interesting phase in the management of cc-TGA. Hopefully, we should be able to define more clearly the indications for different surgical processes used for this condition. This nice paper from your unit helps us and represents an earlier series of patients undergoing anatomical repair. The majority of patients in your group had a Rastelli–Senning or a Rastelli–Mustard type of procedure. Does this reflect the spectrum of morphology you find in your country or surgical selection?

Dr Hiramatsu: I think the spectrum of morphology is a little bit different between Europe and Japan, and I think pulmonary stenosis is more frequent in Japan; and another reason is therapeutic attitude or therapeutic positive indications, I think.

Dr Brawn: Okay. So there is a difference in morphology, probably. Another question is that you note that in your previous publications there is no difference really, or very little difference, between conventional and the anatomical repair. How do you now select for anatomical repair?

Dr Hiramatsu: Recently, in our selection criteria for double-switch operation, the age limit is 15 years old and right ventricular volume must be over 70 or 80% of normal in the arterial and atrial switch operation, and right ventricular volume must be over 130–150% of normal in cases of atrial and Rastelli type of operation.

Dr Brawn: Okay. So you try and make some objective measurements. Only one of the patients had an aortic valve replacement. Can you let us know what the status of the aortic valve was in the other patients? Do you have any problems with aortic valve regurgitation?

Dr Hiramatsu: No. We had only one case of AVR in group II, which means the atrial and arterial switch operation. In other group II patients, we have never experienced significant aortic valve deformity. I think it depends on the age at double-switch operation.

Dr Brawn: Okay. Now, regarding PA banding, eight out of the 18 patients who had a double switch had a PA band to train the left ventricle. Could you give us any insight as to how you decide when to intervene to train the left ventricle or retrain the left ventricle?

Dr Hiramatsu: In our recent criteria or indications for PA banding for LV training, LV pressure must be over 80% of the systemic pressure.

Dr Brawn: Okay. I am curious: do you let some patients with a natural pulmonary valve stenosis go unoperated? Maybe they can live 70 years unoperated. Could you tell us any insight as to how you decide when to intervene to train the left ventricle or retrain the left ventricle?

Dr Hiramatsu: As I mentioned before, 15 years old is the maximum. Dr D. Schranz (Giessen, Germany): Do you let some patients with a natural pulmonary valve stenosis go unoperated? Maybe they can live 70 years unoperated if you have a pulmonary banding or pulmonary stenosis. Do you make this decision in any case for a surgical procedure, or do you let some patients go without any operation?

Dr Hiramatsu: We did not. Dr Schranz: Is there a comment from you maybe, Pascal? Because we can have patients who can live 70 years, maybe, with a balanced circulation when...
Hiramatsu et al. [1] have reported their experience with an anatomic repair of congenitally corrected transposition of the great arteries (ccTGA). This included 90 patients operated on between 1983 and 2010. Seventy-two patients with ccTGA, ventricular septal defect (VSD) and pulmonary stenosis underwent atrial switch plus intraventricular rerouting. Eighteen patients without pulmonary stenosis had atrial switch plus arterial switch. The long-term results were carefully evaluated. Differences between the two groups were noted. Patients without pulmonary stenosis (the double-switch group) had a better outcome at 20 years regarding survival (83% vs 76%, NS), freedom from reoperation (94% vs 78%, P < 0.05) or arrhythmia (79% vs 57%, P < 0.05). The total freedom from all cardiac events (death, re-intervention or reoperation, arrhythmia) was significantly different (60% in the double-switch group vs 40% in the group with pulmonary stenosis). The functional outcome was similar in both groups and, overall, satisfactory. These results should, however, be taken with caution, because of the small number of patients in the double-switch group.

Interestingly, the Birmingham group (another institution with a large experience in this field) has recently reported its own experience between 1991 and 2011 [2]. This included 45 patients with Rastelli-Senning repair and 68 patients with double-switch operation. The results at 10 years were somewhat different. Survival (84% in the double-switch group vs 77% in the Rastelli-Senning group) and freedom from reintervention (50% vs 49%) were similar in both groups. However, freedom from death, transplantation or heart failure was better in the Rastelli-Senning group.

These results may seem contradictory. Actually, I think that they show only how difficult it is to draw out firm conclusions when so many factors are involved. ccTGA is a rare malformation; it is therefore difficult (actually impossible for any individual institution) to collect large cohorts of patients who undergo homogeneous surgical approaches. Very different patients are involved; patients with right ventricular dysfunction and severe tricuspid regurgitation because of Ebstein-like malformation are different from patients with a well-functioning right ventricle and a competent tricuspid valve; patients with an intact ventricular septum who need left ventricular retraining before undergoing a double-switch operation are different from patients with a large VSD and a left ventricle ready to sustain the systemic output. Different surgical options are available and may yield different results; regarding atrial switch, the Senning and Mustard operations or hemi-Mustard procedure with cavo-pulmonary anastomosis may influence the incidence of post-operative arrhythmias; reconstructing the right ventricular outflow tract with or without an extracardiac conduit may greatly modify the need for reoperation. Well-designed large multicentre cohort studies are necessary to address all these issues.

Before such information is available, some considerations must be kept in mind:

(1) As illustrated by the present paper, as well as by the Birmingham’s report, anatomic repair may achieve excellent late results in selected patients but cannot be considered as the optimal option for all patients with ccTGA. It is anatomically impossible in some patients (i.e. severe malformation of the mitral valve, non-committed or restrictive VSD). It entails a significant late morbidity. Of major concern is the risk of