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ORIGINAL ARTICLE

Comparison of the Ross/Ross–Konno aortic root in children before and after the age of 18 months†

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Abstract

OBJECTIVES: Evaluation of long-term results after the Ross/Ross–Konno operation in patients <18 months of age, focusing on pulmonary autograft performance.

METHODS: Retrospective analysis of patients who underwent the Ross/Ross–Konno operation (1991–2011). Data were obtained from hospital records and follow-up was 100% complete.

RESULTS: Between January 1991 and December 2011, 140 patients underwent the Ross/Ross–Konno operation and 22 patients were <18 months of age (male/female: 15/7). The median age was 166 days. 14 patients had a Ross operation and 8 patients a Ross–Konno operation. Presentation at surgery was aortic valve stenosis in 13, regurgitation in 7 and mixed disease in 2. Only 4 patients (18%) had no surgery prior to Ross/Ross–Konno, and among the others the previous most frequent operation was aortic valvotomy (55%). There were 3 early deaths, all in high-risk cases with poor preoperative left ventricular function. At discharge there was no neoaortic regurgitation in 10, trivial in 4 and mild in 5. The median follow-up is 10.8 years (range 0.96–21). There was 1 late death due to progressive ventricular dysfunction 2.4 years after Ross–Konno and mitral valve replacement. Survival for patients <18 months was 81% at 5–10 years (18 patients) and for older patients was 98.2% at 5–10 years. There was no neoaortic regurgitation in 8 patients, mild in another 8 and moderate in 2 with a freedom from moderate regurgitation significantly lower in comparison with older patients (100–80% vs 83.5–73.4% at 5–10 years). Freedom from aortic reoperation for patients <18 months was significantly higher compared with older (100 vs 95.4–84.4% at 5–10 years, $P < 0.04$). Reoperation in the RVOT was higher in younger patients compared with the other (85–64.6% vs 97.2 and 84.7% at 5–10 years $P = 0.02$). *Z*-score aortic root diameter remained constant in the <18-months group compared with significant dilatation in the >18-months patients ($P < 0.01$).

CONCLUSIONS: Ross/Ross–Konno can be invaluable in the younger age group but not without risk in the setting of ventricular dysfunction. Long-term performance of the neoaortic valve is significantly better than in older children, which is related to maintenance of normal root dimensions compared with progressive dilatation in patients undergoing Ross/Ross–Konno at older ages.

Keywords: Congenital heart disease • Aortic valve disease in children • Left ventricular outflow tract obstruction • Ross procedure • Ross–Konno procedure

INTRODUCTION

The first Ross procedure, performed in an adult patient in 1967 [1], introduced the concept of using the pulmonary valve as an autograft to replace the aortic valve. The Konno modification of the Ross consists in adding an incision between the right and left coronary cusps extended through the aortic annulus to the ventricular septum with the aim of enlarging the left ventricular outflow tract (LVOT).

The Ross/Ross–Konno operation is particularly useful in the paediatric and adolescent age groups due to the potential growth of the pulmonary autograft as neoaortic valve, the avoidance of anticoagulation, the scarcity of very small aortic valve prosthesis and the flexible application to managing complex forms of LVOT obstruction [2–6].

However, it is a complex and technically demanding procedure with long-term concerns related to progressive neoaortic root dilatation, associated aortic regurgitation and the need for reoperation for replacing/repairing the autograft and/or the allograft used to reconstruct the right ventricular outflow tract (RVOT) [7–11]. This has led to more conventional valve replacements being considered in older children to avoid the complications of the Ross procedure [12]. Despite these reservations, the Ross operation and Ross–Konno have demonstrated outstanding outcomes in infants and small children where no other surgical options were possible [2–6].

The most dramatic change in aortic root dimension tends to occur early after the Ross procedure, related to the relatively

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thinner and weaker arterial wall of the pulmonary autograft compared with the native aorta.

This study aimed to investigate whether performing the Ross at an earlier age protected against pathological root dilatation due to the lower wall tensions in a smaller aorta and to different adaptive capability of younger patient autograft.

MATERIALS AND METHODS

The study is a cross sectional analysis of all patients who underwent the Ross/Ross–Konno operation between January 1991 and December 2011 at the Birmingham Children's Hospital, UK.

End points were mortality, grade of neoaortic valve regurgitation, aortic root dimension, aortic valve reoperation (aortic valve replacement, aortic root replacement or reduction aortoplasty) and RVOT reoperation.

Pre Ross/Ross–Konno history, demographic data, indications for surgery, type of operation and hospital course were obtained from patient's chart and hospital records. Follow-up data were obtained by reviewing cardiological charts and echocardiograms until December 2012 if patients were followed at the Birmingham Children's Hospital and/or by patient's data retrieved from other cardiology departments if they were followed outside. Follow-up was 100% complete.

Cardiac procedures performed prior to the Ross/Ross–Konno operation were recorded and patients were analysed according to the original valve pathology as well as by valve status at the time of surgery (such as in cases where aortic balloon valvotomy or surgical valvotomy changed the pathological mechanism from stenosis to regurgitation). Main indications for surgery were classified as stenosis, regurgitation and mixed in the case there was an equivalent presence of both.

For each patient followed at the Birmingham Children's Hospital, detailed review of all echocardiograms from the time of the operation to the most recent available was performed. Degree of neoaortic valve regurgitation (classified as none, mild, moderate and severe), aortic stenosis and the presence of left ventricular outflow tract obstruction (LVOTO) were recorded. Aortic root diameters were measured in end-diastole parasternal long axis view considering three levels: (i) aortic annulus (defined by the hinge point of the aortic cusps), (ii) sinuses of Valsalva, (iii) sinutubular junction as previously published guideline for evaluation of the aortic root dimensions [13, 14]. Aortic root dimensions were obtained from all echocardiograms, standardized to body surface area and expressed as *z*-scores [15].

The aim of the study was to investigate whether performing the Ross at an earlier age protected against pathological root dilatation and valve regurgitation; 18 months was considered to be the most appropriate age cut-off for the following reasons. There have been reported evidences, based on human cardiac valve histopathological studies, showing that a profound remodelling on aortic and pulmonary valve take place during the first and second year of life [16]. After this age the remodelling process decreases and almost stops, then semilunar valves lose their adaptive/remodelling capacity [16, 17]. Following these evidences of the remodelling process, the age was tested and 18 months was found as a natural age cut-off between younger and older patient groups.

Data were analysed using statistical programme (StataCorp. 2011. Stata Statistical Software: Release 12. College Station, TX, USA: StataCorp LP). Data were expressed as mean ± standard deviation and as median and range when appropriate. Tests used were Student's *t*-test, χ^2 and Fisher's exact tests; Kaplan–Meier method analysis was performed for event-free and survival curves. Conversion of aortic absolute diameter to *z*-score was done using previous published regression equations for calculation of *z*-scores of cardiac structures infants, children and adolescents [15]. Differences in *z*-scores were analysed with non-parametric regression models.

RESULTS

From January 1991 to December 2011, 140 patients underwent a Ross or Ross–Konno operation at the Birmingham Children's Hospital. There were 90 males (64%) and 50 females (36%). A total of 120 patients underwent a Ross operation (86%) and 20 a Ross–Konno (14%). In the <18-months group, there were 22 patients, with 14 (64%) Ross and 8 (36%) Ross–Konno. In the >18-months group, there were 118 patients with 106 (90%) Ross operations and 12 (10%) Ross–Konno. The Ross and Ross–Konno operations have different distribution within the two groups, and Ross–Konno is more frequent in the <18-months group. In this group median age at surgery was 0.46 years (range: 166 days to 1.38 years) and in the older group was 11.26 years (range: 1.5–35.4 years); among them, there were 8 adult patients that were referred to the unit for previous cardiac surgery at the Birmingham Children's Hospital or because affected by an aortic valve congenital disease.

The aortic valve morphology in the younger patients was bicuspid in 14 patients (64%), indeterminate in 5 and tricuspid in 2; the remaining case was a failure of a previous implanted aortic homograft. In the >18-month group, the aortic valve was mainly bicuspid (66%); among the less frequent aortic valve anomalies, there were 3 patients who underwent Ross/Ross–Konno for dysfunction of previously implanted prosthetic mechanical aortic valve and 1 patient for failure of previously implanted aortic homograft. Two remaining patients had a normal anatomical aortic valve with severe regurgitation in one and severe LVOTO after Senning operation in the other. Detailed aortic valve morphologies for both age groups are listed in Table 1.

The primary aortic valve disease was mainly congenital aortic valve stenosis in 77% of the patients (108/140 patients). However, due to previous interventions, the indications for surgery at the time of Ross procedure were different: mixed aortic valve disease in 65 patients (46%), aortic valve regurgitation in 45 (32%) and pure aortic stenosis in 27 (19%). In the <18-months group, indications were aortic stenosis in 13 (59%), aortic regurgitation in 7 (32%) and mixed in another 2 patients (9%). Indications for surgery in the younger group were significantly different ($\chi^2 = 29.84$; $P < 0.001$) from the older group in which mixed disease (53%) represents the main indication followed by aortic valve regurgitation (32%) and aortic valve stenosis (12%) (Table 1).

The majority of the patients underwent an aortic valve procedure (surgery and/or catheter intervention) before the Ross/Ross–Konno operation. A total of 80 patients (57%) underwent surgery prior to the Ross/Ross–Konno for a total of 134 operations (1.67 surgeries per patient). Aortic surgical valvotomy accounted for 47% of all operations, followed by LVOTO resection (16%). The remaining procedures are listed in Table 2. Among the <18-months group, 18 patients (82%) underwent an operation prior the Ross/Ross–Konno, it was a single procedure in 16 patients and 2 procedures in 2 patients. In the >18-months group 62 (52%), patients had previous surgery; in 44 it was a single operation and in 18 multiple operations (Table 2). In the overall

Table 1: Patient demographic data

	Age <18 months (n = 22)	Age >18 months (n = 118)	Total (n = 140)
Demographic			
Male/female	15/7	75/43	90/50
Age (years)	0.46 (0.19–1.38)	11.26 (1.5–35.4)	9.88 (0.19–35.4)
Weight (kg)	6.45 (3–11.5)	38.5 (8.7–98)	31.1 (3–98)
Height (cm)	63 (48–77)	146 (76–185)	136 (48–185)
Body surface area (m^2)	0.16 (0.08–0.26)	0.84 (0.21–1.78)	0.71 (0.8–1.78)
Aortic primary anatomy			
Bicuspid	14 (64%)	77 (65%)	91 (66%)
Tricuspid	2 (9%)	23 (19%)	25 (18%)
Indeterminate	5 (23%)	12 (10%)	17 (12%)
Normal[a]	–	2	2
Others[b]	1	4	5
Indication for surgery			(χ^2 = 29,84; P = 0.00)
Regurgitation	7 (32%)	38 (32%)	45 (32%)
Stenosis	13 (59%)	14 (12%)	27 (19%)
Mixed	2 (9%)	63 (53%)	65 (46%)
Other	–	3	3

[a]Normal aortic valve comprehends: one case of normal aortic valve with severe regurgitation and another one with normal valve but severe LVOTO after Senning operation.
[b]Others: patients with previously replaced aortic valves, 3 patients had dysfunction prosthetic aortic valve and 2 (1 patient <18 months) had dysfunction of implanted aortic homograft.

Table 2: Surgery before Ross/Ross–Konno operation

	<18-Months group (n = 22)	>18-Months group (n = 118)	Total (n = 140)
Previous operations			
None	4 (18%)	56 (47%)	60 (43%)
1 operation	16 (73%)	44 (37%)	60 (43%)
2 operations	2 (9%)	13 (11%)	15 (11%)
≥3 operations	–	5 (4%)	5 (3%)
Type of operations			
Aortic valvotomy	12	51	63
LVOTO relief	2	19	21
Other	3	18	21
Coarctation repair	4	10	14
VSD closure	2	7	9
Arch repair	2	4	6
Total n operation	25	109	134

Arch repair using patch and that requires bypass through a midline sternotomy; Coarctation repair: coarctation resection and direct anastomosis through a thoracotomy; LVOTO relief: left ventricular outflow tract resection; VSD: ventricular septal defect.

population, 43 patients underwent a cardiac catheter intervention prior to the Ross/Ross–Konno operation. There were a total of 47 cardiac catheter procedures like aortic balloon valvuloplasty (n = 43) and coarctation balloon dilatation (n = 4).

The Ross and Ross–Konno operations were performed using the aortic root replacement technique for implantation of the pulmonary autograft. The technique remained the same during the entire period of the study using two layers of continuous polypropylene suture lines to implant the root. In case of LVOTO, the outflow tract was enlarged using the modified Konno technique. Reconstruction of the RVOT was performed using pulmonary homograft in 121 cases (86%), aortic homograft in 12 (8.5%), Contegra® conduit in 6 cases and Tissuemed® conduit in 1, and the last two conduit types were used when appropriate-sized homografts were not available.

In the <18-months group, the Ross–Konno operation was more frequent (36%) than in the older group (10%) and aortic homograft or Contegra® together were used more frequently (50% vs 6.8%), reflecting the limited availability of adequate size pulmonary homografts. There was no difference in the bypass or cross-clamp times between the two groups, despite the increased incidence of Ross–Konno in the younger patients (Table 3).

Among the younger group, 2 (9%) patients were neonates and underwent the Ross–Konno operation at 6 and 20 days of life. In the first patient, Ross–Konno was used as primary surgical option for severe congenital aortic valve stenosis associated with severe LVOTO. The second patient underwent the Ross–Konno operation after failure of surgical aortic valvotomy for mixed disease of the aortic valve, discrete LVOTO and impossibility to wean from ventilator support. Both patients survived.

The complications experienced were low cardiac output (LCO), cardiac arrest, heart block (transient and permanent) and other minor complication listed in Table 3. In the <18-months group, 31.8% of the patients experienced a complicated postoperative course vs 7.6% of the patients in the >18-months group (P = 0.04). There was no heart block in the younger group in contrast with the older group in which 3 patients (2.8%) developed heart block requiring permanent pacemaker. Two were Ross–Konno procedures and one was a Ross operation.

Two patients in the <18-months group had LCO associated with severe mitral regurgitation. Both presented at surgery with aortic valve stenosis, mitral regurgitation and dilated left ventricle. One patient underwent a Ross operation at the age of 6 months, but the mitral regurgitation did not improve in the postoperative course, and underwent mitral valve replacement after 2 days. The patient died 3 days later for untreatable LCO state due to severe compromised left ventricular function. The second case

Table 3: Operation data and postoperative complications

Operation data	<18-Months group (n = 22)	>18-Months group (n = 118)	Total (n = 140)	
Ross	14 (64%)	106 (90%)	120 (86%)	
Ross–Konno	8 (36%)	12 (10%)	20 (14%)	
Cardiopulmonary bypass (min)	127 (90–323)	141.5 (101–378)	141 (90–378)	P = 0.58
Aortic cross clamp (min)	104 (80–151)	101 (75–202)	102 (75–202)	P = 0.55
Complications				
Chest left open	7	5	12	
Low cardiac output	2[a]	2	4	
Cardiac arrest	3[b]	1	4	
Pacemaker implantation	–	3	3	
Temporary atrioventricular block	–	3	3	
Neurological damage	1		1	
Diaphragm palsy	1		1	
Hospitalization (days)	13.5 (6–86)	7 (5–66)	7 (5–86)	P = 0.00

[a]1 patient had LCO associated with mitral valve regurgitation and underwent mitral valve replacement.
[b]1 patient had cardiac arrest for acute pulmonary homograft disruption and emergency RVOT conduit replacement.

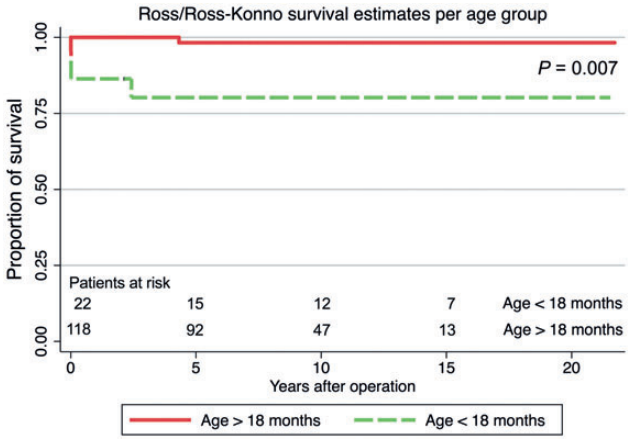

Figure 1: Ross/Ross–Konno survival estimates (Kaplan–Meier) per age group.

underwent a Ross–Konno at 22 days of age and the mitral valve was replaced 3 days after surgery. He was discharged home with no aortic regurgitation but with mildly dilated left ventricle. In the following years, the left ventricular dilatation progressed and he died on the transplant waiting list 2.4 years later.

There were 4 early deaths (2.8%). Three were in the younger age group (13.6%) and 1 on the older age group (0.8%). In the younger age group, all early death presented with left ventricular dilatation and dysfunction; in 2 cases there were associated LVOTO and moderate-to-severe mitral valve regurgitation. The surgery was performed at 5, 7 and 13 months. One early death occurred in the older age group, and the patient was 13 years old with impaired LV function preoperatively who had persistent and untreatable left ventricle dysfunction postoperatively. Era of operation was also important, with a significantly higher early mortality in the first 5 years of the study (13.6%) compared to subsequent years (0.8%) (P = 0.01).

Survival for patients <18-months was 81% at 5 and 10 years and for the >18-months group was 98.2% at 5 and 10 years. Survival curve plotted for age showed a hazard ratio of 22.3 with P = 0.01 for the <18-months group (Fig. 1).

The median hospital stay for the >18-months group was 7 days (range: 5–66 days) compared with 13.5 days (range: 6–86 days) for the younger age group (P < 0.001). Predischarge echocardiogram demonstrated no aortic valve regurgitation in 83 patients (61%) and mild in 53 patients (39%); there were no moderate/severe aortic valve regurgitation. All patients were discharged without clinically significant residual LVOT obstruction and/or RVOT conduit stenosis or regurgitation.

The overall median follow-up duration was 10.8 years (range: 0.96–21 years). There were 2 late deaths, 1 patient for each group, at 2.4 years (the <18-months group) and 4.3 years (>18-months group). The first patient has been previously described and the second patient died at 4.3 years for progressive left ventricular dilatation and dysfunction associated with moderate aortic valve regurgitation.

Among the 134 survivals, 26 (19.4%) underwent reoperation related to the Ross/Ross–Konno; 15 underwent RVOT reintervention, 6 underwent reintervention only in the aortic valve and/or aortic root and the remaining 5 underwent both.

Reinterventions on the aortic root occurred in 11/116 (9.4%) patients, all from the >18-months group, after a median time of 6.2 years (range: 1–12.9 years) from the Ross/Ross–Konno. The reoperations were aortic valve and root replacement in 6, isolated aortic valve replacement in 3 and aortic valve cusps suspension associated with aortic root reduction plasty in the other 2. Curve for aortic valve and root reoperation is shown in Fig. 2. Freedom from aortic valve and root reoperation for the <18-months group is 100% (at 5 and 10 years) and for the >18-months group is 95.4 and 84.4% (at 5 and 10 years) (P = 0.04).

Reintervention on the RVOT was performed in 20 patients; 7 (38%) were in the younger age group compared with 13 (11%) in the older group (P = 0.02). Kaplan–Meier curve for reintervention in the RVOT is shown in Fig. 3. Freedom from RVOT reintervention for the <18-months group is 85 and 64.6%, respectively at 5 and at 10 years, and for the >18-months group 97.2 and 84.7%, respectively at 5 and at 10 years.

The grade of aortic regurgitation among the survivors at the follow-up was none in 47 patients, mild in 60 and moderate in 8. The shift from mild to moderate occurred in 22 patients (17%)

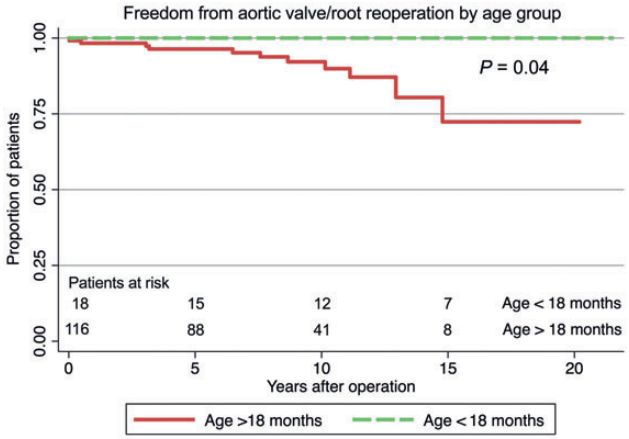

Figure 2: Freedom from aortic valve/root reoperation per age group.

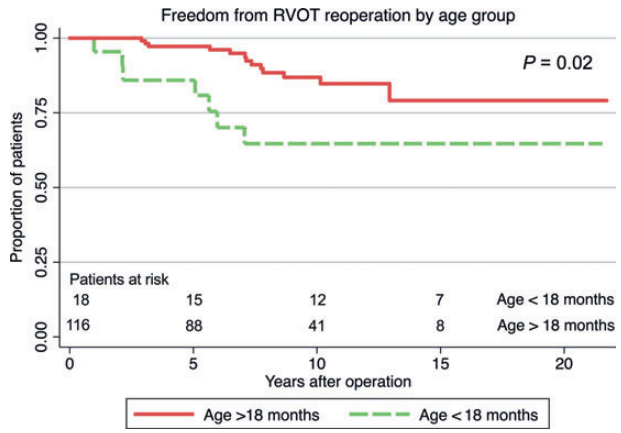

Figure 3: Freedom from RVOT reoperation per age group.

in total, and 2 of them (2/18 = 11%) in the younger group at 9 and 11.7 years after operation. In the >18-months group, 20 patients (20/108 = 18%) had progression of aortic regurgitation from mild to moderate after a mean time of 2.89 ± 2.7 years. Freedom from aortic valve regurgitation more than moderate at 5 and 10 years is 100 and 80% in the <18-months group and 83.5 and 73.4% in the >18-months group ($P = 0.05$).

Aortic root dimensions were followed serially on echocardiogram and standardized to body surface area and expressed as *z*-scores. The results are shown in Fig. 4 and demonstrate a remarkable stability of the aortic root dimension in the younger age group compared with the progressive dilatation seen in the older age group. Mean aortic root *z*-scores in the younger age group at the first control were +2.17 ± 1.46 for the annulus, +2.76 ± 1.45 for the sinuses of Valsalva and +3.15 ± 1.12 for the sinutubular junction. At the last control, mean *z*-score for the annulus was +3.06 ± 1.14, for the sinuses of Valsalva +3.15 ± 0.82 and for the sinutubular junction +2.56 ± 1.16. Progression in the *z*-scores for the three regions of the aortic root was respectively +0.89; +0.39 and −0.59 and was not significant (respectively $P = 0.13$; $P = 0.53$; $P = 0.45$).

In the older group, the aortic root *z*-scores move from +1.17 ± 1.11 to +2.3 ± 1.62 for the annulus, from +2.16 ± 1.5 to +3.3 ± 1.2 for the sinuses of Valsalva and for the sinutubular junction from +2.14 ± 1.27 to +2.91 ± 0.95. Older patients then presented a significant increment in the root *z*-scores for the three root areas respectively of +1.13 ($P = 0.00$), +1.14 ($P = 0.00$) and +0.76 ($P = 0.004$).

DISCUSSION

Ross and Ross–Konno remain a debated surgical options, not only for the technical difficulties of the operation itself, but also for the sacrifice of a good functioning pulmonary valve to replace a dysplastic aortic valve. The advantages are the growth capability of the autograft, the avoidance of anticoagulation and the suitability in smaller patients for whom there are no adequate prosthetic materials available. Long-term disadvantages are aortic root dilatation, aortic valve progressive regurgitation and the reoperation in the LVOT and RVOT. From our experience, we hypothesize that in smaller children, who require a Ross/Ross–Konno operation, the pulmonary autograft may perform better than in older children for the following reasons: different characteristic of pulmonary wall and valve, different adaptive/remodelling capability and different autograft size.

The aortic and pulmonary roots have a similar structure during foetal life regarding semilunar leaflet and vessel wall. The roots during foetal life have an equal content of collagen with the same spatial disposition, similar content of elastin fibres and similar density of cells as reported by Aikawa *et al.* [16], who described the remodelling process that takes place in human semilunar valve from foetal life to adult age. The haemodynamic changes in terms of pressures that occur after birth in the systemic and pulmonary circulation activate a remodelling process that creates the different vessel wall characteristics seen in later life. Semilunar leaflets maintain the capability to remodel their structure in case the mechanical loading conditions change during early and late neonatal age, but this capability progressively decreases during childhood and becomes almost minimal in adulthood. Sing of adaptive capability loss is progressively decreasing number of immature cells in the valve tissue and this is directly related to the aging process. When the pulmonary root is placed in the systemic position, the increased pressure loading and stress stimulates the reactivation of these cells involved in the remodelling process as far as the mechanical equilibrium is restored and only then the remodelling cells return to a quiescent state [17]. Based on this findings, it is possible to suppose that pulmonary root in the young age group (<18 months) has a structure more similar to that of the aortic root with higher number of immature cells and higher content on fibrin, elastin and collagen fibres than the one in the older group (>18 months). Then, because the remodelling process has an inverse relation with age, younger pulmonary autografts have a less extended and a more efficient adaptation when pressure loads change from lower pulmonary values to higher systemic values [16, 17].

In the Ross and Ross–Konno, typically there is a degree of instantaneous dilatation as the aortic cross clamp is removed but the adaptive/remodelling process takes place over the next months as the autograft responds to the new pressure and mechanical load activating the immature cells [17, 18]. The entity of the remodelling process has not been clearly defined yet, but it seems that the younger the patient, the higher the chance that the process is effective and efficient. Other factors are involved in the autograft remodelling capability such as residual lesions, valve competence, loss of vasa vasorum and apoptosis process due to the harvesting process of the autograft. The absolute size of the pulmonary autograft play also an important role, in fact a smaller pulmonary autograft will encounter lower wall tension when

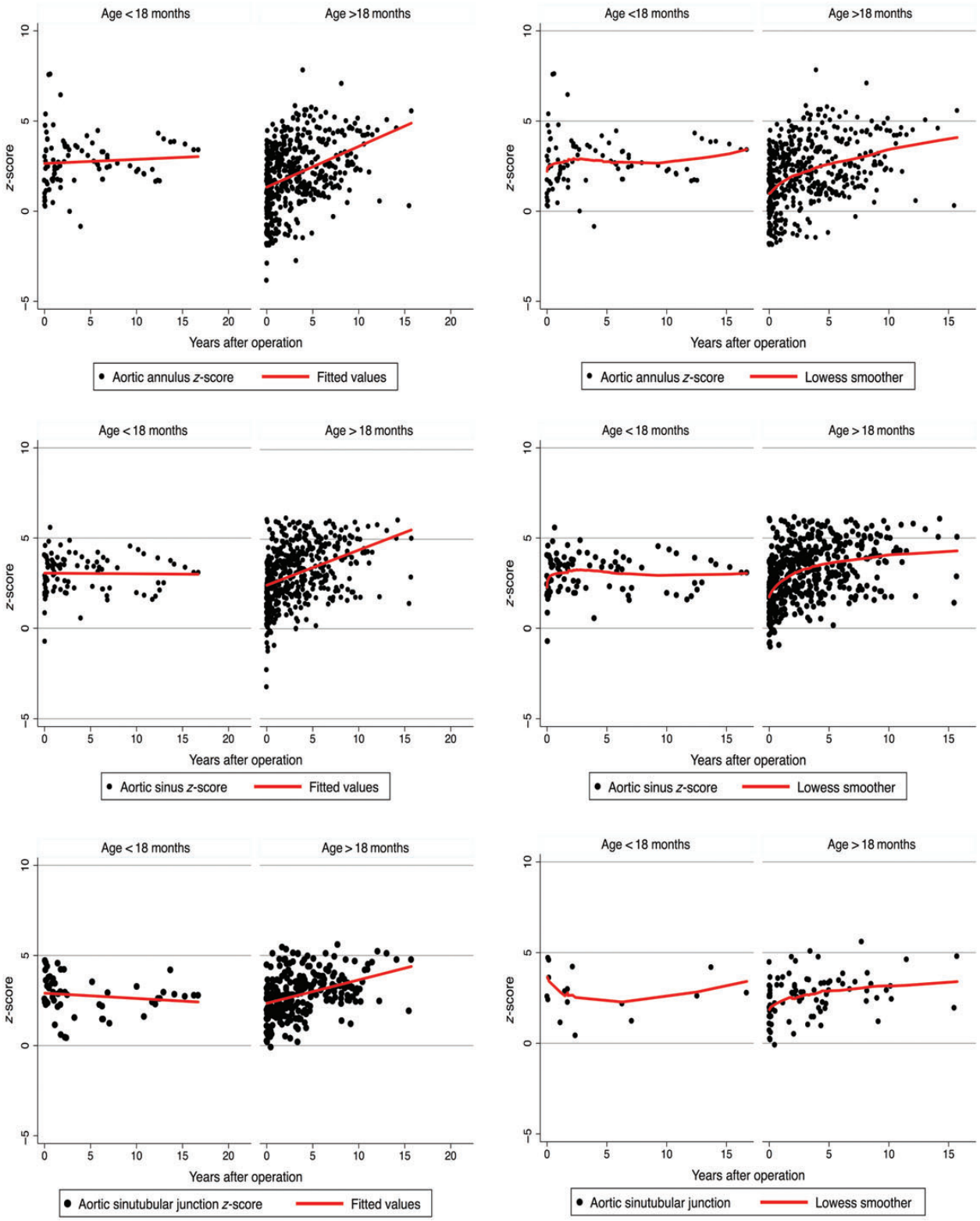

Figure 4: Progression of neoaortic root *z*-scores after Ross/Ross–Konno operation at annular, Valsalva sinuses and sinutubular junction levels per age group. *z*-scores are plotted per single neoaortic region and per age group. Non-parametric estimation models (fitted values—right sided figures, Lowess smoother left-sided figures) reveal higher stability on the neoaortic root in the <18-months group and dilatation in the >18-months group.

placed in the systemic position compared with larger pulmonary autograft based on the physical concept of Laplace's law. The lower wall tension experienced by the smaller pulmonary autograft is probably involved as well in the adaptive changes occurring during the first months following implantation before any dilatation has occurred.

Gradual aortic root dilatation over the years, particularly at the level of the mid-sinuses and at the sinutubular junction [19–21],

has been indicated as the main reason for the development and progression of autograft valve regurgitation.

Patients <18 months of age after a Ross/Ross–Konno operation have no significant progression in the aortic root dimension in our experience (Fig. 4). The aortic root *z*-scores in the younger group remain almost stable with not significant increase (+0.89 in the annulus +0.39 in the sinuses and decreased −0.59 in the sinutubular junction) as recently reported by others [3–5]. Elder *et al*. [3] reported *z*-scores of aortic root, in similar patients for age group, showing some initial dilatation after which the aortic root remained stable and did not progress significantly in the long term.

In contrast, the older age group showed progressive dilatation (increments of *z*-scores of +1.13 for the annulus, +1.14 for the sinuses and +0.76 for the sinutubular junction) reaching mean *z*-score values in the sinuses and sinutubular junction of 3.3 and 2.95. These data suggest that for patients >18 months of age the process is a 'pathological' dilatation rather than a 'natural' adaptation to patient somatic growth. These clinical findings support the histological characteristics and load adaptive capability of the pulmonary autograft suggested and reported in previous studies [3–5].

Performance of the neoaortic valve is excellent in younger patients and 89% had none/mild regurgitation at a follow-up of 10 years and only in 11% the aortic valve competence deteriorated from mild to moderate during the follow-up.

In the older group, 18% of the patients developed progression of aortic regurgitation from none/mild to moderate, and it occurred significantly earlier after the operation (mean time 2.89 years). The older age group did have a higher incidence of preoperative aortic valve insufficiency (considering primary aortic regurgitation and mixed aortic disease) compared with the younger group, predominantly as a sequalae of previous valvotomy or valvuloplasty. However, the primary (i.e. presenting) aortic lesion was the same in both groups and the preoperative *z*-scores of the annulus was also the same in both groups, suggesting that they had comparable aortic root pathology. Furthermore, across the whole series, preoperative aortic insufficiency was not an independent risk factor for the development of late root dilatation.

Various solutions have been proposed to support the aortic root in older patients. Among stabilization techniques, the use of Dacron felt [22] has been proposed to prevent dilatation without preventing obviously aortic sinuses dilatation. Inclusion technique of the autograft in a Dacron vascular tube [23] or in a Valsalva vascular prosthesis [24] offers the advantages of supporting the entire autograft in a stable prosthesis that stops dilatation. These stabilization techniques may offer better result in adults but it is not applicable in the paediatric and young adolescent group where there is a need for maintaining growth capability. A different approach would be to use the subcoronary implantation technique, but again this is possible only in older children and with an adequate-sized LVOT.

Our data suggest that the Ross and Ross–Konno performed in the younger age group (<18 months) may be protective from long-term progressive aortic root dilatation. If ventricular function is well preserved, the results of the Ross, even in this subset age group, have been very encouraging and it raises the question as to whether this should be considered in preference to aortic valve repair techniques, which have often been proposed as a means of delaying the Ross procedure to an older age. This is particularly attractive as an alternative to complex repair techniques such as multiple cusp extensions, where the mid-term results have not been as good as valve replacement and or Ross operation.

Type of reoperation in Ross patients have been reported by several groups [8–11] and vary from extensive valve, root and arch repair/replacement in Mayo Clinic experience [11], to isolated valve replacement without root replacement in the series reported by Alsoufi *et al*. [7]. Different approaches to reoperations are mainly a reflection of the different population characteristics and underlying pathology – for example the Mayo Clinic patients were much older and did not have rheumatic heart disease when compared to the series from Alsoufi. Reoperation on the LVOT can be performed safely, although it is still a complex procedure and usually involves surgery on the aortic valve as well as on the aortic root.

CONCLUSION

Ross/Ross–Konno can be invaluable in the young paediatric patients where there are no other surgical options due to complex aortic valve disease associated with LVOTO. Ross/Ross–Konno operations in the current era present good survival in young populations, as in older patient groups, but left ventricular dilatation and mitral regurgitation still represent significant risk factors for mortality. Encouraging results have been found in the younger age group that show a remarkable stability of the neoaortic root when compared with older children and this may reflect the much lower wall stresses encountered in the smaller size pulmonary autograft, thus allowing the autograft time to adapt to the higher pressure environment without suffering late dilatation. Subsequent enlargement of the root in these patients appears to be somatic growth rather than pathological dilatation. In conclusion, patients <18 months, despite the need for more reoperation in the RVOT, have an excellent outcome in terms of stability of the aortic root, competence of the aortic valve and the avoidance of reoperation for aortic valve and/or aortic root.

Conflict of interest: none declared.

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APPENDIX. CONFERENCE DISCUSSION

Dr R. Jonas *(Washington, DC, USA)*: Dr Lo Rito and colleagues have achieved excellent results in 22 patients, all 18 months of age or less. Your actuarial survival at 10 years was approximately 80%, and there were no reoperations for autograft dilation or autograft regurgitation. Your freedom from reoperation for RV outflow tract obstruction was 65% at 10 years. These results are as good or better than reports from several other centres which have reported on young Ross patients. Some of those reports have described a reintervention-free rate as low as 37% at five years. It's clear from your report that the stability of the neoaortic root is greater in this younger subset than is seen with older paediatric patients and definitely much better than with adults.

The recent report from the German-Dutch Ross registry suggests that nearly 50% of adult patients who have a Ross procedure as a root replacement will require replacement of the autograft by 15 years postoperatively. However, despite the greater stability of the autograft in the aortic position in infants having the Ross procedure, the operation still suffers, as Dr Ebels has just reminded us very eloquently, from the fundamental disadvantage that you are replacing one valve problem with two. And although the autograft may perform better in younger patients, obviously the need for conduit reoperation is a definite impediment to the quality of life for these younger patients.

I want to also point out another potential risk that we discovered earlier this year in Washington because we had been assuring families that we would perhaps be able to extend the life of the homograft conduit on the right side with a Melody valve. We placed a Melody valve in an 8-year-old patient, and within one month, the rigid pre-stent that's now required for the Melody valve had eroded into the somewhat bulbous neoaortic root and created a very large 2 x 3 cm aortopulmonary window. And interestingly, I reported this in Los Angeles at a conference one week ago. When I sat down, the person next to me said, "That's a coincidence, we had exactly the same thing happen at our centre recently." So the Melody valve may not be a good choice where you have a devascularized and dilated neoaortic root sitting next to a very rigid Melody stent.

It's also clear to everybody attending this conference that the landscape for aortic valve replacement is changing rapidly as TAVI becomes widely accepted for adult aortic valve replacement. I just visited the booth for the ValveXchange group who are represented here. They have applied for the CE mark and tell me that it will not be long before it will be possible to exchange leaflets in a surgically implanted stent that can have the leaflets replaced as many times as is required.

So I would suggest that all of these developments should encourage us to preserve the aortic valve whenever possible through a programme of aggressive balloon dilation of the aortic valve early in life. I believe that creating some degree of mild or moderate aortic regurgitation actually helps to grow the small, originally congenitally stenotic aortic annulus, as well as helping to grow the small left heart and to improve the diastolic function of the small left heart. At some point an aortic valve repair is often required, so perhaps if the balloon is done in infancy, perhaps the child gets to five, six, seven years of age. Hopefully then you get something like ten years of growth, and at that point either a ValveXchange type valve or a TAVI type valve can be placed.

So my question for you is, in the setting of this changing landscape for aortic valve replacement options and technology, what do you see as the current indications for a Ross procedure? You're suggesting we should move it to a younger age in your conclusion slide, but who should we be moving to a younger age?

Dr Lo Rito: I think you're right, but I didn't highlight, for brevity of the presentation, that all of these younger patients who required a Ross earlier are patients who have decreased left ventricular function or progressive aortic regurgitation, in the case of the balloon valvotomy, that create a mixed disease, or progressive aortic stenosis or LVOT obstruction. So this is one subset of young patients that we were not able to treat with conventional treatment such as aortic surgical valvotomy. In fact, they didn't have a repeat operation trying to save the aortic valve, but they had just one failing operation before the Ross.

So I think we should not extend the Ross operation to all cases because I think that in the ones in whom we are able to achieve a good result, as you say, with repeated balloon and probably aortic open valvotomy, we should keep following that route because if they reach the age of adolescence, probably it is better for them to have the valve replaced. But in this small subset of patients, I think we should not be scared to do the Ross because actually it is proven to have a good term outcome in relation to the aortic root.

Regarding the question about the RVOT, of course they have more reoperation in the RVOT, but we are doing reoperation on RVOT as well in tetralogy of Fallot, and we are not worried about reoperating on their right ventricular side. So my answer is, I don't think we should be worried about reoperating in the conduit in the Ross as well.

So I may have been provocative with my last question on the last slide, but I think we should not forget this technique, and I think it's very useful for this small subset of young patients and probably something we should not completely forget. And when we have a patient who has this type of clinical pattern, we should not try to push him forward because we are going to lose left ventricular function and this is a price we cannot probably pay for the clinical status of the patient. So I think probably these remain the patients in whom an early Ross procedure is still indicated.