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SUMMARY

Background
The Rastelli procedure is complex surgical procedure with marked morbidity and mortality in the medium and long-term follow-up. These adverse outcomes seem to be more frequent when the ventricular septal defect (VSD) is anatomically remote or noncommitted to the aorta.

Objective
To evaluate the impact of the anatomical location of the VSD on the outcomes of the Rastelli procedure.

Material and Methods
A total of 47 patients were included with a mean follow-up of 6 years after surgery (15 months-14 years). Patients were divided into two groups: group I (committed VSD, n=29) and group II (remote or noncommitted VSD, n=18).

Results
During the immediate postoperative period, reoperations and arrhythmias were more frequent in group II (p=0.05 and p=0.06, respectively).
After a mean follow-up of 3 years (1 day-13 years) following surgery, 22 patients underwent 27 reoperations.
A residual VSD was closed in 11 patients at a mean of 1 month, 8 of which were remote VSD (p=0.007).
Six patients developed subaortic stenosis requiring surgery at a mean of 5 years.
The right ventricle-to-pulmonary artery conduit was replaced in 12 patients at a mean of 5 years.
Immediate postoperative mortality was 6% (3 patients).
Global mortality was 17.2% (n=8); 7 patients had noncommitted VSD (p=0.003).

Conclusions
Remote VSD in patients undergoing the Rastelli procedure is associated with: 1) increased mortality, 2) greater incidence of reoperations in the immediate postoperative period, and, 3) a trend towards greater incidence of arrhythmias in the immediate postoperative period.

Key words
Cardiac Surgery - Double Outlet Right Ventricle - Transposition of the Great Arteries - Pulmonary Stenosis

Abbreviations
MV Mechanical ventilation
SPA Subclavian-pulmonary artery anastomosis
AVB Atrioventricular block
CPB Cardiopulmonary bypass
ASD Atrial septal defect
VSD Ventricular septal defect
DORV Double outlet right ventricle
PE Pulmonary stenosis
AIVR Accelerated idioventricular rhythm
TGA Transposition of the great arteries
RV-PA Right ventricle-pulmonary artery
X Mean
BACKGROUND
In 1969, Giancarlo Rastelli proposed the surgical procedure that bears his name for the treatment of patients with transposition of the great arteries (TGA) with ventricular septal defect (VSD) and pulmonary stenosis (PS). (1-3) This procedure was subsequently used for other congenital heart defects, as double outlet right ventricle (DORV) with pulmonary atresia. (4)

The clear benefit is that this technique preserves the function of both ventricles (biventricular repair) with left ventricular baffling to the aorta. (5) The Rastelli surgery is a complex surgical procedure with marked morbidity and mortality in the medium and long-term follow-up. (6) The most frequent complications are residual VSD, the development of left ventricular outflow tract obstruction, stenosis or insufficiency of the conduit between the right ventricle and the pulmonary artery (RV-PA) (7-9) and ventricular arrhythmias. (10)

Left ventricular baffling to the aorta emerging from the right ventricle is technically easier when the VSD is committed to the great arteries. On the contrary, remote VSD, which is seen in 1/3 of cases, is technically more difficult. In the latter group of patients, either the Nikaidoh procedure (aortic translocation and biventricular outflow tract reconstruction) (7, 11, 12) or the Fontan-Kreutzer procedure (right heart bypass) are valid options.

The anatomical position of the interventricular septum and the presence of abnormal attachments in the tricuspid and or mitral valve to the margin of the VSD where LV baffling to the aorta becomes more difficult, (13-17) are determinant factors to choose the best surgical technique in this group of patients; however, we did not find any references in the bibliography about the outcomes of the Rastelli procedure according to the type of VSD. For this reason, the goal of the present study was to evaluate the impact of the anatomical location of the VSD on the immediate outcomes of the Rastelli procedure and on the short and long-term follow-up.

MATERIAL AND METHODS
We conducted a retrospective study in a cohort of 47 patients undergoing the Rastelli procedure between January 1989 and December 2007 in the Hospital de Pediatría “Prof. Dr. Juan P. Garrahan”.

Patients with congenitally corrected transposition of the great arteries requiring the Rastelli procedure and associated atrial switch operation, DORV and TGA with VSD associated with pulmonary atresia and unsuitable pulmonary vascular tree anatomy, and those lost to follow-up were excluded from the study.

Mean age at surgery was 4 years (15 months - 8 years). Mean weight was 14 kg (6.300-22 kg).

Two anatomical variants were identified: TGA in 24 patients and DORV in 23. Right ventricular outflow obstruction was secondary to pulmonary stenosis in 41 patients and to pulmonary atresia in 6. The following types of VSD were identified: subaortic (2 patients, 49%), subpulmonary (3, 6.4%), doubly committed subarterial (3, 6.4%), muscular (9, 19%) and inlet (9, 19%) defects. Patients were divided into two groups according to the location of the VSD relative to the great arteries: group I (committed VSD, n = 29) and group II (remote or noncommitted VSD (n = 18). Subaortic or subarterial VSDs were considered related to the great arteries. In addition, those perimembranous or muscular defects adjacent to the aorta were also considered committed when left ventricular baffling to the aorta originating in the right ventricle seemed to be feasible after echocardiographic examination. On the contrary, inlet and remote muscular VSDs were considered noncommitted.

The associated defects were patent ductus arteriosus in 16 patients, right aortic arch in 7, atrial septal defect (ASD) in 6, coronary artery anomaly in 6, double superior vena cava system in 7, juxtaposition of the atrial appendages in 3, situs inversus with dextrocardia in 3, straddling tricuspid valve in 3, tricuspid valve dysplasia in 2, vascular ring in 1, and complete atrioventricular canal in 1.

The diagnosis was made on the basis of clinical, radiological, electrocardiographic and, especially, echocardiographic and angiographic findings.

Palliative preoperative procedures included atrial balloon septostomy and subclavian-pulmonary artery anastomosis (SPA). Eleven patients required atrial septostomy, mean age 3.4 months (1 day - 14 months). A total of 42 SPA were performed in 38 patients, mean age 6.4 months (1 day - 30 months): left SPA in 28, right SPA in 6 and bilateral subclavian-pulmonary artery anastomosis in 4 patients.

The characteristics of the study population are described in Table 1.

Surgical technique
All 47 patients underwent median sternotomy, and were placed on cardiopulmonary bypass using bicaval cannulation and cardiopleic solution. Mean cardiopulmonary bypass time was 186.76 min (83-420) and mean aortic cross-clamp time was 91.86 min (44-187).

In all cases, the VSD was baffled with to the aorta with a Dacron patch.

The right ventricular outflow tract was reconstructed using aortic homograft in 18 patients, pulmonary homograft in 22 and pericardial baffle in 7. Additional procedures were used in 32 patients: VSD enlargement in 16, resection of the infundibular septum in 13, closure of ASD in 3, repair of main pulmonary artery and/or pulmonary artery branches in 3, tricuspid valve repair in 2, detachment of tricuspid chordae in 2 and mitral valve repair in 1.

Follow-up
All the patients included in the present study were followed-up since hospital discharge until the end of the study; mean postoperative follow-up was 6 years (15 months-14 years).

All patients were evaluated at our institution and underwent physical examination, chest-X ray, electrocardiogram, color-Doppler echocardiography, exercise stress test and 24-hour Holter monitoring. Twenty patients required cardiac catheterization.

Statistical Analysis
Data were stored using Microsoft Office Excel 2003©. All calculations were performed using Statistix 8.0© software package.

Frequency and/or percentage distribution were established for all the variables in relation with the total number of cases; values were expressed as proportions, mean and standard deviation or median and interquartile range.
RESULTS

Cardiopulmonary bypass time and aortic cross-clamp time

Mean cardiopulmonary bypass time and mean aortic cross-clamp time were 69.21 min (± 55) and 81.68 min (± 20), respectively in group I (committed VSD) and 215.65 min (± 67) and 108.65 min (± 38), respectively, in group II (non-committed VSD) (p = 0.01).
Duration of mechanical ventilation and hospitalization
Mean duration of mechanical ventilation (MV) and hospitalization were 1 day (1-6 days) and 8 days (6-13 days), respectively for group I and 2 days (1.5-4.5 days) and 8 days (5-25 days), respectively for group II (p = 0.15 and p = 0.54, respectively).

Mortality
Three patients (6%) died in the immediate postoperative period (mean 4 days, 3-5 days) due to multiple organ failure. The remaining five deaths occurred at 5 months (mean, 2-17 months), resulting in medium-term mortality of 10.6%. Seven out of 8 patients who died had remote VSD (p = 0.003) (Table 2).

Survival
Overall survival was 82.8% after a mean follow-up of 6 years (15 months-14 years) following corrective surgery. Survival was higher in the group of patients with committed VSD (p = 0.003) (Figure 1).

Arrhythmias
During the immediate postoperative period, 9 patients presented arrhythmias: atrioventricular block (AVB) (n = 4), junctional tachycardia (n = 3) and junctional rhythm (n = 2). Arrhythmias were more frequent in group II (6 patients) compared to group I (3 patients), but this difference was not significant (p = 0.06), and was not related with enlargement of the VSD during surgery (p = 0.64). A definite pacemaker was implanted to 4 patients with AV block; all these patients were in group II (p = 0.14).

During long-term follow-up, one patient had an episode of ventricular tachycardia with left bundle branch block pattern three years after the Rastelli procedure; the anatomic substrate was severe stenosis of the homograft increasing right ventricular pressure to 80% of systemic pressure. The arrhythmia was controlled after homograft was replaced and right ventricular pressure returned to normal levels. Another patient presented accelerated idioventricular rhythm (AIVR) 8 years after corrective surgery. Both patients belonged to group II (p = 0.14).

Reinterventions
Three years (mean, 1 day - 13 years) after corrective surgery, 39 reinterventions were performed: 12

Table 2. Analysis of mortality

<table>
<thead>
<tr>
<th>Patient</th>
<th>Type of VSD</th>
<th>Palliative surgery (number)</th>
<th>Age undergoing Rastelli procedure</th>
<th>Reoperations (number)</th>
<th>Cause of mortality</th>
<th>Age of death (time after surgery)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>I</td>
<td>LSPA - 21 days old</td>
<td>5 years</td>
<td>RV-PA conduit replacement</td>
<td>Uncontrolled infection in infectious endocarditis(atypical mycobacterium, Mycobacterium fortuitumicheloni complex)</td>
<td>7 years (19 months)</td>
</tr>
<tr>
<td>2</td>
<td>II</td>
<td>RSPA - 1 month</td>
<td>6 years</td>
<td>LV-to-aorta baffle revision and replacement of the RV-PA conduit (2)</td>
<td>Uncontrolled infection in infectious endocarditis(Staphylococcus aureus)</td>
<td>6.5 years (9 months)</td>
</tr>
<tr>
<td>3</td>
<td>II</td>
<td>RSPA - 6 months</td>
<td>3 years</td>
<td>Removal of VSD patch + Glenn (1)</td>
<td>Multiorgan failure Low cardiac output syndrome</td>
<td>3 years (3 days)</td>
</tr>
<tr>
<td>4</td>
<td>II</td>
<td>LSPA - 6 months</td>
<td>3 years</td>
<td>Closure of LV-RA communication (1)</td>
<td>Infectious endocarditis (Staphylococcus aureus)</td>
<td>3 years (4 days)</td>
</tr>
<tr>
<td>5</td>
<td>II</td>
<td>LSPA -15 days RSPA - 2 months (2)</td>
<td>– Closure of residual VSD and of LV-RA communication (1)</td>
<td>Infectious endocarditis (Pseudomonas aeruginosa)</td>
<td>(32 days)</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>II</td>
<td>LSPA - 18 months</td>
<td>4 years and 4 months</td>
<td>(0)</td>
<td>Multiorgan failure</td>
<td>4 years and 4 months (5 days)</td>
</tr>
<tr>
<td>7</td>
<td>II</td>
<td>RSPA - 25 months</td>
<td>3 years and 7 months</td>
<td>(0)</td>
<td>Infectious endocarditis (Staphylococcus aureus) with residual VSD</td>
<td>3 years and 9 months (75 days)</td>
</tr>
<tr>
<td>8</td>
<td>II</td>
<td>(0)</td>
<td>6 years and 11 months</td>
<td>– Closure of residual VSD + closure of mitral deft (1)</td>
<td>Severe ventricular dysfunction Low cardiac output syndrome</td>
<td>7 years (51 days)</td>
</tr>
</tbody>
</table>

VSD: Ventricular septal defect. LSPA: Left subclavian-left pulmonary artery anastomosis. RSPA: Right subclavian-right pulmonary artery anastomosis. RV-PA: Right ventricle-pulmonary artery. LV: Left ventricle. LV-RA: left ventricle-right atrium
interventional catheterization procedures in 9 patients and 27 reoperations in 22.

Interventional catheterization procedures
Interventional catheterization procedures included balloon angioplasty of the stenotic homograft in 9 patients, balloon angioplasty of branch pulmonary artery stenosis in 2, stent implant in branch pulmonary arteries in 2, embolization of aortopulmonary collateral vessels in 1, persistent left superior vena cava in 1 and closure of left ventricular-to-right atrial communication in 1 (Figure 2).

Reoperations
Reoperations included closure of VSD, resection of subaortic stenosis and/or LV-to-aorta baffle revision and replacement of the RV-PA conduit. Three patients (6%) were reoperated in the immediate postoperative period: LV-to-aorta baffle revision in 1 and closure of the VSD in 2. All three patients had non-committed VSD (p = 0.05).

One month after the procedure (mean; 1 day - 6 months), 11 patients (23%) required VSD closure. VSD was non-committed in 8 patients (p = 0.007). VSD was secondary to infectious endocarditis in 6 patients.

Six patients (12.8%) developed significant subaortic stenosis requiring surgery after a mean of 5 years (2-9 years). Four of these patients belonged to group II (p = 0.31) (Figure 3).

Fig 1. Survival after the Rastelli procedure in both groups.

Fig 2. Angiography of a patient with DORV who underwent left subclavian-left pulmonary artery anastomosis at the age of 7 months and Rastelli procedure at 3 years and 7 months. Seven months after surgery, the patient required balloon angioplasty of the right pulmonary artery and stent implantation in the left pulmonary artery. The images show the latter procedure.
DISCUSSION

The Rastelli procedure is the conventional surgery for TGA with VSD and PS and DORV with PE. (2, 18) The medium and long-term clinical outcomes show that these patients are not free of complications and reinterventions, (8, 9, 19) which are apparently more frequent when the VSD is anatomically remote or non-committed to the aorta. For this reason, many centers prefer univentricular correction rather than biventricular repair in the presence of this type of VSD.

In this retrospective study, we have compared the outcomes of the Rastelli procedure in patients with committed versus non-committed VSD, confirming that remote VSD has an adverse influence on these outcomes. In fact, in this subgroup of patients of our series, mortality, need of early reinterventions, residual VSD, arrhythmias and the development of subaortic stenosis were higher than in patients with VSD committed to the great arteries.

The surgical procedure is more complex in non-committed VSDs, with higher cardiopulmonary bypass (CPB) time and aortic cross-clamp time, which might explain the greater mortality.

In addition, the technical difficulties related to the procedure produce suboptimal hemodynamic results and higher incidence of reoperations.

In the presence of a non-committed VSD, it is technically more difficult to construct the LV baffle tunnel to the aorta, requiring longer patches and a peculiar intraventricular disposition. These factors explain the greater incidence of residual VSD seen in these patients. Even more, as many of these patients have an inlet muscular VSD, the surgeon has to be very cautious to preserve the bundle of His, probably contributing to residual VSD.

The trend towards greater incidence of arrhythmias and/or AV block in the immediate postoperative period was not related to enlargement of the VSD as an additional surgical procedure, and coincides with the findings of other series (4, 20). We consider that this trend was related to trauma and inflammation of the perinodal area as a consequence of a more complex surgery and to the bundle of His passing anterosuperiorly to an inlet muscular VSD. In fact, the two patients requiring definite pacemaker implantation had an inlet muscular VSD.

As opposed to other series, (4, 21) the presence of arrhythmias in our study in the long-term was non-significant: only 4.2% in 5 years.

The development of subaortic stenosis is a complication described after the Rastelli procedure; in our patients, we found an incidence or 12.7% after a mean follow-up of 5 years, a number that is similar to that previously reported. The incidence of subaortic stenosis was high in patients with remote VSD due to the persistence of a restrictive VSD with insufficient enlargement or an elongated left ventricular outflow tract with anterior orientation; however, this difference was not significant. Probably, this might be the
Table 3. Results of the Rastelli procedure

<table>
<thead>
<tr>
<th>Variable</th>
<th>Committed VSD (n = 29)</th>
<th>Non-committed VSD (n = 18)</th>
<th>OR</th>
<th>95% CI</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aortic cross-clamp time, min (mean-SD)</td>
<td>81.68 (± 20)</td>
<td>108.65 (± 38)</td>
<td>4.33</td>
<td>0.92-20.3</td>
<td>0.01</td>
</tr>
<tr>
<td>−CBP time, min (mean-SD)</td>
<td>169.21 (± 55)</td>
<td>215.65 (± 67)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Days in MV (median-IQR)</td>
<td>1 (1-6)</td>
<td>2 (1.5-4.5)</td>
<td>0.61</td>
<td>0.76-8.6</td>
<td>0.15</td>
</tr>
<tr>
<td>Days of hospitalization (median-IQR)</td>
<td>8 (6-13)</td>
<td>8 (5-25)</td>
<td>0.92</td>
<td>0.61-1.3</td>
<td>0.54</td>
</tr>
<tr>
<td>Arrhythmia in the immediate postoperative period</td>
<td>3 (0)</td>
<td>6 (0)</td>
<td>0.25</td>
<td>0.04-1.59</td>
<td>0.18</td>
</tr>
<tr>
<td>Reoperations</td>
<td>11 (10)</td>
<td>11 (10)</td>
<td>0.61</td>
<td>0.76-8.6</td>
<td>0.12</td>
</tr>
<tr>
<td>Reoperations in the immediate postoperative period</td>
<td>0 (0)</td>
<td>3 (0)</td>
<td></td>
<td></td>
<td>0.05</td>
</tr>
<tr>
<td>Reoperation for closure of VSD</td>
<td>3 (2)</td>
<td>8 (2)</td>
<td>1.93</td>
<td>1.52-3.15</td>
<td>0.007</td>
</tr>
<tr>
<td>Reoperations due to subaortic stenosis</td>
<td>2 (1)</td>
<td>4 (1)</td>
<td>0.25</td>
<td>0.04-1.59</td>
<td>0.18</td>
</tr>
<tr>
<td>Mortality</td>
<td>1 (1)</td>
<td>7 (2)</td>
<td>2.88</td>
<td>1.95-1.62</td>
<td>0.003</td>
</tr>
</tbody>
</table>


consequence of multifactorial causes, such as conal septal hypertrophy and preexistent discontinuity between the mitral valve and the aortic valve producing a fibrous diaphragm, as seen in two of our patients with committed VSD.

Replacement of the RV-PA conduit due to stenosis or insufficiency is inevitable after the Rastelli procedure and independent of the anatomic position of the VSD; it is the consequence of the natural history of homografts. In our series, 25% of our patients presented this complication after 5 years, similar (20, 22 and lower) than the frequency reported by other studies, and without differences between both groups. However, the incidence of reoperations was lower with pericardial conduits.

Since June 2005, we our developing an initial experience with the Nikaidoh procedure, that consists of translocation of the great arteries avoiding the complex baffling of a remote VSD to the aorta.

Seven patients have already been operated on. In all cases, the surgery consisted on aortic translocation, reimplantation of coronary arteries and right ventricle to pulmonary artery connection using aortic (3 patients) or pulmonary (4 patients) homografts.

No deaths occurred with this procedure. None of the patients developed residual VSD or significant outflow tract obstruction of both ventricles. Due to the suboptimal outcomes obtained with the Rastelli procedure and the encouraging preliminary results of the Nikaidoh procedure, we believe that the latter is the surgical technique of choice in patients with remote VSD.

CONCLUSIONS

Remote VSD in patients undergoing the Rastelli procedure is associated with: 1) increased mortality, 2) greater incidence of reoperations in the immediate postoperative period, and, 3) a trend towards greater incidence of arrhythmias in the immediate postoperative period.

In this subgroup of patients with non-committed VSD, the Nikaidoh procedure emerges as the best option in terms of immediate hemodynamic results and of potential complications in the medium and long-term. When this alternative is not possible, univentricular correction should be considered.

RESUMEN

Cirugía de Rastelli: impacto adverso de la comunicación interventricular no relacionada con los grandes vasos en los resultados quirúrgicos

Introducción

La evolución de los pacientes operados con técnica de Rastelli revela que se trata de un procedimiento quirúrgico complejo no exento de morbilidad y mortalidad a mediano y a largo plazos. Las complicaciones serían, aparentemente, más frecuentes cuando la comunicación interventricular (CIV) es anatómicamente remota o no relacionada con la aorta.

Objetivo

Evaluar el impacto de la localización anatómica de la CIV en la cirugía de Rastelli.

Material y métodos

Se incluyeron 47 pacientes con una media (X) de seguimiento postquirúrgico de 6 años (15 meses-14 años). Los pacientes se subdividieron en: grupo I, CIV relacionada con la aorta (n = 29) y grupo II, CIV remota o no relacionada (n = 18).

Resultados

En el posquirúrgico inmediato, las reoperaciones (p = 0.05) fueron más frecuentes en el grupo II, en el cual se observó también una tendencia a mayor frecuencia de arritmias (p = 0.06).

Luego de una X = 3 años (1 día-13 años) posquirúrgicos se realizaron 27 reoperaciones en 22 pacientes.
Se cerró CIV residual en 11 pacientes a una X = 1 mes. En 8 de ellos, la CIV era remota (p = 0,007). Desarrollaron estenosis subaórtica que requirieron cirugía 6 pacientes a una X = 5 años. Se reemplazó el conducto ventrículo derecho-arteria pulmonar en 12 pacientes a una X = 5 años. La mortalidad en el periodo posquirúrgico inmediato fue del 6% (3 pacientes). La mortalidad global fue del 17,2% (n = 8); 7 pacientes tenían CIV no relacionada (p = 0,003).

**Conclusiones**

La CIV remota en la cirugía de Rastelli se asocia con: 1) mayor mortalidad, 2) mayor frecuencia de reoperaciones en el posquirúrgico inmediato y 3) tendencia a mayor frecuencia de arritmias en el posquirúrgico inmediato.

**Palabras clave** > Cirugía cardíaca - Ventrículo derecho con doble salida - Transposición de los grandes vasos - Estenosis pulmonar

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