Results of the double switch operation for congenitally corrected transposition of the great arteries

Brian W. Duncan\textsuperscript{a,*}, Roger B.B. Mee\textsuperscript{a}, C. Igor Mesia\textsuperscript{b}, Athar Qureshi\textsuperscript{b}, Geoffrey L. Rosenthal\textsuperscript{b}, Shivaparakash G. Seshadri\textsuperscript{a}, Geoffrey K. Lane\textsuperscript{b}, Larry A. Latson\textsuperscript{b}

\textsuperscript{a}Pediatric and Congenital Heart Surgery, The Children’s Hospital at The Cleveland Clinic, 9500 Euclid Avenue, Cleveland, OH, USA
\textsuperscript{b}Pediatric Cardiology, The Children’s Hospital at The Cleveland Clinic, 9500 Euclid Avenue, Cleveland, OH, USA

Received 27 September 2002; received in revised form 21 March 2003; accepted 25 March 2003

Abstract

Objective: To determine the outcomes for a program that utilizes the double switch operation as the primary approach for congenitally corrected transposition.

Methods: The records of 46 consecutive patients from a single institution who had undergone a double switch operation from October 1993 to March 2002 were reviewed. The records of 24 patients who were evaluated during the same period and felt not to be candidates for the double switch operation or who are awaiting double switch after pulmonary artery banding were also reviewed.

Results: The median age at operation was 28 months (range 2 months to 16.3 years). Associated defects included ventricular septal defect 40, pulmonic stenosis 13 and pulmonary atresia 16. Twenty-six patients underwent an arterial switch operation combined with a Senning procedure while 20 patients underwent combined Rastelli and Senning procedures. Before the double switch, 12 patients had required pulmonary artery banding and 21 patients had systemic to pulmonary artery shunts. The median duration of stay in the intensive care unit was 3.5 days (range 2–60 days) and the median duration of total hospital stay was 8 days (range 5–60 days). There were no hospital deaths; one patient died 5 months after discharge due to an arrhythmogenic cardiac arrest during a median follow-up of 24 months [long-term survival 98% (95% CI 89–100%).

Conclusions: The double switch operation may be performed with excellent hospital and long-term survival. The theoretical advantages of this procedure which enables the morphologic left ventricle and mitral valve to support a systemic pressure load must be established by careful follow-up of these patients.

\textsuperscript{©} 2003 Elsevier Science B.V. All rights reserved.

Keywords: Congenitally corrected transposition; Double switch operation; Senning procedure; Rastelli procedure; Arterial switch operation

1. Introduction

The combination of atrioventricular discordance and ventriculoarterial discordance, termed congenitally corrected transposition of the great arteries (ccTGA), produces a unique set of management challenges. Atrioventricular discordance results in physiologic correction of the transposed great arteries which, if associated cardiac anomalies are minor or absent, may allow prolonged survival. However, associated anomalies such as ventricular septal defect (VSD), morphologic left ventricular outflow tract obstruction and tricuspid valve abnormalities occur in the majority of these patients [1,2]. Although the presence of these associated conditions influences the clinical presentation of this condition and the type and timing of surgical intervention, the ability of the morphologic right ventricle and the tricuspid valve to withstand a lifetime of exposure to systemic pressure largely determines the ultimate outcome of these patients.

Progressive dysfunction of the tricuspid valve and right ventricle occurs after prolonged exposure to systemic pressure in a substantial percentage of patients who have undergone an atrial switch operation for transposition of the great arteries with increased risk for patients who have an accompanying VSD [3–5]. Similarly, progressive dysfunction of the morphologic right ventricle has been described in a substantial percentage of unoperated patients with ccTGA or in those patients who have undergone ‘classic’ intracardiac repair comprised of VSD closure with operations
on the tricuspid valve and/or the morphologic left ventricular outflow tract [6–14].

We have previously published our approach utilizing the double switch operation as the primary surgical treatment for patients with ccTGA [15–18]. The rationale for this approach is that the morphologic left ventricle and mitral valve are utilized to support the systemic circulation. The aim of the present report is to provide long-term follow-up of patients treated with this approach and to establish the impact of the double switch operation on the function of the right ventricle and tricuspid valve. In addition, refinements that have evolved in patient selection for this procedure will be discussed.

2. Methods

From October 1993 through March 2002, 46 patients underwent the double switch operation at this institution. A review of 22 of these patients has been previously reported [16]. The median age of these 46 patients was 2.3 years (range 2.0 months to 16.3 years) and the median weight was 12 kg (range 3.6–56 kg). Twenty-six patients (56%) underwent a combined Senning procedure and arterial switch operation (S–ASO) while 20 patients (44%) underwent combined Senning and Rastelli procedures (S–R). The median age of the 26 S–ASO patients was 2.0 years (range 2.0 months to 16.3 years) with a median weight of 10.2 kg (range 3.6–56 kg). The median age of the 20 S–R patients was 2.6 years (range 9.9 months to 8.6 years) with a median weight of 12.6 kg (range 6.9–34 kg). Forty-one patients (89%) were situs solitus (S,L,L segmental anatomy) while five patients were situs inversus (I,D,D segmental anatomy). Twenty-nine patients had levocardia, 12 patients had dextrocardia and five patients had mesocardia.

2.1. Associated lesions

Forty patients (87%) had a hemodynamically significant VSD. Twenty-eight patients (61%) had hemodynamically significant obstruction of the morphologic left ventricular outflow tract with 12 cases (26%) of pulmonic stenosis and 16 cases (35%) of pulmonary atresia. Two patients (4%) had significant aorto-pulmonary collateral arteries. Ten patients (22%) had unequivocal Ebstein’s malformation of the tricuspid valve, which was judged to be mild in three, moderate in two and severe in five. Three patients (6.5%) had complete heart block preoperatively.

2.2. Preoperative evaluation

All patients underwent preoperative echocardiography with tricuspid regurgitation graded according to the following scale: none, mild, moderate and severe tricuspid regurgitation. Right ventricular dysfunction was graded as normal, mild, moderate and severe right ventricular dysfunction. Thirty-five patients (81%) underwent preoperative cardiac catheterization to determine the left ventricular pressure, to determine the pulmonary vascular resistance and to fully delineate the cardiac anatomy. Five patients (11%) underwent magnetic resonance imaging to determine the indexed left ventricular mass (median 61 gm/m²; range 54–70 gm/m²) and left ventricular function [19].

2.3. Previous operative procedures

Thirty-four patients (74%) had undergone surgery prior to the performance of the double switch operation. Twelve patients (26%) underwent prior pulmonary artery banding for the control of congestive heart failure in six patients and for retraining of the left ventricle in six patients. One patient with a type B interruption of the aortic arch underwent arch repair and pulmonary artery banding as the initial procedure. One patient with complete heart block underwent pacemaker insertion at the time of pulmonary artery banding. Pulmonary artery banding to retrain the left ventricle was first performed a median of 19.4 months (range 6.3–64 months) prior to double switch: two patients required no band revisions, one patient required one band revision, two patients required two band revisions and one patient required three band revisions. The techniques for pulmonary artery banding for left ventricular retraining and the careful follow-up necessary to determine that the left ventricle has been suitably prepared for the double switch operation have been previously described [15–18].

Twenty-one patients (46%) underwent a prior systemic to pulmonary artery shunt (20 systemic arterial to pulmonary artery shunts and one superior vena cava-pulmonary anastomosis). One patient with associated total anomalous pulmonary venous return and LPA stenosis underwent an initial repair of the anomalous pulmonary venous drainage, patch enlargement of the left pulmonary artery and a systemic to pulmonary artery shunt as the initial procedure. One particularly complex patient with combined ccTGA, tetralogy of Fallot and pulmonary atresia underwent five procedures including initial placement of a central (Melbourne) shunt and staged unifocalizations prior to undergoing a S–R procedure and has been previously reported [20]. One patient underwent isolated repair of aortic coarctation prior to double switch.

2.4. Operative technique

The operative techniques for both the S–ASO and S–R have been described previously [15,16]. The Senning procedure is performed first. The Shumacker modification, utilizing in situ pericardium for the completion of the pulmonary venous atrium, was utilized when there was discordance of the atrial situs and the position of the ventricular apex or if the amount of atrial tissue appeared...
Inadequate to create a chamber for pulmonary venous return [21]. Patients with concordance of the atrial situs and the position of the ventricular apex are those with S,L,L segmental anatomy and levocardia or I,D,D segmental anatomy and dextrocardia. Patients with atrial situs and ventricular apical discordance include those with S,L,L segmental anatomy and dextrocardia or I,D,D segmental anatomy and levocardia. Nineteen of the 46 patients (41%) had a Shumacker modification of the Senning portion of the procedure. A VSD was closed in patients undergoing S–ASO via the right atrium for S,L,L patients and via the left atrium for I,D,D patients according to the technique described by de Leval et al. [22] to minimize the risk of complete heart block. For patients who underwent a S–R, the VSD was closed through the incision in the morphologic right ventricle and a 14–22 mm Hancock-valved Dacron conduit was used to reconstruct the right ventricular outflow tract in all but a single patient in whom an aortic homograft (20 mm) was utilized.

2.5. Patients not undergoing the double switch operation

During the same period, 24 patients presented for evaluation who have not undergone a double switch operation (Table 1). Nine of these patients underwent pulmonary artery banding to condition the morphologic left ventricle in anticipation of an eventual arterial switch operation. Two of these patients subsequently died after pulmonary artery banding while seven remain as candidates for eventual double switch. Fifteen patients were treated with an approach other than the double switch operation including four patients who have been treated with a single ventricle approach, six patients who have undergone a classic two-ventricle repair and five patients who were treated with heart transplantation.

2.6. Statistical analysis and institutional approval

Comparisons between preoperative and postoperative amounts of tricuspid regurgitation were made using the Wilcoxon signed ranks test. Survival and reoperation-free survival were demonstrated using Kaplan Meier curves. This study was approved by the Institutional Review Board of The Cleveland Clinic Foundation.

3. Results

3.1. In-hospital results

There was no 30-day or hospital mortality. The mean cardiopulmonary bypass times and aortic cross-clamp times were 201 ± 49 and 104 ± 21 min, respectively for the S–ASO and 192 ± 47 and 101 ± 20 min, respectively for the S–R. Patients were intubated for a median of 2 days (range 1–21 days), were monitored in the intensive care unit for a median of 3.5 days (range 2–60 days) and were hospitalized for a median of 8 days (range 5–60 days). There were 27 complications in 16 patients (Table 2). One patient with I,D,D segmental anatomy, pulmonic stenosis, VSD and a small left ventricle developed circulatory failure after a S–R which required support with a ventricular assist device. The patient subsequently was weaned from the device but suffered a cardiac arrest soon thereafter. This patient was emergently placed on cardiopulmonary bypass, the double switch was taken down and a Fontan procedure was performed due to the persistent inability of the small left ventricle to support the systemic circulation. This same patient had significant watershed cerebral infarcts from which there has been nearly complete recovery. Three patients who did not have complete heart block prior to surgery required pacemaker implantation after the double switch, two of whom have subsequently returned to sinus rhythm. The two children requiring tracheostomy have subsequently had full recovery allowing decannulation. The child who experienced acute tubular necrosis required hemodialysis for 20 days postoperatively, however, this patient subsequently had return of normal renal function.

3.2. Long-term outcomes

Intermediate-term follow-up is significantly incomplete and available for only 37 of the original 46 patients (80%) at a median of 24 months (range 6 weeks to 103 months) after the double switch. The nine patients without follow-up were referred from outside of the US; further information is not available for these patients. There was one late death in a 7 year-old with S,L,L segmental anatomy, a perimembranous VSD and severe pulmonary stenosis. The patient underwent a S–R which was complicated by postoperative renal failure and persistent pleural effusions. The patient had Ebsteinoid malformation of the tricuspid valve and moderate tricuspid regurgitation preoperatively which progressed to severe tricuspid regurgitation postoperatively. The patient’s renal failure and effusions resolved and the patient was discharged to home 27 days postoperatively where the patient suffered a sudden cardiac arrest, presumably due to a primary arrhythmogenic event, 5 months after surgery. All but one of the survivors for whom follow-up is available are in NYHA functional category I while a single patient is in category II.

Three patients required late operative intervention. One patient had complete heart block requiring pacemaker implantation 10 weeks after the double switch. One patient developed significant mitral regurgitation that was treated with mitral annuloplasty 5 years after surgery. One patient who had undergone left ventricular retraining and a subsequent S–ASO developed progressive left ventricular dysfunction in the postoperative period. This patient’s left ventricular function was no more than mildly depressed in the immediate postoperative period but deteriorated to severe dysfunction over a period of several months. The

intermediate postoperative period but deteriorated to severe dysfunction over a period of several months.
patient underwent successful cardiac transplantation 21 months after the double switch operation. The Kaplan Meier survival and reoperation-free survival curves for these 46 patients are shown in Fig. 1.

Follow-up echocardiography was available for 32 patients at a median follow-up of 26 months (range 2–103 months). Tricuspid regurgitation decreased significantly in these patients after double switch (P < 0.001 for preoperative versus postoperative tricuspid regurgitation) (Fig. 2). In addition, right ventricular function was abnormal in more than 20% of patients preoperatively while after the double switch two patients have abnormal right ventricular function (6%) (Fig. 3). All but two patients with available follow-up have normal left ventricular function after the double switch operation; one patient has mild left ventricular dysfunction and the patient discussed above with severe left ventricular dysfunction requiring transplantation.

### 4. Discussion

For the majority of patients with ccTGA and associated
lesions such as VSD, obstruction of the left ventricular outflow tract or Ebstein’s malformation of the tricuspid valve, surgical therapy will be required. However, the ability of the morphologic right ventricle and tricuspid valve to provide lifelong support of the systemic circulation is uncertain, even if associated lesions are mild or absent. For this reason, the double switch operation has been advocated for the treatment of patients with ccTGA with concomitant treatment of hemodynamically significant associated conditions. After the double switch operation the morphologic left ventricle/mitral valve support the systemic circulation while the right ventricle/tricuspid valve support the pulmonary circulation. This approach has obvious theoretical appeal, however, the double switch operation is a complex and technically demanding operation. Since long-term survival is possible in some ccTGA patients even without operation, it is imperative that the results of the double switch be regularly reviewed to ensure that the benefits outweigh the risks. This report details the experience over a decade at an institution that employs the double switch operation as the primary surgical approach for patients with ccTGA.

4.1. Patient selection

General guidelines for selection of patients for either a S–ASO or a S–R have been reported previously [16]. The requirements for a S–ASO are that there is no more than mild morphologic left ventricular outflow tract obstruction at the end of the procedure and that the systolic left ventricular pressure is more than 70% of the systolic systemic pressure in children and closer to 100% in adolescents prior to surgery. Mild or dynamic pulmonic stenosis does not preclude a S–ASO and may confer some advantage by preconditioning the left ventricle. The requirements for a S–R are the presence of two usable ventricles and satisfactory branch pulmonary arteries without high pulmonary vascular resistance and a VSD which is committable to the aorta [16].

Fig. 4 summarizes the decision process for management of an infant who presents with ccTGA and unrestricted pulmonary blood flow who is presumed to be a candidate for a S–ASO. The size of the VSD and the presence of symptoms of congestive heart failure contribute to initial decision-making. For patients with a large VSD and congestive heart failure, timing of operation is determined by the presence of atrial situs and ventricular apical concordance or discordance. In general, if the patient’s condition allows it is preferable to defer a double switch operation until beyond early infancy in patients with atrial situs and ventricular apical discordance because the Senning portion of the procedure is technically more difficult in the youngest patients with this anatomy. Patients who possess unrestricted pulmonary blood flow and atrial situs apical discordance should undergo a pulmonary artery band to control pulmonary blood flow which allows a S–ASO to be performed when the patient is beyond early infancy when the Senning is technically easier. For patients with a large VSD and congestive heart failure, it is advisable to perform a double switch operation within the first 3 months of life if there is concordance of the atrial situs and the position of the ventricular apex. Early operation in these children prevents the development of complications related to congestive heart failure and irreversible pulmonary vascular obstructive disease.

For patients with unrestricted pulmonary blood flow and either a restrictive VSD or an intact ventricular septum, the presence or absence of congestive heart failure determines the next point in the decision process (Fig. 4). If these patients demonstrate significant congestive heart failure, it is usually due to the presence of tricuspid regurgitation. The amount of tricuspid regurgitation determines the next decision point: if tricuspid regurgitation is mild to moderate placement of a pulmonary artery band will further reduce the volume load on the ventricles, shift the ventricular...
septum toward the right ventricle, which will decrease the amount of tricuspid regurgitation and retrain a left ventricle operating at low pressures. For patients with severe tricuspid regurgitation early S–ASO should be strongly considered regardless of atrial situs ventricular apical concordance or discordance provided the morphologic left ventricular pressure is at least 70% of systemic pressure. Otherwise a pulmonary artery band is required or if the patient is in extremis an urgent S–ASO may be performed with postoperative left ventricular retraining with a left ventricular assist device. Patients with a restrictive VSD or an intact ventricular septum without congestive heart failure may be followed closely until right ventricular failure or tricuspid regurgitation develop. At that time these patients will require a pulmonary artery band to retrain the left ventricle. The guidelines for left ventricular retraining are now well established [15,16,18]. Patients with ccTGA and no significant associated lesions who remain free of congestive heart failure may continue to be followed as long-term survival without operative intervention is possible in this patient group.

Fig. 5 demonstrates the management approach to patients with ccTGA and restricted pulmonary blood flow. In considering the best approach to these patients it should be kept in mind that some milder forms of obstruction to pulmonary blood flow due to subvalvar disease will be amenable to resection of muscle or accessory atrioventricular valve tissue in the left ventricular outflow tract and a S–ASO procedure. Patients with a small pulmonary valve and unresectable subpulmonic obstruction should undergo a S–R procedure. The decision tree begins with an assessment of the severity of obstruction to pulmonary blood flow (Fig. 5). In general, patients with mild to moderate pulmonic stenosis will demonstrate one of three physiologic states: cyanotic patients with limited pulmonary blood flow, patients with relative balance between the systemic and pulmonary circulations and patients in whom the degree of pulmonic stenosis is mild with relatively high pulmonary blood flow. Patients with moderate limitation of pulmonary blood flow who become significantly cyanotic as young infants may be managed initially with a systemic to pulmonary artery shunt followed by a S–R when they outgrow the shunt, ideally at around 18 months of age. Patients with mild to moderate pulmonic stenosis with a pulmonary blood flow to systemic blood flow ratio of less than 2:1 and normal pulmonary artery pressures often present with a balanced circulation without significant cyanosis or evidence of pulmonary overcirculation. These patients may be followed until evidence of tricuspid regurgitation and right ventricular failure supervene and in the presence of atrial situs ventricular apical concordance, a S–R may then be performed at any age. In patients with discordance of the atrial situs and the ventricular apex, operation is easier in a somewhat older child which may justify waiting until beyond 1 year of age if there is mild tricuspid regurgitation. Some patients with pulmonic stenosis who are not amenable to resection and S–ASO may demonstrate high pulmonary blood flow (pulmonary to systemic flow ratio above 3:1) despite a 50–60 mm gradient across the left ventricular outflow tract. These patients usually come to S–R earlier due to more rapid development of tricuspid regurgitation with early right ventricular failure and even progressive increases in pulmonary vascular resistance. In these patients who are not amenable to a subpulmonic resection and a S–ASO, a S–R should be performed at the time these complications occur.

For patients who present with severe pulmonic stenosis or for patients who present with pulmonary atresia, a systemic to pulmonary artery shunt may be performed in the neonate. A 5 mm polytetrafluoroethylene interposition graft

![Fig. 3. Right ventricular function before and after double switch operation.](image)

![Fig. 4. Paradigm for managing patients with ccTGA and unrestricted pulmonary blood flow.](image)
performed through a thoracotomy between the subclavian artery and the pulmonary artery provides initial limitation to pulmonary blood flow that increases gradually as the patient grows. A S–R may be performed in the patient with atrial situs and ventricular apical concordance when the patient outgrows the shunt. Repeated shunt procedures would only be considered in the patient with atrial situs and ventricular apical discordance who outgrows the shunt early in an attempt to defer S–R until the child is approximately 18 months of age.

Both Figs. 4 and 5 attempt to convey the general management principles for the double switch operation; however, the importance of an individualized approach for each patient with ccTGA cannot be overemphasized. Most importantly, careful follow-up of these patients, noting subtle changes in their physiologic status such as early signs of tricuspid regurgitation or right ventricular enlargement, are required to determine the optimal timing of intervention.

4.2. Patients not suitable for the double switch operation

Rigidly applied contraindications do not exist for the double switch operation and the same highly individualized approach should be taken for any patient with ccTGA who is not an ideal candidate. However, general principles can be applied that suggest a given patient is a poor candidate for this complex procedure. Patients who present beyond adolescence with advanced right ventricular or especially biventricular failure would rarely be offered a double switch operation. The chances for successful ventricular retraining diminish in postpubescent individuals especially if ventricular function is already significantly compromised. Patients who present beyond adolescence with isolated tricuspid regurgitation and relatively well preserved ventricular function may occasionally be suitable for classic intracardiac repair with tricuspid valve repair or replacement and procedures to treat a VSD or left ventricular outflow tract obstruction. Older patients with advanced right ventricular or bi-ventricular dysfunction should probably be considered for heart transplantation.

Patients with a previously closed, large VSD have a poor outlook probably due to the same factors that lead to poor outcomes for patients with TGA–VSD: these patients often have poor tricuspid valve function and a non-contractile portion of the septum due to the VSD patch. Patients with significant dysfunction of the morphologic left ventricle and mitral valve do poorly when these structures are required to support the systemic circulation after a double switch operation. Finally, significant dysrhythmias and complete heart block confer less chance of long-term survival. While rhythm disturbances alone may not preclude a double switch operation, the presence of rhythm abnormalities may serve to disqualify an otherwise marginal candidate. Again, all of these considerations should be highly individualized: an older patient who presents with elevated pressures in the left ventricle due to resectable subpulmonic obstruction may tolerate retraining quite well and be an acceptable candidate while a patient of the same age with low left ventricular pressures might be expected to do poorly with retraining.

4.3. In-hospital outcomes

Despite its technical complexity, the double switch operation can be performed with low in-hospital mortality. In the present study, all patients survived to hospital discharge after the double switch operation in keeping with a number of reports which have also demonstrated low hospital mortality after the double switch [23,24]. Several factors are important contributors to good hospital outcomes. High flow cardiopulmonary bypass is utilized for all cases small enough to warrant a blood prime (<18 kg) with α-blockade established by administration of phenoxybenzamine (1 mg/kg) [16]. The Shumacker modification, utilizing in situ pericardium to enlarge the pulmonary venous atrium, greatly facilitates the performance of the Senning procedure in patients that possess atrial situs and ventricular apical discordance, or when the pulmonary venous atrium is small [21]. VSD closure is performed with sutures placed on the morphologic right ventricular side of the septum as described by de Leval et al. [22]. In the present series, this resulted in a low incidence of postoperative complete heart block (three patients) with
eventual recovery of sinus rhythm in two of these patients. Finally, use of peritoneal drainage catheters in all patients and low dose peritoneal dialysis whenever urinary output falls or hyperkalemia supervenes in the early postoperative period minimizes postoperative edema and the consequences of capillary leakage.

4.4. Preservation of right ventricular and tricuspid valve function and long-term outcomes

The use of the double switch operation is predicated on the assumption that long-term outcomes will be better with the morphologic left ventricle supporting the systemic circulation. Long-term follow-up of patients with ccTGA demonstrates a high incidence of failure of the systemic morphologic right ventricle and progressive tricuspid regurgitation. Piran et al. [10] demonstrated symptomatic congestive heart failure in 32% of patients with a mortality of 25% in patients with ccTGA when followed into the third and fourth decades. In a multi-institutional study, Graham et al. [11] found that 25% of patients with ccTGA had symptoms of congestive heart failure by 45 years of age and if significant associated lesions were present, two-thirds developed symptomatic heart failure. Prieto et al. [9] found after 20 years of follow-up 30% of patients had significant right ventricular failure while more than 40% developed severe tricuspid regurgitation; 26% died or required cardiac transplantation at an average age of 25 years.

If sensitive measures of ventricular function are used, an even larger percentage of ccTGA patients can be shown to have subclinical evidence of limited cardiovascular reserve. Hornung demonstrated that a significant percentage of ccTGA patients had perfusion defects during exercise stress testing with radionuclide imaging. Sixty-two percent had ejection fractions below 55% while a quarter of these patients had ejection fractions below 40% [7]. Fredriksen et al. [6] found that the aerobic capacity of patients with ccTGA, as determined by VO₂max was only 30–50% of that demonstrated by controls. These patients also demonstrated a blunted chronotropic response to exercise.

Of particular importance to the present study are long-term results for patients who have undergone a classic intracardiac repair of VSD closure, left ventricle to pulmonary artery conduits and/or tricuspid valve surgery [8,12,25,26]. These patients represent the true alternative surgical group to the patients in the present study who have undergone a double switch operation. Sano et al. [8] demonstrated a mortality of 17% by 10 years after classic repair with a 71% incidence of significant tricuspid regurgitation and a 50% incidence of right ventricular dysfunction. Termignon et al. [12] found a 30% mortality after 10 years of follow-up in patients who had undergone a classic repair.

In the present study, with follow-up that now extends to more than 8 years, tricuspid regurgitation was significantly reduced after the double switch operation. Prior to surgery, one-half of the patients demonstrated moderate to severe tricuspid regurgitation; after surgery, more than one-half of the patients had no tricuspid regurgitation while only 13% of the patients had moderate to severe tricuspid regurgitation. Even if tricuspid regurgitation is severe preoperatively, concomitant tricuspid valve repair is only rarely needed at the time of the double switch operation with substantial improvement in function of the tricuspid valve usually occurring when exposed to the lower pressures in the pulmonary circulation. After surgery, right ventricular function was judged to be normal in all but two patients. In addition, nearly all of the patients in the present series demonstrated normal left ventricular and mitral valve function after intermediate-term follow-up. Comparing these results to earlier reports from this center, these good outcomes for tricuspid valve and right ventricular function appear to be stable [16]. With the right ventricle and tricuspid valve no longer exposed to systemic pressure in the pulmonary circuit, continued good function may be anticipated.

References

Appendix A. Conference discussion

Dr A. Dodge-Khatami (Amsterdam, The Netherlands): Your group from Cleveland has presented in the last years their extensive experience on these patients and set guidelines as to how to go about these patients. And I from Cleveland has presented in the last years their extensive experience on these patients and set guidelines as to how to go about these patients. And I

from Cleveland has presented in the last years their extensive experience on these patients and set guidelines as to how to go about these patients. And I

And I noticed again here, your patient group goes up to 16 years of age. In those guidelines, basically what came out is that patients older than 16, who are mostly minimally symptomatic or asymptomatic, have uncertain long-term outcomes. Should they still be double switched or are they double switchable? What would be the current recommendations and what would you do with an older asymptomatic patient who comes in with CCTGA, would you necessarily always recommend a double switch?

Dr Mesia: This is a very important point. I think over the last few years we have come to understand that older patients are not always suitable candidates for double switch, particularly when we are retraining the left ventricle. A few of these patients, depending on the specific associated lesions, already present with high pressure in the morphologic left ventricle and good ventricular function. Those are the patients who are probably good candidates for double switch.

From the group of older patients who undergo placement of pulmonary bands for retraining, a number of them go on to develop left ventricular dysfunction. Even after double switch, and with proven good left ventricular function before surgery.

There are several factors that influence when an older patient is a suitable candidate for a double switch. When to do it? It just depends on the associated lesions and how they respond to left ventricular retraining.

Dr B. Williams (Toronto, Canada): It seems to me that many patients with congenitally corrected transposition don’t present until they’re adults, and yet your upper age limit was 16. Is there an age limit that would preclude doing this approach?

Dr Mesia: We say, as a general rule, that the approach has to be individualized. Having said that, as a general rule, with post-adolescence patients, we are particularly careful in moving them forward to a double switch. A number of them undergo pulmonary band placement, not necessarily for retraining but for symptomatic treatment. We have at least 5 patients who have improved dramatically after pulmonary band placement. In these group of patients the question is, should we move forward to a double switch or not? I don’t think we have the answer for this question yet. However, it’s very important to be very careful with older patients.

Dr D. Metras (Marseille, France): I have a question concerning the cardiac rhythm of these patients in the mid- and long-term. We have a more modest experience, but it looks like we have more postoperative junctional rhythms in the mid-term than in the regular Mustard or Senning done for transposition, probably due to some particular arrangement of the bundles in the atrium. What are your results in terms of cardiac rhythm?

Dr Mesia: There was only one patient who had a significant junctional tachycardia after the procedure. However, there were 3 patients who developed complete AV block after double switch. During follow-up, 2 out of those 3 patients who needed pacemaker implantation recovered sinus rhythm function. However, we haven’t seen, so far, many problems with tachyarrhythmias. There was only one patient that I remember.

Dr Metras: You mean that except one, all your patients have a stable, long-term sinus rhythm?

Dr Mesia: At latest follow-up, yes.

Dr C. Schreiber (Munich, Germany): Could you please clarify why you excluded 24 patients? You were talking primarily about 70 patients which you had enrolled.

Dr Mesia: The patients who were excluded were actually from the study, are reviewed a little further in the paper. Seven patients underwent pulmonary band placement and are waiting for double switch, or are in the process of getting morphologic left ventricular retraining. Five patients developed significant left ventricular dysfunction and were referred for cardiac transplantation. There were 5–6 patients who underwent conventional surgical repair. That’s the group of older patients who came to us for consideration of double switch, but they were too old for left ventricular retraining. There were a couple of tricuspid valve replacements and a couple of VSD closures. I don’t remember the details. Four to five patients were patients that were thought to have two good sized ventricles, but eventually underwent single ventricle repair, particularly for having VSDs that were uncommitted or difficult to commit.

Dr F. Hart (Munich, Germany): There is still controversy regarding the length and the extent of the retraining period of the LV. Can you comment on that?

Dr Mesia: Again, it’s very difficult to comment on that question when there’s limited experience. By and large, our approach with a ventricle that
is deconditioned, has been to first place a pulmonary band to achieve no more than 50% to 60% systemic pressures. Eventually, we either let the patients outgrow the size of the band and, therefore, gradually increase the pressures in the left ventricle, or like in 3 out of the 6 patients, they came back for adjustment of the pulmonary band to try to achieve at least 80% systemic pressures. So, when they achieve 80% systemic pressures. That’s the time when we think the patient is ready, as long as he has a good left ventricular function and about 80% systemic pressure. We have a protocol tailored to each individual patient. We use echocardiography and catheterization data. We also look at the indexed left ventricular mass, by MRI. It seems that the indexed left ventricular mass should increase during the first few months after placement of pulmonary band. However, if it increases too much, then that’s probably a risk factor for failure.

**Dr G. Stellin (Padova, Italy):** In light of your beautiful results (obviously, zero mortality is fantastic), what’s your current approach now for the neonate with a corrected transposition and intact ventricle septum? Would you band the pulmonary artery in the neonatal period or in the early infancy in order to prevent left ventricle deconditioning?

**Dr Mesia:** I had a graph with an algorithm showing our way of approaching each patient on an individual basis in the presence or absence of a VSD. For a neonate with an intact ventricular septum and no tricuspid regurgitation, probably we would just observe. This approach is because patients with that particular type of anatomic set-up can survive for several years without any clinical problems. If the patient has significant tricuspid regurgitation, probably that will be an indication for double switch or to band the patient earlier, rather than later to retrain the left ventricle, even in the absence of right ventricular dysfunction. If the patient has a large VSD with congestive heart failure, that would be an indication to proceed with a double switch or banding. Again, this is a very individualized approach depending on the particular type of anatomy and physiology that we’re dealing with.

**Dr Stellin:** In other words, would you band the pulmonary artery in the presence of an “Ebstenoid type” tricuspid valve with significant regurgitation, in early infancy?

**Dr Mesia:** We would do it, yes. However, if there were no tricuspid regurgitation, we would probably watch the patient until either he develops some change in the right ventricular function or tricuspid regurgitation.

**Dr J. Moll (Lodz, Poland):** I would like to ask the Cleveland Clinic experience. Because I have much smaller experience in double switch and much worse experience. And in all of our patients we had elevated systemic venous pressure to keep output. And it was in every patient. And one of those patients we even catheterized because I thought that I made some mistakes in the Senning procedure, because I have less experience in the Senning. We didn’t find any gradient. And it was in every patient we found that this systemic venous pressure was elevated to keep correct cardiac output. Did you find something like that?

**Dr Mesia:** Yes. We had a number of patients with significant diastolic dysfunction. Even in the presence of borderline normal or good systolic function, we had elevated venous pressures and prolonged hospitalization with significant effusive complications from high venous pressures. All of these patients eventually recovered. There was no patient that I remember that had structural obstruction of the systemic or pulmonary venous baffles. I think these were particularly older patients, not infants or newborns.

**Dr Moll:** With older patients, usually older patients. And another question, is there any trick in the Senning procedure, because this Senning is a little bit different because anatomy of atrium is different from TGA, this in corrected transposition. It looks for me different. Is there some trick to do it better?

**Dr Mesia:** I know Dr Mee’s approach, has been to pay attention to concordance or discordance of the atrial situs and ventricular apex. He uses the Shumacker modification of the Senning for patients with atrial situs apical discordance. For these patients with discordant atrial situs and ventricular apex, we try to get them by until they are little older. We think that by the age of 18 months, probably, technically, it will be easier. I know Dr Mee has paid significant attention to this matter.

**Dr H. Lindberg (Oslo, Norway):** You had 6 patients that were retrained with a pulmonary artery band. What were the interval between the banding and the procedure and what criteria do you use for a successful retraining?

**Dr Mesia:** Out of those 6 patients who underwent pulmonary band placement, the median time interval between placement of the first band and a double switch was about 26 months. Again, we do this very carefully. There is catheterization involved, there is MRI, and there is echocardiography. Each patient gets the band placed in the OR under careful TEE monitoring. I know that there’s no patient that goes from a normal left ventricular low pressure to more than 50%–60% systemic at one time. We pay particular attention to the shifting of the ventricular septum to the midline seen in TEE in the OR. There has to be very good left ventricular function before the patient leaves the OR. The patient stays in the hospital for a few days for monitoring of the left ventricular function. If there is any question about deterioration of left ventricular function after banding, the patient returns to the OR for reloosening within the same hospitalization. We try to achieve no less than 50% systemic pressures the first time, but depending on how the patient responds in the OR we may want to go higher. The interval between band re-adjustment(s) is usually of no less than 6 months. After a successful banding and retraining we expect at least 80–85% systemic pressures, normal systolic function and an indexed left ventricular mass of no less than 60–80 gm/m² before doing a double switch.