Ross operation early and mid-term results in children and young adults

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Abstract

Objective: The Ross procedure has been cited as the procedure of choice for young patients requiring aortic valve replacement. However, potential for reintervention requirement in both left and right ventricular outflow tracts can be a source of concern. The aim of the present study was to describe our experience with this procedure.

Methods: A retrospective chart review of all the patients who underwent the Ross procedure in a single institution was performed. National death registry records were used for late mortality.

Results: Eighteen Ross procedures between May 2003 and May 2018 were performed. The median age of the cohort was 15 [interquartile range (IQR): 12-18] years. The pulmonic conduit was a homograft in 11 patients, Labcor in 5 patients, Contegra in 1 patient, and Medtronic Freestyle Valve in 1 patient. There were three early deaths. The median follow-up of 15 hospital survivors was 11 (IQR: 3–14) years. Any late mortality was not observed. In the two surviving patients with infective endocarditis, there was no recurrent infective endocarditis. Freedom from reintervention was 80% at 8 years and onward. Any risk factors associated with reintervention could not be identified. However, freedom from autograft dilatation at 10 years was 45%.

Conclusion: Autograft failure is a potential problem in the long-term follow-up of Ross patients. Freedom from reintervention was satisfactory, and the type of pulmonic conduit did not affect the mid-term outcomes. In patients with infective endocarditis, the Ross procedure has a low recurrence rate, but it might have an increased risk of mortality. *(Anatol J Cardiol 2019; 22: 21-5)*

Keywords: congenital, Ross-Konno, left ventricular outflow tract obstruction, autograft, aortic valve stenosis

Introduction

Replacement of the aortic valve using a pulmonary autograft was first reported by Donald Ross in 1967 (1). The Ross procedure has many advantages in young patients who require aortic valve replacement (AVR). After the Ross procedure, the patient does not require anticoagulation, and the diameter increase of the autograft over time can match the somatic growth in pediatric patients. There have been reports favoring the Ross procedure as the procedure of choice in young patients (2, 3). However, other reports have held a more cautious perspective (4, 5). A recent review comparing Ross, mechanical AVR, and homograft implantation concluded that all the valve substitutes had suboptimal results (6).

In the Ross procedure, the aortic valve is replaced by the patient's own pulmonary valve. Right ventricular outflow tract (RVOT) in turn is reconstructed using a right ventricle to pulmonary artery (RV–PA) conduit. On the left ventricular outflow tract (LVOT), a major concern is dilatation of the autograft leading eventually to aortic insufficiency. The necessity of using an RVOT conduit is unique to the Ross procedure among various methods of AVR. The conduit becomes stenotic over time due to the patient's growth or calcification. As a result, the potential for reintervention requirement in both LVOT and RVOT can be a source of concern. The Ross procedure has been performed since 2003 in children and young adults. The aim of the present study was to describe our experience of the Ross procedure in a single center.

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Methods

A retrospective chart review of all the patients who underwent the Ross procedure in a single institution was performed. The operations were performed through a standard median sternotomy under cardiopulmonary bypass (CPB). Repeated doses of blood cardioplegia were used. After cardioplegic arrest, the aorta and the pulmonary artery were transected. Buttons of the coronary ostia were harvested. The aortic leaflets were excised. The pulmonary root was harvested as an autograft. The autograft was implanted into the LVOT with running sutures. In cases with annular stenosis, the Ross–Konno procedure was performed by anterior enlargement of the aortic root. Autograft was implanted to the LVOT, and the coronary buttons were transferred. After the aortic anastomosis, the aortic clamp was removed. RVOT was reconstructed using either a homograft or a heterograft based on availability.

The patients were followed up with routine echocardiographic examinations. Postoperative follow-up data were collected from the follow-up echocardiography and cardiac catheterization data. In patients who did not have any hospital records within the last year, mortality status was checked through national death registry records.

Statistical analysis was performed using IBM SPSS Statistics software version 21 (SPSS Inc., Chicago, IL, USA). Categorical variables were expressed as number (percentage). Continuous variables were represented as mean (standard deviation) or median [interquartile range (IQR)], as appropriate. Kaplan–Meier was used for survival analysis. Chi-square or Fisher's exact test was used for intergroup comparisons of categorical variables. Mann-Whitney U test was used for continuous variables. A p value of 0.05 was accepted as statistically significant.

Results

A total of 18 Ross procedures between May 2003 and May 2018 were performed. The median age of the cohort was 15 (IQR: 12–18) years. The study included 8 male patients. The indication for the operation was aortic stenosis in 12 patients, infective endocarditis in 3 patients, aortic insufficiency in 1 patient, and mechanical valve dysfunction in 2 patients. There were 3 patients who were previously operated, 2 with AVR and 1 with subaortic ridge resection. Additionally, 5 patients had a history of aortic balloon valvuloplasty. Out of 16 patients with native aortic valves, aortic valve was bicuspid in 7 patients and tricuspid in 9 patients.

There were 10 Ross-Konno and 8 isolated Ross procedures. The RV–PA conduit was a homograft in 11 patients, Labcor in 5 patients, Contegra in 1 patient, and Medtronic Freestyle Valve in 1 patient.

The median length of stay in the hospital was 11 (IQR: 5–16) days. Atrioventricular block was not observed in our cohort. There were three early deaths. Any factors associated with mortality could not be identified. The first mortality recorded was from a 6-year-old male patient. He was under treatment for acute lymphoblastic leukemia in another center when he was diagnosed with infective endocarditis. He had a 19×14 mm vegetation on the aortic valve and aortic stenosis with a peak gradient of 40 mm Hg. He also had multiple brain abscesses refractory to medical treatment and surgical drainage. He underwent the Ross procedure and was extubated on postoperative day 1. However, at the night of postoperative day 1, he developed sudden cardiac arrest. Resuscitation was unsuccessful.

The second mortality was that of an 11-year-old female patient. On preoperative echocardiography, she had a tunnel type subaortic stenosis with a peak systolic gradient of 134 mm Hg. Aortic annulus was 9 mm (z: –4.70). There was severe left ventricular hypertrophy. Additionally, there was mitral stenosis with a mean gradient of 12 mm Hg. In the operation, mitral valvuloplasty was performed in addition to the Ross–Konno procedure. The mitral valve was exposed through a transseptal incision. The supramitral membrane was resected, and the papillary muscles were divided. The posterior leaflet was augmented using a pericardial patch. After the CPB, the left atrial pressure was increased, and patch augmentation of the anterior mitral valve was performed with a second period of cardioplegic arrest. She was followed up under mechanical ventilation. On postoperative day 1, she had a sudden cardiac arrest. She was resuscitated and placed on extracorporeal membrane oxygenation (ECMO) support. She died on postoperative day 3 due to multiorgan failure on ECMO support.

The third mortality was that of a 13-year-old male patient. He had a bicuspid aortic valve with a valvular peak systolic gradient of 80 mm Hg. He had a history of aortic balloon valvuloplasty at the age of 3 and another one at the age of 6. He underwent a standard Ross–Konno procedure. He was weaned off mechanical ventilation on the day of the operation. On postoperative day 3, he had acute respiratory distress, followed by cardiopulmonary arrest. He was resuscitated and placed on ECMO support. He died on postoperative day 4 due to multiorgan failure under ECMO support.

The median follow-up of 15 hospital survivors was 11 (IQR: 3–14) years. No late mortality was observed. In the two surviving patients with infective endocarditis, there was no recurrent infective endocarditis for the 3 years and 5 years, respectively, they were followed up. One patient underwent balloon angioplasty of the RV–PA conduit stenosis 3 years after the operation, and one patient underwent balloon angioplasty of both the RV–PA conduit and autograft anastomosis at the LVOT 8 years after the operation. Freedom from reintervention was 80% at 8 years and onward (Fig. 1). No risk factors associated with RVOT or LVOT reintervention could be identified.

There were eight Ross–Konno patients who were hospital survivors. The median preoperative, early postoperative, and late postoperative LVOT gradients of these patients were 110 (IQR: 95–154) mm Hg, 0 (IQR: 0–7) mm Hg, and 5 (IQR: 0–13) mm Hg, respectively. There was a significant difference between the

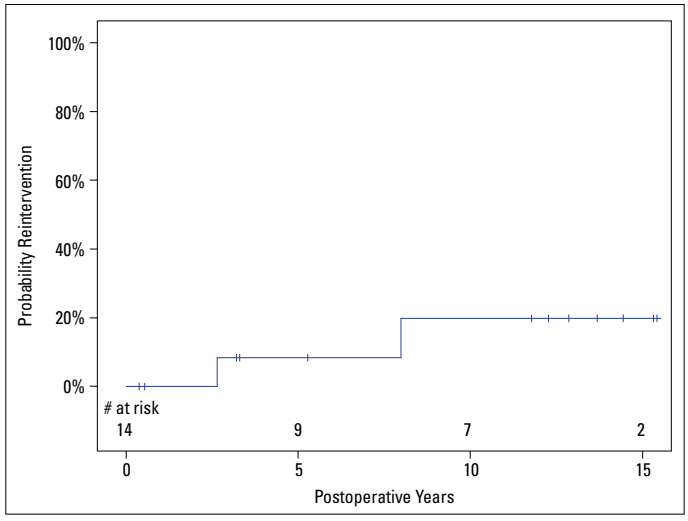

Figure 1. Kaplan–Meier curve for the probability of reintervention (both left and right ventricular outflow tracts) of the cohort

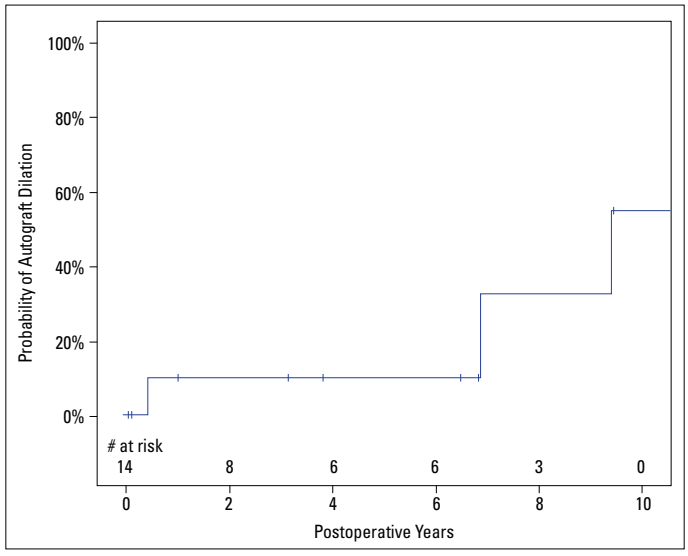

Figure 2. Kaplan–Meier curve for the probability of autograft dilation of the cohort

Table 1. Insufficiency of the aortic and the conduit valve data at the latest echocardiographic follow-up

	PI	AI
None	6	1
Trivial	1	5
Mild	4	6
Mild–moderate	2	2
Moderate	2	1

AI - aortic insufficiency; PI - insufficiency at the right ventricle to pulmonary artery conduit valve

preoperative and early postoperative LVOT gradients of the patients (p=0.02). There was no significant difference between the early and late postoperative gradients.

On the latest echocardiographic examination, the median right ventricle outflow tract gradient was 26 (IQR: 21–30) mm Hg. The degree of insufficiency at the aortic and RV–PA conduit valves at the latest echocardiographic follow-up is shown in Table 1. There were two patients with moderate pulmonary insufficiency and one patient with moderate aortic insufficiency. Other patients had less than moderate valvular insufficiency. There were no patients who required autograft intervention for autograft failure within the study period. However, freedom from autograft dilatation at 10 years was 45% (Fig. 2).

Discussion

The outcomes of the Ross procedure in a single center were described. Our cohort mostly consisted of patients who required aortic root enlargement and who had infective endocarditis. The Ross–Konno procedure effectively reduced the LVOT gradient in this cohort. Other patients who underwent the isolated Ross procedure due to non-infective indications were all hospital survivors. Early mortality was most commonly observed in patients with comorbidities. On follow-up, freedom from reintervention was 80% after 8 years. The present study could not demonstrate any effect of the type of RV–PA conduit on the freedom from reintervention. Although there were no reoperations for autograft failure, more than half of the patients showed signs of autograft dilatation at 10 years.

In our cohort, the most common indication was aortic stenosis in accordance with the literature (7-9). In patients with LVOT obstruction who required AVR, the Ross–Konno procedure was our procedure of choice. Our results showed that the reduction in LVOT gradient was significant after surgery. In the long-term follow-up of these patients, the reduction in LVOT gradient was durable. On the other hand, autograft failure is a potential problem after the Ross procedure. In this cohort, there were no reoperations for autograft failure within the study period. However, more than half of the cohort had aortic root dilatation at 10 years. Reoperations for autograft failure have been reported to be between 81% and 89% (10, 11) at 10 years postoperatively. Although in the adult population various techniques for autograft support using prosthetic material have been reported (12-14), in small children, fixing the small aortic root and limiting the growth potential are not desirable. Inclusion cylinder technique and its modifications (15, 16) can be promising in this age group as they offer autologous support for the autograft.

The longevity of the RV–PA conduit is an important issue in patients after the Ross procedure. In our cohort, there were two patients who required reintervention for conduit stenosis. Freedom from reintervention at 8 years and onward was 80%. These values agree with the previous studies (17, 18). No association between the type of RV–PA conduit and freedom from reintervention could be demonstrated. In adult Ross procedures, homografts have shown excellent long-term performance (17). However, there is no consensus in the literature whether the

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choice of the RV–PA conduit affects the reintervention rate in the pediatric population. Although homografts have been cited as the conduit of choice by many studies (17, 19), other studies, similar to our findings, found no association between the RV–PA conduit choice and RVOT reintervention rate (7, 18). Homografts have been used in approximately half of the patients in our cohort. Although our approach is to use homografts in the Ross procedure, where available, we do not consider the lack of availability of the homografts as a contraindication for the Ross procedure in the pediatric population.

In our cohort, 3 out of 18 patients underwent the Ross procedure due to infective endocarditis. Low recurrence rates have been reported with the Ross procedure in adult patients with infective endocarditis (20, 21). However, data on pediatric patients with infective endocarditis are limited. Two of the hospital survivors had infective endocarditis. These patients were under follow-up, one for 3 years and the other one for 5 years, and they did not have recurrent infective endocarditis. This finding is similar to the previous studies (20, 21). One of the three patients with infective endocarditis died. This patient had a concomitant brain abscess. He was extubated on postoperative day 1 but had a sudden cardiac arrest at the night of the same day. The cause of this sudden cardiac arrest could not be definitively identified. Infective endocarditis has been previously reported as a risk factor for mortality in Ross patients (22). Our hypothesis is although the recurrence rate is low, caution is advisable when selecting patients with infective endocarditis for the Ross procedure.

The mortality rate of the Ross procedure in children has been reported to be between 0.83% and 13% (6, 19). When only neonates and infants are taken into consideration, these values become 11.76% and 21.62% (6). Our cohort has a higher early mortality rate of 17%. The higher rate of mortality of this cohort can be attributed to patient selection. Our cohort involves patients with infective endocarditis and patients who require additional interventions. Owing to the small sample size of the cohort, no risk factors associated with mortality could be identified. However, some inferences can be drawn from the review of the clinical courses of three patients who died in the hospital. The first mortality is of the patient with infective endocarditis mentioned above. The second mortality was of a patient who underwent concomitant mitral valve surgery. Owing to the failure of mitral valve repair at the first attempt, a second period of aortic cross-clamp was required. We can speculate that additional intracardiac repair that leads to increased ischemic and CPB times during the Ross procedure, which already involves the manipulation of aortic and pulmonary valves, has been detrimental for the patient. After this experience, we decided not to perform the Ross procedure in patients who require additional intracardiac interventions. Our current strategy is to perform the Ross/Ross–Konno procedure as an isolated procedure. The third mortality was related to acute respiratory distress on postoperative day 3. This patient had a normal echocardiographic examination in the early postoperative period. The sudden onset of cardiac failure common to all three deaths of this cohort implies arrythmia as the cause of death.

Study limitations

Our study has limitations. The present study is limited by its retrospective nature. The number of cases is limited, and we could not identify any risk factors associated with mortality or reintervention rates. The median follow-up of 11 years can limit the evaluation of the reintervention rates at both RVOT and LVOT.

Conclusion

The reduction in LVOT gradient was effective and durable in this cohort. However, autograft failure is a potential problem in the long-term follow-up with more than half of the patients having echocardiographically demonstrated aortic root dilatation in 10 years. Freedom from reintervention was satisfactory after the Ross procedure, and the type of RV–PA conduit did not affect the mid-term outcomes. In patients with infective endocarditis, the Ross procedure has a low recurrence rate, but it might have an increased risk of mortality. Mortality was seen in patients with concomitant morbidities; therefore, our current approach is to avoid the Ross procedure in such patients if possible.

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