

# Valve-Sparing Aortic Root Replacement by Congenital Heart Surgeons: A Single Center Experience.

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February 9, 2022

## Abstract

**Background** Aortic root dilatation and aortic insufficiency can occur in patients with prior conotruncal defect surgery, the Ross procedure, and connective tissue disease (CTD). Valve-sparing aortic root replacement (VSRR) is an excellent choice for these young patients. We present the outcomes of young patients undergoing VSRR by congenital heart surgeons at a single center. **Methods** A single center retrospective chart review from April 2008 – April 2021 was performed. Patients with aortic root and valve pathology who underwent VSRR were identified. A total of 49 patients were identified by using the hospital surgical database. Three VSRR techniques were utilized during the study period; aortic valve-sparing root remodeling (Group 1, 7 patients), aortic valve-sparing root reimplantation (Group 2, 32 patients), and a modified root remodeling procedure utilizing a geometric annuloplasty ring (Group 3, 10 patients). Patient characteristics, post-VSRR echocardiogram studies and need for intervention, and survival were analyzed. **Results** Long-term survival after VSRR is excellent with only one death related to cardiac causes. Valve degeneration remains a concern with 13 patients (26%) requiring subsequent aortic valve replacement over the study period. Risk factors for aortic valve reintervention were the aortic root remodeling technique, mild or greater immediate post-operative aortic insufficiency and higher post-operative mitral insufficiency. Follow-up was significantly longer in Group 1 patients compared to Groups 2 and 3. **Conclusions** Valve-sparing aortic root replacement is safely performed by congenital heart surgeons in a heterogeneous patient population. Valve degeneration remains a concern and greater than trivial post-operative aortic insufficiency should prompt further attempts at valve repair or replacement.

## Valve-Sparing Aortic Root Replacement by Congenital Heart Surgeons: A Single Center Experience.

**Running Head:** VSRR by Congenital Heart Surgeons

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**Presentation:** The Society of Thoracic Surgeons, Virtual Format, January 31, 2021.

**Word count:** 4261

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## **Classifications: 20**

### **Background**

Aortic root dilatation and aortic insufficiency can occur in patients with prior conotruncal defect surgery, the Ross procedure, and connective tissue disease (CTD). Valve-sparing aortic root replacement (VSRR) is an excellent choice for these young patients. We present the outcomes of young patients undergoing VSRR by congenital heart surgeons at a single center.

### **Methods**

A single center retrospective chart review from April 2008 – April 2021 was performed. Patients with aortic root and valve pathology who underwent VSRR were identified. A total of 49 patients were identified by using the hospital surgical database. Three VSRR techniques were utilized during the study period; aortic valve-sparing root remodeling (Group 1, 7 patients), aortic valve-sparing root reimplantation (Group 2, 32 patients), and a modified root remodeling procedure utilizing a geometric annuloplasty ring (Group 3, 10 patients). Patient characteristics, post-VSRR echocardiogram studies and need for intervention, and survival were analyzed.

### **Results**




Long-term survival after VSRR is excellent with only one death related to cardiac causes. Valve degeneration remains a concern with 13 patients (26%) requiring subsequent aortic valve replacement over the study period. Risk factors for aortic valve reintervention were the aortic root remodeling technique, mild or greater immediate post-operative aortic insufficiency and higher post-operative mitral insufficiency. Follow-up was significantly longer in Group 1 patients compared to Groups 2 and 3.

### **Conclusions**

Valve-sparing aortic root replacement is safely performed by congenital heart surgeons in a heterogeneous patient population. Valve degeneration remains a concern and greater than trivial post-operative aortic insufficiency should prompt further attempts at valve repair or replacement.

### **Graphical/Visual Abstract (second page)**

## 49 patients undergoing valve-sparing aortic root replacement (2008-2021) in a single congenital heart center

Procedure Types	Follow-Up Duration (yrs)	Incidence of Aortic Valve Reintervention	
			
Remodeling n=7	14 ± 4	100%	➤ Long-term survival is excellent in all groups
Reimplantation n=32	5 ± 3	19%	➤ Long-term valve degeneration 26%
Modified Reimplantation n=10	0.2 ± 0.2	0%	➤ 100% reintervention rate for the remodeling technique, all prior Ross patients
			➤ Risk factors for valve degeneration were the remodeling technique and mild or greater postoperative AI

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## Background

Aortic root and valve disease can present in young patients, who seek care at congenital heart centers. Aortic root aneurysm can occur in patients with prior conotruncal defect surgery, the Ross procedure, and connective tissue diseases.<sup>1, 2</sup> When left untreated, aortic root dilatation in these patients may lead to the development of aortic insufficiency, heart failure, dissection, rupture, and death.<sup>3</sup> Due to the young age of these patient populations, congenital heart surgeons must be familiar with the management and repair of aortic root and valve disease.

Valve-sparing aortic root interventions are appealing to minimize future valve interventions and/or systemic anticoagulation. However, their outcomes are poorly defined due to the heterogeneity and complexity of this population. In this retrospective, single-center study, we describe the outcomes of three different VSRR procedures and determine the factors associated with the need for subsequent reintervention.

## Methods

### PATIENTS AND METHODS

This study was deemed exempt by the University of Michigan Institutional Review Board and the need for Informed Written Consent was waived due to its retrospective nature.

#### Data Source

We identified patients who underwent valve-sparing aortic root surgery at C.S. Mott Children's Hospital, University of Michigan Medical Center, for the treatment of neoaortic dilatation after the Ross procedure, aortic dilatation secondary to connective tissue disease, or aortic dilatation after prior conotruncal heart surgery between the years 2008 and 2021.

#### Patient Population

Patients who underwent valve-sparing aortic root surgery were identified. We then collected preoperative, operative, and postoperative characteristics from institutional and regionally-shared medical records of each patient.

## Definitions

### *Operative Techniques and Clinical Management*

Patients were divided into three groups according to the type of valve-sparing root replacement technique utilized; Group 1 patients received a root remodeling (Yacoub) procedure, as described elsewhere. Group 2 patients underwent the reimplantation technique (David procedure).<sup>17</sup> Group 3 patients underwent placement of a trileaflet aortic annuloplasty ring (HAART 300, Biostable), leaflet plication, and root remodeling procedure. The details of aortic annuloplasty ring sizing and placement have been previously reported.<sup>18</sup> The root remodeling procedure was then performed using a Valsalva graft that was sized approximately 5 mm more than the size of the ring that was implanted. The left coronary button was then reimplanted, followed by the distal anastomosis of the graft to the ascending aorta. Cardioplegia was then infused to pressurize the root, the site for the right coronary button was determined, and the right coronary artery was implanted.

### *Outcomes*

Patient survival, postoperative aortic insufficiency (AI) grade determined by echocardiogram or cardiac MRI, and need for aortic valve reintervention were determined for each patient.

### *Statistical Analysis*

Data analysis was performed using SPSS 27 software (Armonk, NY). Data are presented as median for continuous data and number (%) for categorical data. Univariate comparisons across different groups of surgical techniques were performed using the  $\chi^2$  test for categorical data and ANOVA. A p-value < 0.05 (2-tailed) was considered statistically significant.

## Results

We identified 49 patients who underwent a valve-sparing root replacement during our study period. Twenty-four patients had a history of the Ross procedure, 17 patients had a history of connective tissue disease, and eight patients had a history of congenital heart defect. Group 1 was comprised of seven patients who underwent the aortic root remodeling technique, Group 2 was comprised of 32 patients who underwent the aortic root reimplantation technique, and Group 3 was comprised of ten patients who underwent an aortic root remodeling technique with a rigid aortic annuloplasty ring. All seven patients in Group 1 had undergone a prior Ross procedure. Group 2 and Group 3 were patients with a history of connective tissue disease, prior conotruncal heart surgery, congenital heart disease, or a prior Ross procedure. Mean follow-up was significantly different between groups ( $p < 0.001$ ). Group 1 and Group 2 patients had 7- and 14-year follow-up respectively and Group 3 had 1-year follow-up. The traditional aortic root remodeling technique has been largely abandoned in our institution and replaced with the aortic root remodeling technique with rigid annuloplasty ring since 2018. Aortic root reimplantation has been largely replaced as well, but is still used for selected patients based on surgeon preference and patient anatomy. Patient demographics were similar among groups with a median age of 19 years (range 8-32) and 87% male sex (table 1). There were no significant differences between the cardiopulmonary bypass and aortic cross-clamp times between groups 1 and 2; however, cardiopulmonary bypass times and aortic cross-clamp times were significantly higher in group 3 ( $p < 0.001$ ). Group 1 and 3 were more likely to have a history of the Ross procedure ( $p = 0.003$ ), and group 2 was more likely to have a history of connective tissue disease ( $p = 0.21$ ). Group 1 was more likely to require subsequent aortic valve replacement, with all Group 1 patients requiring aortic valve replacement over the study period ( $p < 0.001$ ). Group 3 did not have any required valve reinterventions during the study period. Group 3 was also more likely to have undergone aortic valve leaflet plication as part of their procedure ( $p = 0.004$ ). There were no significant differences between aortic annulus measurements preoperatively ( $p = 0.741$ ), but there was a significant difference in postoperative aortic annulus measurement ( $p = 0.01$ ). Preoperative AI grades were similar between groups and were 2, 1.7, and 2.4 respectively ( $p = 0.2$ ). However, immediate postoperative AI grades were significantly different between groups: 2.2, 1.2, and 1.2 respectively ( $p = 0.3$ ). The most recent AI grades in patients from group 2 and 3 who did not require

subsequent aortic valve replacement were similar ( $p = 0.9$ ). Survival was good in all groups with one early mortality due to hemorrhage and one late mortality from malignancy.

Thirteen patients required aortic valve replacement after their valve-sparing root replacement during the study period. According to procedure type, all seven patients (100%) in Group 1 required valve replacement, six patients (19%) in Group 2 required aortic valve replacement, and no patient required valve replacement in Group 3 ( $p < 0.001$ ). Average time to aortic valve replacement after valve-sparing aortic root replacement between Group 1 and Group 2 was 4.7 and 3.4 years, respectively ( $p = 0.6$ ). History of connective tissue disease, prior Ross procedure, concomitant procedures, and prior sternotomy were not associated with increased risk for reintervention. Patients who required subsequent valve reintervention had significantly longer follow-up ( $p < 0.001$ ), larger postoperative aortic annulus measurements ( $p = 0.012$ ), and longer cardiopulmonary bypass ( $p = 0.01$ ) and aortic cross clamp times ( $p = 0.003$ ). There was no significant difference in preoperative AI grades; however, patients requiring future aortic valve replacement had significantly higher immediate postoperative AI grades ( $p < 0.001$ ). Six of seven patients with greater than mild aortic insufficiency on immediate postoperative echocardiogram required subsequent aortic valve replacement ( $p < 0.001$ ). Postoperative mitral regurgitation was also higher in patients who eventually required aortic valve replacement ( $p = 0.05$ ).

## Conclusions

Aortic root dilatation, with or without resultant aortic insufficiency, is commonly encountered in congenital heart surgery. The optimal management of these young patients is challenging for clinicians due to their heterogeneity and often complex surgical histories. The development of standardized guidelines for surgical interventions for these patients is also challenging due to their relative heterogeneity and infrequency in comparison to adult aneurysmal disease and connective tissue aneurysmal disease. Current evidence also suggests that the complications of dissection and rupture are less common than in adults with *de novo* aortic root dilatation.<sup>9</sup> Many clinicians with experience treating these patients, including the authors of this report, advocate for the consideration of surgical intervention when aneurysms reach 5-5.5 centimeters or when there are concomitant surgical lesions that require interventions, such as aortic insufficiency causing significant heart dysfunction or concomitant right sided heart disease.<sup>10, 11</sup>

Most patients in our center have a history of connective tissue disease, a prior Ross procedure, unrepaired congenital heart disease, or prior congenital heart surgery. The Bentall procedure with either a mechanical or bioprosthetic valve has been the historical treatment of choice for patients with aortic root dilatation with or without aortic insufficiency. The Bentall procedure offers a straight-forward surgical technique and excellent long-term results with the use of mechanical valves; however, the late complications of bleeding, valve reintervention, and thrombosis remain concerning.<sup>12</sup> The Bentall procedure with the use of bioprosthetic valved conduits offers patients the avoidance of systemic anticoagulation and reasonable long-term freedom from reintervention. A recent study by Chirichilli demonstrated a freedom from reoperation at 16 years of 74.7%. While these results are impressive and makes this an appealing choice for older patients, the likelihood of reintervention for patients with decades of life expectancy is a near certainty. For these reasons, valve-sparing root replacement is an appealing treatment strategy and may be underutilized.<sup>13-15</sup>

Optimal technique for valve-sparing root replacement is another area of debate. Extensive literature has been published on the indications and efficacy of valve-sparing aortic root replacements in adult patients with aortic root dilatation; however, less literature is available guiding clinicians on the optimal indications for intervention in patients with a history of prior congenital heart surgery with only small case series published with mixed results.<sup>7, 16-19</sup> Although other less common surgical techniques have been described, valve-sparing root replacement techniques can be grouped as either aortic root type reimplantation or aortic root type remodeling. The aortic root replacement technique is generally believed to be superior to the aortic root remodeling technique as it provides circumferential support of the aortic root and basal ring. The aortic root remodeling technique replaces the aortic sinuses but does not provide support to the intercommissural triangles and basal ring. Numerous studies have shown the aortic root reimplantation technique to have lower rates of reoperation and recurrent aortic insufficiency<sup>16</sup>. Like our results with the aortic root remodel-

eling procedure, a study by Roubertie and colleagues examining results of valve-sparing operations in 23 children with connective tissue disease found that the aortic root reimplantation technique was superior to the aortic root remodeling technique. They observed a nearly 50% reintervention rate for patients treated with the aortic root remodeling technique.<sup>16</sup> The largest series examining the results of valve-sparing aortic root replacement in children was published by Fraser et al in 2018 and examines the results of 100 consecutive pediatric patients.<sup>20</sup> Most patients in this series had a history of connective tissue disease (90%) with the remaining patients having a history of congenital heart surgery. Like our results comparing the reimplantation and remodeling techniques, they observed superior freedom from reintervention with the aortic root reimplantation technique compared to the aortic root remodeling technique. Late complications of aortic insufficiency and aneurysm formation remained a concern.<sup>20</sup>

Despite the seemingly clear advantage of the aortic root reimplantation technique, it is technically more challenging to perform an adequate external aortic root dissection, especially in patients who have undergone prior arterial switch operations or the Ross procedure. The aortic root remodeling technique obviates the need for extensive external root dissection, but durability is concerning. The use of a rigid geometric subannular annuloplasty ring can standardize aortic valve repair and aortic root replacement techniques, and its utilization during the aortic root remodeling technique provides annular and intercommissural triangle stabilization while avoiding the extensive external aortic root dissection necessary for the reimplantation technique.

This study examined the outcomes of valve-sparing aortic root replacement in a congenital heart center. Over our study period we utilized three different valve-sparing techniques, (1) the traditional aortic root remodeling technique, (2) the aortic root reimplantation technique, and (3) a modified aortic root remodeling technique with the use of a rigid annuloplasty ring. The traditional aortic root remodeling technique (Group 1) was only utilized in patients with aortic root dilatation after the Ross procedure and was associated with a 100% reintervention rate over our study period, whereas the aortic root reimplantation group (Group 2) and the remodeling group with annuloplasty ring (Group 3) had 19% and 0% respectively. These results should be interpreted with caution as the average follow-up was significantly longer in Group 1 (17 years) compared to Group 2 (8 years) or Group 3 (0.7 years). Nevertheless, the average length of time to aortic valve reintervention in Group 1 and Group 2 was 5 and 3 years respectively. The largest risk for aortic valve replacement after valve-sparing root surgery in our study, as well as other studies, was higher degrees of postoperative aortic insufficiency. The immediate postoperative degree of aortic insufficiency was less in Group 3 ( $p = 0.03$ ) which suggests that this technique will have acceptable durability. Similar to other studies, our findings suggest that the traditional aortic root remodeling procedure should be avoided due to poor durability.

This study does have significant limitations. First, the technique utilized over the study period was subject to surgeon preference which could impact the overall results. Secondly, there are significant differences in the follow-up duration between the three groups and only short-term follow-up is available for Group 3 patients.

In summary, valve-sparing aortic root replacement can be safely performed for a variety of indications by congenital heart surgeons. However, there is continued risk for valve failure over time. The traditional aortic root remodeling technique, when applied to patients with dilatation following the Ross procedure, should be avoided as this cohort of patients had a 100% failure rate over our study period. The aortic root reimplantation technique has acceptable results; however, there was a 20% rate of valve failure over our study period. The aortic root remodeling technique combined with the use of geometric ring annuloplasty has good short-term results; however, longer follow-up is needed to fully delineate its durability and effectiveness in comparison to the aortic root reimplantation technique.

**Table 1.** Stats by procedure. Group 1 = Aortic Root Remodeling 2 = Aortic Root Reimplantation. Group 3 = Aortic Root Reimplantation with rigid annuloplasty ring.

<b>Variable Median + std or n (%)</b>	<b>Total n=49</b>	<b>Group 1 n=7</b>	<b>Group 2 n=32</b>	<b>Group 3 n=10</b>	<b>p-value</b>
Age, years	19 ± 6	19 ± 6	19 ± 6	21 ± 4	0.674
Male sex	43 (87)	7 (100)	27 (84)	9 (90)	0.506
Weight at Operation, kg	71 ± 25	67 ± 20	71 ± 29	72 ± 16	0.923
Postop Length of Stay, days	5 ± 2	6 ± 3	6 ± 2	4 ± 1	0.156
Follow-Up, years	5 ± 5	14 ± 4	5 ± 3	0.2 ± 0.2	< 0.001
Follow-Up to Last Known Alive, years	8 ± 6	17 ± 3	8 ± 3	0.7 ± 0.5	< 0.001
Valve Reintervention	13 (27)	7 (100)	6 (19)	0 (0)	< 0.001
Time to Rein- tervention, years	4 ± 4	5 ± 5	3 ± 3	-	0.606
CTD	18 (37)	0 (0)	16 (50)	2 (20)	0.021
Death	2 (4)	1 (14)	1 (3)	0 (0)	0.307
Time to Death, years	5 ± 8	11	0	-	-
Follow-Up to Most Recent Echo, years	5 ± 5	10 ± 8	5 ± 4	0.4 ± 0.4	< 0.001
Prior Ross Procedure	25 (51)	7 (100)	11 (34)	7 (70)	0.003
Leaflet Manipulation	15 (31)	3 (43)	5 (16)	7 (70)	0.004
Etiology					
Congenital	21 (49)	0 (0)	15 (47)	6 (60)	0.509
Bicuspid	14 (33)	1 (100)	9 (28)	4 (40)	
Acquired	7 (16)	0 (0)	7 (22)	0 (0)	
Other	1 (2)	0 (0)	1 (3)	0 (0)	
Redo	28 (57)	7 (100)	14 (44)	7 (70)	0.016
Sternotomy					
Concomitant Procedure	31 (63)	4 (57)	18 (56)	9 (90)	0.145
Preop AI (0 = none, 1 = trivial, 2 = mild, 3 = moderate, 4 = severe)	1.9 ± 1	2.4 ± 9.7	2.1 ± 0.9	2.4 ± 9.7	0.203
Greater Than Trivial Preop AI	34 (69)	5 (71)	21 (66)	8 (80)	0.685
Greater Than Mild Preop AI	16 (33)	4 (57)	7 (22)	5 (50)	0.083

Variable Median $\pm$ std or n (%)	Total n=49	Group 1 n=7	Group 2 n=32	Group 3 n=10	p-value
Preop MR (0 = none, 1 = trivial, 2 = mild, 3 = moderate, 4 = severe)	0.86 $\pm$ 0.68	0.71 $\pm$ 0.49	0.88 $\pm$ 0.7	0.9 $\pm$ 0.74	0.835
Postop AI (0 = none, 1 = trivial, 2 = mild, 3 = moderate, 4 = severe)	1.4 $\pm$ 0.9	2.2 $\pm$ 1.0	1.3 $\pm$ 0.9	1.2 $\pm$ 0.6	0.030
Greater Than Trivial Postop AI	23 (47)	6 (85)	14 (44)	3 (30)	0.064
Greater Than Mild Postop AI	6 (12)	4 (57)	2 (6)	0 (0)	< 0.001
Postop MR (0 = none, 1 = trivial, 2 = mild, 3 = moderate, 4 = severe)	1.0 $\pm$ 0.8	1.1 $\pm$ 0.4	1.1 $\pm$ 0.9	0.8 $\pm$ 0.4	0.586
Most Recent AI (0 = none, 1 = trivial, 2 = mild, 3 = moderate, 4 = severe)	1.5 $\pm$ 0.9	-	1.5 $\pm$ 1.0	1.6 $\pm$ 0.8	0.926
Most Recent Greater Than Trivial AI	20 (50)	-	15 (50)	5 (50)	1.000
Most Recent Greater Than Mild AI	7 (18)	-	6 (20)	1 (10)	0.471
Most Recent MR (0 = none, 1 = trivial, 2 = mild, 3 = moderate, 4 = severe)	1.2 $\pm$ 0.6	-	1.2 $\pm$ 0.9	1.2 $\pm$ 0.6	0.921
Perfusion Time, min	197 $\pm$ 57	160 $\pm$ 20	177 $\pm$ 41	282 $\pm$ 28	< 0.001
Cross-Clamp Time, min	143 $\pm$ 52	108 $\pm$ 23	123 $\pm$ 28	230 $\pm$ 24	< 0.001
Preop Aortic Annulus, mm	28 $\pm$ 5	31 $\pm$ 6	26 $\pm$ 5	29 $\pm$ 4	0.064



Variable Median + std or n (%)	Total n=49	Group 1 n=7	Group 2 n=32	Group 3 n=10	p-value
Postop Aortic Annulus, mm	22 ± 3	26 ± 6	22 ± 2	21 ± 2	0.027

**Table 2.** Stats by reintervention. Group 1 = no reintervention. Group 2 = reintervention.

Variable Median + std or n (%)	Total n=49	Group 1 n=36	Group 2 n=13	p-value
Age, years	20 ± 6	20 ± 5	17 ± 7	0.121
Male sex	36 (73)	31 (86)	12 (92)	0.559
Prior Procedure				
David	32 (65)	26 (72)	6 (46)	< 0.001
Yacoub	7 (14)	0 (0)	7 (54)	
Modified Yacoub	10 (20)	10 (28)	0 (0)	
Weight at Operation, kg	71 ± 25	73 ± 25	65 ± 25	0.317
Postop Length of Stay, days	5 ± 2	5 ± 2	6 ± 3	0.235
Follow-Up, years	5 ± 5	3 ± 4	11 ± 5	< 0.001
Follow-Up to Last Known Alive, years	8 ± 6	5 ± 4	13 ± 5	< 0.001
CTD	18 (37)	14 (39)	4 (31)	0.603
Prior Ross Procedure	25 (51)	17 (47)	8 (62)	0.376
Leaflet Manipulation Etiology	15 (31)	11 (31)	4 (31)	0.989
Congenital	21 (49)	17 (47)	4 (57)	0.584
Bicuspid	14 (32)	11 (31)	3 (43)	
Acquired	7 (16)	7 (19)	0 (0)	
Other	1 (2)	1 (3)	0 (0)	
Redo Sternotomy	28 (57)	20 (56)	8 (62)	0.709
Concomitant Procedure	31 (63)	25 (69)	6 (46)	0.135
Death	2 (4)	1 (3)	1 (8)	0.443
Time to Death, years	5 ± 8	0	11	-
Follow-Up to Most Recent Echo, years	5 ± 5	4 ± 4	9 ± 6	0.002
Preop AI (0 = none, 1 = trivial, 2 = mild, 3 = moderate, 4 = severe)	1.9 ± 1.0	2.0 ± 1.1	1.8 ± 0.8	0.625
Greater Than Trivial Preop AI	34 (69)	26 (72)	8 (62)	0.474

Variable Median + std or n (%)	Total n=49	Group 1 n=36	Group 2 n=13	p-value
Greater Than Mild Preop AI	16 (33)	12 (33)	4 (31)	0.866
Preop MR (0 = none, 1 = trivial, 2 = mild, 3 = moderate, 4 = severe)	0.9 ± 0.7	0.8 ± 0.6	1.0 ± 0.8	0.38
Postop AI (0 = none, 1 = trivial, 2 = mild, 3 = moderate, 4 = severe)	1.4 ± 0.9	1.1 ± 0.7	2.2 ± 0.6	< 0.001
Greater Than Trivial Postop AI	23 (47)	12 (33)	11 (85)	0.001
Greater Than Mild Postop AI	6 (12)	0 (0)	6 (46)	< 0.001
Postop MR (0 = none, 1 = trivial, 2 = mild, 3 = moderate, 4 = severe)	1.0 ± 0.8	0.8 ± 0.7	1.4 ± 0.9	0.05

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