Surgical solutions for complex aortic root pathology

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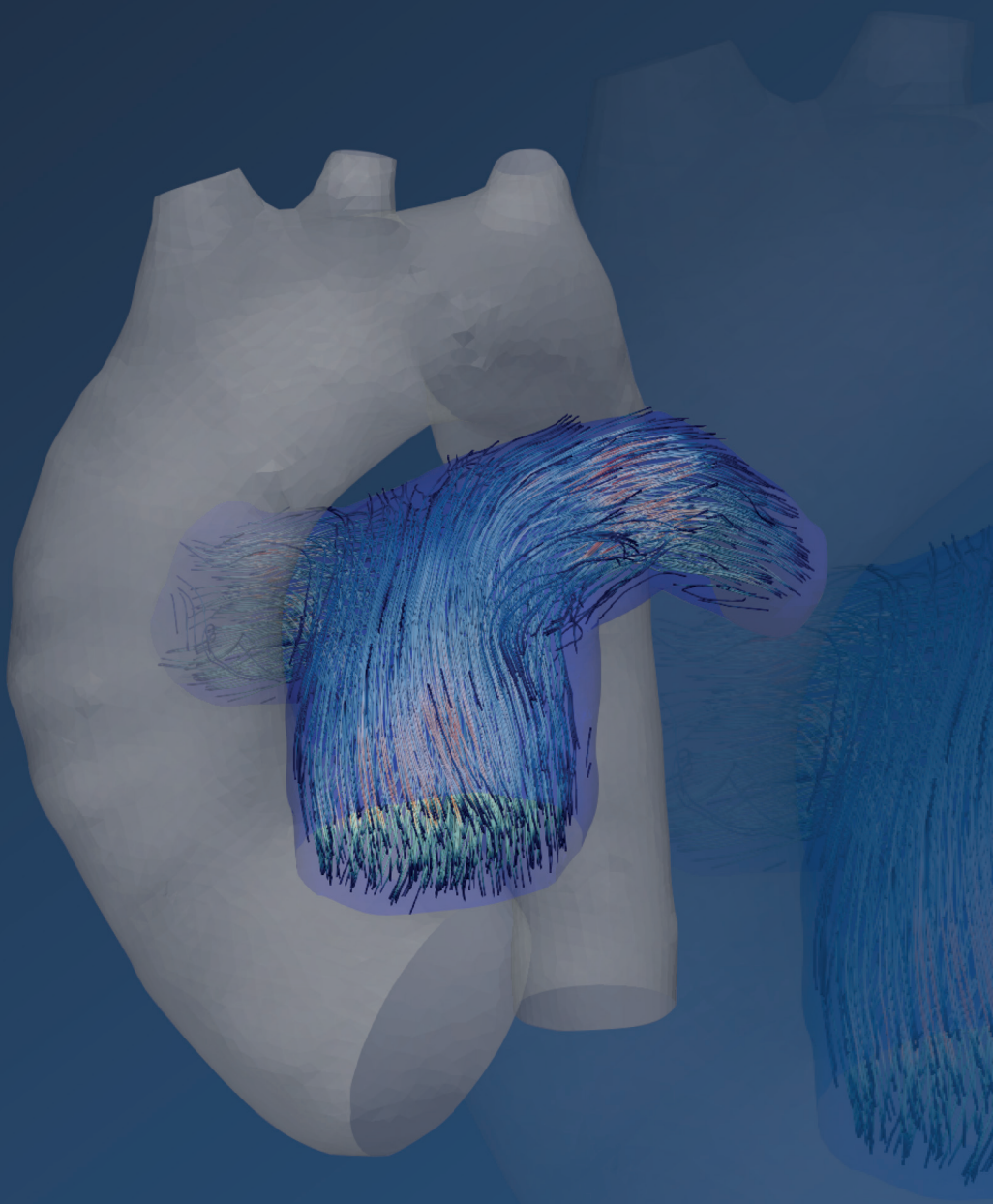
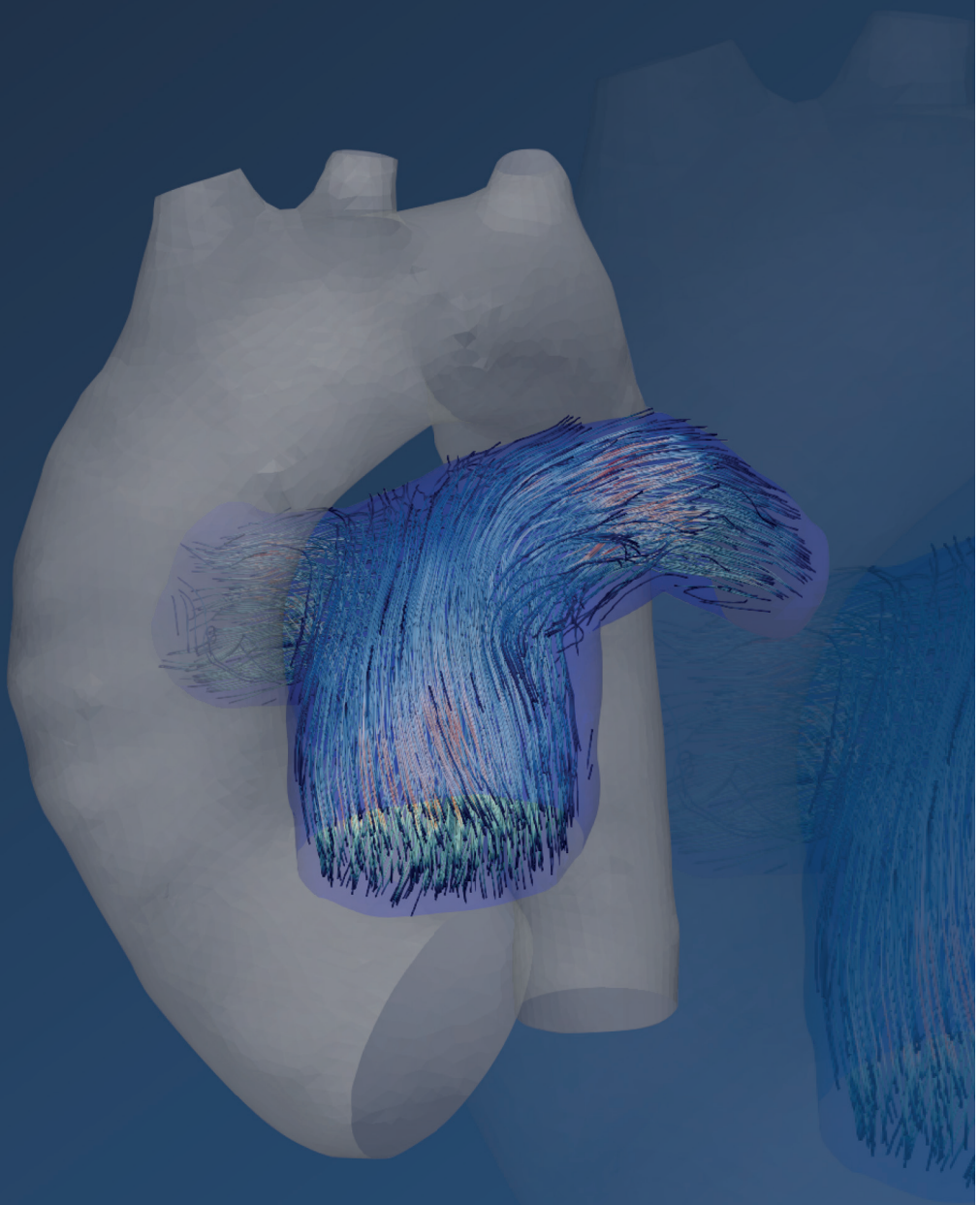


PART 1

THE PULMONARY AUTOGRAFT



TWENTY-YEAR EXPERIENCE WITH THE ROSS-KONNO PROCEDURE

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CHAPTER 2

ABSTRACT

OBJECTIVES

The Ross–Konno procedure is a last resort for patients with complex multilevel left ventricular outflow tract obstruction (LVOTO) often having other cardiovascular anomalies. It is typically preceded by multiple surgeries. Literature is scarce on long-term follow-up series. Therefore, we have reviewed our 20-year experience with this procedure in order to provide insights in patients' outcomes and to optimize patient selection.

METHODS

Between January 1995 and December 2014, 48 patients underwent the Ross–Konno procedure. The median age at operation was 12.8 months (range, 11 days to 31 years). Twenty-two (46%) patients were under 1 year of age. Forty-four (92%) patients had undergone a total of 82 previous procedures. Eleven (23%) patients had concomitant surgery, predominantly mitral valve (n = 5) and aortic arch surgery (n = 5).

RESULTS

The median follow-up time was 4.3 years (range, 0–20 years). There were 6 (12.5%) early deaths and 4 (8.3%) late deaths. Estimated overall survival at 5, 10 and 15 years was 83, 79 and 70%, respectively. Poor LV function was a risk factor for early mortality (odds ratio = 9.5; 95% confidence interval = 1.4–63.7; P = 0.020). Twelve patients required a total of 29 procedures in 17 reoperations. Five patients required reoperation for autograft failure at a median of 14 years (range, 5–15 years) postoperatively. Estimated freedom from all causes of reoperation at 5, 10 and 15 years was 82, 55 and 30%, respectively. All patients had complete and durable relief of LVOTO. At latest follow-up, 5 patients had a sinus of Valsalva Z -score of 5 or greater. One patient had Grade II autograft insufficiency.

CONCLUSIONS

The Ross–Konno procedure is a durable solution for multilevel LVOTO in a highly complex patient population with high incidence of previous procedures. High early mortality rates in patients with impaired left ventricular function emphasize the

importance of patient selection. Freedom from reoperation shows a continuous attrition rate. Reoperation for autograft failure may occur late after the Ross–Konno procedure.

INTRODUCTION

The Ross–Konno operation is a complex procedure for severe or multilevel (sub) aortic stenosis. It consists of using the patients' pulmonary valve as a neoaortic autograft (Ross procedure), combined with the opening of a narrowed left ventricular outflow tract (LVOT) by incising the outflow septum. Patients in need of a Ross–Konno procedure often have accompanying congenital anomalies, such as coarctation of the aorta (CoA), an interrupted aortic arch (IAA) and ventricular septal defects (VSDs). Careful patient selection is important in predicting the success of the procedure, especially in patients with a borderline left ventricle (LV) and/or endocardial fibroelastosis (EFE). The capability of the autograft to grow with the child is one of the main advantages of the Ross–Konno procedure in infants and children. Downsides of the Ross–Konno operation are the need for reoperations for the right ventricle to pulmonary artery (RV–PA) conduit and possible dilatation of the autograft root.

Literature is scarce on long-term follow-up series of the Ross–Konno operation. The aim of this study was to review our 20-year experience with the Ross–Konno procedure in a large cohort of patients to evaluate outcomes and optimize patient selection.

MATERIALS AND METHODS

Patients and data collection

The Ethics Committee of the Leiden University Medical Center approved this retrospective observational study and waived the need for patients' or parental informed consent. All patients who underwent the Ross–Konno procedure between January 1995 and December 2014 were identified. Only patients requiring a generous interventricular incision to augment the LVOT were included in this study. Young patients with a Ross operation in whom the septum was slightly incised to make the autograft fit into the smaller aortic annulus were excluded. Data were collected from medical records, including patient charts, operative reports and echocardiographic examinations.

Surgical technique

Median sternotomy was performed in all patients. Cardiopulmonary bypass was achieved with bicaval cannulation and mild hypothermia (deep hypothermia in

case of aortic arch reconstruction). Antegrade cold crystalloid cardioplegia with external myocardial cooling was administered. Both coronary arteries were excised and mobilized, and the aortic valve and sinuses of Valsalva were removed. The pulmonary autograft was excised with a muscular tongue of the right ventricular (RV) anterior wall. The Konno incision was made in the interventricular septum starting under the commissure between the left and right coronary leaflets, towards the VSD if present (resecting the outflow septum). This incision was extended as far as necessary for full relief of LVOT obstruction (LVOTO). Fibrous and/or muscular LVOTO was resected or enucleated. The pulmonary autograft was placed in aortic position, typically closing the interventricular incision using the RV anterior wall tongue (in 2 early patients, the septal incision was closed with a patch). Care was taken to precisely position the RV anterior wall tongue as a minor rotation could lead to some degree of aortic insufficiency. Both coronary artery buttons were reimplanted into the autograft and the distal anastomosis with the ascending aorta was made. The RV–PA connection was restored, using a homograft or bovine jugular vein conduit. Running sutures were universally used, with sutures at the base of the root being reinforced with a thin strip of pericardium in all patients. Reinforcement of the sinotubular junction anastomosis was mainly used in older children.

Echocardiographic data

For each patient, echocardiographic data were collected to examine ventricular and valvular functions as well as neoaortic dilatation. Preoperative and most recent (or before reoperation) measurements of the LVOT (minimal diameter), aortic annulus, sinus and sinotubular junction diameters were retrospectively collected and analysed. Echocardiographic *Z* -scores of annular and sinus of Valsalva diameters were based on data from Pettersen *et al* . and Roman *et al*., respectively[1, 2].

Statistical analysis

Continuous variables are expressed as mean ± standard deviation (SD) or median and (interquartile) range, as appropriate. Categorical variables are reported as numbers and percentages. Early mortality was defined as death within 30 days after the Ross–Konno operation or during the same hospital admission. Overall survival and freedom from reoperation were estimated using the Kaplan–Meier

method and differences in survival curves between subgroups of patients were tested using the log-rank test. Estimates of overall survival and freedom from reoperation are expressed as percentage with 95% confidence interval (CI). Univariable binary logistic regression analysis was performed to identify risk factors for early mortality. Univariable Cox regression analysis was performed to examine risk factors for late mortality and reoperation. The predictor variables considered were age at operation and preoperative poor LV function, EFE and aortic annular size. The non-parametric Mann–Whitney *U* -test was used to evaluate the difference in neoaortic dilatation between patients aged under or above 1 year at operation. A *P* -value of less than 0.05 (two-sided) was considered statistically significant. Survival analysis was performed using GraphPad Prism 6 (GraphPad Software, Inc., La Jolla, CA, USA). All other analyses were performed with SPSS 20 (IBM Corp., Armonk, NY, USA).

RESULTS

Patient population

Forty-eight (73% male) patients underwent the Ross–Konno procedure. The median age at operation was 12.8 months, ranging from 11 days to 31 years (interquartile range, 3.7 months to 9.4 years). Twenty-two patients were under 1 year of age, 9 of which were under 3 months of age and 3 were younger than 1 month at the time of surgery. One patient was older than 18 years. Almost half of the patients (48%) had associated cardiac or aortic anomalies. Two patients had a bicuspid pulmonary valve at the time of surgery. Five patients had a preoperative poor LV function and important EFE. Patient characteristics and initial diagnoses are presented in Table 1.

Table 1: Patient characteristics and initial diagnoses

Patient characteristics	***n* (%)**
Male sex	35 (73)
Age at operation in months (median; range)	12.8 months (11 days–31 years)
<1 year of age	22
<3 months	9
<1 month	3
Aortic annulus *Z* -score (mean ± SD)	−1.7 ± 2.3
Initial diagnosis	
AoS/LVOTO	25 (52)
IAA–VSD	11 (23)
AoS + CoA + arch hypoplasia (±VSD)	10 (21)
AVSD	2 (4)
HOCM	1 (2)
Bicuspid aortic valve	24 (50)
Bicuspid pulmonary valve	2 (4)
AR after balloon dilatation	7 (15)
Mitral valve stenosis/insufficiency	5 (10)
Poor LV function	5 (10)
Important EFE	5 (10)

AoS: aortic valve stenosis; AR: aortic regurgitation; AVSD: atrioventricular septal defect; CoA: coarctation of the aorta; EFE: endocardial fibroelastosis; HOCM: hypertrophic obstructive cardiomyopathy; IAA: interrupted aortic arch; LV: left ventricle; LVOTO: left ventricular outflow tract obstruction; SD: standard deviation; VSD: ventricular septal defect.

There were a total of 82 previous interventions in 44 patients. Nineteen procedures were percutaneous, namely foetal balloon valvuloplasty (*n* = 1), balloon valvuloplasty (*n* = 15) and balloon dilatation of the CoA (*n* = 3). One patient had an intrauterine balloon valvulotomy of the aortic valve at a gestational age of 24 + 4 weeks, dilating the aortic annulus from 2.5 to 2.9 mm. Two days after birth, another balloon valvuloplasty was performed. Eventually, at the age of 16 days, the Ross–Konno procedure was performed. One 31-year-old patient required a Ross–Konno procedure because of LVOTO after coarctation repair twice, resection of a subaortic stenosis and balloon valvuloplasty of a bicuspid aortic valve. The aortic annulus of this patient had a diameter of 12 mm and the autograft had a diameter of 25 mm. In 3 patients, balloon valvuloplasty resulted in significant aortic regurgitation (AR). A complete overview of previous procedures is presented in Table 2.

Table 2: Previous procedures

Procedure	*n* (%)
Percutaneous balloon dilatation	19 (40)
Aortic valve repair	11 (23)
Subaortic stenosis repair	18 (38)
Aortic arch repair	16 (33)
IAA + VSD repair	8 (17)
VSD closure	6 (13)
AVSD repair	2 (4)
Hybrid Norwood	3 (6)
Mitral valve replacement	2 (4)
Mitral valve repair	1 (2)
Tricuspid valve repair	1 (2)
Pacemaker implantation	2 (4)
Operation for constrictive pericarditis	1 (2)
Aortopexy	1 (2)

AVSD: atrioventricular septal defect; IAA: interrupted aortic arch; VSD: ventricular septal defect.

Operative data and complications

Operative details are presented in Table 3. The septal incision was closed using the RV free wall tongue attached to the pulmonary autograft in all but 2 early patients in whom a xenopericardial patch was used. Eleven patients had concomitant procedures, namely, aortic arch repair (n = 5), mitral valve surgery (n = 5) and ascending aorta replacement (n = 1). In most patients (69%), the RV–PA connection was restored, using a Contegra (Medtronic, Minneapolis, MN, USA) bovine jugular vein graft. Three patients required postoperative extracorporeal membrane oxygenation (ECMO) support because of low cardiac output. ECMO could be weaned in 2 patients who survived. There were 6 (12.5%) early deaths, of whom 2 were neonates and 4 were infants. Preoperative poor LV function with or without EFE was present in 4 out of the 6 early deaths. The 2 neonatal patients in this group had severe LV dysfunction and important EFE (Table 4). Univariable binary logistic regression analysis identified poor LV function (odds ratio = 9.5; 95% CI = 1.4–63.7; P = 0.020) as the only risk factor for early mortality.

Table 3: Operative details

Operative details	*n* (%)
Emergency setting	3 (6)
Cross-clamp time in minutes (median; range)	147 (80–305)
Concomitant procedures	11 (23)
Aortic arch repair	5 (10)
Ascending aorta replacement	1 (2)
Mitral valve repair	2 (4)
Mitral valve replacement	1 (2)
Mitral valve rereplacement	2 (4)
RV–PA conduits	
Bovine jugular vein conduit	33 (69)
Cryopreserved pulmonary homograft	11 (23)
Cryopreserved aortic homograft	4 (8)
Complications	
LCO requiring ECMO	3 (6)
Permanent pacemaker	2 (4)

ECMO: extracorporeal membranous oxygenation; LCO: low cardiac output; RV–PA: right ventricle to pulmonary artery.

Table 4: Mortality causes

Patient no.	Age at operation	Time between surgery and death	Diagnosis	Previous surgery	Concomitant procedures	Cause of death
1	11 days	0 days	AoS, CoA, arch hypo-plasia, VSD, poor LVF, EFE	AVP, aortic arch repair	None	LV failure
2	123 days	0 days	IAA, VSD, severe LVOTO, poor LVF	IAA + VSD repair, aortopexy	None	LV failure
3	4 months	0 days	IAA, VSD, LVOTO	Hybrid Norwood	Arch repair	Massive lung bleeding
4	3 months	6 days	IAA, VSD, LVOTO	Hybrid Norwood	Arch repair	Postop ECMO, thrombus in neoaortic root and MI
5	54 days	10 days	IAA, VSD	IAA + VSD repair	None	LV failure

Patient no.	Age at operation	Time between surgery and death	Diagnosis	Previous surgery	Concomitant procedures	Cause of death
6	26 days	24 days	AoS, poor LVF, EFE	Balloon dilatation resulting in AR	None	Septic shock
7	15 months	6 months	CoA, iAVSD	CoA + AVSD repair, MVR	None	Unexplained (no autopsy)
8	16 years	7 months	AoS, AR, DSAS	DSAS removal	None	Pulmonary embolism during balloon dilatation of the RV–PA conduit
9	74 days	1.1 year	AoS, severe EFE	Balloon dilatation resulting in AR	None	Sepsis, cardiac and pulmonary insufficiency
10	37 days	12.7 years	Unknown syndrome, AoS, CoA, arch hypoplasia, VSD	AVP, aortic arch + VSD repair	None	Recurrent pneumonia/ empyema, with secondary heart failure

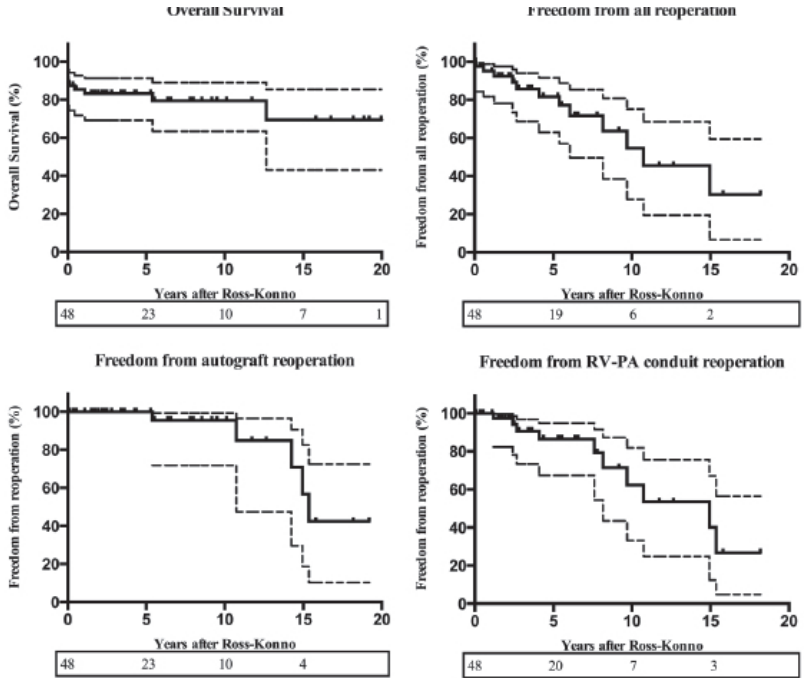
AoS: aortic stenosis; AR: aortic regurgitation; AVP: aortic valve plasty; CoA: coarctation of the aorta; DSAS: discrete subaortic stenosis; ECMO: extracorporeal membrane oxygenation; EFE: endocardial fibroelastosis; IAA: interrupted aortic arch; iAVSD: incomplete atrioventricular septal defect; LV: left ventricle; LVF: left ventricular function; MI: myocardial infarction; LVOTO: left ventricular outflow tract obstruction; MVR: mitral valve replacement; RV–PA: right ventricle to pulmonary artery; VSD: ventricular septal defect.

Follow-up

Follow-up was complete, with recent follow-up for all alive patients. Median follow-up for the total study population was 4.3 years (range, 0–20 years).

A total of 10 (20.8%) patients died, including early deaths. Causes of mortality for the whole series are presented in Table 4. Estimated overall survival at 5, 10 and 15 years was 83 (95% CI: 69–91%), 79 (95% CI: 63–89%) and 70% (95% CI: 43–85%), respectively (Fig. 1). There were 4 (8.3%) late deaths. The linearized occurrence rate of late mortality was 1.4% per patient-year. No risk factor for late mortality could be identified.

Figure 1:



Kaplan–Meier curves for estimated overall survival (upper left), freedom from all reoperation (upper right), freedom from autograft reoperation (lower left) and freedom from right ventricular to pulmonary artery conduit reoperation (lower right). Dashed lines denote 95% confidence intervals. Numbers under the curves depict numbers at risk. RV–PA: right ventricle to pulmonary artery.

Table 5: Reoperation procedures

Reoperation procedures	***n* (%)**
RV–PA conduit change	10 (21)
Second RV–PA conduit change	3 (6)
Pulmonary autograft replacement	5 (10)
Mitral valve repair	2 (4)
Mitral valve replacement	1 (2)
Tricuspid valve repair	3 (6)
Residual VSD closure	1 (2)
Aortic arch rerepair	2 (4)
Ascending aorta plasty	1 (2)
Ostium plasty of LCA	1 (2)

LCA: left coronary artery; RV–PA: right ventricle to pulmonary artery; VSD: ventricular septal defect.

Table 6: Autograft reoperations

Patient no.	Age at operation	Years between surgery and reoperation	Indication for reoperation	Reoperation procedure
1	3 months	5.4	AR	Mechanical Bentall
2	17 years	10.7	Autograft dilatation (49 mm), PS, MS	Stentless aortic bioprosthesis, PVR (homograft), MVP
3	5 years	14.2	Autograft dilatation (45 mm), PS	Mechanical Bentall, PVR (homograft)
4	10 years	14.9	Autograft dilatation (50 mm), AR, PR, MR, TR	Stentless aortic bioprosthesis, PVR (homograft), MVP, TVP
5	10 years	15.3	AR, PS	Mechanical Bentall, PVR (bovine jugular vein)

AR: aortic regurgitation; MR: mitral regurgitation; MS: mitral stenosis; MVP: mitral valve plasty; PR: pulmonary regurgitation; PS: pulmonary stenosis; PVR: pulmonary valve replacement; TR: tricuspid regurgitation; TVP: tricuspid valve plasty.

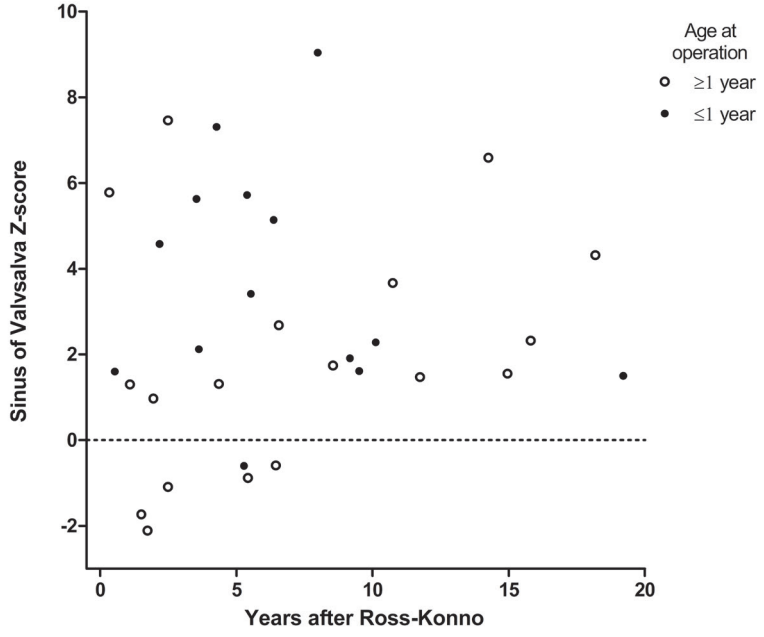
Twelve patients required a total of 29 procedures in 17 reoperations. Reoperative procedures are presented in Table 5. Freedom from any reoperation, autograft reoperation and RV–PA conduit reoperation curves are presented in Fig. 1. Estimated freedom from all causes of reoperation was 82 (95% CI: 63–92%), 55 (95% CI: 28–75%) and 30% (95% CI: 7–59%) at 5, 10 and 15 years, respectively. Five patients required reoperation for autograft failure at a median of 14 years (range, 5–15 years) postoperatively (Table 6). One of these patients was under the age of 1 year at the time of the Ross–Konno procedure. This patient had a bicuspid pulmonary autograft which became insufficient after 5.4 years and had to be replaced with a mechanical prosthesis. Three patients had dilatation of the pulmonary autograft resulting in AR and underwent redo surgery. Two of them received a mechanical Bentall procedure and 1 patient received a stentless bioprosthetic aortic root replacement. One patient had aortic root dilatation without AR and underwent a Bentall procedure and RV–PA conduit change. For autograft reoperation, estimated 5-, 10- and 15-year freedom from reoperation rates were 100, 96 (95% CI: 72–99%) and 57% (95% CI: 19–83%), respectively. The

linearized occurrence rate of autograft reoperation was 1.8% per patient-year. No risk factor for autograft reoperation could be identified. Estimated freedom from RV–PA conduit reoperation at 5, 10 and 15 years was 87 (95% CI: 67–95%), 62 (95% CI: 33–82%) and 40% (95% CI: 12–67%), respectively. Freedom from RV–PA conduit reoperation curves was not significantly different for patients with a homograft versus patients with a bovine jugular vein conduit (log-rank test: $P = 0.806$).

Echocardiographic follow-up

At latest (or before reoperation) echocardiographic follow-up (median follow-up time = 4.4 years; range, 1 month to 19 years), 4 patients had mild (Grade I) AR and 1 patient had moderate (Grade II) AR. All other patients had none or trivial AR.

Figure 2:



Autograft sinus of Valsalva *Z* -scores at latest echocardiographic follow-up. No association between operation before the age of 1 year and a reduced incidence of autograft dilatation could be demonstrated.

Reliable measurements of the autograft sinus of Valsalva at last follow-up could be obtained for 32 patients. The sinus of Valsalva *Z* -scores for patients younger

versus those above 1 year of age at the time of the Ross–Konno procedure are shown in Fig. 2. The *Z* -scores were not significantly different between both age groups. Left ventricular function was good in all but 4 patients who had moderate impairment of the LV function. Three patients had a moderate RV dysfunction; all other patients had good RV function. Mean gradient over the RV–PA conduit was 27 ± 15 mmHg. Two patients had severe pulmonary regurgitation (PR) which was tolerated well, 2 patients had moderate to severe PR, 4 had moderate PR and all other patients had none to mild PR.

DISCUSSION

This study describes 48 patients who underwent the Ross–Konno procedure for severe multilevel LVOTO. All but 1 patient were under 18 years of age at the time of surgery, with half of all patients being under 3 years of age. The vast majority of patients had undergone one or more procedures prior to the Ross–Konno operation. The Ross–Konno procedure was and also should be a last resort operation. We believe that the Ross–Konno procedure is safer in older patients and that, whenever possible, this operation should be avoided in the neonatal or early infancy period.

Damage to the first septal branch of the left anterior descending coronary artery is extremely rare as the septal incision usually does not interfere with this artery. However, there are several other technical challenges associated with the Ross–Konno procedure. First, the use of the attached tongue of the RV anterior wall typically directs the positioning of the autograft on top of the LV. Care should be taken to obtain a perfect fit of this autograft 'tongue' into the interventricular incision as the failure to do so may result in postoperative autograft insufficiency. In the present series, neoaortic valve regurgitation mostly occurred much later in the follow-up. Second, especially in neonates and infants with a small aorta, coronary artery reimplantation can be difficult, as the distance between both coronary ostia has to be much greater when implanted in the pulmonary autograft. In neonates and small children in our series, the right coronary artery had to be directed to the non-coronary sinus in several patients. Lastly, coronary kinking has to be avoided at all costs.

The Ross–Konno procedure remains a high-risk procedure. In our series, the early mortality rate was 12.5%. This is consistent with a recent report by Vergnat *et*

al. [3], reporting an early mortality rate of 10.2%. We found preoperative poor LV function to be a risk factor for early mortality, which was also found by Vergnat *et al*. Mitral valve disease in our series, however, was predominantly valve insufficiency, whereas in the report by Vergnat and his group it was mainly valve stenosis. This difference might be due to patient selection or chance. Others have reported more positve early mortality outcomes. Aszyk *et al.* described their experience with 16 patients under the age of 1 year undergoing the Ross–Konno procedure. They reported no early mortality [4]. Maeda *et al.* [5] reported 1 early death in 24 patients operated under the age of 1 year.

Deciding which patient is eligible for the Ross–Konno procedure can be very difficult, especially in patients with borderline LV and EFE. We had a learning curve in selecting patients for either biventricular repair, i.e. the Ross–Konno procedure, or univentricular palliation. In our series, a preoperative poor LV function with or without EFE was present in 3 of the 6 early deaths. Most of our early deaths with preoperative LV impairment were in the earlier years. In retrospect, 2 patients with both poor LV function and EFE would nowadays not have been selected for Ross–Konno surgery. We now believe that the Ross–Konno procedure can be performed even when the LV function is seriously depressed. However, this statement is only valid when the LV dysfunction is caused by aortic valve stenosis and/or AR. In these cases, the LV dysfunction appears to be reversible following the Ross–Konno operation. On the other hand, in our experience, when the LV dysfunction is (also) present because of some degree of LV hypoplasia, especially when combined with more than mild EFE, results are much worse and one should probably refrain from the Ross–Konno procedure. Our experience with EFE resection combined with a Ross–Konno operation is however limited; others have reported better outcomes [6]. It should be noted though, that the grey area for choosing between biventricular repair, i.e. Ross–Konno, or univentricular management, i.e. Fontan circulation, remains broad and the diagnostic criteria are not always clear. In 3 patients that eventually underwent the Ross–Konno procedure, we first performed a hybrid Norwood procedure (ductal stenting and bilateral pulmonary artery banding) to allow the LV to grow, postponing the decision for uni- or biventricular correction. Two of these patients died (Table 4). The third patient was doing well 3 months after surgery.

Estimated freedom from autograft reoperation in our study was 100, 96 and 57% at 5, 10 and 15 years, respectively. This is consistent with other reports about autograft reoperation after Ross or Ross–Konno procedures in the same age group [3 , 7–9]. Autograft reoperation was indicated by either neoaortic root dilatation or regurgitation. It has been postulated that surgery early in life might protect against autograft dilatation. Lo Rito *et al.* [7] have shown the neoaortic root to be more stable in children operated on at an age younger than 18 months when compared with patients operated on at an age of over 18 months. We were not able to demonstrate such association between younger age (i.e. operation before the age of 1 year) and more stable autograft diameters. In our series, only 1 out of 22 patients operated on under 1 year of age needed later autograft replacement. This patient had a dysplastic bicuspid pulmonary valve at the time of the Ross–Konno procedure and this bicuspid pulmonary autograft developed valve insufficiency and was replaced by a mechanical prosthesis 5.4 years after the Ross–Konno procedure. In one other patient with a bicuspid pulmonary valve, the autograft showed complete normal function 5 years after the Ross–Konno procedure. Some patients in whom the autograft was dilated showed no aortic insufficiency in our series. This has been observed also in a study by others on autograft dilatation in patients after the Ross procedure [10]. Finally, without any exception, the Ross–Konno operation resulted in complete and durable relief of LVOTO. There were no residual or recurrent LVOT gradients.

Limitations

This is a retrospective study. Patient selection and operative care have improved over the years with likely improved outcomes. Although this is a relatively large patient cohort, in the survival analyses, the numbers of patients at risk after 10 years of follow-up were too small to allow for reliable estimates. Small patient numbers also hampered the analyses for potential risk factors for mortality and reoperation. The *Z* -score reference formulas for the preoperative and latest echocardiographic data were necessarily retrieved from two sources. For latest echocardiographic aortic sinus diameter, the *Z* -score formula for children was used in our patients under 18 years of age; the *Z* -score formula for adults till 40 years was used in our adult population.

CONCLUSION

The Ross–Konno operation is a valuable procedure that can fully and durably relieve multilevel LVOTO in a highly complex patient population with often multiple associated anomalies and previous interventions. High early mortality rates in patients with impaired LV function emphasize the importance of adequate patient selection. Freedom from reoperation shows a continuous attrition rate, most often for RV–PA conduit replacement. Reoperation for autograft failure may occur late after the Ross–Konno procedure.

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