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Repair of transposition of the great arteries, ventricular septal defect and left ventricular outflow tract obstruction

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Abstract

Objectives: This study was undertaken to compare the outcomes of the Lecompte procedure and Rastelli repair in the transposition of the great arteries (TGA) with ventricular septal defect (VSD) and left ventricular outflow tract obstruction (LVOTO) and to determine the risk factors associated with unfavorable events. **Methods**: Over a 12-year period (April 1990–October 2002), 35 patients underwent complete repair for TGA, VSD, and LVOTO. Twenty-five patients (71%) underwent the Lecompte modification, and mean age and weight were 23.4 ± 18.2 months and 10.2 ± 3.0 kg. Ten patients (29%) underwent the Rastelli operation, and mean age and weight were 39.1 ± 36.1 months and 13.8 ± 6.8 kg. **Results**: One early death (3%) occurred after the Lecompte procedure and no late death. The mean follow-up was 5.9 ± 3.8 years. Eight patients in the Rastelli group (80%) underwent a late reoperation for obstruction of the extracardiac conduit, and in four of these patients, a reoperation for LVOTO was concomitantly required. Reoperation was also required in six patients of the Lecompte group (25%); five for right ventricular outflow tract obstruction (RVOTO) including one for LVOTO and two for VSD leakage, and one for mitral regurgitation and left pulmonary artery stenosis. The interval prior to reoperation ranged from 1.6 to 11.1 years, with a mean of 5.7 ± 3.1 years. The actuarial figures for freedom from reoperation at 5 and 10 years were 40.0 ± 15.5 and 26.7 ± 15.0% after the Rastelli operation and 95.7 ± 4.3 and 63.5 ± 12.6% after the Lecompte procedure ($P = 0.02$). Multivariate analysis by Cox regression analysis revealed that the risk factors of RVOTO were a younger age at operation, the Rastelli operation, and ductus ligation during the operation. **Conclusions**: The Lecompte procedure and Rastelli repair provide satisfactory early and late results. However, substantial late morbidity is more associated with conduit obstruction, and LVOTO in Rastelli repair rather than Lecompte procedure.

Keywords: Transposition of the great arteries; Rastelli; Lecompte

1. Introduction

Over the past three decades the operation described by Gian Carlo Rastelli and his colleagues [1,2] in 1969 has been considered the procedure of choice for the surgical repair of transposition of the great arteries (TGA) associated with ventricular septal defect (VSD) and left ventricular outflow tract obstruction (LVOTO). In the original description, the procedure involved baffling of the VSD to the aorta and connection of the right ventricle to the pulmonary arteries with a homograft conduit. This operation was the first described with the theoretic advantage of incorporating the left ventricle as the systemic ventricle for correction of TGA. It was widely applied to treat this subset of patients, and continues to be the most common surgical option.

However, the Rastelli operation is not ideal for at least four reasons: (1) it is not feasible in some patients because of unfavorable intracardiac anatomy; (2) the use of an extracardiac prosthetic conduit for reconstruction of the pulmonary outflow tract needs a subsequent reoperation to change this conduit; (3) it delays complete correction until

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a larger conduit can be replaced; and (4) homografts are not always available because of limited supply, especially in Asian countries.

In an effort to overcome these limitations, Lecompte and colleagues [3] introduced a new surgical technique that includes extensive resection of the conal septum and direct reimplantation of the pulmonary trunk on the superior margin of the right ventricular infundibulotomy without a prosthetic conduit. Using this procedure the size of the VSD and abnormal attachment of the tricuspid chordae on the conal septum are not limiting factors. However, some anatomic contraindications remain, for example, remote VSD or multiple VSDs, which make the construction of a left ventricle to aorta tunnel impossible; hypoplasia of one of the ventricles; and diffuse hypoplasia of the pulmonary arteries [4].

Since 1990, both procedures have been practiced in our unit. We reviewed our entire 12-year experience involving the surgical treatment of TGA/VSD/LVOTO. Our purpose was to describe the outcomes of the Lecompte procedure and Rastelli repair in the TGA with VSD and LVOTO and to determine the risk factors associated with unfavorable events.

2. Materials and methods

2.1. Patients' profile

During the 12-year period (April 1990–October 2002), 35 patients underwent a Rastelli repair or Lecompte procedure for TGA with VSD and LVOTO. The male to female ratio was 21:14 and age at operation ranged from 4 months to 11 years (mean 27.9 ± 25.1 months). Thirty-three patients (94%) were less than 5 years of age, and weight at operation ranged from 5.7 to 32.5 kg (mean 11.2 ± 4.7 kg). Seventeen patients (49%) weighed less than 10 kg. Weights in the Lecompte group were lower than the Rastelli group (Table 1).

Table 1
Patients' profile

	Lecompte group	Rastelli group	*P*-value
Sex (male:female)	13:12	8:2	0.25
Age (months)	23.4 ± 18.2	39.1 ± 36.1	0.10
Body weight (kg)	10.2 ± 3.0	13.8 ± 6.8	0.04
PA (%)	3 (12)	3 (30)	0.32
Distal PS (%)	4 (16)	0 (0)	0.30
VSD			0.69
PMTE (%)	9 (38)	3 (38)	
PMOE (%)	7 (29)	3 (38)	
PMIE (%)	7 (29)	1 (13)	
Muscular (%)	1 (4)	1 (13)	

PA, pulmonary atresia; PS, pulmonary stenosis; VSD, ventricular septal defect; PMTE, perimembranous trabecular extension; PMOE, perimembranous outlet extension; PMIE, perimembranous inlet extension.

Table 2
Palliative procedure

	Lecompte group	Rastelli group	*P*-value
BAS (%)	2 (8)	2 (20)	0.56
BT shunt (%)	15 (60)	5 (50)	0.71
Interval (years)	1.8 ± 1.1	1.9 ± 0.9	0.82

BAS, balloon atrial septostomy; BT, Blalock-Taussig.

Preoperative diagnosis was dextro- or levo-TGA in 20 (57%), and an anteroposterior relation in 15 (43%). VSD was perimembranous trabecular extension in 12 (34%), perimembranous outlet extension in 10 (29%), perimembranous inlet extension in 8 (23%), muscular in 2 (6%), undetermined in 3 (9%), and multiple in 2 (6%). Associated anomalies were pulmonary atresia in 6 (17%), distal pulmonary artery stenosis in 4 (11%), major aorto-pulmonary collateral arteries in 2 (6%), tricuspid valve septal chordal insertion onto the ventricular septal crest in 4 (11%), and Rastelli type C of atrioventricular septal defect (AVSD) in 1 (3%) (Table 1). Twenty patients (57%) had a previous Blalock-Taussig shunt. Four (11%) had balloon atrial septostomy during cardiac catheterization. Other palliations were unifocalization of the major aorto-pulmonary collateral arteries in 2 (6%), and palliative right ventricle to the pulmonary artery conduit in 1 (3%). The mean time interval from the first palliation to definitive repair was 1.8 ± 1.0 year (2 days–4.1 years) (Table 2).

2.2. Surgical procedures

Twenty-five patients underwent complete repair with the Lecompte modification of the REV (*réparation*à l'*étage ventriculaire*) procedure, whereas ten patients underwent the Rastelli operation. The choice between these two options was dictated by anatomic conditions. We preferred the Rastelli procedure in pulmonary artery distorsion, multiple pulmonary artery stenosis, hypoplastic pulmonary artery, or elevated pulmonary vascular resistance, and the Lecompte procedure in favorable anatomy, or in early age. In the Lecompte procedure, the ascending aorta in TGA, VSD and LVOTO is large and unlike the repair of simple TGA, is not reimplanted posteriorly. The presence of branch pulmonary artery stenosis in combination with free pulmonary regurgitation may lead to severe postoperative right ventricular failure. Therefore, the use of the Lecompte procedure was preferred for patients with nearly normally sized ascending aorta in dextro-TGA or for patients with side-by-side great vessels. Additional procedures were VSD extension in 22 (63%), ductus ligation in 22 (63%), atrial septal defect (ASD) closure in 25 (71%), distal pulmonary artery angioplasty in 4 (11%), tricuspid annuloplasty in 1 (3%), and mitral annuloplasty and mitral valvuloplasty in 1 (3%).

2.2.1. Lecompte procedure (n = 25, 23.4 ± 18.2 months of age)

All patients were operated on by median sternotomy. Cardiopulmonary bypass was conducted at moderate systemic hypothermia (28 °C). The mean cardiopulmonary bypass time was 174 ± 33 min and aortic crossclamp time was 88 ± 20 min. In two patients, total circulatory arrest and deep hypothermia was used. Before bypass, the branches of pulmonary artery were mobilized sufficiently beyond the pericardial reflection and previous shunts were controlled. The Lecompte procedure was performed in the same manner as described previously [5]. Resection of the conal septum or VSD extension was undertaken in all patients. This resection enlarges even a small restrictive VSD and constructs a straighter left ventricular outflow tract, which helps to prevent obstruction. After intraventricular baffling, the pulmonary artery was pulled down to the superior margin of the right ventriculotomy, without translocating the pulmonary artery anterior to the aorta, so called the Lecompte maneuver [6]. Right-sided translocation of the main pulmonary trunk was performed in seven patients, which depended on the initial position of the great arteries. In cases involving the abnormal insertion of the tricuspid tensor apparatus (papillary muscle in 2, septal leaflet chordae in 1), we transferred them onto the right side of the patch after division, instead of using the flap technique [7]. Finally, the right ventricular outflow tract (RVOT) was reconstructed with an anterior patch, after a monocusp valve was inserted along the margin of the ventriculotomy. A monocusp valve was constructed in 23 patients (92%), using either glutaraldehyde treated autologous pericardium in 18 patients, or Gore-Tex membrane (W. L. Gore and Associates, Flagstaff, AZ) in five patients. Glutaraldehyde treated autologous pericardium was also used for the RVOT hood in these 18 patients, but not in the five patients in whom a Gore-Tex patch was used. Additional operative procedures were VSD extension in 15 (60%), ductus ligation in 17 (68%), ASD closure in 19 (76%), distal pulmonary artery angioplasty in 2 (8%), tricuspid annuloplasty in 1 (4%), and mitral annuloplasty and mitral valvuloplasty in 1 (4%) (Table 3).

Table 3
Operative procedure

	Lecompte group	Rastelli group	*P*-value
Additional procedure			
VSD extension (%)	15 (60)	7 (70)	0.71
Ductus ligation (%)	17 (68)	5 (50)	0.44
ASD closure (%)	19 (76)	6 (60)	0.34
Others (%)	4 (16)	2 (20)	1.00
CPB times (min)	174 ± 33	154 ± 25	0.09
ACC times (min)	88 ± 20	87 ± 16	0.85

VSD, ventricular septal defect; ASD, atrial septal defect, CPB, cardiopulmonary bypass; ACC, aortic crossclamp.

2.2.2. Rastelli procedure (n = 10, 39.1 ± 36.1 months of age)

All procedures were performed using conventional extracorporeal techniques. The mean cardiopulmonary bypass time was 154 ± 25 min and aortic crossclamp time was 87 ± 16 min. In cases involving an abnormal insertion of the tricuspid tensor apparatus (septal leaflet chordae in 1), it was transferred onto the right side of the patch after division. Polystan valved conduits (Polystan, Vaerløse, Denmark) were used in the reconstruction of pulmonary ventricular outflow tracts. Graft sizes were 20 mm in 2 (20%), 18 mm in 7 (70%), and 16 mm in 1 (10%). Proximal anastomosis was supplemented in some patients with a hood of bovine pericardium or Gore-Tex vascular patch to make the conduit curvature smooth. There were no technical problems encountered in placing conduits. Additional operative procedures were VSD extension in 7 (70%), ductus ligation in 5 (50%), ASD closure in 6 (60%), and distal pulmonary artery angioplasty in 2 (20%). No differences were observed between two groups in terms of additional procedures and cardiopulmonary bypass data (Table 3).

2.3. Statistical analysis

Statistical analyses were performed using SPSS version 10.0 software (SPSS, Inc., Chicago, IL, USA). All results were expressed as mean ± SD, and the value of P less than 0.05 was considered statistically significant. The significance of differences between two groups was assessed by using the unpaired Student's t-test, the χ^2 test or Fisher's exact test. Actuarial data were analyzed using Kaplan–Meier formulae. Differences between groups were evaluated by a log-rank test. Early mortality was defined as death occurring within 30 days of the operation. The follow-up status of patients was determined by retrospective review of hospital records or by telephone interviews.

3. Results

3.1. Early results

3.1.1. Early mortality

There was one early death (3%). This female patient was diagnosed as having TGA, Rastelli type C of AVSD, and LVOTO. She underwent the Lecompte procedure, and developed a complete atrioventricular block after weaning from the cardiopulmonary bypass. The cause of death was low cardiac output syndrome due to residual LVOTO and mitral regurgitation after repair of an unbalanced AVSD.

3.1.2. Early morbidity

Four patients (11%) underwent reoperation due to bleeding. Chylothorax occurred in three patients (9%), junctional ectopic tachycardia in one (3%), complete atrioventricular block in one (3%), pneumonia in one (3%), and sepsis in one (3%).

Table 4
Causes of late reoperation

Cause	Lecompte group ($n = 24$) (%)	Rastelli group ($n = 10$) (%)	P-value
RVOTO	5 (21)	8 (80)	0.05
LVOTO	1 (4)	4 (40)	0.02
Others	3 (13)	0 (0)	0.02
Total	6 (25)	8 (80)	0.54

RVOTO, right ventricular outflow tract obstruction; LVOTO, left ventricular outflow tract obstruction.

3.2. Late results

None of the 34 early survivors was lost from follow-up, which ranged from 2 months to 12 years, with a mean of 5.9 ± 3.8 years.

3.2.1. Late mortality

There was no late mortality.

3.2.2. Late reoperation

Eight in the Rastelli group (80%) underwent reoperation for obstruction of the extracardiac conduit: in four of them reoperation for LVOTO was concomitantly required. Reoperation was required in six patients of the Lecompte group (25%): five for right ventricular outflow tract obstruction (RVOTO) with one for LVOTO and two for VSD leakage, and one for mitral regurgitation and left pulmonary artery stenosis. Reoperations for RVOTO in the Rastelli group were more frequently required than Lecompte group ($P = 0.05$) and reoperations for LVOTO in the Rastelli group were more frequently required than Lecompte group ($P = 0.02$) (Table 4). The interoperative interval ranged from 1.6 to 11.1 years, with a mean of 5.7 ± 3.1 years. Types of late reoperation were RVOT reconstruction with a Polystan valved conduit in 6, homograft interposition in 3, stenotic valve excision in 3, and pulmonary valve replacement in 1 and subaortic muscle resection for LVOTO in 5, mitral valvuloplasty and mitral annuloplasty in 1, left pulmonary artery angioplasty in 1, and permanent pacemaker insertion in 1.

The actuarial values for freedom from reoperation at 5 and 10 years were 40.0 ± 15.5 and 26.7 ± 15.0% after the Rastelli operation, and 95.7 ± 4.3 and 63.5 ± 12.6% after the Lecompte procedure ($P = 0.02$). (Fig. 1). Multivariate analysis by Cox regression analysis revealed that the risk factors for RVOTO were a younger age at operation, the Rastelli operation, and ductus ligation during the operation.

4. Discussion

4.1. Early morbidity and mortality

The surgical treatment for TGA, VSD and LVOTO has evolved dramatically over the past 40 years, and currently the procedures to treat this anomaly have an operative mortality of less than 5%. In the present study, one hospital death occurred among the 35 patients (3%). The cause of death was low cardiac output syndrome due to residual LVOTO and mitral regurgitation after repair of an unbalanced AVSD. We believe that the morphologic risk factors for early mortality are AVSD and double-outlet right ventricle with subpulmonic VSD or remote VSD because of a long baffle, small left ventricle and restrictive VSD, and

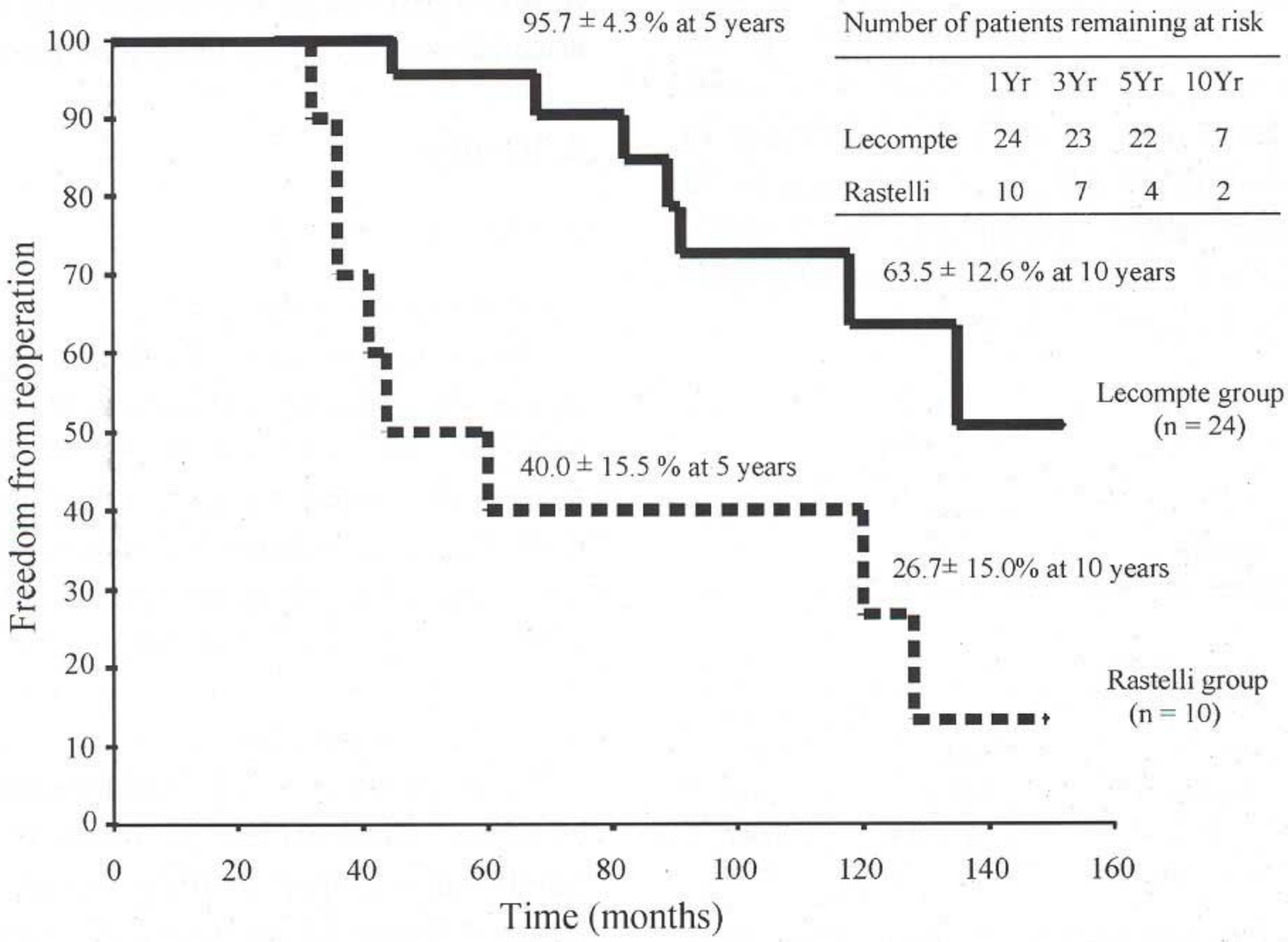

Fig. 1. Freedom from reoperation.

the presence of tricuspid valve straddling or abnormal attachments, which prevents a proper intraventricular tunneling. In our series, the morphologic risk factor for early mortality was a Rastelli type C of AVSD (1 mortality in 1 patient). However, the presence of a tricuspid valve straddling or abnormal attachments (no mortality in 4 patients, 11%) was not morphologic risk factor for early death. Under such circumstances, it is necessary to divide the chordae and reattach them to the VSD baffle. Niinami and colleagues [8] have described a technique to avoid the complications of tricuspid malinsertion in this subset. We did not use the flap technique [7], but directly transferred these tensor apparatuses to the patch after division. Echocardiography revealed mild regurgitation in all patients immediately postoperatively and at long term follow-up. Therefore, we believe that this is a reliable alternative for the transfer of the abnormal tension apparatus.

The use of a prosthetic conduit and a right-sided conduit may be related to the potential problem of coronary compression by the prosthetic valve ring [9]. The relation of the conduit with the sternum is crucial in a Rastelli repair. Right-sided conduits may be more affected by sternal compression since the position of the right ventricular infundibulum is more anterior [10]. We used a flexible conduit, and three patients had an L-transposed aorta in the Rastelli group, necessitating a right-sided conduit. We did not apply the Lecompte maneuver in the Lecompte group, and could bring directly the main pulmonary artery to the right ventricle. We decided not to use the Lecompte maneuver in the Lecompte procedure because orthotopically posterior-positioned pulmonary arteries reduce the possibility of the pulmonary outflow tract obstruction as seen in arterial switch operations [11], and we wanted to simplify the procedure and reduce the risk of bleeding by avoiding the transection of aorta and reanastomosis. Tension-free anastomosis was accomplished by sufficient dissection of the pulmonary arterial branches beyond the pericardial reflection. Consequently, it is important to decide whether the main pulmonary artery will be translocated on the right or the left of the aorta. Therefore, we believe the essential point of this procedure is the translocation of the pulmonary artery. In our series, seven cases involved translocation of the main pulmonary artery to the right of the aorta, based on the initial position of the great arteries. We did not experience any sternal compression or mortality due to right-sided conduit or translocation of the pulmonary artery.

4.2. Right ventricular outflow tract obstruction

In our study, younger age at operation, Rastelli operation, and ductus ligation during operation were the risk factors of RVOTO. Eight in the Rastelli group (80%) underwent reoperation for obstruction of the extracardiac conduit. Somatic overgrowth sometimes makes conduit replacement inevitable. The decision to reoperate is influenced by symptoms, transconduit gradient, and dysfunction of the pulmonary ventricle [12,13]. The earliest indication of conduit failure is development of tricuspid insufficiency [12]. A number of studies showed that symptoms might be absent despite moderate obstruction [14]. Conduit replacement should be performed if significant pulmonary ventricular dysfunction develops. The late results of heterograft valve conduits [15] are similar to those of homografts [16]. Several prosthetic valved conduits may be used to connect the right ventricle to the pulmonary artery. We used a Polystan valved conduit because we believe a flexible conduit is probably better in small children. However, the reoperation free rate was found to be extremely low, which may be a major limitation to use this conduit. The long-term durability of extracardiac valved conduits is not yet ideal. Creation of viable pulmonary artery autografts has been tried through tissue engineering by endothelialization with autologous cells [17].

One of the most important considerations, when evaluating the effectiveness of a Lecompte procedure, is the fate of the RVOT. We experienced RVOTO in 25% of patients after the Lecompte procedure. Vouhé and associates [4] reported that RVOTO occurs in 26% of patients, with a mean follow-up of 55 months. Therefore, there has been much concern about the potential for growth of the native pulmonary trunk and changes of the monocusp valve with time. Lecompte and coworkers [3] emphasized in their first report of this procedure that pulmonary regurgitation should be prevented by a reliable method of inserting a valve in the pulmonary outflow tract. In all cases, we placed a monocusp valve expecting this valve to improve the immediate postoperative result by preventing a sudden hemodynamic change from a pressure-loaded right ventricle to a volume-loaded ventricle. Pulmonary regurgitation was insignificant in all patients. However, among five patients who had reoperations for residual pulmonary stenosis, four had severe calcification of the monocusp valve. In five recent cases, we used Gore-Tex membrane, which is thought to have a water-repellent nature and makes calcification less likely [18,19]. Analyzing our results, it appears that the long-term fate of the reconstructed RVOT depends primarily on the monocusp valve, which is prone to progressive calcification, degeneration, and subsequent valvular dysfunction over time, although this type of repair has an advantage in terms of the potential for the growth of the patient's own native pulmonary artery. Recent reports include investigations into the need for valves [20], the preservation of the native pulmonary valves [21], and the use of alternative materials for monocusp valves [18,19].

4.3. Left ventricular outflow tract obstruction

In the Lecompte group, resection of the conal septum or VSD extension was undertaken in all patients. This resection enlarges even a small restrictive VSD and forms a straighter left ventricular outflow tract, which helps

prevent obstruction. The technique of VSD baffling for the Rastelli procedure has evolved during the course of our experience, and enlargement of the VSD is undertaken in 70% of patients. In our study, we underwent LVOTO postoperatively in 5 patients (15%): four in the Rastelli group and one in the Lecompte group. Rychik and associates [22] have postulated that in TGA, as in double-inlet left ventricle, the VSD or bulboventricular foramen becomes restrictive after the biventricular repair or the Fontan operation because the left ventricular volume is often increased before the correction. However, resection of the anterosuperior margin of the defect carries a potential increased risk for atrioventricular block and scar tissue as a substrate for arrhythmia. The presence of hypertension in the right ventricle would be another factor for late LVOTO, since right ventricular hypertension will produce septal hypertrophy and leftward septal displacement. The tunnel shape of the left ventricular outflow tract after a repair may be affected by right ventricular hypertension.

4.4. Late failure, mortality, and arrhythmia

The cause of the late death is left ventricular failure and sudden death [23]. Abnormal left ventricular wall stress was demonstrated in patients after a repair [24]. The old patients at operation were affected by hypoxia, obstruction, and volume overloading over a prolonged period. Other factors are a large prosthetic component as a VSD baffle and right ventricular hypertension [25]. The cause of late sudden death is late arrhythmia by right bundle branch block that may progress to bifascicular and complete atrioventricular block, scar tissue, and right ventricular hypertension due to RVOTO.

In conclusion, the Lecompte procedure and Rastelli repair provide satisfactory early and late results at this length of follow-up. Substantial late morbidity is more associated with conduit obstruction, and LVOTO in Rastelli repair rather than Lecompte procedure. The Lecompte procedure allows early, complete anatomic repair and reduces the need for late reoperation because of its obviation of an extracardiac conduit and the potential growth of the pulmonary artery. However, late obstruction related to the monocusp valve is a problem that remains to be solved.

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