Long-term results of the REV (réparation à l’étage ventriculaire) operation

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Objective: Despite its innovative features, the réparation à l’étage ventriculaire (REV) procedure has not gained large popularity in the treatment of transposition of the great arteries, ventricular septal defect, pulmonary stenosis, and related anomalies, and thus the Rastelli operation remains the preferred type of repair. We try to obviate the alleged lack of long-term results that has been suggested to explain this reluctance to change.

Methods: We reviewed a series of 205 patients who underwent the REV procedure between 1980 and 2003.

Results: Hospital mortality was 12% (24 patients). Ten of 181 early survivors residing in distant countries could not be traced for follow-up. There were 13 late deaths (2 of noncardiac causes). Overall survival and freedom from any reoperation at 25 years were 85% and 45%, respectively, as determined by using the Kaplan–Meier method. Reoperation was commonly required because of recurrent right ventricular outflow tract obstruction (36 patients) and rarely by left ventricular outflow tract stenosis (3 patients). By using cumulative method analysis, at the 25-year follow-up interval, the probability of being alive without reoperation was 45%, that of reoperation for right ventricular outflow tract obstruction was 33%, and that of reoperation for left ventricular outflow tract obstruction was 5%. Clinical status is excellent: 137 patients are in New York Heart Association class I (87%), and 131 patients are free of arrhythmias.

Conclusions: These results represent a considerable improvement over those of the Rastelli operation in terms of survival and need for reoperation for right or left ventricular outflow tract obstruction. (J Thorac Cardiovasc Surg 2011;142:336-43)

Anomalies of the ventriculoarterial connection with subpulmonary stenosis are often indicated with acronyms, such as transposition of the great arteries (TGA), double-outlet right ventricle (RV), double-outlet left ventricle (LV), and Taussig–Bing anomaly, and are treated classically by using the Rastelli operation.1 In our clinical practice, for the sake of simplification, we have chosen to group all these anomalies under the term malposition of the great arteries, taking as a unifying element the essential need for surgical repair to establish a connection between the LV and the aorta and an unobstructed pathway from the RV to the pulmonary artery. The operation described by Lecompte and colleagues2 in 1980, termed réparation à l’étage ventriculaire (REV), by including the reconnection of the aortic valve with the LV through a ventricular septal defect (VSD) endorses the principle of the Rastelli operation. Here, though, similarity between the 2 procedures ends: resection of the conal septum (to avoid the spiral shape of the LV-to-aorta tunnel) and direct reimplantation of the pulmonary trunk on the RV (to prevent the need for a complete tubular prosthesis) are unique features of the REV procedure. By using this technique, anatomic repair could be an option for a greater number of patients.

The early results of the REV procedure have been reported in detail.3-8 Because it has been claimed that its long-term results are unclear, a study was undertaken to fill this vacuum of scientific knowledge.

MATERIALS AND METHODS

Two hundred five patients with malposition of the great arteries, VSD, and left ventricular outflow tract obstruction (LVOTO) underwent operations performed by 2 of the authors (Y.L. and P.V.) between 1980 and 2003; the median age at the time of the REV procedure was 1.7 years (range, 0.3–16 years), and 93 (45%) patients were younger than 2 years. Twenty-four patients died during the operation and are excluded from further analysis. One hundred eighty-one (88%) patients survived and are the subject of this study; their median age at the time of the operation was 1.8 years (range, 0.35–16 years).

Diagnosis

Along with our terminology,9 all patients had complex forms of malposition of the great arteries. In detail, typical TGA was present in 142 patients, a double-outlet RV was present in 27 patients, and a double-outlet LV was present in 6 patients. In 6 patients the intracardiac anatomy could not be further classified because of lack of subaortic or subpulmonary coni, unusual position and size of the VSD, and bizarre spatial relationship between the great arteries.

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Associated anomalies were found in 78% of the patients (Table 1): 42 patients had an anomaly of the tricuspid valve, 16 patients had an anomaly of the mitral valve, and 6 patients had an anomaly of both atrioventricular valves. Conal septal anatomy was unusual in 16 patients (bilateral coni, 10 patients; no coni, 6 patients), and 26 patients did not have a well-developed muscular structure interposed between the semilunar valves.

### Previous Operation

One hundred thirteen procedures were performed in 97 patients before the REV procedure (54%). Most operations were concerned with improving total or effective pulmonary flow. Three patients without native LVOTO were found to have abnormalities of the pulmonary valve that precluded an arterial switch operation. Pulmonary artery banding was performed as a stage toward the REV procedure.

### Technique

The technical aspects of the procedure have been extensively described and remained relatively uniform throughout the entire experience. Resection of the conal septum, conal septal flap, and the French (Lecompte) maneuver were performed as indicated by the anatomy of the conal septum and tricuspid valve and the spatial relation of the great arteries. In 6 patients with absent subarterial coni no resection was required. A pericardial patch or a composite pericardial–Dacron patch was used to construct the intracardiac tunnel. The technique of right ventricular outflow tract (RVOT) repair was less consistent (Table 2); several materials were adopted for the patch and for the monocusp valve. In a limited number of patients, the segment of native aorta, excised concomitantly with the French (Lecompte) maneuver, was included in the pulmonary trunk–right ventricular anastomosis with the aim of reducing tension.

### Follow-up

Ten of these patients residing in distant countries could not be traced after hospital discharge. Therefore information on survival and clinical status was available for 171 early survivors (follow-up 94.5% complete).

The mean follow-up interval was 12.3 ± 7.1 years. Patients were seen in the outpatient clinics of 2 institutions (Hôpital Necker, Paris, France, and Institut Jacques Cartier, Massy, France). Alternatively, local cardiologists or pediatricians were contacted. A few patients were traced and contacted directly by telephone or mail.

### Reoperation

All surgical procedures involving the ventricular outlets, septa, and atrioventricular conduction abnormalities were considered reoperations. Interventional procedures, such as balloon dilatation of the RVOT, dilatation or stenting of the pulmonary arteries, or VSD device occlusion were also counted as reoperations.

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### TABLE 1. Associated malformations in early survivors

<table>
<thead>
<tr>
<th>Malformation</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td>49 (27%)</td>
</tr>
<tr>
<td>Tricuspid valve anomaly</td>
<td>42</td>
</tr>
<tr>
<td>Mitral valve anomaly</td>
<td>13</td>
</tr>
<tr>
<td>Mitral and tricuspid valve anomaly</td>
<td>6</td>
</tr>
<tr>
<td>Multiple VSDs</td>
<td>16</td>
</tr>
<tr>
<td>Restrictive VSD</td>
<td>6</td>
</tr>
<tr>
<td>Pulmonary atresia</td>
<td>6</td>
</tr>
<tr>
<td>Pulmonary arterial branch stenosis</td>
<td>21</td>
</tr>
<tr>
<td>Coronary anomaly</td>
<td>35</td>
</tr>
<tr>
<td>Left atrial appendage juxtaposition</td>
<td>19</td>
</tr>
<tr>
<td>Situs inversus</td>
<td>4</td>
</tr>
<tr>
<td>Dextrocardia</td>
<td>1</td>
</tr>
<tr>
<td>IVC interruption, azygos continuation</td>
<td>1</td>
</tr>
<tr>
<td>Hypoplastic RV</td>
<td>1</td>
</tr>
</tbody>
</table>

VSD, Ventricular septal defect; IVC, inferior vena cava; RV, right ventricle.

### Statistical Analysis

Survival and event-free survival estimates were obtained by using the Kaplan–Meier and cumulative incidence methods. The Cox proportional hazard method was used to define the association of a variable or a combination of variables with survival or adverse events.

### Informed Consent

This study was approved by the institutional review board, and informed consent was obtained from each participant.

### RESULTS

Overall survival at a 25-year follow-up interval was 85% by using the Kaplan–Meier method. At the same time interval, survival free of any reintervention was 45% (Figures 1 and 2).

The cumulative incidence method permits the evaluation of a single event at a given time provided that other events have not occurred. Figure 3 shows an analysis of 5 events at the 25-year follow-up interval: death, reoperation-free survival, and reoperation for right ventricular outflow tract obstruction (RVOTO), LVOTO, residual VSD, or other causes. The probability of reoperation-free survival, although decreasing from time zero (ie, the REV procedure) is the highest (45%), followed by reoperation for RVOTO, LVOTO, residual VSD, or other causes (33%, 5%, 6%, and 6.5%, respectively).

### TABLE 2. Management of RVOTO

<table>
<thead>
<tr>
<th>Patch</th>
<th>No. of patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dacron/pericardium</td>
<td>100</td>
</tr>
<tr>
<td>Other</td>
<td>56</td>
</tr>
<tr>
<td>Autologous aortic patch</td>
<td>21</td>
</tr>
<tr>
<td>Monocusp valve</td>
<td></td>
</tr>
<tr>
<td>Heterologous pericardium</td>
<td>101</td>
</tr>
<tr>
<td>Autologous pericardium</td>
<td>49</td>
</tr>
<tr>
<td>Other</td>
<td>7</td>
</tr>
<tr>
<td>Valved conduit</td>
<td>2</td>
</tr>
</tbody>
</table>

RVOTO, Right ventricular outflow tract.
Thirteen (7.6%) patients died late after the REV procedure. Causes of death are indicated in Table 3. Six deaths (4 sudden, 1 at reoperation for caval thrombosis, and 1 after a prolonged episode of supraventricular tachycardia) were caused by or triggered by an arrhythmia.

Reoperation

LV–aorta tunnel. Three patients required reoperation for relief of LVOTO; 2 of them, as mentioned above, died. One patient had an aneurysmal patch causing right ventricular obstruction; the aneurysm was resected, and the patch was replaced. In another patient the tunnel patch calcified and fractured twice, causing a recurrent VSD. The patch was completely replaced each time.

Recurrent/residual VSD. Ten patients required 12 procedures to close a recurrent or residual VSD. In one case a residual VSD was closed by means of transcatheter device implantation. Three patients received a concomitant tricuspid annuloplasty, and 3 received a pulmonary branch arterioplasty.

RVOTO/pulmonary regurgitation. This was the most frequent cause of reintervention. Table 4 indicates the procedures performed in 36 patients. The most frequently required type of reoperation was replacement of a stenotic RVOT patch with another monocusp valve–bearing patch. A valved conduit was deemed necessary in 6 patients only (3 of whom underwent reoperations elsewhere). On the whole, in 5 patients (14% of those undergoing reoperations) the aim of surgical intervention was to control pulmonary regurgitation and improve the function of a dilated RV.

Right ventricular dilatation was severe in 1 child, who did not survive reoperation at another institution.

Several variables were analyzed for their possible effect on the need for reoperation on the RVOT. Three were found to increase independently the risk for reoperation: lower age at the time of the REV procedure (as a continuous variable), use of a monocusp valve made of autologous pericardium, and use of an autologous aortic segment to extend the RVOT patch. This aortic segment was often found to be calcified at reintervention.

The hazard function of reoperation for RVOTO is shown in Figure 4. The curve has a constant hazard phase that

suggests that the probability of reoperation does not increase over time.

Clinical Status
Most late survivors are asymptomatic (New York Heart Association class I, 137 [87%] patients). Nineteen (12%) patients are mildly symptomatic, with arrhythmias, dyspnea on major effort, hypertension, or noncardiac complaints (New York Heart Association class II). Two patients are severely symptomatic: 1 has important neurologic sequelae after cardiac arrest at reoperation, and 1 awaits transplantation after myocardial infarction during the REV procedure.

Thirty-one patients regularly attend school at their age level, and 11 have permanent employment. Thirty-eight patients are actively involved in sports. Five women had 6 successful pregnancies and deliveries.

Reinvestigation
An effort test was carried out in 31 patients. Results were normal in 27 (87%) patients and submaximal for age in 4 patients, without evidence of arrhythmia.

Arrhythmias. One hundred thirty-one (72% of traced patients) patients were free of rhythm disturbances. Fourteen patients had a permanent pacemaker implanted at the time of the REV procedure for complete atrioventricular block; 5 subsequently recovered normal sinus rhythm, and 1 died of pacemaker dysfunction. Episodes of supraventricular tachycardia/flutter occurred in 7 patients, ventricular tachycardia occurred in 3 patients, and sporadic ventricular extrasystole occurred in 7 patients. One patient has first-degree atrioventricular block. Three further patients required a permanent pacemaker during follow-up.

One patient received an implantable cardioverter to treat ventricular tachycardia, and 2 had successful ablation of atrial arrhythmias. In 1 patient 3 attempts at ablation failed to control an incessant atrial tachycardia; he died of acute myocardial dysfunction.

Echocardiographic analysis. Bidimensional echocardiographic data were available for 157 patients (92% of traced patients); they are summarized in Figure 5. In 10 (6%) patients a systolic subaortic gradient was recorded, averaging 16.3 ± 10.8 mm Hg. Aortic and mitral regurgitation were uncommon and usually mild: 1 patient with a major preoperative mitral anomaly has moderate mitral regurgitation. Measurements of left ventricular function (ejection fraction and shortening fraction) were available for 54 patients; they were within normal limits in 49 (91%) patients, moderately decreased in 4 patients (ejection fraction, 41–45%), and severely decreased in 1 patient (awaiting heart transplantation).

Invasive re-evaluation. Repeat catheterization was performed in 41 patients. In 18 cases the study was prompted by suspicion of a residual defect and heralded reoperation; 2 patients have abnormal RVOT pressure and await reoperation. Catheterization data were within normal limits for 21 (51%) patients. However, 2 patients subsequently died, one at 116 months (arrhythmia and caval thrombosis) and the other suddenly at 2 months from the study. Three further patients underwent reoperations 59, 74, and 86 months after a normal study result.

Imaging by means of magnetic resonance or computed tomographic scanning was performed on 23 patients. Results were normal in 17 (74%) patients. It was part of re-evaluation toward repeat surgical intervention in 2 cases and demonstrated residual defects in 4 (3 cardiac defects and 1 thoracic defect).

DISCUSSION
Misconceptions have limited the acceptance of the REV procedure throughout the surgical constituency. It has been deemed to be technically challenging, and poorly known
long-term results. Nonetheless, from the early days, the REV procedure was applied to all sorts of anomalies of ventriculoarterial connection in addition to “classic” TGA. Patients with associated defects considered as contraindications or risk factors for the classic Rastelli operation (a total of 61 patients in this study) were not denied repair. Age constraint was not especially felt, unlike with the Rastelli operation. Our inference is that a comparison between the results described here and those of the Rastelli operation in the recent literature could be biased against the REV procedure; still, overall survival and survival free of reoperation are considerably worse than in the present series (Table 5).11-15

Dearani and coworkers14 focused on the problem of reoperation for recurrent RVOTO. Their late incidence of LVOTO was low because of liberal use of VSD enlargement. Ideal age at repair remained unspecified. There was a high prevalence of late cardiac/sudden deaths (38/160 [24%] early survivors), but the authors believed that the Rastelli operation remains the preferred repair for this subset of patients. Conversely, Kreutzer and colleagues11 consider their late results with the Rastelli operation disappointing in comparison with atrial, arterial, and even Fontan repairs. Recurrent RVOTO and LVOTO, as well as sudden death, constituted serious problems. They infer that the Rastelli operation should be reserved for patients with severe “mechanical” native LVOTO, whereas patients with “dynamic” (or resectable) LVOTO16 are perhaps better served by an arterial switch operation and VSD closure in the first year of life. We do not disagree with this line of thought but note that a large percentage of patients with TGA or malposition of the great arteries, VSD, and LVOTO have complex forms of obstruction not amenable to resection and marked pulmonary valve hypoplasia17-20 or even pulmonary atresia. A recent anatomic study20 brings support to the hypothesis of Kreutzer and colleagues11; unfortunately, early and late results in patients with pulmonary valve hypoplasia at the arterial switch operation seem worse than expected from an arterial switch operation with a “normal” native pulmonary valve. Emani and associates21 reviewed their results with 3 techniques in patients with TGA and LVOTO: arterial switch operation alone, arterial switch operation and LVOT repair, and non–arterial switch operation repair (Rastelli or Nikaidoh procedures). The isolated arterial switch
operation and Rastelli operation had the highest incidence of reintervention for recurrent/residual LVOTO.

Reoperation caused by LVOTO was rare in our series (3 patients) because a large LV–aorta connection was created as a result of the resection of the conal septum and the use of a shorter and straight patch. Conversely, anterior VSD enlargement is usually recommended for the Rastelli operation.11,12,14

Reoperation for RVOTO was necessary in a certain number of our patients, some of whom required multiple procedures. Even so, comparison of our data with those of the Rastelli operation clearly demonstrates a much lower prevalence of this complication (Table 5). Moreover, the hazard function of reoperation for RVOTO after the REV procedure remains constant over time. We were unable to find similar data regarding the Rastelli operation or conduit operations in the literature, but intuitively, the instantaneous risk of reoperation in this category of patients will increase with time because of a worsening patient–conduit mismatch and progressive conduit degeneration.

In this series lower age at the time of the operation as a continuous variable (with a cutoff point at 22 months) and 2 technical modifications stood out as independent risk factors for repeat operations because of RVOTO. Although faulty techniques must be abandoned, the ideal age at the time of the operation remains elusive. Ninety-three (45%) patients in this series were younger than 2 years at the time of the operation.

The problem is also unsolved with the Rastelli operation. It has been suggested that an early Rastelli operation would achieve better long-term results by avoiding the negative effects of long-standing cyanosis and ventricular overload. A trend toward higher early surgical mortality and the concern for multiple reoperations might discourage most units from adopting this policy, as has occurred in the past.

Unlike patients with tetralogy of Fallot,22 pulmonary insufficiency caused by the time-limited function of the monocusp valve has not emerged as a frequent cause for reoperation. In 30 of 33 patients who underwent reoperations performed by our group, the original patch was simply replaced by a monocusp valve bearing a polytetrafluoroethylene patch tailored by a ring-reinforced tubular prosthesis (PTFE stented tubular graft; W.L. Gore & Associates, Inc, Flagstaff, Ariz). On a mean follow-up time interval of more than 12 years, patients having undergone the REV procedure seem to tolerate pulmonary insufficiency remarkably well. We can only speculate on the reason for such a favorable course. The straight, short tunnel patch (producing a limited bulge in the right ventricular cavity) and the resection of the conal septum, rather than VSD anterosuperior enlargement, might favorably affect both left and right ventricular function. Left ventricular function is not as good after a Rastelli operation.22

Dissatisfaction with the Rastelli operation23 recently revived interest in the operation originally described by Bex and coworkers in 198024 and adopted by Nikaidoh in 1984.15 This complex procedure, termed “aortic translocation” borrows steps from the arterial switch operation. Rastelli operation, REV procedure, and Ross operation. It is logical to anticipate complications inherent to all these procedures, such as coronary distortion, aortic valve regurgitation, and recurrent RVOTO. Uncertainty still exists about whether to translocate the coronary artery or arteries, and at times this has been done at the completion of the procedure because of evidence of myocardial ischemia.

Moderate aortic insufficiency was reported by Morell25 in 3 of 11 survivors; one of them eventually required aortic valve replacement.26 Moderate aortic insufficiency was also observed in 1 of 11 patients by Bautista-Hernandez and colleagues,27 and mild aortic insufficiency was found in 8 of 18 survivors by Yeh and associates.28

In these 3 series the median follow-up intervals were 33 months, 59 months, and 11 years, respectively. On the whole, 15 reoperations for conduit replacement were required on 40 early survivors. Atrial and ventricular arrhythmias also occurred, requiring a maze procedure in one case and an implantable defibrillator in another. The authors were compelled to make this choice by the need to solve the problem of LVOTO after the Rastelli operation. In our study of a large series of patients with a long follow-up interval, we have shown that LVOTO rarely occurs.

In a recent article Hu and coworkers26 compared the Rastelli operation, the REV procedure, and the Bex–Nikaidoh

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**TABLE 5. Overall survival and prevalence of reoperation for recurrent RVOTO: Comparison with literature data**

<table>
<thead>
<tr>
<th>Source</th>
<th>Procedure</th>
<th>Twenty-year overall survival</th>
<th>Fifteen-year freedom from RVOT reoperation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Great Ormond Street Hospital, London (1981), 13</td>
<td>Rastelli</td>
<td>58%*</td>
<td>1</td>
</tr>
<tr>
<td>Children’s Hospital, Boston (2000), 11</td>
<td>Rastelli</td>
<td>52%</td>
<td>20%</td>
</tr>
<tr>
<td>Mayo Clinic, Rochester (2001), 14</td>
<td>Rastelli</td>
<td>59%</td>
<td>29%</td>
</tr>
<tr>
<td>German Heart Center (2007), 13</td>
<td>Rastelli</td>
<td>58%</td>
<td>32%</td>
</tr>
<tr>
<td>University of Texas, Dallas (2007), 15</td>
<td>Bex–Nikaidoh</td>
<td>95%</td>
<td>64%</td>
</tr>
<tr>
<td>This series, 171 patients (1980–2003)</td>
<td>REV procedure</td>
<td>83%</td>
<td>75%</td>
</tr>
</tbody>
</table>

RVOTO: Right ventricular outflow tract obstruction. *Eight-year survival. [Including early mortality (= 10%). | Including early mortality (= 7%). |

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procedure performed randomly on 30 patients. They modified both the REV procedure and the Bex–Nikaidoh procedure by adding total pulmonary valve translocation to the original techniques. All patients (n = 11) undergoing the modified Bex–Nikaidoh procedure survived, whereas 2 patients died after the REV procedure and 1 after the Rastelli operation.

The follow-up interval is less than 12 months in each surgical group; there were no reoperations or sudden deaths. One patient in the Bex–Nikaidoh group has mild aortic insufficiency, and 9 have a degree of pulmonary insufficiency, as do 5 after the REV procedure. It is apparent, even with such a short follow-up interval, that the modification of the technique has not yielded the desired result of avoiding pulmonary regurgitation. Moreover, 1 patient in the Bex–Nikaidoh group shows early evidence of aortic regurgitation. In our opinion neither the Bex–Nikaidoh procedure nor its ingenious modifications have yet produced clear evidence of results superior to those of the REV procedure.

Limitations of the Study

The nature of the study is retrospective. Although a large series of patients was analyzed, emphasis was put on the use of a unique procedure rather than on patient diagnosis. Patients in some anatomic subgroups could have a better probability of early and long-term survival than those with classic TGA who were treated with the Rastelli operation in the compared series. Clinical status was generally good, but complete noninvasive imaging and functional studies were performed in a limited number of late survivors.

CONCLUSIONS

Although of enormous historical value as the first successful step toward anatomic repair of TGA, the value of the classic Rastelli operation is presently reduced by the recognition that alternative options exist. We describe the excellent results obtained with the REV procedure, which obviates many contraindications of the Rastelli operation. Because its performance does not contemplate a prosthetic conduit, it can be safely applied to infants and small children, thus avoiding multiple palliative steps. Recurrent LVOTO is exceptional, and recurrent right ventricular obstruction is less common and shows a constant hazard curve. Ventricular function and clinical status are excellent in the majority of survivors. If a search for alternatives is at all justified, we strongly suggest that survival, incidence of complications, need for reoperation, and ventricular performance of “new” procedures be compared with those of the REV procedure and not with the Rastelli operation.

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References


