

Central Illustration: Performing the Ross Procedure on Children - Determination of the Optimal Timing and Surgical Indications-2026

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ABSTRACT

Background: The Ross procedure in the surgery of congenital heart defects is an indispensable method of prosthetics of the aortic valve, used in cases where the use of other prostheses is associated with a high risk of fatal complications. This operation is especially relevant at an early age, since small-diameter prostheses with similar biological and hemodynamic characteristics are currently unavailable. However, this operation is one of the most difficult prosthetic options, for which it is necessary to clearly determine the indications, the optimal age of the operation and the expected duration of an uncomplicated course.

Objective: To systematize the experience of performing Ross/Ross-Konno surgery in children.

Materials and Methods: A total of 68 patients were examined, 53 boys and 15 girls aged 3.49 [2.15;6.17] years at the time of surgery (weight 15[12.4;20.5]); Ross surgery was performed on 56 patients, Ross-Konno surgery on 12 patients) for the period from 2006 to 2025. The median follow-up was 3.6 [1.2;7.2] years.

Results: It is shown that young children (up to 3 years old) They have the highest freedom from autograft dysfunction, however, they are most at risk of RV-PA conduit dysfunction due to its small size. Preschool and school-age children have a high degree of freedom from RV-PA-related complications (since most used large-diameter RV-PA conduits), however, the autograph is more susceptible to dysfunction, which occurs mainly by the mechanism of dilation under systemic pressure.

Conclusion: In our opinion, the Ross procedure is most relevant in early and preschool age, when a small prosthesis is required, and more and more alternatives to prosthetics appear at school age and older, which requires detailed study.

Keywords: Aortic Valve Malformations; Ross Procedure; Allografts

Introduction

Surgery of congenital malformations of the aortic valve is a very wide field in which a large number of possible operations are available [1]. Among all the operations, the Ross procedure is the most difficult and traumatic option. Due to the high degree of expected surgical trauma during the Ross procedure, it is especially important to

determine the appropriate indications and examine the results in different age categories [1]. The main advantage of the Ross procedure is that the aortic valve prosthesis is a pipeline that can be called "ideal" in terms of its size and hemodynamic characteristics [1-4]. In pediatric cardiac surgery, the Ross procedure is used in patients who require aortic valve replacement but cannot be matched with a suitably sized artificial or biological prosthesis (i.e. a small mammalian aortic

xenograft or reduced adult aortic allograft). The procedure is thus indispensable in very young children (1-3 years old), and in older patients also remains an important option [4]. Collection and systematization of the results of Ross procedures performed on patients of different age categories is of great practical importance for the plan-

ning of management tactics for patients with aortic malformation and the consideration of possible alternative prosthetic options. The aim of the study is to retrospectively describe the medium-term results of the Ross procedure when performed on children of different age groups (Figure 1).



Figure 1.

Materials and Methods

This study retrospectively reviews the medical histories of patients who underwent aortic valve prosthetic surgery with pulmonary autograft (a total of 68 patients, 53 boys and 15 girls, with a mean age of 3.49 [2.15; 6.17] years, with a mean weight of 15 kg. [12.4; 20.5] at the time of surgery; The Ross procedure was performed on 56 patients and the Ross-Konno procedure was performed on 12 patients) from 2006 to 2025. The median follow-up age was 3.6 [1.2; 7.2] years.

Research Endpoints

We divided the total group of patients into 3 age groups to study the surgical results in each of them in detail:

1. Early childhood group (up to 3 years old), N=27
2. Preschool age group (4-7 years old), N=28
3. School-age group (7-18 years old), N=13

We evaluated overall survival, the freedom from significant complications related to the conduit in the pulmonary position (RVOT-related complications) and the conduit in the aortic position (RVOT-related complications), the freedom from dysfunction and freedom from reoperation. RVOT conduit dysfunction was defined as an increased gradient at the level of the valve, distal or proximal anastomosis greater than 40 mm Hg or valve regurgitation with a severity level greater than Grade II. Autograft dysfunction was defined as the development of valve regurgitation with a severity level greater than Grade II.

Statistical Methods

For descriptive statistics of quantitative variables, median values with interquartile range were used, while absolute values of occurrence, with the frequency of occurrence expressed as a %, were used to describe categorical variables. The Kaplan-Meier method was used to estimate survival and freedom from RVOT- and autograft-related complications, and survival curves were compared using the log-rank method. Single and multivariate analyses were applied. The two independent samples were compared using the nonparametric Mann-Whitney U-test for quantitative variables and Pearson's chi-square test for categorical variable. The Wilcoxon test was used to compare related samples. The significance level used was <0.05, and Bonferroni correction was calculated as <0.0167 and used to compare the three age groups with each other.

Results

Preoperative Characterization of Patients (Table 1)

The mean age of surgery in the total group was 3.5[2.1; 6.2] years. The most frequent indication for surgery was bicuspid aortic valve with predominant clinical valve stenosis. In 56% of cases the Ross procedure was performed as reintervention aortic valve surgery, most commonly after transluminal balloon valvuloplasty (TLBVP). Moreover, in 22 cases the Ross procedure was performed urgently in case of critical stenosis of the tricuspid aortic valve (in 7 cases) or after TLBVP that resulted in the development of total BAV insufficiency (in 15 cases). In most cases, despite severe aortic valve stenosis, the Z-score of the fibrosus ring was within normal limits, and the cause of stenosis was commissural fusion of the flaps. In 12 out of 68 patients there was a significant narrowing of the fibrosus ring, therefore the enlargement was performed using the Konno technique in 11 patients and the Manouguian technique was performed for one patient (Table 1).

Table 1: Preoperative characterization of patients.

	All (N=69)	Early childhood(up to 3 years)(N=27)	Preschool (3 to 7 years old)(N=28)	School (from 7 years old) (N=13)	p-value (with Bonferroni correction)
Anthropometric characteristics of patients					
Age of patient when surgery performed, years	3.5[2.1;6]	1.6[1.3;2.4]	4.4[3.6;5.4]	9.9[9;13]	-
Weight at the time of surgery, kg	15[13;20]	12[10;13]	17[15;19]	44[35;50]	-
Indication for surgery					
AV stenosis	48 (70%)	16 (59%)	19 (68%)	13 (100%)	0.06
Total AV insufficiency after TLBVP AV, N (%)	15 (22%)	7 (26%)	8 (29%)	0 (0%)	0.08
Combined AV defect	4 (6%)	3 (11%)	1 (3%)	0 (0%)	0.58
IE AV, N (%)	1 (2%)	1 (4%)	0 (0%)	0 (0%)	0.54
Anamnesis					
Revision surgery	38 (55.9%)	15 (55.6%)	17 (60.7%)	6 (46.2%)	0.68
Previously performed interventions					
Transluminal balloon valvuloplasty, N (%)	31 out of 38 (81.6%)	12 out of 15 (80.0%)	14 out of 17 (82.4%)	5 out of 6 (83.3%)	0.92
AVp, N (%)	4 out of 38 (10.5%)	1 out of 15 (6.7%)	2 out of 17 (11.8%)	1 out of 6 (16.7%)	0.64
TLBVP + aortic valvuloplasty, N (%)	3 out of 38 (7.9%)	2 out of 15 (13.3%)	1 out of 17 (5.9%)	0 out of 6 (0.0%)	0.38

Survival Rate

The actuarial survival rate for the entire follow-up period (19 years) in the overall group was 94.2% (5-year-96%, 10-year-94.2%). There were four fatalities. Of these, 3 were in the early postoperative period and 1 in the long-term period. In the schoolchildren group, the three-year survival rate was 100%, in the early childhood group, the 5-year survival rate was 93% and the 10-year survival rate was 89%. In the preschool group, the 5-year and 10-year survival rates were 96% (Figure 2). When comparing Kaplan-Meier curves, no statistically significant differences were obtained (P=0.6). All 3 hospital deaths

were related to impaired coronary blood flow due to stenosis of the reimplanted coronary arteries in the autograft. All three patients underwent Ross procedure without enlarging the fibrous ring. The ages of surgery were 1, 2, and 4 years. One fatal outcome occurred in reintervention on a RVOT conduit in a young patient (1.74 years old with an implanted Contegra conduit). The early postoperative period was complicated by an acute hemorrhagic cerebral circulation disorder in the right middle cerebral artery basin, which was the main link of thanatogenesis (Table 2). The medium-term results are presented in (Table 1).

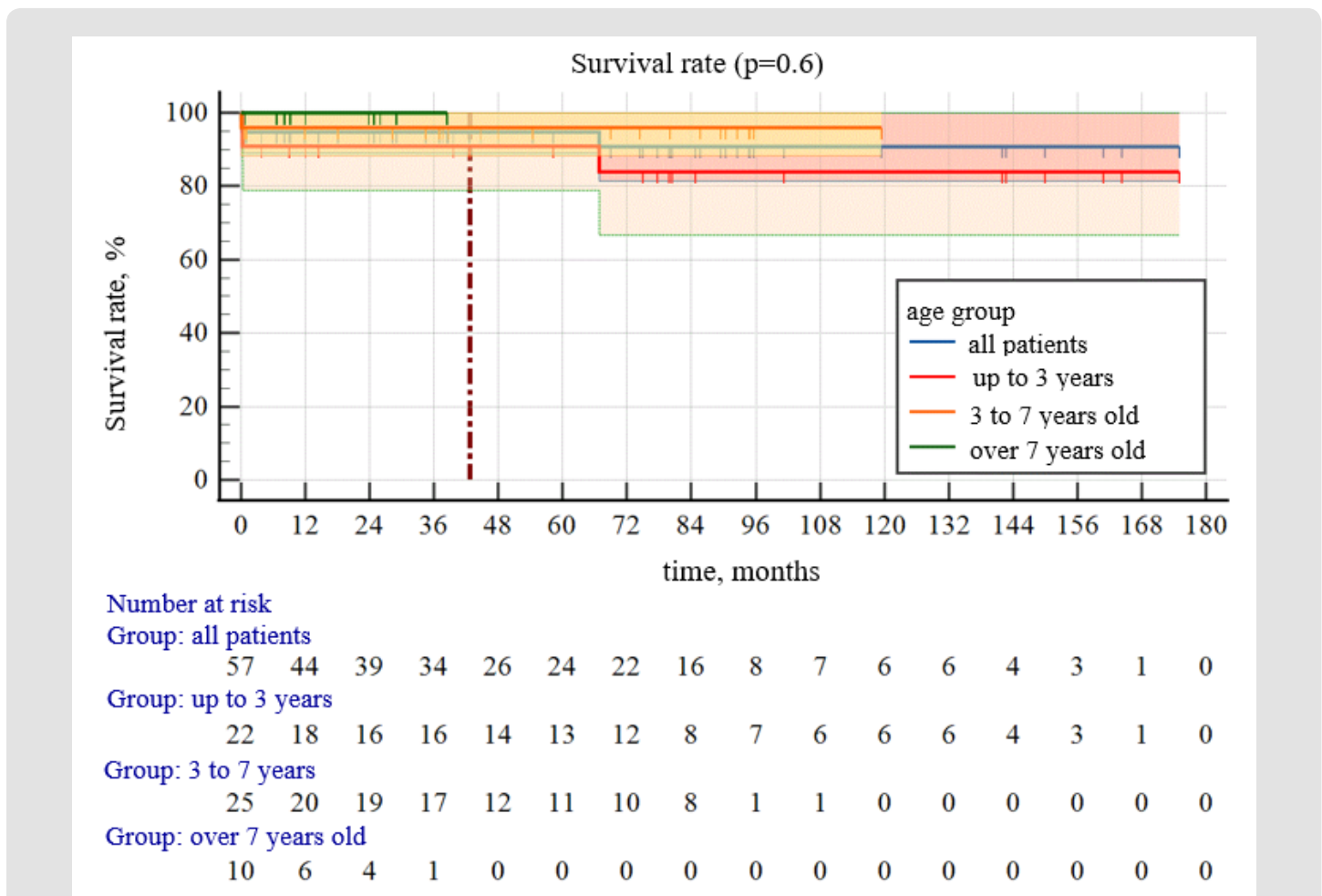


Figure 2: Actuarial Survival. Early childhood is from 1 to 3 years old, preschool is from 3 years old to 7 years old, and school age is from 7 years old. The dotted line indicates the median follow-up in the total group. Actuarial survival curves were not statistically significantly different when compared using the log-rank method (p=0.6).

Table 2: Mid-term results of surgery. *- comparison of results up to 3 years after surgery. Significance level with Bonferroni correction = 0.0167 (comparing 3 age groups).

On discharge	All (N=68)		Early childhood (up to 3 years)(N=27)		Preschool (3 to 7 years old) (N=28)		School (from 7 years old) (N=13)		p-value	
	Medium-term period	On discharge	Medium-term period	On discharge	Medium-term period	On discharge	Medium-term period	On discharge		
Follow-up time, months	-	42.6 [14.4;85.7]	-	76.3 [20.75;131.8]	-	51.2[32.8;89.7]	-	17.9[8.3;25.7]	0.00194	
Overall survival rate, %	-	94%	-	88.9% (24 out of 27)	-	96.5% (27 out of 28)	-	100% (13 out of 13)	0.38294	
Autograft Indicators										
Time of AV assessment, months	-	54.4[25.1;92]	-	78.8 [41.9;131.8]	-	69[37.3;91.5]	-	17.9[8.3;25.7]	0.00193	
Aortic fibrosus ring, mm	15[12;16;52]	19[17;22]	12[12;13]	18[16.8;22.3]	15.5[14;16.75]	19[17;20]	20[19;23]	22[20;22.5]	-	
	<0.001		0.012		<0.001		>0.05			
Maximum transaortic gradient, mm Hg.	10[7;14]	9[8;12]	10[7;14]	8[6;11]	10[8;14]	11[8;15]	7[7;9;5]	8[8;11]	0.52163	
	0.77		0.85		0.9		0.6			
Autograph dysfunction, N (%)	-	8 out of 68(10.3%)	-	2 of 27(7.4)	-	3 out of 28(10.7%)	-	3 out of 13(23%)	0.00011	
AK reoperations (open), N (%)	-	2 of 68 (2.9%)	-	2 out of 27(7.4%)	-	0 out of 28(0%)	-	0 out of 13(0%)	0.93106	
RVOT conduit Indicators										
Time taken to evaluate the RVOT conduit, months	-	38.4[12;85.7]	-	58.2[10;109]	-	47.9[28.3;89.5]	-	10.6[5;25]	0.00546	
Peak transprosthetic gradient, mm Hg.	9[7;12]	30[15;41]	10[7;12.5]	40[20.5;50]	9[7;12]	27[18;40]	7[6;8]	12.5[7;25]	0.00481	
	<0.001		<0.001		<0.001		<0.001			
Conduit dysfunction, N (%)	-	23 out of 68 (33.8%)	-	15 out of 27 (55.6%)	-	8 out of 28 (28.6%)	-	0 out of 13(0%)	0.63927	
Reoperations on the RVOT, N (%)	-	11 out of 68(16.2%)	-	7 out of 27(25.9%)	-	4 of 28 (14.3%)	-	0 out of 0 (0%)	0.53306	
Reinterventions, N (%)	Endo-vascular	-	6 out of 11(54.5%)	-	5 out of 7(71.4%)	-	1 in 4 (25%)	-	0 out of 0 (0%)	-
	Hybrid	-	3 out of 11 (27.3%)	-	1 in 7 (14.3%)	-	2 out of 4 (50%)	-	0 out of 0 (0%)	
	Open	-	2 out of 11 (18.2%)	-	1 in 7 (14.3%)	-	1 in 4 (25%)	-	0 out of 0 (0%)	

Dysfunction of the RVOT Conduit During Ross/Ross-Konno Surgery

The overall freedom from RVOT conduit dysfunction was 68% at 5 years and 35% at 10 years (Figure 3). The freedom from RVOT conduit dysfunction in school-aged patients was 100% over 3 years (with a median follow-up of school-aged patients of 10.6[5; 25] months).

The freedom from RVOT conduit dysfunction after 5 years was 90% in preschoolers and 65% in the early childhood subgroup (p=0.04 when comparing early childhood and preschool curves). From year 5 to year 7 of follow-up, survival curves differ in favor fewer instances of dysfunction in preschoolers (compared with early adulthood). After year 7, the freedom from dysfunction curves in the early childhood and preschool patients overlap.

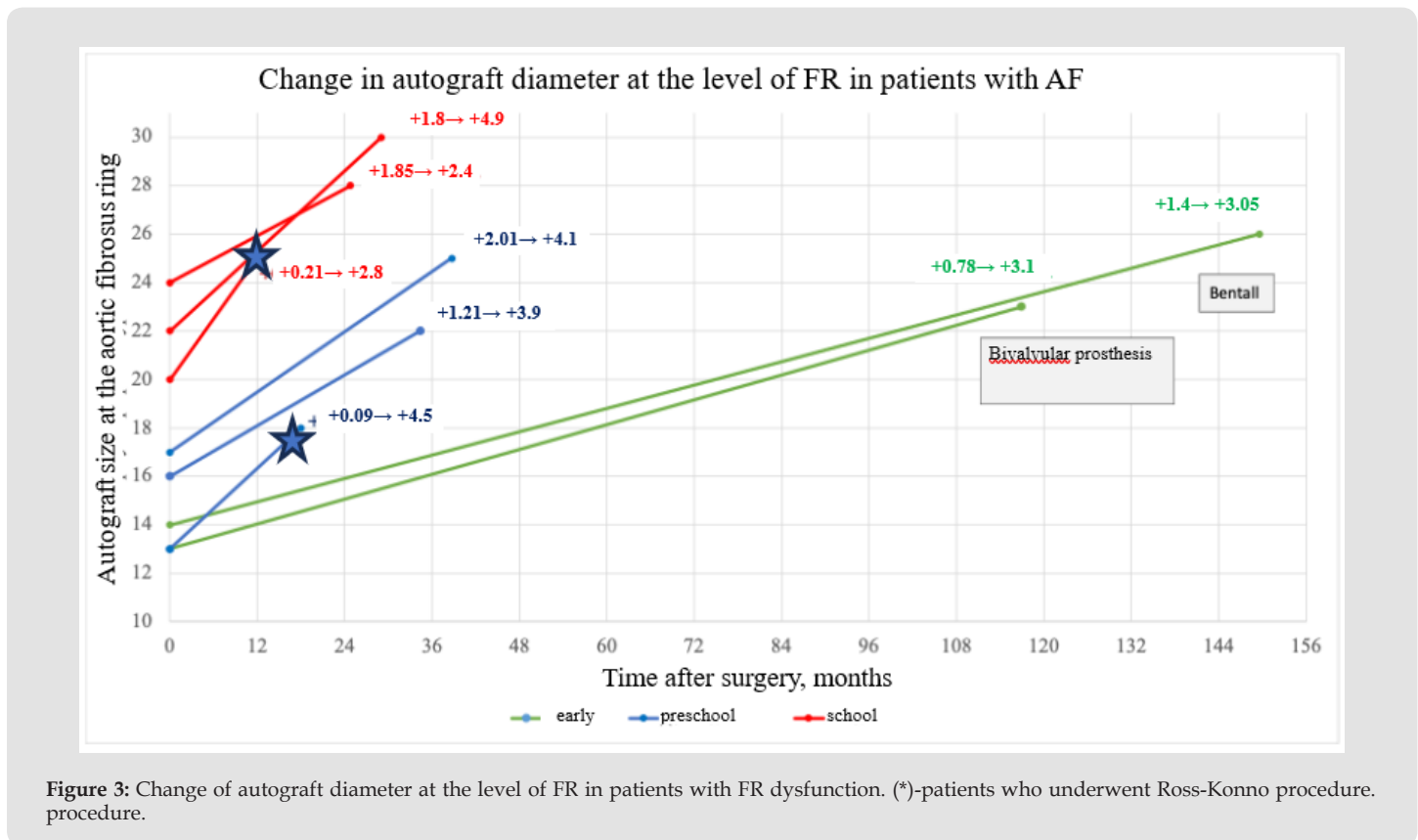


Figure 3: Change of autograft diameter at the level of FR in patients with FR dysfunction. (*)-patients who underwent Ross-Konno procedure.

Autograft Dysfunction in Ross/Ross-Konno Procedure

The overall freedom from autograft dysfunction was 85% at 5 years and 73% at 10 years. For ages over 7 years, 35% for 3 years, for ages 3 to 7 years, 85% for 5 and 10 years, and for ages under 3 years, 100% for 5 and 85% for 10 years. During the follow-up period, 8 cases of autograft dysfunction were identified (6 cases after Ross procedure and 2 cases after Ross-Konno procedure performed in patients aged 5 and 10) (Figure 3). In 6 cases there was a pronounced

insufficiency of the prosthesis, as evidenced by the presence of aortic root dilatation. In 2 other cases, autograft dysfunction was moderately insufficient, as evidenced by presence of marked dilatation of the aortic root in the area of the sinotubular ring. Autograft expansion at the level of the aortic fibrous ring was noted in all cases. Moreover, whereas initially the Z-score of the FR autograft for the aortic position was 0 to +2, at the time of dysfunction the Z-score of the AF was +2.4 (early childhood age group) to +4.9 (Figure 4).

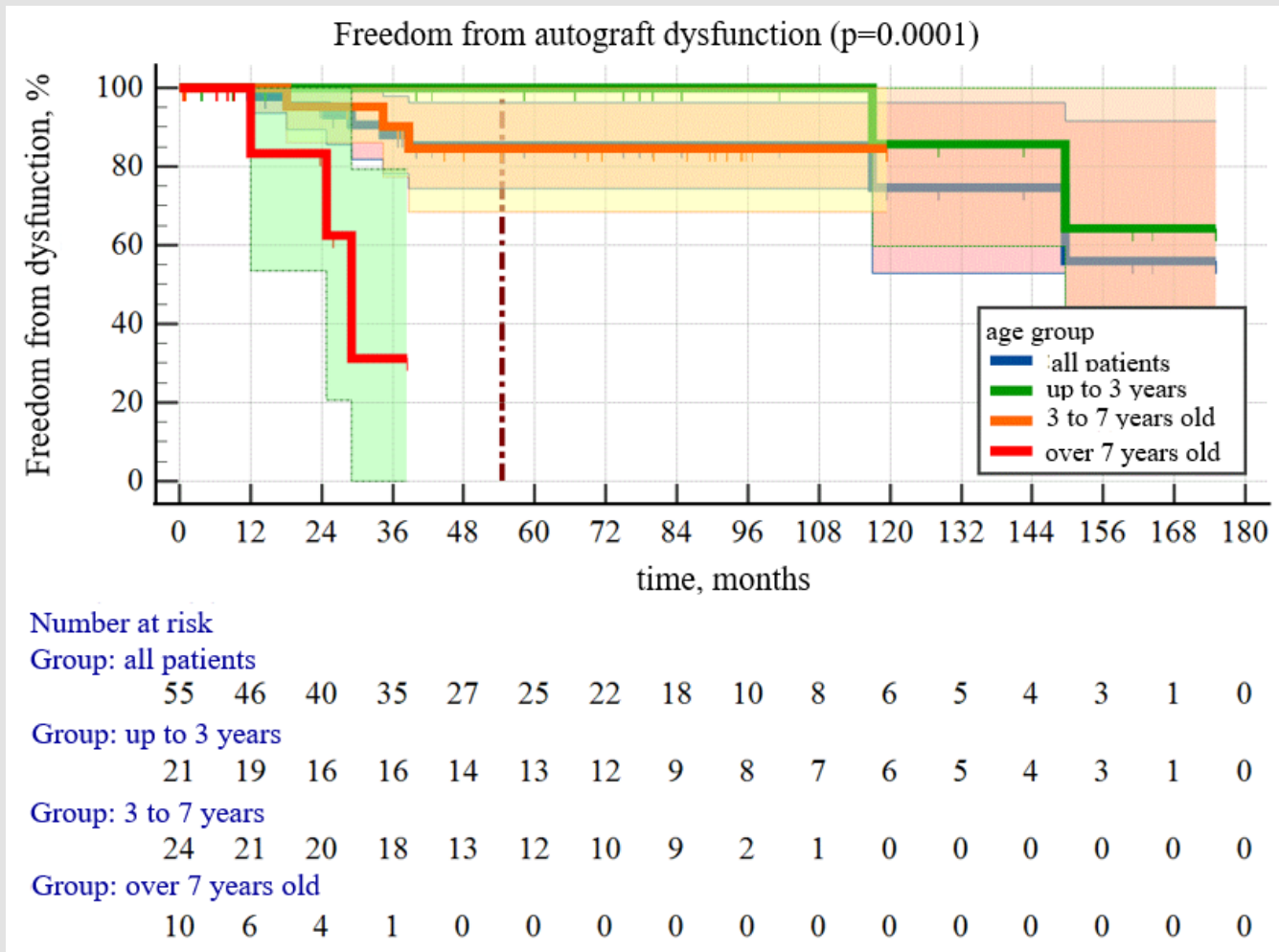


Figure 4: Freedom from autograft insufficiency greater than Grade 2. Early childhood is from 1 to 3 years old, preschool is from 3 years old to 7 years old, and school age is from 7 years old. The dotted line indicates the median follow-up in the total group. When comparing curves, the significance level is 0.0001.

The most significant dilatation was noted in two patients after performance of the Ross-Konno procedure, +5 mm at 1 year after surgery. Patients with detected autograft dysfunction are currently being observed, and a strategy for further management of their condition is being determined. Two patients underwent reintervention, as described below in the relevant section. When assessing the level of freedom from autograft dysfunction, it was found that there was an age-dependent difference in Kaplan-Meier curves as early as 1 year after surgery (significance level at log-rank comparison=0.0001). Patients in the early childhood age group had the longest autograft life without the development of significant dilatation, with school-age patients showing the earliest onset of dysfunction (Figure 4). Because we found 2 cases of early autograft dysfunction after performance of the Ross/Konno procedure, we performed a subanalysis to compare this type of intervention with results of the standard Ross procedure without enlarging the fibrosus ring.

Subanalysis: A Comparative Assessment of Freedom from Autograft Dysfunction following Performance of the Ross and Ross-Konno Procedures

The freedom from autograft dysfunction varied depending on the type of surgery performed on the patient. 8 patients aged 1 to 4 years (2[1.3; 5.03] years) underwent the Ross-Konno procedure. This procedure was chosen because they had a true narrow fibrous ring, which had a Z-score of -0.74[-1.52;0.07], which did not correspond to the size of the autograft, which had a Z-score for aortic position of +0.85[+0.2; +1.8]. Despite the fact that the patients' fibrosus ring size was within 1-2 standard deviations of the normal, we performed the enlargement because it was impossible to implant in the aortic position an autograft which had a seating size 3-4 mm larger than the annulus fibrosus. From the total patient base, we selected 23 similar clinical cases, comparable in age and autograft size, but on whom the

Ross procedure was performed. In these patients, the Z-score of the the aortic fibrosus ring was +1.06[0.34;1.95], and the Z-score of the autograft FR was +1.38[0.97;1.92]. The seating diameter of the autograft almost exactly corresponded to the dimensions of the FR, so there was no need to perform the enlargement of the FR.

When comparing age- and body weight-matched patients who underwent the Ross-Konno (N=8) and Ross (N=23) procedures, it was found that after the Ross procedure, the freedom from autograft dysfunction rate was 93% after 5 years, while for patients who underwent the Ross-Konno procedure the absence of autograft dysfunction rate was 63% at 5 years (the difference in the Kaplan-Meier curves was statistically significant) (Figure 5). The main mechanism for the

increased frequency of autograft dysfunction after the Ross-Konno procedure was the more pronounced dilatation of the autograft (+38% vs. +13% after Ross procedure) at the level of the aortic fibrosus ring due to the fact that the basal annulus of the aortic valve was crossed during the enlargement of the fibrosus ring, and a flap of the right ventricular myocardium was used as a patch for annulus expansion (Figure 6). It is likely that the right ventricular muscle tissue, which performs the function of dilating the fibrosus ring, and thus fact that the fibrous ring was open, were factors in the loss of aortic root support function. Under these conditions, the autograft dilated significantly faster under systemic pressure conditions, which caused its dysfunction.

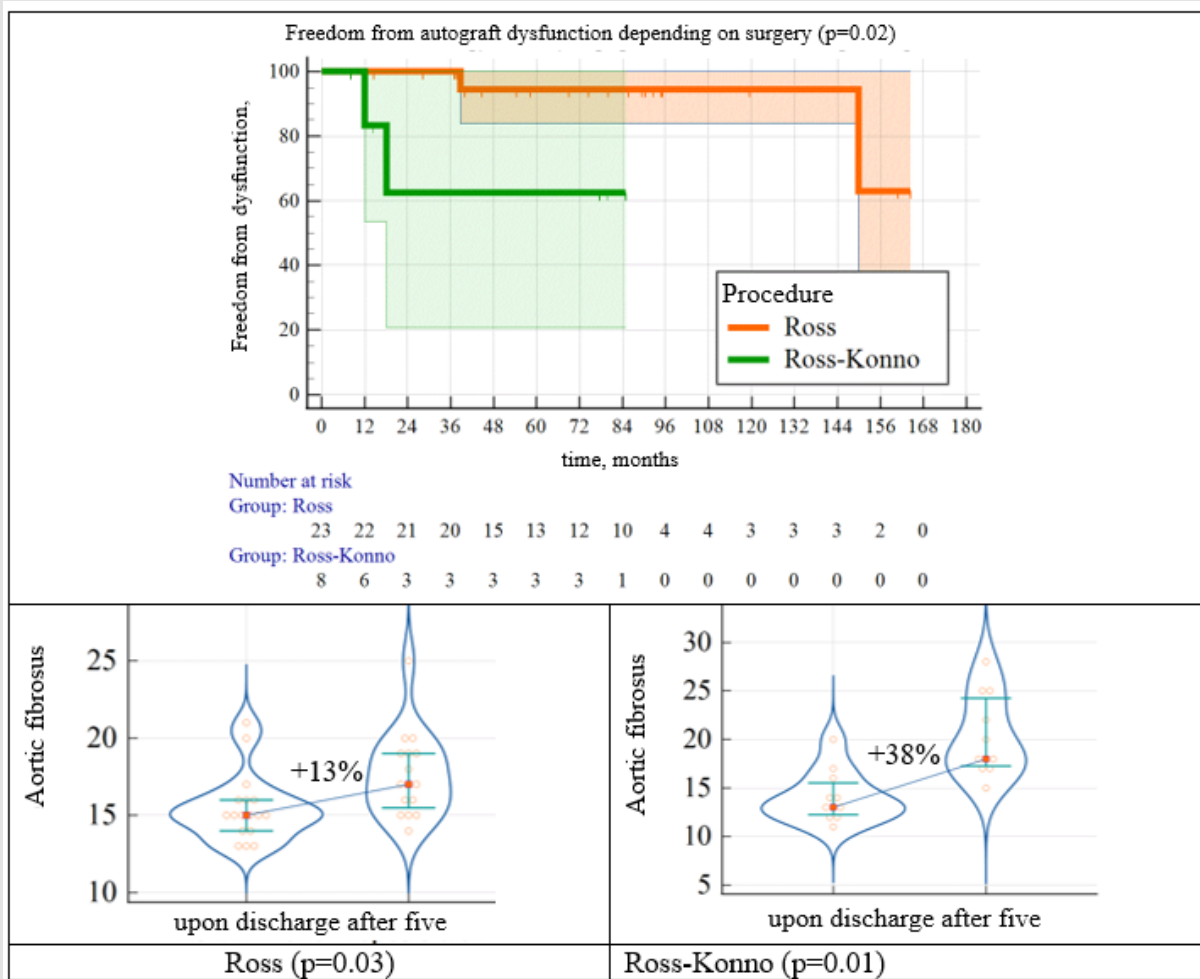


Figure 5: Comparison of freedom from autograft dysfunction and changes in the aortic fibrosus ring dilatation following Ross and Ross-Konno procedures.

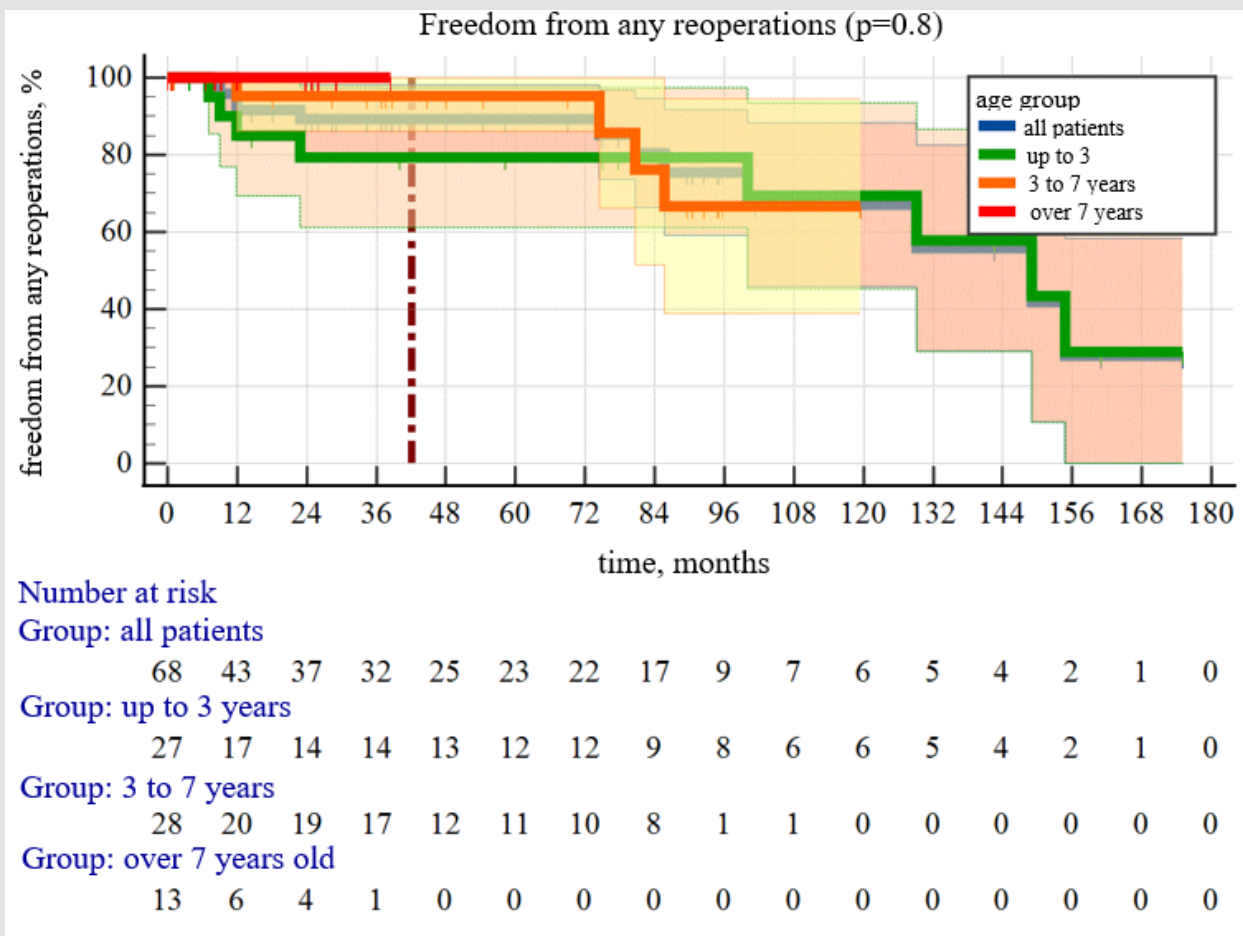


Figure 6: Freedom from any reoperations.

Certainly, our observation regarding earlier onset of dysfunction and more rapid dilatation of the autograft with the Ross-Konno operation requires further study. Although the performance of Konno enlargement of the FR AV was not a confirmed risk factor for autograft dysfunction.

Reinterventions After Performance of the Ross/Ross-Konno Procedure

Freedom from revision surgery in the overall group was 80% at 5 years and 65% at 10 years. Revision surgery was performed on a total of 12 out of 68 patients: 10 patients were treated for the dysfunction of the RVOT conduit without intervention on the autograft, 1

patient was underwent revision surgery for aneurysmal dilatation of the autograft and development of severe neo-aortic valve insufficiency without intervention on the RVOT conduit, and 1 patient underwent replacement of both aortic autograft and RVOT conduit due to dysfunction of both conduits (Figure 6).

Reoperations on Autograft

In the overall group, the actuarial rate for the freedom from autograft reoperations at 5 years was 100% and at 10 years was 85%. In 2 cases repeat surgery was performed 11 and 12 years after the Ross procedure, both in patients who had undergone the Ross procedure in early childhood (Figure 7).

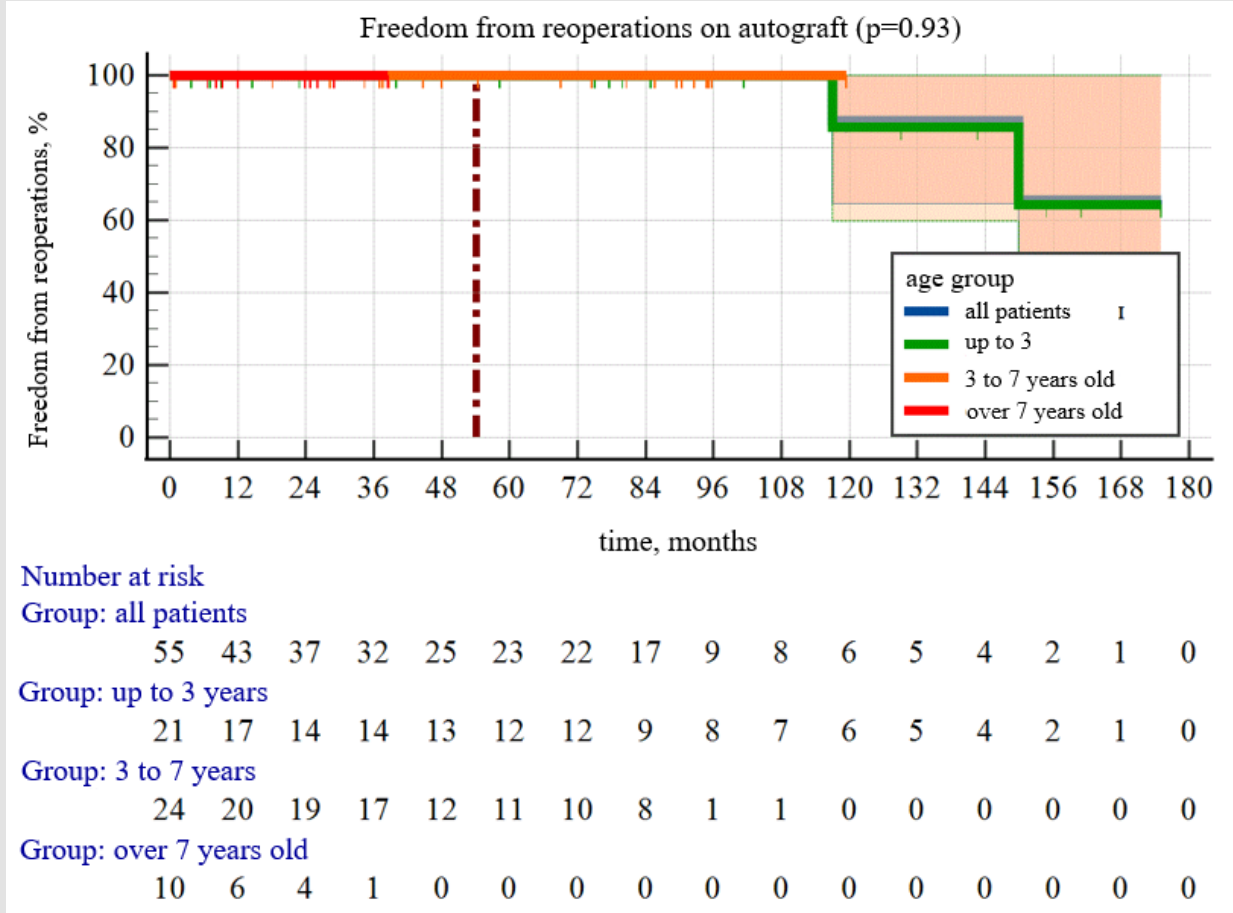


Figure 7: Freedom from reoperations on AV. Early childhood is from 1 to 3 years old, preschool is from 3 years old to 7 years old, and school age is from 7 years old. The dotted line indicates the median follow-up in the total group. Significance level for comparison by log-rank method = 0.93.

Reoperations on the RVOT

A total of 11 interventions were performed on the RVOT conduit. Endovascular operations aimed at angioplasty of stenosis in the area of distal anastomosis of the conduit with the pulmonary artery trunk were used most frequently (in 6 of 11 cases). Hybrid interventions were performed in 3 cases - pulmonary artery angioplasty was performed as the first stage, and after that, revision implantation of the RVOT conduit prosthesis was performed. In all cases, replace-

ment with a pulmonary allograft was performed. In 2 cases, only open intervention was performed to eliminate stenosis at the level of the valve. Freedom from reoperations on RVOT was 75% over the entire follow-up period. In the over 7 years group, 100% for 3 years, in the 3 to 7 years group, 97% for 5 years and 65% for 10 years, and in the under 3 years group, 80% for 5 and 10 years. Kaplan-Meier curves were not significantly different (p=0.93) due to the difference in median follow-up between groups (Figure 8).

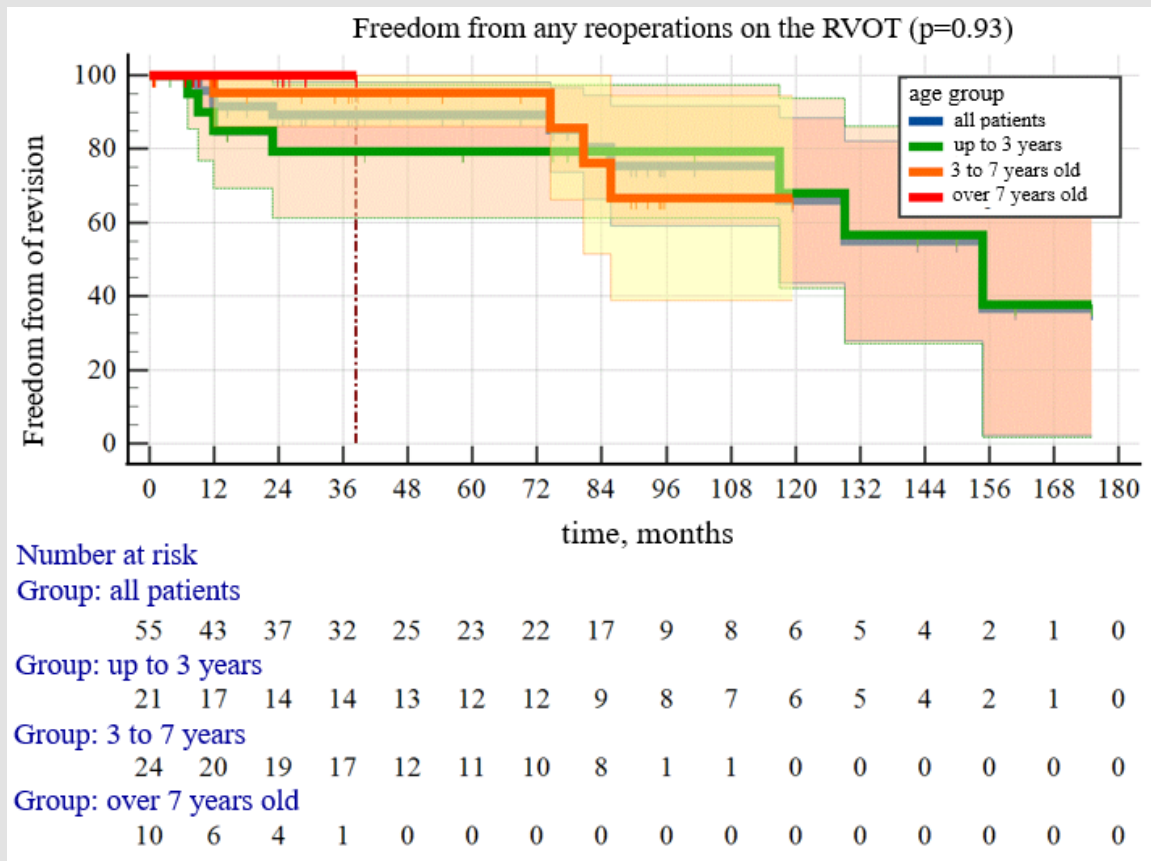


Figure 8: Freedom from any revision surgery on the RVOT valve. Level of significance at median follow-up of 3-4 years = 0.93.

Thus, where the Ross procedure is performed at an earlier age we can expect a longer period with no complications in the functioning of the autograft, but in this case we are forced to use much smaller RVOT conduits (in our case - from the bovine jugular vein), which in combination with the most intensive growth of the organism at an early age leads to early “outgrowing” of the conduit. However, in patients younger than 7 years, the Ross procedure is justified in terms of the long-term absence of complications in the autograft function and the lack of alternative small-diameter prostheses with similar hemodynamic characteristics.

Discussion

Indications for the Ross Procedure

In our department, Ross procedure was often performed in cases where there were urgent indications in the form of critical aortic valve stenosis or total insufficiency after TLBVP (Table 1), when the only treatment option was prosthesis, but an artificial aortic valve prosthesis of a suitable diameter was not available. In such cases, the

autopulmonary conduit was ideal from the point of view of service life and hemodynamic characteristics, and was therefore used. In addition to an ideal prosthesis-patient match, an additional advantage of the Ross procedure performed at an early age is the greater level of freedom from autograft dysfunction (compared with the preschool and school age groups), the absence of autograft calcinosis, which provides easier reoperation, and the absence of calcinosis transfer to the coronary artery buttons.

Choice of a RVOT Conduit

It cannot be overlooked that when the Ross procedure is performed in early life the need for small-diameter conduits in the pulmonary position remains a disadvantage. The most readily available conduits, the Contegra conduit, used by us, was not prone to a high incidence of prosthetic endocarditis (although this problem is highly relevant for Contegra [5]) and generally provided a reasonably good functional life, which was limited mainly by rapid “outgrowing”, which is also inevitable for any other small-diameter “non-growing” conduit

[6-11]. In global scientific literature there are almost no systematic comparisons of the results of using different conduits when performing the Ross procedure [11-14]. However, a significant problem with the use of the Contegra conduit is its structural degeneration and tendency to calcinosis involving the distal anastomosis and pulmonary artery region, which is less susceptible to pulmonary allograft [7].

According to our subanalysis of the freedom from dysfunction of the RVOT conduits following the use of pulmonary allograft or Contegra (for the purposes of the analysis, we selected 10 prostheses of each type with comparable diameters ranging from 18 to 20 mm, all implanted at an early age) it appeared that the pulmonary allograft

demonstrated a greater level of absence of dysfunction, probably because of greater resistance to structural changes (Figure 9). Due to the difficulties encountered in reprothesizing Contegra, we almost completely abandoned the use of this type of conduit when conducting the Ross procedure, a decision which was based on global data on the lower degree of calcification of pulmonary allografts. We also have high hopes for bicuspidalized pulmonary allografts, which may provide better surgical outcomes in young children and be less affected by calcinosis, which will greatly simplify revision implantation of the prosthesis [7] (Figure 10).

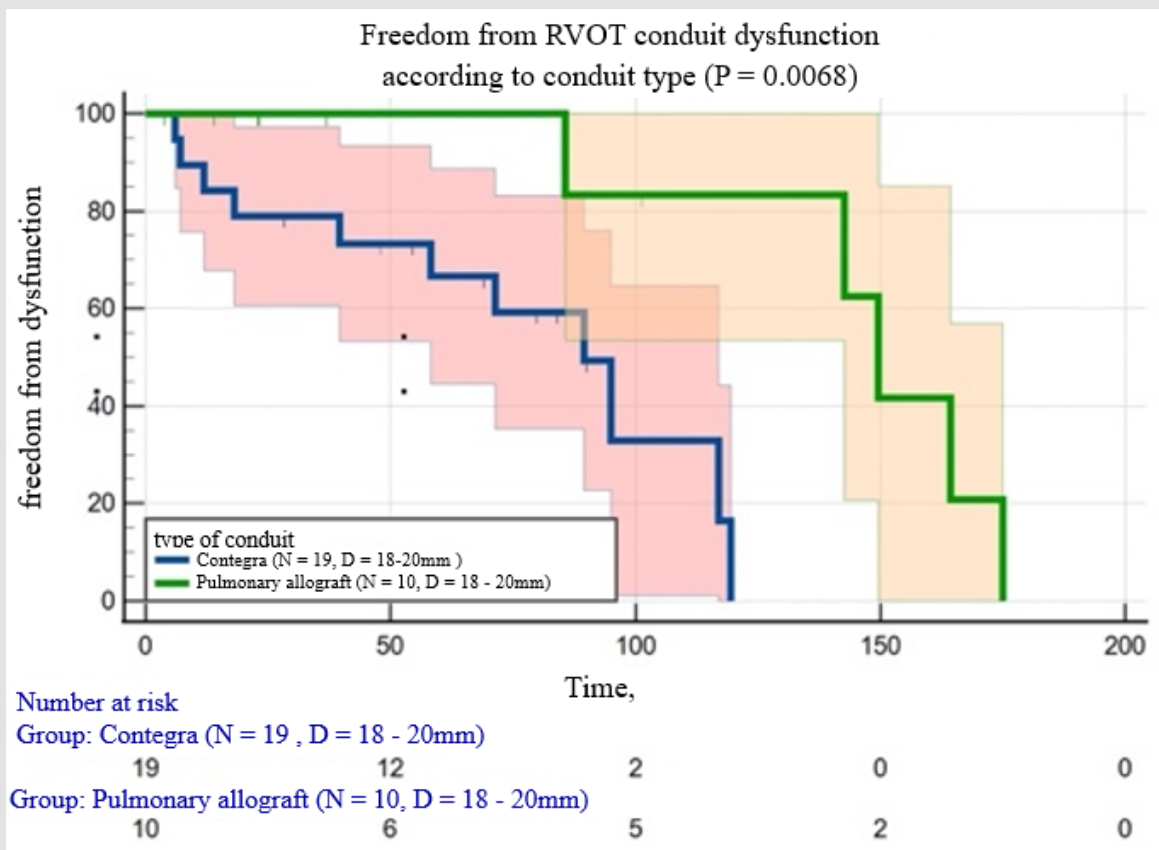


Figure 9: Freedom from of Contegra and pulmonary allograft conduit dysfunction. Significance level 0.007.

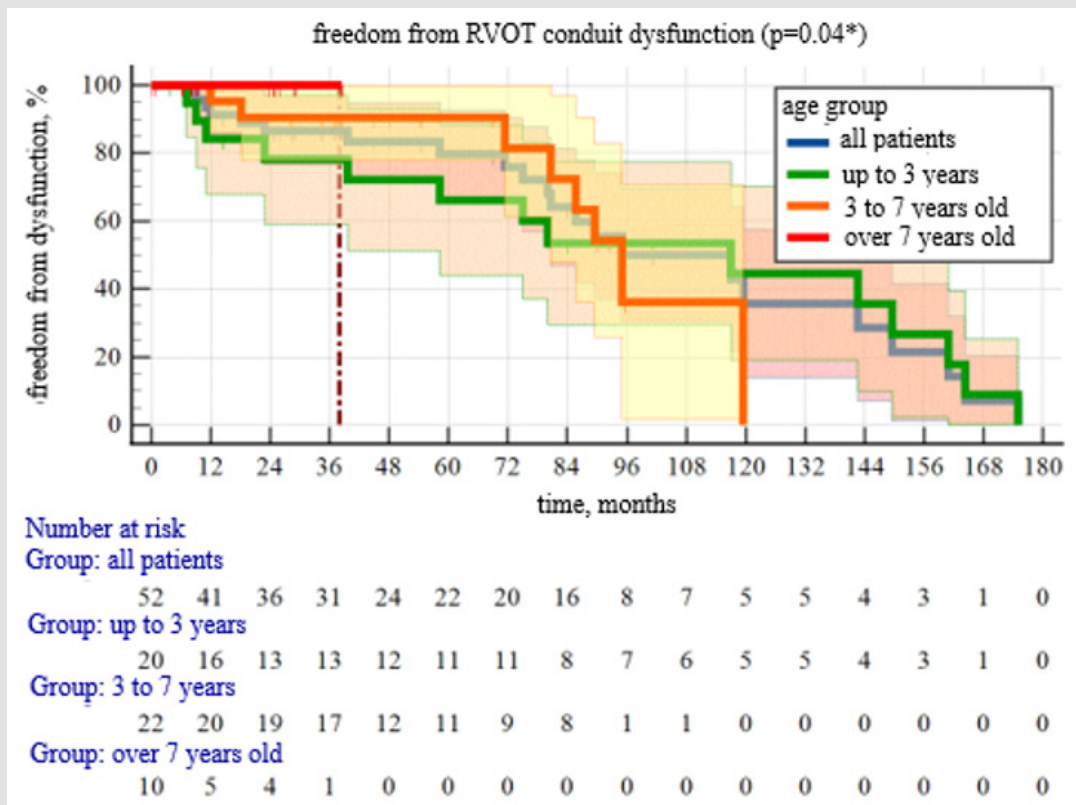


Figure 10: Freedom from RVOT conduit dysfunction as a function of age group. Early childhood is from 1 to 3 years old, preschool is from 3 years old to 7 years old, and school age is from 7 years old. The dotted line indicates the median follow-up in the total group. When the curves were compared by log-rank method, the significance level was 0.639, but when they were adjusted to a median follow-up of 38 months, the significance level was 0.04.

Conclusion

Our experience with the Ross procedure shows that young children (under 3 years of age) have the highest level of freedom from autograft dysfunction, but are most at risk of dysfunction of the RVOT conduit due to its small size. Preschool and school-aged children have a high level of freedom from complications related to RVOT conduit, (because in the majority of cases large-diameter RVOT conduits are used), but the autograft is more susceptible to dysfunction, mainly through the mechanism of dilatation under systemic pressure conditions. In our opinion, the Ross procedure is most appropriate in the early childhood and preschool period, when a small-sized prosthesis is required. In school-age and older patients more prosthetic alternatives are now becoming available, and these need to be studied in detail.

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Conflict of Interest

All authors declare that they have no conflict of interest.

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