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Long-term outcomes of Ross procedure in pediatric patients: A single-center experience

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ABSTRACT

Objectives: This study aimed to share a single-center experience with the Ross procedure in pediatric patients and to demonstrate that the need for reoperations due to autograft failure can be minimized with appropriate surgical technique.

Patients and methods: This single-center retrospective study analyzed 18 pediatric patients (16 males, 2 females; median age: 3 years; range, 4 months to 8 years) who underwent the Ross operation between January 2002 and January 2024. Surgical techniques, patient demographics, and outcomes were assessed. Statistical analysis was conducted using the Kaplan-Meier method for overall survival and freedom from reoperation.

Results: Early mortality was 5.5%, primarily due to sepsis and multiple organ failure. The 10-year survival rate was 94%, demonstrating favorable long-term outcomes. Two (11.1%) patients required reoperation for conduit dysfunction, with a 15-year freedom from conduit reintervention rate of 77%.

Conclusion: The Ross procedure in pediatric patients showed low mortality and morbidity rates, with excellent long-term survival outcomes. Effective surgical techniques and modifications were crucial in preventing autograft dysfunction and reducing reoperation rates. The study challenges the notion that the Ross procedure leads to the development of two valve diseases, emphasizing its significance as a preferred option for aortic valve replacement in children.

Keywords: Aortic valve disease, autograft growth, conduit dysfunction, pediatric cardiology, Ross procedure.

Children with severe aortic valve disease often require surgical intervention to restore valve function and prevent complications. Available surgical options include aortic valve repair, mechanical or biological valve replacement, and the Ross procedure. The Ross procedure involves replacing the diseased aortic valve with the patient's own pulmonary valve, while a valved conduit is used to reconstruct the right ventricular outflow tract (RVOT).[1] This technique has gained popularity in pediatric and adolescent patients due to its superior hemodynamic profile, elimination of the need for long-term anticoagulation, and the ability of the autograft to grow with the patient, potentially reducing the need for future reoperations.[2]

Another emerging alternative in pediatric aortic valve surgery is the Ozaki procedure, which involves the reconstruction of the aortic valve using autologous pericardium tailored to create new leaflets. The Ozaki technique offers advantages, such as avoiding prosthetic material and preserving native aortic root geometry. However, its long-term durability in children remains under investigation, and it may not provide the same growth potential as the Ross procedure.[3]

Despite these advantages, concerns persist regarding the potential for autograft insufficiency and degeneration of the conduit used for RVOT reconstruction over time, particularly in younger patients. This has led to the perception that the

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Ross procedure may "convert one valve disease into two valve diseases."[4]

Nevertheless, it is important to contextualize this view. While conduit reintervention may be required, the overall outcomes of the Ross procedure, including freedom from autograft dysfunction and excellent long-term survival, have challenged this perception.[5] This study aimed to share our single-center experience with the Ross procedure in pediatric patients and to demonstrate that with appropriate surgical technique, the need for reoperations due to autograft failure can be minimized.

PATIENTS AND METHODS

This retrospective study included 18 pediatric patients (16 males, 2 females; median age: 3 years; range, 4 months to 8 years) who underwent the Ross operation at the Ege University Faculty of Medicine Department of Pediatric Cardiovascular Surgery between January 2002 and January 2024. Data were obtained from hospital records, national medical databases, and telephone interviews with patients followed up at external centers. Inclusion criteria were as follows: *(i)* diagnosis of severe aortic valve disease requiring surgical replacement, *(ii)* age below 18 years at the time of surgery, and *(iii)* complete availability of operative and follow-up data. Patients with incomplete records or follow-up shorter than 12 months were excluded. Written informed consent was obtained from the parents and/or legal guardians of the patients. The study protocol was approved by the Ege University Faculty of Medicine Ethics Committee (Date: 25.01.2024, No: 2024-2174 24-1.1T/53). The study was conducted in accordance with the principles of the Declaration of Helsinki.

Follow-up included scheduled outpatient visits and regular transthoracic echocardiography evaluations. For patients not followed at our institution, follow-up status and echocardiographic data were obtained via structured telephone interviews with patients or their guardians. Early mortality was defined as death occurring within 30 days of the operation or before hospital discharge.

Surgical technique

All patients were operated on under cardiopulmonary bypass (CPB) with aortobicaval cannulation via median sternotomy. Myocardial arrest was achieved with blood cardioplegia administered through the aorta or directly into the coronary ostia following aortotomy, depending on the presence of aortic insufficiency.

Aortotomy was performed 1 cm above the coronary ostia. The coronary artery buttons were excised and preserved. Aortic valve leaflets were resected. The pulmonary autograft was harvested after transecting the pulmonary artery at the bifurcation level. With a Hegar dilator placed through the right ventricle, an incision was made 5 to 10 mm below the pulmonary valve in the infundibulum. Care was taken to avoid injury to the first septal artery during autograft harvesting.

In cases of significant mismatch between the aortic annulus and pulmonary autograft, plication of the noncoronary and left coronary trigones was performed using pledgeted pericardial sutures. In patients with severely hypoplastic aortic annulus, the Ross procedure was combined with a Konno enlargement.

The pulmonary autograft was implanted into the native aortic position using interrupted 4/0 nonabsorbable multifilament sutures (Figure 1). The coronary buttons were reimplanted into the neoaortic root using running prolene sutures. The distal aortic anastomosis was then completed using running sutures.

The RVOT was reconstructed using either pulmonary homografts, aortic homografts, bovine jugular vein conduits (Contegra; Medtronic, MN, USA), or porcine pericardial conduits (BioConduit®; BioIntegral Surgical Inc., Mississauga, Canada). The distal anastomosis was carefully fashioned to prevent stenosis. The conduit size was selected to match patient anatomy, with a moderately large size (conduit Z-score from 0 to + 2).

All patients were evaluated intraoperatively with transesophageal echocardiography after weaning from CPB. Postoperative patients were evaluated with transthoracic echocardiography both before and after discharge and at regular intervals.

Statistical analysis

Jamovi version 2.5.2 was used for all statistical analyses. Categorical variables were presented as frequencies and percentages. Continuous variables were expressed as median (range). Survival and

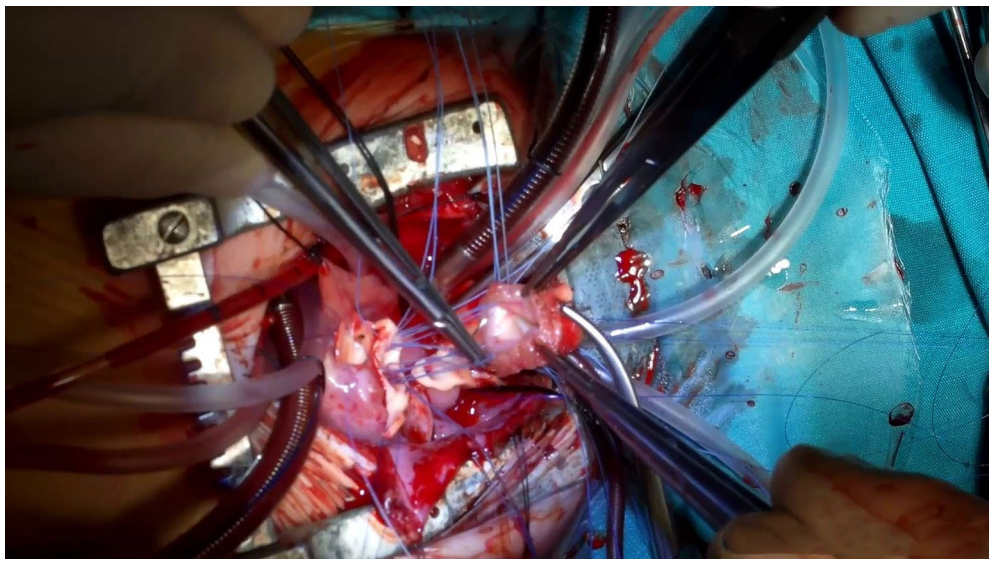

Figure 1. An intraoperative image of the youngest patient, a four-month-old male patient, showing implantation of a pulmonary autograft into the aortic annulus with separated sutures. The yellow circle indicates the pulmonary autograft, the blue oval shape indicates the aortic annulus, the blue star indicates the excised button-shaped left coronary, and the yellow star indicates the right coronary ostium.

freedom from reoperation were estimated using the Kaplan-Meier method. Reintervention was defined as any surgical procedure involving the RVOT conduit or autograft. A p value <0.05 was considered statistically significant.

RESULTS

The median weight was 13 kg (range, 4.6 to 25 kg). Four (22%) patients were infants. Eight (44%) patients had a bicuspid aortic valve, and one patient had Shone complex. Eleven patients had isolated aortic stenosis, three had isolated regurgitation, and four had mixed lesions.

Twelve (66.6%) patients had a history of balloon valvuloplasty, and one had prior surgical commissurotomy. Two (11.1%) patients required preoperative mechanical ventilation and inotropic support, both undergoing urgent surgery.

Two patients had left ventricular outflow tract obstruction. One of these patients underwent the Ross-Konno procedure, and the other underwent subaortic discrete membrane resection. In a case with annulus-pulmonary autograft mismatch, trigonal plication was performed. No reinforcement technique was used for autograft support.

For RVOT reconstruction, pulmonary homografts were used in 14 patients, aortic homografts in two, Contegra conduit in one, and porcine pericardial conduit in one patient. Conduit sizes ranged from 14 to 22 mm. The distal anastomosis was fashioned to avoid stenosis. Three patients underwent concomitant atrial septal defect closure, and one had subaortic discrete membrane resection. Mean cross-clamp and CPB times were 153±18 and 171±17 min, respectively. No intraoperative mortality occurred. Demographic and operative data of the patients are summarized in Table 1.

Two patients required prolonged ventilation (>24 h), both of whom had preoperative mechanical ventilation. One patient underwent delayed sternal closure at 36 h postoperatively. Early mortality occurred in one high-risk patient (two-year-old weighing <10 kg) who developed sepsis and multi-organ failure on postoperative Day 21. No late mortality was observed. The five-year survival rate was 94% (Figure 2).

The mean follow-up duration was 83.8±65 months (range, 12 months to 10 years). Two (11.1%) patients underwent reoperations due to conduit dysfunction (one with pulmonary homograft at year seven and another with porcine pericardial conduit at year three). No autograft reintervention

Table 1
Demographic, echocardiographic, and operative data of patients (n=18)

Variables	n	%	Mean±SD	Median	Range
Age (year)				3	4 months - 8 years
Weight (kg)				13	4.6-25
Sex					
Male	16	88.9			
Female	2	11.1			
Bicuspid aortic valve	8	44.4			
Shone complex	1	5.5			
Dominant aortic valve lesion					
Stenosis	11	61.1			
Insufficiency	3	16.7			
Mixed lesion	4	22.2			
Previous balloon valvuloplasty	12	66.6			
Previous surgical commissurotomy	1	5.5			
Preoperative mechanical ventilation	2	11.1			
Urgent surgery	2	11.1			
Concomitant surgery	4	22.2			
Atrial septal defect closure	3	16.7			
Subaortic discrete membrane resection	1	5.6			
Ross-Konno procedure	1	5.6			
Aortic annulus trigone plication	1	5.6			
Cardiopulmonary bypass time (min)			171±17		
Cross-clamp time (min)			153±18		
Conduits used for RVOT reconstruction					
Pulmonary homograft	14	77.8			
Aortic homograft	2	11.1			
Contegra	1	5.6			
Porcine pericardial conduit	1	5.6			
Conduit sizes (mm)					14-22
Postoperative prolonged ventilation	2	11.1			
Delayed sternal closure	1	5.6			
Early mortality	1	5.6			
Late mortality	0	0			
Reoperation due to conduit failure	2	11.1			
Autograft dysfunction/reintervention	0	0			
Mean follow-up duration			83.8±65		12 months - 10 years

SD: Standard deviation; RVOT: Right ventricular outflow tract.

or dysfunction was observed. Five-year freedom from reoperation was 90% (Figure 3). Due to the small cohort with longer follow-up, 10- and 15-year Kaplan-Meier estimates were not statistically reliable and were therefore not reported.

All surviving patients showed appropriate growth of the pulmonary autograft in serial echocardiograms, with preserved Z-score values and no significant autograft insufficiency. None had aneurysmal dilation or stenosis.

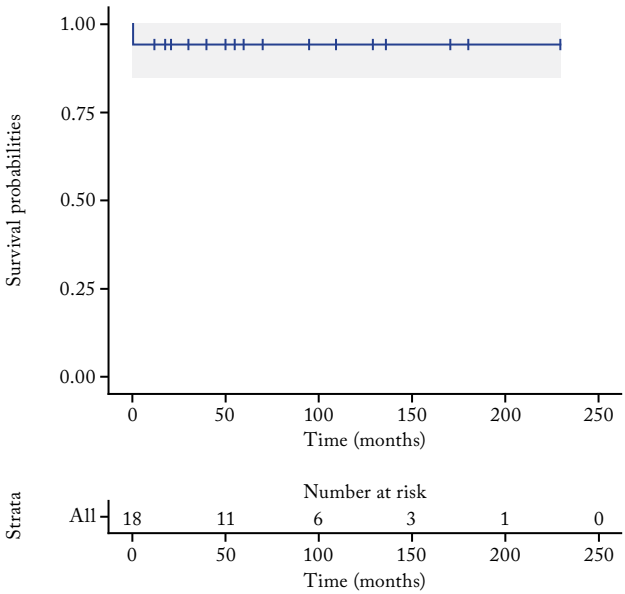

Figure 2. Kaplan-Meier plot showing patient survival.

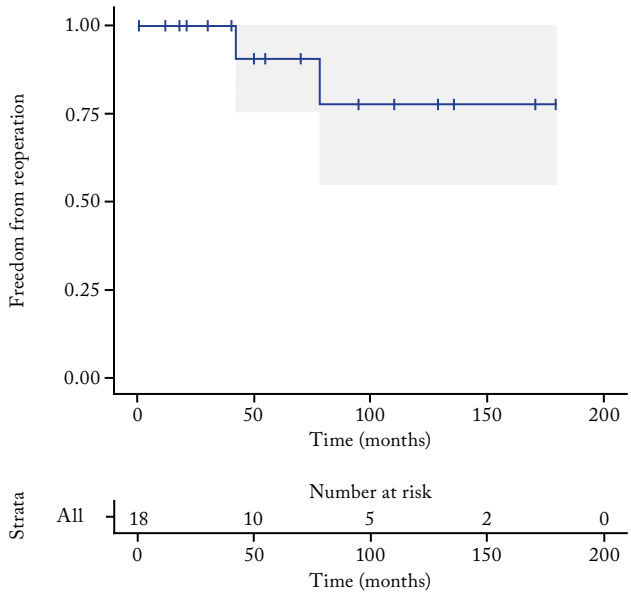

Figure 3. Kaplan-Meier plot showing freedom from reintervention.

DISCUSSION

The Ross procedure offers several advantages over traditional prosthetic valve replacements in pediatric patients, including optimal hemodynamics, avoidance of anticoagulation, and potential for somatic growth. These characteristics are particularly valuable in the growing child.[5,6] Our study contributes to the evidence supporting the use of the Ross procedure in children by presenting a favorable early outcome, low reoperation rates, and preserved autograft function during mid-term follow-up.

Our observed early mortality rate (5.5%) is consistent with previous pediatric series, where rates have ranged from 0 to 16% depending on age and risk profile.[7,8] The single mortality in our cohort occurred in a critically ill infant who underwent emergency surgery after prior balloon valvuloplasties and presented with organ dysfunction. These risk factors are well-documented predictors of poor outcome.[4,9]

Importantly, there were no cases of late mortality. Five-year survival was 94%, which aligns with the outcomes reported in recent systematic reviews and registry studies in pediatric cohorts.[10]

No autograft dysfunction or reoperation was observed in our series. This is a noteworthy finding, as prior studies have reported autograft failure rates of approximately 1% per year.[9] Several factors may account for our favorable results, including the use of precise surgical techniques such as interrupted sutures at the proximal anastomosis to preserve geometry, avoidance of reinforcement that may limit growth, and inclusion technique rather than full root replacement. However, the absence of autograft-related complications must be interpreted cautiously, as some patients were followed at external centers and limited follow-up beyond 10 years reduces long-term reliability. We acknowledge that the possibility of unreported dysfunction due to loss to follow-up remains.

To prevent conduit degeneration, homografts were preferentially used. Nevertheless, two patients required conduit replacement due to degeneration-one with a pulmonary homograft at year seven, and the other with a porcine pericardial conduit (BioIntegral Surgical Inc., Mississauga, Canada) at year three. The early failure of the latter is consistent with literature describing an inflammatory response and premature degeneration associated with this material.[11] Our five-year freedom from reintervention was 90%, comparable to prior reports of 79 to 90%.[12,13]

Aortic root dilatation or aneurysm formation did not occur in any of our patients during follow-up.

We attribute this to the use of subcoronary implantation and the natural anatomical integration of the pulmonary autograft. Reinforcement techniques, while sometimes effective in adults, are not routinely used in children due to concerns over growth limitation.[9,14,15]

Regarding alternative techniques, the Ozaki procedure has emerged as a promising reconstructive option in pediatric aortic valve disease. It offers avoidance of prosthetic materials and preserves native aortic root dynamics. However, long-term durability in children remains uncertain, and it lacks the growth potential offered by the Ross procedure. Further comparative studies are needed to define its role.[3,16,17]

Intraoperative care to preserve the first septal branch during autograft harvest is essential, as injury may compromise myocardial perfusion. We emphasize that meticulous dissection was performed in all cases to prevent such complications.

No patient in this series required extracorporeal membrane oxygenation support, including the single early mortality case. Given the late deterioration and progression to multi-organ failure, extracorporeal membrane oxygenation was not initiated due to the poor likelihood of recovery and ongoing septic complications.

This study had several limitations. It was limited by a small sample size and single-center retrospective design. The follow-up period varied significantly between patients, and longer-term outcomes beyond 10 years were available for only a few patients, limiting statistical validity for long-term survival and reoperation estimates. Additionally, the absence of a comparison group and potential loss to follow-up in some cases restricts the generalizability of the findings. Despite these limitations, our results support the use of the Ross procedure as a favorable option for pediatric aortic valve replacement when performed by experienced surgeons.

In conclusion, this study demonstrated that the Ross procedure can be performed in pediatric patients with low mortality and morbidity. With appropriate surgical technique, the autograft can function well and grow with the patient, minimizing the need for reoperation. Our findings suggest that conduit-related reinterventions are manageable and do not outweigh the significant benefits of using the patient's own tissue. While vigilance in long-term follow-up remains essential, the perception that the Ross procedure converts one valve disease into two may be overly pessimistic in the context of properly selected and managed pediatric patients.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: Idea/concept, references and fundings, materials: O.N.T.; Design, literature review: O.N.T., Y.A.; Control/supervision, critical review: Y.A., R.E.L.; Data collection and/or processing, analysis and/or interpretation, writing the article: M.A., O.N.T.

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