The Ross procedure in patients aged less than 18 years: The midterm results

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Objective: This study reviews a single-center experience with the Ross procedure in infants and young children.

Methods: From November 1993 to March 2012, 55 children aged less than 17 years underwent a Ross procedure. The patients ranged in age from 2 days to 17 years (median, 5.9 years). Thirteen patients were infants, and 18 patients were preschool children. The predominant indication for the Ross procedure was aortic stenosis. Twenty-seven patients (49%) with left ventricular outflow tract obstruction underwent a modified Ross–Konno procedure. Twenty-five patients (45%) had undergone 40 previous cardiac procedures. Preoperatively, 3 patients showed severe left ventricular dysfunction, with 2 of the patients requiring intubation and inotropic support. Concomitant procedures were performed in 11 patients (20%). Nine patients underwent mitral valve surgery, and 2 patients underwent subaortic membrane resection.

Results: Patients were followed up for a median of 66 months (range, 3 months to 17 years). Overall survival at 1, 2, 5, and 10 years was 84.9%. Hospital mortality rate was 13% (7/55 patients). All deaths occurred in neonates or infants, except 1 who was aged less than 4 years. Freedom from reoperation for autograft failure was 100% at 1 year, 96.7% at 5 years, and 73.7% at 10 years. During follow-up, 7 patients required a reoperation on the autograft for dilatation and severe aortic insufficiency. Freedom from reoperation for the right ventricular outflow tract replacement was 56.1% at 10 years.

Conclusions: The low rate of autograft failure demonstrates that the Ross procedure is an attractive option for the management of aortic valve disease and complex left ventricular outflow tract obstruction in the pediatric population. However, alternative options must be considered in adolescents and young adults. (J Thorac Cardiovasc Surg 2014;147:383-8)

The optimal operative management of aortic valve disease in children and young adults remains controversial. The Ross operation, first described more than 40 years ago, is aimed at the treatment of aortic valve disease and based on autograft transplantation of the native pulmonary valve into the aortic position and reconstruction of the pulmonary outflow tract with an homograft.[1] The Ross operation allows growth of the autograft and avoidance of anticoagulation therapy.[2] However, because the need for multiple reinterventions for the replacement of the right ventricular outflow tract (RVOT) substitute and neoaortic root dilatation leading to aortic regurgitation have been reported,[3-5] one could argue that the Ross procedure might turn a single-valve disease into a double-valve pathology.[6]

The goal of this study was to report a single-institution experience with the Ross operation in neonates, infants, children, and young adults and to describe the incidence of pulmonary autograft dilation and insufficiency, as well as the need for reintervention on the autograft or homograft.

MATERIALS AND METHODS

The Committee on Clinical Investigation at Bambino Gesú Children's Hospital approved the study and waived the requirement for written informed consent.

Subjects

Between November 1993 and March 2012, all patients aged less than 18 years who underwent a Ross procedure at the Bambino Gesù Children's Hospital were included. All patients underwent surgery with the standard technique of cardiopulmonary bypass with bicaval cannulation and moderate hypothermia. Myocardial protection was achieved by intermittent infusion of blood cardioplegia.

A standard technique of complete root replacement with coronary transfer was used for the Ross procedure. No pledgets were used. In case of left ventricular outflow tract (LVOT) obstruction, a modified Ross–Konno procedure was performed. The ventriculoplasty incision was patched by an extension of attached infundibular free wall muscle harvested with the autograft. A pulmonary/aortic homograft or a heterograft was used for RVOT reconstruction.

The primary outcome measure was the need for late reintervention. The secondary outcome variables were represented by 28-day mortality and late survival, late right ventricle to pulmonary artery (RV-PA) conduit, and autograft failure. For the purpose of this analysis, RV-PA failure was defined as the presence of insufficiency or stenosis of the conduit with a mean gradient of at least 30 mm Hg and right ventricular pressure at least 2/3 to

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Abbreviations and Acronyms
CI = confidence interval
LVOT = left ventricular outflow tract
RVOT = right ventricular outflow tract
RV-PA = right ventricle to pulmonary artery

respect the systemic arterial pressure; autograft failure was defined as neoaortic root dilatation with z score changes from hospital discharge to follow-up or changes in neoaortic valve insufficiency grade from discharge to follow-up.

The immediate postoperative result was assessed by transesophageal echocardiography in all patients after surgery. Further echocardiography studies were obtained preoperatively, postoperatively, and annually thereafter. Autograft valve function, homograft or RVOT conduit valve function, and left ventricular function were assessed by M-mode, 2-dimensional echocardiography and color flow Doppler.

Statistical Analysis

STATA version 11.1 data analysis and statistical software (StataCorp LP, College Station, Tex) was used for statistical analysis. Continuous variables are reported as median (range), and categoric variables are reported as number (proportion). The t test and Wilcoxon rank-sum test were used for continuous variables as appropriate; the chi-square and Fisher exact tests were used for categoric variables as appropriate. Survival curves for freedom from RV-PA, autograft failure, and late survival were obtained by use of the Kaplan–Meier method, and comparisons were performed with the log-rank test. The continuous variables examined included age and weight at treatment, which were dichotomized at a cutoff of 1 year and 10 kg, respectively, to explore the effect of younger age and low body weight on outcomes. Additional multivariate survival analyses for the separate end points of RV-PA and autograft failure or late survival were performed by means of Cox proportional hazards multiple regression models. The selection of independent variables for the model was based on statistical significance in univariable testing. All comparisons were 2-sided.

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RESULTS

A total of 55 patients underwent the Ross operation at the Ospedale Pediatrico Bambino Gesù during the study period. Demographic and selected operative characteristics for these patients are summarized in Table 1. Median age and weight at surgery were 5.9 years (range, 2 days-17 years) and 18 kg (range, 3.5-110), respectively. Follow-up information was available for 74% of the 55 patients. Median follow-up time was 66 months (range, 3 months to 17 years).

The preoperative hemodynamic indication for the Ross procedure was represented by a combination of aortic stenosis and regurgitation for 31 patients (56%). In 27 patients (51%) with LVOT obstruction, a modified Ross–Konno procedure was performed. In 2 patients with Shone's anomaly, concomitant mitral valve surgery was required.

Twenty-five patients (47%) had undergone a total of 40 previous cardiac procedures. These procedures are listed in Table 1. Three patients (5.5%) showed severe preoperative left ventricular dysfunction; of those, 2 patients required preoperative mechanical ventilation and inotropic support.

Whenever possible, a pulmonary homograft (31 patients, 51%) was preferentially used to reconstruct the RVOT. An aortic homograft was used in 3 patients (6%), and a heterograft was used in 23 patients (42%). The median diameter of the replaced pulmonary valve was 19 mm (range, 10-26 mm).

Concomitant procedures were performed in 11 patients. A concurrent mitral valve surgery was performed in 9 patients (17%); of those, 6 patients underwent mitral repair and 3 patients underwent valve replacement. The remaining 2 patients underwent subaortic membrane resection. Two patients who underwent the Konno–Ross operation required insertion of a permanent pacemaker for a complete atrioventricular block.

Twenty-Eight–Day Mortality and Late Survival

Seven patients (13%) died within 28 days from surgery. Three patients were neonates, 3 patients were infants, and 1 patient was a preschool child. Of those patients, 1 showed signs of aortic valve endocarditis with severe insufficiency and died of multiple organ failure. Six patients underwent a Ross–Konno procedure, with concomitant mitral valve surgery in 2. Two neonates with critical aortic valve stenosis underwent urgent Ross operation for severe aortic insufficiency after balloon valvuloplasty.

At the univariate level, age less than 1 year, weight less than 10 kg at surgery, and longer cardiopulmonary bypass time were associated with 28-day mortality (Table 2). Because the number of events was low, it was not possible to seek an independent association between demographic variables and 28-day mortality by using a multivariable logistic regression model.

Overall survival at 1, 2, 5, and 10 years was 84.9% (range, 72.1%-92.2%) (Figure 1). We were not able to identify any independent association between perioperative characteristics and late survival (Table 3).

Late Reintervention

Overall freedom from all-cause reoperation was 97.6% (range, 84.6%-99.7%), 86.7% (range, 70.7%-94.3%), and 48.1% (range, 27.2%-66.2%) at 1, 5, and 10 years, respectively (Figure 2).

Reoperations on the Autograft

Overall freedom from reoperation for autograft failure was 100%, 96.7% (range, 78.6%-99.5%), and 73.7% (range, 47.2%-88.4%) at 1, 5, and 10 years, respectively. Six patients (11%) required reoperation on the autograft; the main mechanism was represented by severe aortic insufficiency in 5 patients and aortic root dilatation in 1 patient. One patient required simultaneous reoperation of both the