CHD

Pulmonary root translocation in malposition of great arteries repair allows right ventricular outflow tract growth

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Objective: Optimal surgical treatment of patients with transposition of the great arteries (TGA), ventricular septal defect (VSD), and pulmonary stenosis (PS) remains a matter of debate. This study evaluated the clinical outcome and right ventricle outflow tract performance in the long-term follow-up of patients subjected to pulmonary root translocation (PRT) as part of their surgical repair.

Methods: From April 1994 to December 2010, we operated on 44 consecutive patients (median age, 11 months). All had malposition of the great arteries as follows: TGA with VSD and PS (n = 33); double-outlet right ventricle with subpulmonary VSD (n = 7); double-outlet right ventricle with atrioventricular septal defect (n = 1); and congenitally corrected TGA with VSD and PS (n = 3). The surgical technique consisted of PRT from the left ventricle to the right ventricle after construction of an intraventricular tunnel that diverted blood flow from the left ventricle to the aorta.

Results: The mean follow-up time was 72 ± 52.1 months. There were 3 (6.8%) early deaths and 1 (2.3%) late death. Kaplan-Meier survival was 92.8% and reintervention-free survival was 82.9% at 12 years. Repeat echocardiographic data showed nonlinear growth of the pulmonary root and good performance of the valve at 10 years. Only 4 patients required reinterventions owing to right ventricular outflow tract problems.

Conclusions: PRT is a good surgical alternative for treatment of patients with TGA complexes, VSD, and PS, with acceptable operative risk, high long-term survivals, and few reinterventions. Most patients had adequate pulmonary root growth and performance. (J Thorac Cardiovasc Surg 2012;143:1292-8)

Transposition of the great arteries (TGA) with ventricular septal defect (VSD) and left ventricular outflow tract (LVOT) obstruction has a prevalence of 20% in the subgroup of neonates with TGA and associated VSD.[1] Although this heart malformation is relatively rare, there is much debate regarding the best procedure for its treatment. Since 1969, several surgical techniques have been developed and used for anatomic repair of this defect: the Rastelli operation, published in 1969[2]; the REV (réparation a l'étage ventriculaire) or Lecompte procedure (1981)[3]; aortic translocation, first used by Nikaidoh[4] (1984), who revived an idea proposed by Bex, Lecompte, and Baillot[5] (1980) for TGA with intact ventricular septum; and the Metras procedure[6] (1997), which is a modification of the Lecompte operation. All of these techniques have some limitations, mainly related to patch augmentation or conduit insertion in the right ventricular outflow tract (RVOT).

With the aim of overcoming these limitations and assuring pulmonary valve competency, since April 1994 we have used pulmonary root translocation (PRT) as an alternative surgical approach for TGA with VSD and pulmonary stenosis (PS) and for selected cases of double-outlet right ventricle with subpulmonary VSD.[7] More recently, we extended its use as part of the double switch procedure in patients with congenitally corrected TGA, PS, and VSD.[8]

The objective of this study was to evaluate the clinical outcome and the RVOT performance in the long-term follow-up of patients after PRT.

PATIENTS AND METHODS

Patients

We reviewed the prospectively collected clinical and echocardiographic data of 44 consecutive pediatric patients who underwent PRT as part of a procedure to repair their complex congenital heart disease from April 1994 to December 2010. The included patients had malposition of the great arteries as follows: TGA with VSD and PS (n = 33); double-outlet right ventricle with subpulmonary VSD (n = 7); double-outlet right ventricle with atrioventricular septal defect (n = 1); and congenitally corrected TGA with VSD and PS (n = 3). The mean patient age was 24 ± 36 months, with a median age of 11 months (range, 1 month to 11 years). Twenty-eight patients were male and 16 were female. Ten (13.6%) patients had previous surgical procedures: systemic–pulmonary shunt had been performed in

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Abbreviations and Acronyms

LVOT = left ventricular outflow tract
PRT = pulmonary root translocation
PS = pulmonary stenosis
REV = réparation a l'étage ventriculaire
RVOT = right ventricular outflow tract
TGA = transposition of the great arteries
VSD = ventricular septal defect

6 patients and pulmonary banding in 4 patients. This study was approved by the Ethics Committee at the Hospital Beneficencia Portuguesa de São Paulo (CAE-0023.0.360.000-07), and informed consent was obtained from the patients' parents.

Surgical Technique

We[8] previously reported the surgical details of PRT for different anatomic variations of TGA. Figure 1 depicts the main steps for repairing TGA, VSD, and PS. The intact pulmonary root with valve was translocated to the left side of the aorta for connection to the right ventricle without a Lecompte maneuver. The VSD was routinely enlarged by resection of the conal septum except in 6 patients with a large VSD extending to the conal septum. Commissurotomy and probe dilation of the pulmonary valve was used in borderline-sized pulmonary valves that we judged not to require patch enlargement. At least 40% of the pulmonary root circumference was sutured directly to the right ventricular wall. In 3 patients with a diagnosis of congenitally corrected TGA with VSD and PS, the Senning procedure, atrial switch repair, was added on.

Regarding the enlargement of the pulmonary root, we adopted a strategy that was similar to that described for the repair of tetralogy of Fallot.[9] We enlarged the pulmonary annulus when the pulmonary annulus Z-score was less than –3, performing the intraoperative measurement with a Hegar dilator, or, when the postrepair systolic gradient exceeded 35 mm Hg, using catheter measurements after cardiopulmonary bypass. In the latter situation, we put the patient back on the pump, removed 1 or 2 interrupted sutures, opened the pulmonary root at the commissural site, and performed a small enlargement with a bovine pericardial valved patch.

Echocardiographic Studies

Each patient was subjected to an echocardiographic study protocol consisting of a preoperative study, an early postoperative study, and follow-up studies scheduled either annually or according to the patient's needs. The diameters of the aortic, pulmonary, tricuspid, and mitral annuli were measured, as was the peak velocity of the right and left ventricular outlets. The degree of pulmonary insufficiency and right ventricular function were determined qualitatively by the pediatric echocardiographer. The diameter measurements were expressed in millimeters. The left ventricular function was evaluated using ejection fraction calculated by the Teicholz method. Insufficiency of the pulmonary and aortic valves was scored as absent, mild, moderate, or severe. The RVOT and LVOT were evaluated by measuring the peak pressure gradients across the pulmonary and aortic valves.

Clinical Follow-up

The patients were followed up after the operation, and the patient's local physician was contacted in case of death. A final follow-up of all patients was conducted from January to March 2011 by telephone or by outpatient evaluation. The functional status according to the New York Heart Association class and the use of medicines to treat heart failure and/or arrhythmias were recorded.

Data Analysis

Descriptive data for continuous variables are presented as mean ± standard deviation or as median with range; categorical variables are presented as relative frequencies. The outcome variables were defined as time from the PRT procedure to events (death, reoperation for RVOT obstruction, LVOT obstruction, residual VSD, or pulmonary balloon dilatation procedure) and recent clinical status. The probability of survival and event-free survival was estimated according to the Kaplan-Meier method and included hospital mortality. Pulmonary valve diameter and gradient changes over time were analyzed using longitudinal data analysis. The exact time of the assessment and all available data were used in these analyses. The χ^2 method was applied to compare categorical variables. Analyses were performed using SPSS software version 17.0 (SPSS, Inc, Chicago, Ill)

RESULTS

There were 3 (6.8%) hospital deaths. The first patient was an 8-year-old boy with a long history of cyanosis who died of heart failure, and the second patient was a 6-month-old boy who had a good hemodynamic result after the operation but died of gram-negative sepsis. The third patient was a 36-day-old baby who underwent surgery 1 day after being intubated and transfused owing to severe hypoxemia. He could not be weaned from cardiopulmonary bypass and died of multiple organ failure despite the use of cardiopulmonary support with extracorporeal membrane oxygenation for 3 days. All 3 deaths occurred in patients with TGA, VSD, and PS who were operated on between 1994 and 2005. There were no hospital deaths or late deaths in the 26 subsequent patients.

The mean aortic crossclamp time was 140 ± 34 minutes, and the mean cardiopulmonary bypass time was 195 ± 42 minutes. The median stay in the intensive care unit was 13 days (range, 2-61 days), and the median hospital stay was 24 days (range, 12-115 days). Atrioventricular block occurred in 2 patients, necessitating permanent pacing.

Longitudinal Clinical Outcome

Long-term follow-up information was obtained for all patients. The mean follow-up time was 72 ± 52.1 months. The survival and reintervention-free survival curves, which include the hospital deaths, showed a 92.8% ± 3.8% survival at 1, 5, 10, and 12 years of follow-up, an 88% ± 4.9% reintervention-free survival at 1 year, and an 82.9% ± 5.9% survival at 2, 5, 10, and 12 years of follow-up (Figure 2). One patient who had progressive postoperative RVOT obstruction died suddenly 72 days after the operation. The follow-up showed that 7 patients needed a total of 8 reinterventions after the PRT. These reinterventions were required owing to residual VSD in 2 patients, LVOT obstruction (left ventricular–aortic tunnel stenosis) in 1 patient, RVOT aneurysm in 2 patients (1 owing to endocarditis and 1 owing to in situ pericardial flap dilation), and RVOT obstruction (pulmonary stenosis) in 2 patients. The reoperations for residual VSD closure and resection of muscular obstruction in the LVOT occurred in the initial 6 months