

Biventricular Repair in Patients with Transposition of the Great Arteries, Ventricular Septal Defect and Pulmonary Stenosis: Rastelli, Nikaidoh or REV Procedures?

Cirugía de reparación biventricular en pacientes con transposición de los grandes vasos, comunicación interventricular y estenosis pulmonar: ¿Rastelli, Nikaidoh, REV?

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ABSTRACT

Background: Biventricular repair of transposition of the great arteries with ventricular septal defect and pulmonary stenosis includes the Rastelli operation, the REV (*reparation a l'etage ventriculaire*) and the Nikaidoh procedure. The treatment of choice is still controversial due to the anatomic variability of this condition and to the suboptimal results of the different surgical techniques proposed.

Objectives: To evaluate the results of biventricular repair procedures performed in our hospital in patients with transposition of the great arteries with ventricular septal defect and pulmonary stenosis. 2. To compare morbidity and mortality with the Rastelli operation and the Nikaidoh procedure in the subgroup of patients with non-committed ventricular septal defect.

Methods: Between 1991 and 2015, 76 patients operated on in our center underwent the Rastelli operation [n=60 (78.9%)], the Nikaidoh procedure [n=13 (17%)] and the REV procedure [n=3 (4%)].

Results: In the immediate postoperative period, 24 patients presented ventricular dysfunction, 18 had arrhythmias and 11 developed subaortic stenosis. None of the patients presented signs of significant aortic regurgitation. During a mean follow-up of 9.9 years (\pm 6 years), 52 patients developed dysfunction of the right ventricle-to-pulmonary artery conduit, 14 patients presented left ventricular outflow tract obstruction, 12 patients had arrhythmias and 1 patient developed moderate aortic regurgitation. Long-term survival at 5 and 10 years was 96%, and 92% at 15 years. Fifty-five percent of the patients required reintervention at 6.2 years (\pm 5), particularly due to dysfunction of the right ventricle-to-pulmonary artery conduit (77%). Nine patients died, 7 in the immediate postoperative period. Mortality was associated with non-committed ventricular septal defect ($p=0.02$), ventricular dysfunction ($p=0.02$), arrhythmias ($p=0.01$) and reoperations ($p=0.0000$) in the immediate postoperative period. In the group of patients with non-committed ventricular septal defect, the Rastelli operation was associated with higher mortality ($p=0.01$) and subaortic obstruction in the immediate and late postoperative periods ($p=0.04$ and $p=0.01$, respectively), compared with the Nikaidoh procedure.

Conclusions:

- Patients undergoing the Rastelli operation, the Nikaidoh procedure and the REV procedure have favorable long-term survival.
- Reinterventions are common (55%), particularly due to dysfunction of the right ventricle-to-pulmonary artery conduit.
- In patients with non-committed ventricular septal defect, the Nikaidoh procedure emerges as a better therapeutic option than the Rastelli operation.

Key words: Heart Defects, Congenital - Cardiac Surgical Procedures/methods - Heart Septal Defects, Ventricular/surgery - Transposition of Great Vessels - Pulmonary Stenosis - Postoperative Period

RESUMEN

Introducción: La corrección biventricular de la transposición de los grandes vasos con comunicación interventricular y estenosis pulmonar incluye las cirugías de Rastelli, REV (*reparation a l'etage ventriculaire*) y Nikaidoh. El tratamiento de elección aún es controversial, dada la gran variabilidad anatómica de esta entidad y los resultados subóptimos de las diferentes técnicas quirúrgicas propuestas.

Objetivos: Evaluar los resultados de las cirugías de corrección biventricular realizadas en nuestro hospital en los pacientes con transposición de los grandes vasos con comunicación interventricular y estenosis pulmonar. 2. Comparar las cirugías de Rastelli y Nikaidoh en términos de morbimortalidad en el subgrupo de pacientes con comunicación interventricular no relacionada con la aorta.

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Material y métodos: Se incluyeron 76 pacientes operados durante el período 1991-2015 en nuestro centro con técnica de Rastelli [$n = 60$ (78,9%)], Nikaidoh [$n = 13$ (17%)] y REV [$n = 3$ (4%)].

Resultados: En el posquirúrgico inmediato, 24 pacientes tuvieron disfunción ventricular, 18 arritmias y 11 estenosis subaórtica. Ninguno presentó insuficiencia aórtica significativa.

En la evolución, con un tiempo medio de seguimiento de 9,9 años (± 6 años), 52 pacientes desarrollaron disfunción del conducto ventrículo derecho-arteria pulmonar, 14 pacientes obstrucción al tracto de salida ventricular izquierdo, 12 pacientes arritmias y 1 paciente insuficiencia aórtica moderada.

La sobrevida alejada a los 5 y 10 años fue del 96% y a los 15 años, del 92%.

El 55% de los pacientes requirieron reintervención a los 6,2 años (± 5), principalmente por disfunción del conducto ventrículo derecho-arteria pulmonar (77%).

Murieron 9 pacientes, 7 de ellos en el posquirúrgico inmediato. La mortalidad se asoció con comunicación interventricular no relacionada con la aorta ($p = 0,02$) y con disfunción ventricular ($p = 0,02$), arritmias ($p = 0,01$) y reoperaciones ($p = 0,0000$) en el posquirúrgico inmediato.

En el grupo de pacientes con comunicación interventricular no relacionada con la aorta, la cirugía de Rastelli presentó mayor mortalidad ($p = 0,01$) y obstrucción subaórtica en el posquirúrgico inmediato ($p = 0,04$) y alejado ($p = 0,01$) que la de Nikaidoh.

Conclusiones:

- Los pacientes con cirugía de Rastelli, Nikaidoh y REV tienen buena sobrevida alejada.
- Las reintervenciones son frecuentes (55%), principalmente por disfunción del conducto ventrículo derecho-arteria pulmonar.
- En los pacientes con comunicación interventricular no relacionada con la aorta, la cirugía de Nikaidoh surge como mejor opción terapéutica que la de Rastelli.

Palabras clave: Cardiopatías congénitas - Procedimientos quirúrgicos cardíacos/métodos - Defectos del tabique interventricular/cirugía - Transposición de grandes vasos - Estenosis pulmonar - Período posoperatorio

Abbreviations

IQR	MV Mechanical ventilation	LVOTO	Left ventricular outflow tract obstruction
AVB	Atrioventricular block	REV	Reparation a l'étage ventriculaire
VSD	Ventricular septal defect	TGA	Transposition of the great arteries
DORV	Double outlet right ventricle	VT	Ventricular tachycardia
PS	Pulmonary stenosis	RV	Right ventricle
VPB	Ventricular premature beats	RV-PA	Right ventricle-pulmonary artery
IQR	Interquartile range	LV	Left ventricle

INTRODUCTION

Transposition of the great arteries (TGA) with ventricular septal defect (VSD) and pulmonary stenosis (PS) is an uncommon congenital heart defect that can be repaired with several surgical techniques.

The method described by Giancarlo Rastelli in 1969, known as the Rastelli operation, has been the conventional treatment of this congenital heart defect for many decades. (1, 2) Nowadays, the Rastelli operation is a surgical procedure with low short-term mortality, (3) but with marked mid and long-term morbidity and mortality. (3-13)

In 1982, Lecompte proposed the reparation a l'étage ventriculaire (REV), a technique that reduces the incidence of left ventricular tract obstruction, but generates significant residual pulmonary regurgitation. (14-18)

In 1984, Hisashi Nikaidoh published a new surgical technique: "aortic translocation and biventricular outflow tract reconstruction". (19) This procedure is presented as a promising therapeutic option, (20-32) particularly for patients with unfavorable anatomy to undergo the Rastelli procedure, as non-committed VSD, (5) restrictive VSD, overriding of the atrioventricular valves, hypoplastic right ventricle and some coronary artery anomalies. (19, 33)

Today, the treatment of choice for TGA with VSD and PS is still controversial due to the anatomic variability of this condition and to the suboptimal results of the different surgical techniques proposed. (34-38)

In the present study, we analyze the mid and long-term surgical outcomes of the Rastelli, Nikaidoh and REV procedures, and also compare the morbidity and mortality of the Rastelli operation and the Nikaidoh procedure in the complex subgroup of patients with TGA and double outlet right ventricle (DORV) with non-committed VSD and PS.

METHODS

We conducted a retrospective study in a cohort of 76 patients with TGA associated with VSD and PS or DORV with TGA like physiology, VSD and PS undergoing biventricular surgery in our center between 1991 and 2015.

Patients were excluded if they had congenitally corrected TGA with VSD and PS requiring double switch plus atrial switch, or had pulmonary atresia or unsuitable pulmonary vascular tree anatomy.

The following anatomical variants were identified: TGA with VSD and PS [$n=42$ (55%)] and DORV with PS and TGA like physiology [$n=34$ (45%)].

All patients had moderate or severe left ventricular outflow tract obstruction (LVOTO) secondary to PS ($n=69$) or pulmonary atresia ($n=7$).

The anatomical location of the VSD was subaortic in 32 patients (42.1%), subpulmonary in 4 (5.3%), doubly committed in 7 (9.2%), of the inlet in 19 (25%) and muscular in 14 (18.4%).

In 33 patients (43.4%) the VSD was non-committed with the aorta and was restrictive in 9 patients (11.8%).

The most commonly associated congenital heart defects were tricuspid valve anomalies (10 cases), consisting of straddling in 5, overriding in 3 and tricuspid valve dysplasia in 2 cases; coronary artery anomalies (7 cases); pulmonary branch anomalies (4 cases); hypoplastic right ventricle (RV) (3 cases); situs inversus with dextrocardia (3 cases); atrial septal defect (2 cases); multiple VSDs (2 cases), double VSD (1 case), complete atrioventricular canal defect (2 cases) and right-sided aortic arch (2 cases).

The diagnosis was made on the basis of clinical, radiological and electrocardiographic findings, and on echocardiographic, angiographic and computed tomography angiography results.

Eighty-four percent of the patients underwent interventional cardiology procedures (16 patients) or palliative surgeries (62 patients) before the operation.

Surgical technique

All the patients included in this study underwent a biventricular repair surgery: Rastelli operation (n=60), Nikaidoh procedure (n=13) or REV (n=3).

Median age at the moment of surgery was 3.7 years [interquartile range (IQR) 25%-75%: 2.4 months-4.8 years; range: 2.4 months-10.8 years] and mean weight was 13.8 kg (\pm 4.2Kg; range: 4.5-24 kg).

Follow-up

All the patients were followed-up in our hospital with a mean follow-up of 9.9 years after surgery (\pm 6 years; range: 6 months-24 years).

All patients were assessed by means of physical examination, chest-X ray, electrocardiogram, color-Doppler echocardiography, tissue-Doppler echocardiography, exercise stress test and 24-hour Holter monitoring. Some patients also underwent stress echocardiography, cardiac magnetic resonance imaging, multislice computed tomography and/or cardiac catheterization.

The diagnosis of heart valve stenosis or regurgitation was made according to echocardiographic findings and following the current guidelines (39, 40); only moderate and severe valve diseases were considered significant.

Functional capacity was assessed using the New York Heart Association classification.

Left ventricular (LV) function was determined by Doppler echocardiography (M-mode, two-dimensional and tissue-Doppler) and magnetic resonance imaging was used to evaluate right ventricular function.

Statistical analysis

Data were stored using Microsoft Office Excel®2013. All the calculations were performed using Statistix 8.0 software package.

Qualitative variables were expressed as absolute values or percentages, while quantitative variables with normal and non-Gaussian distribution were expressed as means and standard deviation or medians and IQR, respectively.

Continuous variables were compared using Student's t test or the Mann-Whitney test, as applicable. Fisher's exact test or the chi square test was used to compare proportions.

A p value <0.05 was considered statistically significant. Mid and long-term survival was estimated using the Kaplan-Meier method.

Ethical considerations

The study was approved by the institutional Ethics Committee according to valid regulations for observational studies and following the recommendations of the Declaration of Helsinki.

RESULTS

Median cardiopulmonary bypass time was 182 min (IQR 25%-75%: 149-220) and mean aortic cross-clamp time was 101.5 min (IQR 25%-75%: 75-131.75)

Patients were hospitalized for a median of 9 days (IQR 25%-75%: 3-17, median inotropic support requirement was 5 days (IQR 25%-75%: 2-9) and median mechanical ventilation was 2 days (IQR 25%-75%: 1-7).

Overall mortality was 11.8%. Nine patients died, 7 in the immediate postoperative period.

Surgical mortality was 9.2% (7 patients) but in the second half of the period (since 2003) mortality decreased to 2.6%.

In-hospital mortality occurred at a median of 17 days after surgery (IQR 25%-75%: 1.5-88 days) and was secondary to severe isolated ventricular dysfunction in 1 patient and in the rest of the patients (n=6) was due to arrhythmias (n=3): ventricular tachycardia in 1, complete atrioventricular block (AVB) in 1, and nodal tachycardia in 1; residual defects (n=3): residual VSD in 2, LVOTO and hypoplastic RV in 1, and sepsis or infective endocarditis (n=4).

Long-term mortality was 2.9%. Two patients died at 1.5 and 11.9 years after surgery due to uncontrolled infective endocarditis and leukemia.

At univariate analysis, mortality was associated with VSD non-committed with the aorta (p=0.02), reoperations (p=0.0000), ventricular dysfunction (p=0.02) and arrhythmias (p=0.01) in the immediate postoperative period.

Long-term survival at 5 and 10 years was 96%, and 92% at 15 years (Figure 1 A).

Ninety-seven percent of the surviving patients are in functional class I with good ventricular function.

In the immediate postoperative period, 34.2% of patients (n=26) presented ventricular dysfunction with favorable response to medical treatment, except for one case that required ventricular assist device for 72 hours.

During follow-up, 97% of the patients presented good ventricular function with a mean shortening fraction of 36.8% (\pm 4.87) and mean ejection fraction of 68.33% (\pm 4.58).

Arrhythmias

In the immediate postoperative period, 18 patients presented arrhythmias: junctional tachycardia in 8, AVB in 8, atrial flutter in 1 and ventricular premature beats (VPB) in 1 patient.

One patient presented first-degree AVB and 7 patients had complete AVB. Complete AVB was transient in 6 patients and a definite pacemaker was implanted in only 1 patient.

Junctional tachycardia was successfully treated with medical therapy in all cases.

The incidence of arrhythmias at the long-term outcome was 19.4% (n=13) at a median time of 8.5 years after surgery (IQR 25%-75%: 5.7-11).

The following arrhythmias were recorded: ventricular tachycardia (VT) in 4 patients, ventricular fibrillation in 1, non-sustained VT in 1, atrial tachycardia in 1, accelerated His bundle rhythm in 1, frequent VBPs in 3 and frequent supraventricular extrasystoles in 2.

Ventricular fibrillation occurred during a diagnostic cardiac catheterization performed 6 months after the Rastelli operation in a patient with residual VSD and left ventricular dilation with severe dysfunction.

Ventricular tachycardia occurred at a median of 8.5 years after surgery (IQR 25%-75%: 2.9-10.5). A hemodynamic substrate [right ventricular pressure overload and/or volume overload due to severe dysfunction of the right ventricle to pulmonary artery (RV-PA) conduit] was identified in 3 patients, requir-

ing an interventional cardiology procedure (n=1) and surgery (n=2). Implantable cardioverter defibrillator therapy was indicated in 3 patients.

Non-sustained VT and atrial tachycardia were identified in 24-hour Holter monitoring in asymptomatic patients and were managed with medical treatment.

Left ventricular outflow tract

Left ventricular outflow tract obstruction was a complication observed only in patients undergoing the Rastelli operation (Figure 2).

Eleven patients (14.4%) presented subaortic stenosis in the immediate postoperative period with a mean gradient of 30 mm Hg (± 13.27). Two of them required reoperation for LV-to-aorta baffle revision, one at 12 hours and the other 15 days after surgery.

During follow-up, subaortic stenosis affected 14 patients (20.8%) with a mean gradient of 38 mm Hg (± 25.19), which was significant in 8 patients (11.9%).

This late complication was the reason for reoperation in 9 patients at a mean time of 6.5 years after surgery (± 2.1). The remaining 5 patients are under control. Three of them have residual stenosis and subaortic stenosis developed in 2 during the long-term follow-up. All the patients remain stable with

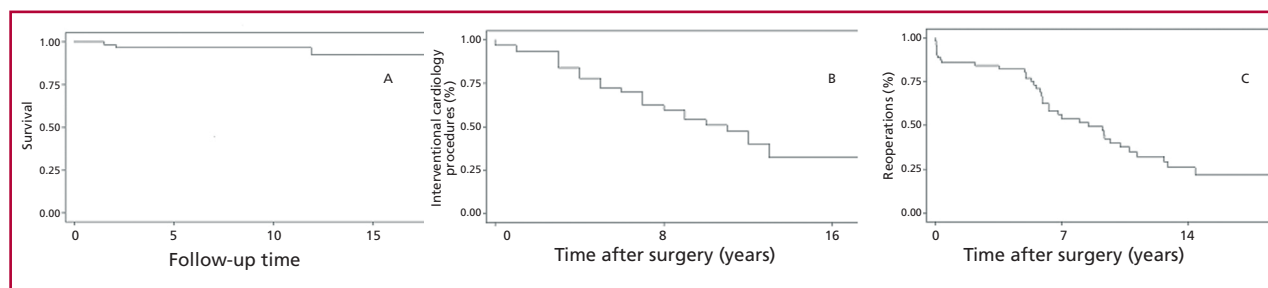


Fig. 1. Kaplan-Meier curves. A. Long-term survival. B. Interventional cardiology procedures. C. Reoperations.

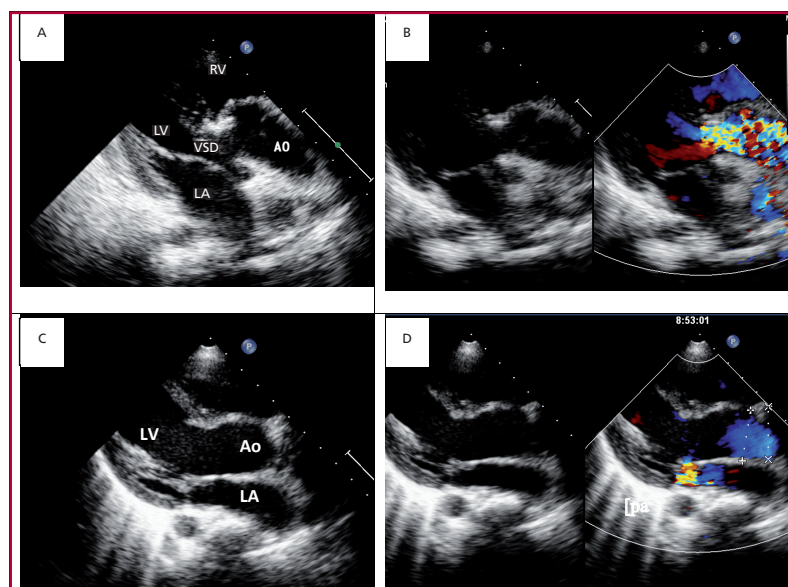


Fig. 2. Echocardiographic images (in the left parasternal long axis view) of a patient with transposition of the great arteries with ventricular septal defect and pulmonary stenosis, undergoing the Rastelli operation (A and B) and the Nikaidoh procedure (C and D). In A, the left ventricular outflow tract is misaligned with an elongated baffle patch from the ventricular septal defect to the aorta that emerges from the right ventricle and generates turbulent flow revealed by color-Doppler echocardiography (B). A better alignment of the left ventricular outflow tract with the aorta is seen in C, with laminar flow by color-Doppler echocardiography (D). LA: Left atrium. Ao: Aorta. RV: Right ventricle. LV: Left ventricle. VSD: Ventricular septal defect.

mild stenosis after a mean follow-up time of 11 years (± 6.8) after surgery.

Significant aortic regurgitation did not occur in the immediate postoperative period. During follow-up, only one patient who underwent the Nikaidoh procedure developed moderate aortic regurgitation (1.6%),

Right ventricular outflow tract

Fifty-two patients developed RV-PA conduit dysfunction: 40 with the Rastelli operation, 10 with the Nikaidoh procedure and 2 with REV (p 0.58).

Thirty-eight patients presented significant stenosis of this conduit with a mean gradient of 62.78 mm Hg (± 16.8), which was severe in 22 patients.

Significant insufficiency of the RV-PA conduit was seen in 33 patients, which was severe in 10.

Seventy-five patients required reoperations due to severe dysfunction of the RV-PA conduit: 32 patients underwent an interventional cardiology procedure (balloon dilation) and 43 patients were reoperated to replace the conduit.

Reinterventions

Fifty-five percent (42 patients) of the patients required reinterventions (interventional cardiology procedures and/or surgery) after a mean of 6.2 years (± 5).

Freedom from interventional cardiology procedures occurred in 72%, 51% and 32% of the patients at 5, 10 and 15 years, respectively (Figure 1B).

A total of 42 interventional cardiology procedures were performed in 29 patients at a mean time of 6.5 years (± 4.5).

The main indication [32 procedures (76.2%)] was severe stenosis of the RV-PA conduit, requiring balloon angioplasty (Figure 3).

The rest of the procedures were: balloon angioplasty or stent implantation of pulmonary artery branches (n=3), stent implantation in the right pulmonary branch + embolization of the left superior vena cava (n=1), embolization of aortopulmonary collaterals (n=2), LV-right atrium closure (n=1) and radiofrequency catheter ablation (n=3).

Freedom from reoperation occurred in 76%, 37% and 21% of the patients at 5, 10 and 15 years, respectively (Figure 1 C).

Fifty-five reoperations were performed in 39 patients after a mean time of 6.05 years after surgery (± 4.7 ; range: 0-18.8 years).

Among the patients undergoing reoperations, 12 required a second reoperation, 3 a third reoperation and 1 patient required 4 reinterventions at a mean time of 7 (± 5.4), 7.5 (± 4.9), and 10.2 years after surgery, respectively. One patient is scheduled for implantable cardioverter defibrillator insertion.

Reoperations are detailed in Table 1.

Subgroup of patients with non-committed ventricular septal defect

Thirty-three patients had non-committed VSD; 20 underwent the Rastelli operation and 13 the Nikaidoh procedure.

The immediate and long-term outcomes of this subgroup are summarized in Table 2.

Among these 33 patients, those undergoing the Rastelli operation had greater mortality (p=0.01) and greater incidence of subaortic stenosis in the immediate (p=0.04) and long-term (p =0.01) follow-up period than those who underwent the Nikaidoh procedure.

DISCUSSION

The surgery of choice for TGA with VSD and PS and for DORV with PS and TGA like physiology is still controversial. Several surgical techniques have been proposed for biventricular repair, as the Rastelli operation, the REV procedure and the Nikaidoh procedure.

In the present study, we corroborated that these three surgical techniques provide favorable long-term survival, as most of the patients are asymptomatic, with good functional class and preserved ventricular function.

In the long-term, the Rastelli operation, the Nikaidoh procedure and the REV procedure clearly demonstrate to be surgical repair techniques but not corrective procedures, with development of complications and need of reinterventions.

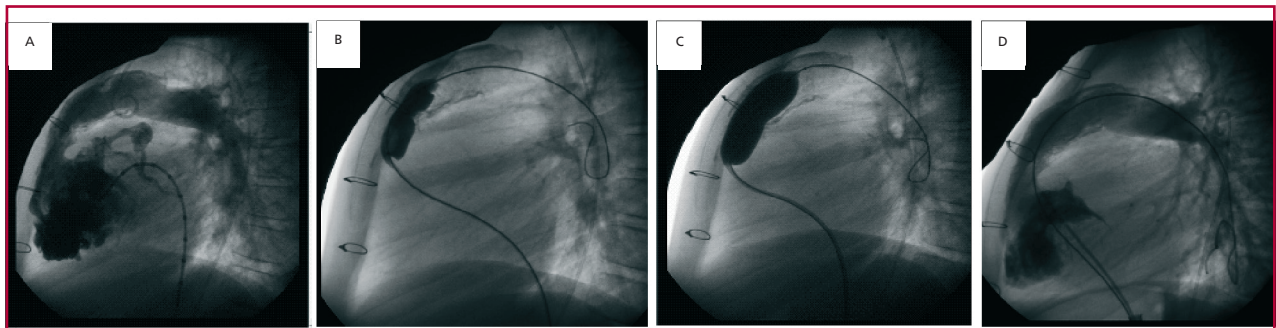


Fig. 3. Interventional cardiology procedure in a patient who developed severe stenosis of the right ventricle-pulmonary artery homograft 5 years after the Rastelli operation. The stenosis is observed in A. B and C. Stenosis dilation using balloon catheter. D. Angiography after effective dilation.

<p>Reoperations indicated (total n=55)</p> <p>On the right side (n=33)</p> <ul style="list-style-type: none"> – RV-PA conduit replacement (23) – RV-PA conduit replacement + VSD closure + mitral valve repair + tricuspid valve repair (1) – RV-PA conduit replacement + punctiform VSD closure + tricuspid valve repair (2) – RV-PA conduit replacement + VSD closure (2) – RV-PA conduit replacement + tricuspid valve repair (1) – RV-PA conduit replacement + mitral valve replacement (2) – RV-PA conduit replacement + mitral valve annuloplasty (1) – Subpulmonary resection + VSD closure (1) <p>On the left side (n=3)</p> <ul style="list-style-type: none"> – Baffle enlargement (1) – Baffle enlargement + Glenn cavo-pulmonary shunt (1) – Resection of subaortic membrane (1) <p>On both outflow tracts (n=11)</p> <ul style="list-style-type: none"> – RV-PA conduit replacement + resection of subaortic membrane (5) – RV-PA conduit replacement + resection of subaortic membrane + LV-Ao baffle revision (1) – RV-PA conduit replacement + VSD enlargement (1) – RV-PA conduit replacement + LV-Ao baffle revision: patch replacement (TV and patch vegetations) (1). – RV-PA conduit replacement + RPAB repair + replacement of the ascending aorta (1) – RV-PA conduit replacement + resection of subaortic membrane and myomectomy (1) – RV-PA conduit replacement + closure of the aneurysm mouth (1) <p>Others (n=8)</p> <ul style="list-style-type: none"> – Closure of residual VSD (3) – Closure of VSD with LV-RA shunt + tricuspid valve repair (1) – Pacemaker implant (1) – ICD implant (2) – Mitral valve replacement with mechanical prosthesis.
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Table 1. Reoperations on the 76 patients operated

RV-PA: Right ventricle-pulmonary artery. VSD: Ventricular septal defect. MV: Mitral valve. LV-Ao: Left ventricle-aorta. TV: Tricuspid valve. RPAB: Right pulmonary artery branch. LV-RA: Left ventricle-right atrium. ICD: Implantable cardioverter defibrillator.

Dysfunction of the RV-PA conduit was the most common adverse event and the main cause of reintervention. Although in the Nikaidoh procedure the aortic translocation to a posterior position leaves more room for the RV-PA conduit connection, avoiding exposure to sternal anterior compression, in our series we did not find significant differences in RV-PA conduit durability with this technique. This result evidences that dysfunction of the RV-PA conduit responds not only to anatomical factors but also to the natural history of homografts.

Subaortic stenosis was the cause of reintervention in 11% of our population undergoing the Rastelli operation, while no cases of LVOTO were reported after the Nikaidoh procedure, in agreement with previous reports. (4, 9, 24, 27, 28) This confirms one of the main foundations of aortic translocation, which is to achieve a better LV-aorta alignment, avoiding long intracardiac tunneling to reduce the risk of subaortic obstruction.

Aortic regurgitation was not a significant adverse event in our series, as opposed to other groups that reported this complication in the outcome of the Ni-

kaidoh procedure. (20) As this procedure preserves the aortic root and the morphologically normal left semilunar valve, the incidence of significant aortic regurgitation is lower than the one described in patients operated on with arterial switch or the Ross procedure.

Arrhythmias were common in our series as in previous reports (4) and secondary to hemodynamic and electrical substrates, confirming the importance of a thorough follow-up in this complex group of patients, where electrocardiogram, color-Doppler echocardiography, 24-hour Holter monitoring and exercise stress test should be routine complementary studies.

Surgical mortality decreased during the second period (learning curve) and long-term mortality was low and attributable to comorbidities rather than to surgery. Mortality was significantly higher in the group of patients with VSD non-committed with the aorta and was also associated with arrhythmias, ventricular dysfunction and reoperations in the immediate post-operative period. The adverse effect of remote VSD on morbidity and mortality related to the Rastelli operation is reaffirmed, as already published by our center,

Table 2. Postoperative results in the subgroup of patients with non-committed ventricular septal defect (n = 33)

Variable	Rastelli surgery (N = 20)	Nikaidoh procedure (N = 13)	p
CPB time, minutes	208.9 (±64.5)	225.3 (±64.2)	0.50
Aortic cross-clamp time, minutes	124.4 (±30.5)	169.1 (±51.04)	0.004
Immediate postoperative period			
– Days in MV	14.1 (±24.4)	10.6 (±13.3)	0.65
– Days with inotropic support	10 (±12.3)	12.5 (±12.7)	0.58
– Days of hospitalization	19.9 (±19.9)	23.2 (±31)	0.72
– Surgical mortality	6	0	0.02
– Ventricular dysfunction	9	6	0.94
– Arrhythmias	8	2	0.31
– LVOTO	5	0	0.04
– Significant aortic valve regurgitation	0	0	
– Interventional cardiology procedures	2	0	0.23
– Reoperations	7	1	0.07
Long-term postoperative period			
– Follow-up, years	9.06 (±6.05)	6.6 (±3.2)	0.24
– Long-term mortality	1	0	0.41
– Ventricular dysfunction	1	1	0.90
– Arrhythmias	3	4	0.47
– LVOTO	6	0	0.01
– Significant aortic valve regurgitation	0	1	0.26
– Significant stenosis of the RV-PA conduit	10	6	0.24
– Significant insufficiency of the RV-PA conduit	11	9	0.85
Overall mortality	7	0	0.01
Interventional cardiology procedures	7	3	0.46
Reoperations	11	4	0.17

CPB: Cardiopulmonary bypass. MV: Mechanical ventilation. LVOTO: Left ventricular outflow tract obstruction. RV-PA: Right ventricle-pulmonary artery.

(5) which motivated a change in the therapeutic strategy of patients with this anatomic variant, initiating our experience with the Nikaidoh procedure. (22)

This subgroup of patients with non-committed VSD has also deserved a specific analysis related to the type of surgery. In patients undergoing the Nikaidoh procedure it was demonstrated that despite this was a very demanding technique with longer cardiopulmonary bypass time, the results were similar to those of the Rastelli operation in terms of ventricular dysfunction, days of hospitalization and requirements of MV and inotropic support, while the Rastelli operation presented higher morbidity and mortality and greater incidence of subaortic stenosis in the immediate and long-term follow-up than the Nikaidoh procedure.

Study limitations

These findings should be confirmed with a larger number of patients during a longer follow-up period.

CONCLUSIONS

The surgical procedures for biventricular repair of patients with TGA, VSD and PS offer good long-term survival rate despite the presence of adverse events and need for reinterventions. In this subgroup of patients with noncommitted VSD, the Nikaidoh procedure

emerges as the best option. Patients who survive to these surgeries of biventricular repair should undergo a thorough and periodical follow-up.

Conflicts of interest

None declared. (See authors' conflicts of interest forms on the website/Supplementary material).

REFERENCES

- Rastelli GC. A new approach to anatomic repair of transposition of the great arteries. *Mayo Clin Proc* 1969;44:1-12.
- Lewis Backer C, Mauvroudis C. The Rastelli operation. *Operative Techniques in Thorac Cardiovasc Surg* 2003;8:121-30. <http://doi.org/fbtb9k>
- Sato Y, Ayusawa M, Komatsu S, Matsumoto N, Ichikawa M, Yoda S, et al. MCDT evaluation of a double outlet right ventricle after the Rastelli procedure. *Int J Cardiol* 2007;117:430-2. <http://doi.org/d3ddjb>
- Kreutzer C, De Vive J, Oppido G, Kreutzer J, Gauvreau K, Freed M, et al. Twenty-five year experience with Rastelli repair for transposition of the great arteries. *J Thorac Cardiovasc Surg* 2000;120:211-23. <http://doi.org/ffjk4w>
- Villalba CN, Woloszyn MI, Mouratian MD, Villa A, Di Santo M, Álvarez L, et al. The Rastelli Procedure: Adverse Impact in Surgical Outcomes of a Ventricular Septal Defect Noncommitted to the Great Arteries. *Rev Argent Cardiol* 2010;78:315-22.
- Dearani J, Danielson G, Puga F, Mair DD, Schleck CD. Late results of the Rastelli operation for transposition of the great arteries. *Se-*

- min Thorac Cardiovasc Surg Pediatr Card Surg Annu 2001;4:3-15. <http://doi.org/b76t>
7. Vouhé P, Tamisier D, Leca F, Ouaknine R, Vernant F, Neveux JY. Transposition of the great arteries, ventricular septal defect, and pulmonary outflow tract. *J Thorac Cardiovasc Surg* 1992;103:428-36.
 8. Brown J, Ruzmetov M, Okada Y, Vijay P, Turrentine MW. Surgical results in patients with double outlet right ventricle: a 20-year experience. *Ann Thorac Surg* 2001;72:1630-5. <http://doi.org/bxgbmq>
 9. Hörer J, Schreiber C, Dworak E, Cleuziou J, Prodan Z, Vogt M. Long-term results after the Rastelli repair for transposition of the great arteries. *Ann Thorac Surg* 2007;83:2169-75. <http://doi.org/bts4kz>
 10. Villagra F, Quero-Jimenez M, Maitre-Azcarate MJ, Gutierrez J, Brito JM. Transposition of the great arteries with ventricular septal defect: Surgical considerations concerning the Rastelli operation. *J Thorac Cardiovasc Surg* 1984;88:1004-11.
 11. Fujii Y, Kotani Y, Takagaki M, Arai S, Kasahara S, Otsuki S, et al. The impact of the length between the top of the interventricular septum and the aortic valve on the indications for a biventricular repair in patients with a transposition of the great arteries or a double outlet right ventricle. *Interact Cardiovasc Thorac Surg* 2010;10:900-5. <http://doi.org/d6gqbn>
 12. Navabi M., Shababian R, Kiani A, Rahimzadeh M. The effect of ventricular septal defect enlargement on the outcome of Rastelli or Rastelli-type repair. *J Thorac Cardiovasc Surg* 2009;138:390-6. <http://doi.org/dkr25q>
 13. Cleveland DC, Kirklin JK, Pavnica JW, Tresler MA, Kukreja M, Dabal RJ, et al. Late left ventricular outflow tract obstruction following the Rastelli operation: expectations out to 20 years. *World J Pediatr Congenit Heart Surg* 2016;7:605-10. <http://doi.org/b76v>
 14. Lecompte Y, Neveux JY, Leca F, Zannini L, Tu TV, Dubois Y, et al. Reconstruction of the pulmonary outflow tract without a prosthetic conduit. *J Thorac Cardiovasc Surg* 1982;84:727-33.
 15. Borromee L, Lecompte Y, Batisse A, Lemoine G, Vouhé P, Sakata R, et al. Anatomic repair of anomalies of ventriculoarterial connection associated with ventricular septal defect. II. Clinical results in 50 patients with pulmonary outflow tract obstruction. *J Thorac Cardiovasc Surg* 1988;95:96-102.
 16. Lim HG, Kim WH, Lee JR, Kim YJ. Twenty-five years' experience of modified Lecompte procedure for the anomalies of ventriculoarterial connection with ventricular septal defect and pulmonary stenosis. *J Thorac Cardiovasc Surg* 2014;148:825-31. <http://doi.org/f6f5cn>
 17. Weyand K, Haun C, Blaszczyk H, Goetz-Toussaint N, Photiadis J, Sinzobahamvy N, et al. Surgical treatment of transposition of great arteries with ventricular septal defect and left ventricular outflow tract obstruction: Midterm results. *World J Pediatr Congenit Heart Surgery* 2010;1:163. <http://doi.org/dr6wzq>
 18. Di Carlo D, Tomasco B, Cohen L, Vouhé P, Lecompte Y. Long-term results of the REV (réparation à l'étage ventriculaire) operation. *J Thorac Cardiovasc Surg* 2011;142:336-43. <http://doi.org/b6fkzk>
 19. Nikaidoh H. Aortic translocation and biventricular outflow tract reconstruction. A new surgical repair for transposition of the great arteries associated with ventricular septal defect and pulmonary stenosis. *J Thorac Cardiovasc Surg* 1984;88:365-72.
 20. Morell VO, Jacobs JP, Quintessenza JA. Aortic translocation in the management of transposition of the great arteries with ventricular septal defect and pulmonary stenosis: results and follow-up. *Ann Thorac Surg* 2005;79:2089-93. <http://doi.org/fbv6h>
 21. Morell V, Jacobs P, Quintessenza J. The role of aortic translocation in the management of complex transposition of the great arteries. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2004;7:80. <http://doi.org/dg24z7>
 22. Villalba C, Mouratian M, Lafuente MV, Irman J, Saad A, Barretta J, et al. The Modified Nikaidoh Procedure: Initial Experience and Short and Mid-term Outcomes. *Rev Argent Cardiol* 2014;82:493-9.
 23. Hu S, Xie Y, Li S, Wang X, Yan F, Li Y, et al. Double-root translocation for double-outlet right ventricle with noncommitted ventricular septal defect or double-outlet right ventricle with subpulmonary ventricular septal defect associated with pulmonary stenosis: an optimized solution. *Ann Thorac Surg* 2010;89:1360-5. <http://doi.org/bh87wm>
 24. Bautista Hernandez V, Marx G, Bacha E, del Nido P. Aortic root translocation plus arterial switch for transposition of the great arteries with left ventricular outflow tract obstruction. *J Am Coll Cardiol* 2007;49:485-90. <http://doi.org/c9c4jg>
 25. Hu SS, Li SJ, Li SJ, Liu ZG, Li Y, Wang L. The double-root translocation technique. *Operative Techniques in Thorac and Cardiovasc Surg* 2009;14:35-44. <http://doi.org/fr8htm>
 26. Sayin O, Ugurlucan L, Saltik L, Sungur Z, Tireli E. Modified Nikaidoh procedure for transposition of the great arteries, ventricular septal defect and left ventricular outflow tract obstruction. *Thorac Cardiovasc Surg* 2006;54:548-66. <http://doi.org/cr85g6>
 27. Yeh T, Ramaciotti C, Leonard SR, Roy L, Nikaidoh H. The aortic translocation (Nikaidoh) procedure: midterm results superior to Rastelli procedure. *J Thorac Cardiovasc Surg* 2007;133:461-9. <http://doi.org/ck7wnc>
 28. Raju V, Myers PO, Quinonez LG, Emani SM, Mayer JE Jr, Pigula FA, et al. Aortic root translocation (Nikaidoh procedure): Intermediate follow-up and impact of conduit type. *J Thorac Cardiovasc Surg* 2015;149:1349-55. <http://doi.org/f7b9jh>
 29. Valverde I, Gomez G, Gonzalez A, Suarez-Mejias C, Adsuar A, Coserria JF, et al. Three-dimensional patient-specific cardiac model for surgical planning in Nikaidoh procedure. *Cardiol Young* 2015;25:698-704. <http://doi.org/f65fd6>
 30. Sakamoto K, Ota N, Murata M, Tosaka Y, Ide Y, Tachi M, et al. Modified Nikaidoh procedure with double-root translocation in a 1-year-old boy. *Ann Thorac Surg* 2014;97:1055-7. <http://doi.org/b76w>
 31. Lee HP, Bang JH, Baek JS, Goo HW, Park JJ, Kim YH. Aortic root translocation with arterial switch for transposition of the great arteries or double outlet right ventricle with ventricular septal defect and pulmonary stenosis. *Korean J Thorac Cardiovasc Surg* 2016;49:190-4. <http://doi.org/b76x>
 32. Kramer P, Ovroutski S, Hetzer R, Hübner M, Berger F. Modified Nikaidoh procedure for the correction of complex forms of transposition of the great arteries with ventricular septal defect and left ventricular outflow tract obstruction: mid-term results. *Eur J Cardiothorac Surg* 2014;45:928-34. <http://doi.org/f5x2dz>
 33. Huhta JC, Edwards WD, Danielson GK, Feldt RH. Abnormalities of the tricuspid valve in complete transposition of the great arteries with ventricular septal defect. *J Thorac Cardiovasc Surg* 1982;83:569-76.
 34. Emani S, Beroukhir R, Zurakowski D, Pigula F, Mayer J, del Nido P, et al. Outcomes after repair for d-transposition of the great arteries with left ventricular outflow tract obstruction. *Circulation* 2009;120(11 Suppl):S53-8.
 35. Bierbach B, Arenz C, Suchowskyj P, Schroth S, Blaszczyk J, Asfour B, et al. Current mid-term outcome with an integrated surgical strategy for correction of d-transposition of the great arteries with ventricular septal defect and left ventricular outflow tract obstruction. *Eur J Cardiothorac Surg* 2016;50:617-25. <http://doi.org/b76z>
 36. Al-Jughiman MK, Al-Omar MA, Van Arsdell GS, Morell VO, Jacobs ML. D-Transposition of the Great Arteries with Ventricular Septal Defect and Left Ventricular Outflow Tract Obstruction (D-TGA/VSD/LVOTO): A survey of perceptions, preferences, and experience. *Pediatr Cardiol* 2015;36:896-905. <http://doi.org/b762>
 37. Prêtre R. Editorial comment: Arterial switch, reparation a l'étage ventriculaire, Rastelli or Nikaidoh? *Eur J Cardiothorac Surg* 2013;44:1094-5. <http://doi.org/b763>
 38. Lecompte Y. Letters to the Editor: Rastelli repair for transposition of the great arteries: Still the best choice? *J Thorac Cardiovasc Surg* 2002;123:192-3.
 39. Baumgartner H, Hung J, Bermejo J, Chambers J, Evangelista A, Griffin B, et al. Echocardiographic assessment of valve stenosis: EAE/ASE recommendations for clinical practice. *Eur J Echocardiogr* 2009;10:1-25. <http://doi.org/bkps2r>
 40. Lancellotti P, Tribouilloy C, Hagendorff A, Moura L, Popescu BA, Agricola E, et al. European Association of Echocardiography. European Association of Echocardiography recommendations for the assessment of valvular regurgitation. Part 1: aortic and pulmonary regurgitation (native valve disease). *Eur J Echocardiogr* 2010;11:223-44. <http://doi.org/dj2fsx>