

Valve-sparing aortic root replacement and remodeling with complex aortic valve reconstruction in children and young adults with moderate or severe aortic regurgitation

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Objectives: The durability of valve-sparing aortic root procedures with aortic regurgitation due to leaflet disease is questioned. Here, we review our experience in combined aortic root and valve reconstruction in children and young adults.

Methods: All valve-sparing aortic root procedures from 2000 to 2012 were reviewed, and patients with aortic valve repair beyond resuspension were included. Root procedures were classified as replacement with reimplantation, root remodeling, or aortic annular and sinotubular junction stabilization. The primary end point was structural valve deterioration, a composite of aortic valve reoperation and/or moderate or greater regurgitation at follow-up.

Results: Thirty-four patients were included during the study period. The surgery consisted of reimplantation in 13 patients, remodeling in 16 patients, and annular and sinotubular junction stabilization in 5 patients. Valve repair consisted of leaflet procedures in 26 patients and subannular reduction in 15 patients. During a median follow-up of 4.2 months (range, 2 weeks–8 years), there were 5 reoperations for aortic valve replacement due to aortic regurgitation, and 2 patients presented with moderate or greater regurgitation. Freedom from structural valve deterioration was $70.1\% \pm 10.3\%$ at 1 year and remained stable thereafter, although it was significantly worse in the reimplantation group ($P = .039$). A more severe degree of preoperative aortic regurgitation ($P = .001$) and smaller graft to aortic annulus ratio ($P = .003$) were predictors of structural valve deterioration.

Conclusions: Valve-sparing root and valve reconstruction can be done with low operative risk and allows valve preservation in most patients. These data should question the assumption that reimplantation is superior when associated with complex valve reconstruction. (J Thorac Cardiovasc Surg 2014;147:1768–76)

Aortic root dilation is uncommon in children, and is mostly related to congenital heart disease and its repair or to connective tissue disorders.^{1–3} Aortic root replacement or remodeling is indicated to avoid the risk of progressive valvular incompetence, aneurysm rupture, and dissection, although these are extremely rare events and criteria for surgical management have not been defined in the pediatric population.⁴

Valve-sparing aortic root replacement with reimplantation has obvious advantages over composite graft or

homograft root replacement and aortic root remodeling, and is progressively being introduced for root replacement in children. Although long-term data in adults have shown excellent results,⁵ outcomes following valve-sparing aortic replacement with reimplantation for children with an aortic root aneurysm are sparse and the literature has suggested that valve-sparing procedures, especially root remodeling, may have a higher failure rate.²

Aortic regurgitation or anomalous leaflet anatomy usually precludes valve-sparing aortic root procedures for aneurysmal disease. However, avoiding valve replacement in a growing child, or at least delaying it as long as possible, is desirable particularly because aortic valve repair in children has shown excellent long-term results.^{6–9} Just as the indications for valve-sparing root replacement are expanding in adults to include patients with aortic valve insufficiency^{10,11} or even as an adjunct to valve repair to increase the durability of aortic valve repair in selected adults,^{12–14} interest in combined aortic valve reconstruction and aortic root replacement in pediatric populations has emerged. The objective of our study was to review our initial experience in combined aortic root replacement or remodeling with complex aortic valve reconstruction.

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Abbreviations and Acronyms

STJ = sinotubular junction

SVD = structural valve deterioration

METHODS**Study Design**

We retrospectively reviewed all patients who underwent aortic valve-sparing root procedures at our institution between 2000 and 2012. Patients with moderate or severe aortic regurgitation before repair, and who underwent aortic root procedures and aortic valve repair beyond resuspension, were included. Root procedures were classified as root replacement with reimplantation (David procedure) or root remodeling (Yacoub procedure or other techniques other than a David reimplantation procedure). The primary end point was structural aortic valve deterioration (SVD), a composite of aortic valve reoperation and/or moderate or greater aortic regurgitation at follow-up. Clinical or treatment variables were recorded to determine predictors SVD. All patients underwent follow-up to death or June 2012. The study was approved by the Boston Children's Hospital Institutional Review Board and individual patient consent was waived.

Surgical Technique

Cardiopulmonary bypass with moderate systemic hypothermia was used in all patients. Myocardial protection consisted of antegrade magnesium-lidocaine blood cardioplegia. The surgical techniques used for aortic root and valve repair were chosen by the operating surgeon based on the mechanisms of regurgitation and aortic root pathology. A dilated annulus was usually reduced by placing either a subannular felt strip sutured to the left ventricular outflow tract with externalized sutures, or a cut ring of tube graft placed externally and below the coronary arteries, sutured using interrupted pledgeted sutures, or a David valve-sparing aortic root replacement with reimplantation. Individually dilated sinuses of Valsalva were plicated; resected and the aortic root reapproximated primarily or replaced with a tongue of graft; or the entire root was entirely replaced either with a David valve-sparing aortic root replacement with reimplantation or Yacoub aortic root remodeling, splitting a tube graft to accommodate the 3 aortic commissures. The techniques used were variable and adapted to the underlying disease and surgeon preference.

Statistical Analysis

Statistical analyses were performed with SPSS software (version 21, IBM-SPSS Inc, Armonk, NY). Data are presented as mean \pm standard deviation or median (range) where appropriate. Regarding the aortic dimensions, the following dimensions were used and normalized using *z* scores: the aortic annulus, aortic root (measured as the largest dimension of the sinuses of Valsalva), and sinotubular junction (STJ). Continuous variables were analyzed with 1-way analysis of variance or the Kruskal-Wallis test when appropriate, and categorical variables using the χ^2 test or Fisher exact test. Actuarial estimates were calculated using the Kaplan-Meier method and differences between curves were assessed by the log-rank test. All statistical tests were 2-tailed and $P < .05$ was taken as significant.

RESULTS**Valve-Sparing Aortic Root Procedures**

Among 48 valve-sparing aortic root procedures conducted during the study period, 34 patients presented with moderate or severe aortic regurgitation before repair

and their repair included aortic or neo-aortic valve reconstruction and were included in the study.

Aortic root procedures consisted of a valve-sparing aortic root replacement following a David V-type reimplantation (reimplantation group) in 13 patients, valve-sparing aortic root remodeling (remodeling group) in 16 patients, and functional annular stabilization by subannular reduction and STJ stabilization with a tube graft in 5 patients. All included procedures were on the systemic semilunar valve and root; that is, aortic for patients with normally related great vessels, or the neo-aortic root and valve for patients with dextro-transposition of the great arteries after an arterial switch operation. Root remodeling procedures ($n = 16$) consisted of Yacoub-type aortic root replacement of all 3 sinuses of Valsalva in 5 patients (31.3%) and sinus resection or reduction in 11 patients (68.8%; 6 single sinus, 3 double sinus, and 2 triple sinus resection or reduction). A portion of these patients have been reported previously.¹⁵

Demographics

Patient baseline characteristics are summarized in Table 1. The mean age at surgery was 15.4 ± 8.7 years. Nine patients had connective tissue disorders (3 Marfan syndrome, 4 Loeys-Dietz syndrome, and 2 unspecified); these patients underwent more reimplantation procedures (6 out of 9; 67%), compared with those without connective tissue disorders (7 out of 21; 33%; $P = .11$). Twenty-one patients had previously repaired congenital heart disease. Three patients had coronary anomalies: 1 with single coronary ostium, 1 with intramural right coronary artery, and 1 patient with the left main coronary artery compressed between the neo-aortic and neopulmonary roots after an arterial switch operation, along with the circumflex artery from the right coronary artery. There were no significant differences in baseline characteristics between groups; in particular, there was no difference in the proportion of patients with conotruncal anomalies.

Aortic Valve Reconstruction

Aortic valve repair consisted of leaflet procedures in 26 patients and subannular reduction in 15 patients. Twenty-five patients had annular stabilization. Techniques of aortic root and valve repair are detailed in Table 2. Although there were not any significant differences in valve repair techniques, more patients in the reimplantation group had STJ and aortic annulus stabilization (11 out of 13) than in the remodeling group (9 out of 16; $P = .1$), as well as aortic root replacement (13 out of 13 vs 5 out of 16; $P < .001$) and ascending aortic replacement (13 out of 13 vs 8 out of 16; $P = .003$). The graft size used did not differ significantly between groups (median, 26 mm; $P = .53$).

Outcomes and Predictors of SVD

There were no early deaths. At predischARGE echocardiography, 19 patients had no or trivial aortic regurgitation

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