Long-Term Survival and Reintervention After the Ross Procedure Across the Pediatric Age Spectrum

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Background. **There are limited data regarding long-term outcomes after the Ross procedure in children. We evaluated mortality and reintervention in a large pediatric cohort.**

Methods. **A retrospective analysis of all patients aged younger than 18 years who underwent the Ross procedure at our institution (1991 to 2013) was conducted. Kaplan-Meier curves and Cox proportion hazard models were used to evaluate long-term outcomes and associated risk factors.**

Results. **Included were 240 consecutive patients undergoing a Ross/Ross-Konno procedure: 18% infants, 48% children, and 33% adolescents. Infants were more likely to have complex left heart disease ($p = 0.005$). Overall survival to hospital discharge was 96%; infants had the highest mortality (18%). Long-term survival status was known for 99.6% (median follow-up, 10.7 years). Overall 15-year survival was 87% (lowest in infants, 72%; $p = 0.003$). Reintervention status was known in 87%. Overall 15-year freedom from any left ventricular outflow tract reintervention was 59%; 85% still had their autograft valve at the latest follow-up. Left ventricular outflow tract reintervention was uncommon in infants (n = 2). Overall 15-year freedom from right ventricular outflow tract reintervention was 53%, and was lower in infants (19%) than in children (51%) and adolescents (76%; $p < 0.0001$).**

Conclusions. **Outcomes after the Ross procedure in children vary by age. Infants more commonly have complex left heart disease and experience higher mortality but have excellent long-term autograft durability. Children and adolescents have higher rates of left ventricular outflow tract reintervention, whereas infants are at highest risk of right ventricular outflow tract reintervention.**

(Ann Thorac Surg 2015;99:2086–95)

The Ross procedure is commonly used in children requiring aortic valve replacement [1, 2]. Advantages include autograft growth potential, excellent flow characteristics, and avoidance of anticoagulation [3, 4]. The operation offers the possibility of life-long durability of the autograft and is particularly useful in patients with a small aortic root or multilevel left ventricular outflow tract (LVOT) obstruction [4–6]. Several disadvantages of the Ross procedure have also been described, including aortic root dilatation and progressive aortic insufficiency (AI) requiring autograft reintervention [7–9]. In the pediatric population, right-sided conduit reinterventions are expected [8].

Most literature on Ross outcomes focuses on adult patients [10–12]. Studies including pediatric patients have had limitations, including small sample sizes, analysis of mixed cohorts of adults and children, and a focus on midterm outcomes [9, 13–16]. Long-term outcomes and an understanding of the effect of age at operation remain unclear. Although more frequent conduit reinterventions may be expected in infants, further study of the durability of the autograft over time and long-term survival in this cohort is needed.

The purpose of the present study was to evaluate long-term survival and need for reintervention after the Ross procedure in a large pediatric cohort. We also sought to describe these outcomes across different pediatric age groups and assess factors associated with long-term outcome.

Material and Methods

The University of Michigan Institutional Review Board approved this study, with waiver of consent.

Study Population

A retrospective review of all patients aged younger than 18 years undergoing a Ross or Ross-Konno procedure at the University of Michigan Congenital Heart Center

Accepted for publication Feb 12, 2015.

*Recipient of the 2014 Congenital Heart Surgery Presidents Award.

Presented at the Sixty-first Annual Meeting of the Southern Thoracic Surgical Association, Tucson, AZ, Nov 5–8, 2014.

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0003-4975/$36.00
http://dx.doi.org/10.1016/j.athoracsur.2015.02.068

Table 1. Patient and Procedural Characteristics (N = 240)

Variables[a]	Overall (n = 240)	Age at Ross Procedure			*p* Value
		Infant (n = 44)	Child (n = 116)	Adolescent (n = 80)	
Patient characteristics					
Male sex	169 (70.4)	29 (65.9)	82 (70.7)	58 (72.5)	0.74
Indication for repair					
AS	60 (25.0)	21 (47.7)	28 (24.1)	11 (13.8)	0.0002
AI	43 (17.9)	3 (6.8)	18 (15.5)	22 (27.5)	
AS/AI	137 (57.1)	20 (45.5)	70 (60.3)	47 (58.8)	
Etiology					0.048
Congenital	222 (92.5)	43 (97.7)	112 (96.6)	67 (83.8)	
Bicuspid aortic valve	164 (68.3)	29 (65.9)	81 (69.8)	54 (67.5)	N/A
Acquired	17 (7.1)	0 (0.0)	6 (5.2)	11 (13.8)	
Other	16 (6.7)	6 (13.6)	5 (4.3)	5 (6.3)	
Any previous intervention	177 (73.8)	33 (75.0)	102 (87.9)	42 (52.5)	<0.0001
Balloon valvuloplasty	98 (40.8)	25 (56.8)	51 (44.0)	22 (27.5)	
Surgical valvotomy	43 (17.9)	1 (2.3)	30 (25.9)	12 (15.0)	
Aortic valve repair	11 (4.6)	1 (2.3)	10 (8.6)	0 (0.0)	
Aortic valve replacement	10 (4.2)	1 (2.3)	5 (4.3)	4 (5.0)	
Coarctation/IAA repair	42 (17.5)	4 (9.1)	26 (22.4)	12 (15.0)	N/A
Sub-aortic stenosis resection	33 (13.8)	1 (2.3)	25 (21.6)	7 (8.8)	
VSD repair	22 (9.2)	2 (4.5)	16 (13.8)	4 (5.0)	
ASO	2 (0.8)	0 (0.0)	2 (1.7)	0 (0.0)	
Konno procedure	3 (1.3)	1 (2.3)	2 (1.7)	0 (0.0)	
Mitral valve operation	4 (1.7)	1 (2.3)	1 (0.9)	2 (2.5)	
AVSD	4 (1.7)	1 (2.3)	2 (1.7)	1 (1.3)	
Other	17 (7.1)	4 (9.1)	11 (9.5)	2 (2.5)	
Diagnosis					
Isolated aortic valve disease	152 (63.3)	20 (45.5)	72 (62.1)	60 (75.0)	0.005
Complex left heart disease	88 (36.7)	24 (54.5)	44 (37.9)	20 (25.0)	
LV dysfunction (any)	43 (17.9)	20 (45.5)	13 (11.2)	10 (12.5)	<0.0001
Mitral stenosis					
None	175 (72.9)	25 (56.8)	89 (76.7)	61 (76.3)	
Mild	42 (17.5)	7 (15.9)	21 (18.1)	14 (17.5)	0.005
Moderate or greater	23 (9.6)	12 (27.3)	6 (5.2)	5 (6.3)	
Endocardial fibroelastosis	24 (10.0)	17 (38.6)	6 (5.2)	1 (1.3)	<0.0001
Shone syndrome	17 (7.1)	8 (18.2)	7 (6.0)	2 (2.5)	0.004
Procedural characteristics					
Outpatient at operation	170 (70.8)	11 (25.0)	91 (78.4)	68 (85.0)	<0.0001
Any concomitant procedure	110 (45.8)	34 (77.3)	48 (41.4)	28 (35.0)	<0.0001
Modified Konno/Konno procedure	78 (32.5)	30 (68.1)	39 (33.6)	9 (11.3)	N/A
Aortic arch repair	16 (6.7)	14 (31.8)	1 (0.9)	1 (1.3)	
Mitral valve operation	4 (1.7)	3 (6.8)	0 (0.0)	1 (1.3)	
Aortic annulus plication	8 (3.3)	1 (2.3)	1 (0.9)	6 (7.5)	
Weight at operation, kg	28.2 (13.6–49)	4.4 (3.3–6.2)	23.8 (17–30.1)	58 (47.6–68)	<0.0001
Cardiopulmonary bypass time, min	149 (133–180)	157 (132–200)	145 (126–169)	158 (138–187)	0.02
Aortic cross-clamp time, min	100 (87–119)	97 (85–133)	96 (84–111)	109 (93.5–121)	0.01
Circulatory arrest	18 (7.5)	15 (34.1)	2 (1.7)	1 (1.3)	<0.0001
Size of pulmonary autograft, mm	19.1 ± 4.8	11.5 ± 2.2	18.7 ± 2.9	23.4 ± 2.5	<0.0001
Initial size of aortic valve annulus, mm	17.7 ± 7.0	7.8 ± 3.3	16.9 ± 4.4	24.0 ± 4.2	<0.0001

(*Continued*)