Pediatric valve-sparing aortic root replacement: Approaching perfection

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Central Message

Aortic root replacement using the reimplantation valve-sparing technique appears to be reproducible in complex pediatric patients with resultant excellent outcomes.

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Over the past 3 decades, aortic root replacement surgery has evolved with much attention given to preserving the aortic valve when possible. Valve-sparing operations including the reimplantation and remodeling procedures, along with modifications, have gained support with the techniques being expanded to treat aneurysmal aortic disease in children. The benefits of conserving the valve are clear: avoidance of complications associated with anticoagulation therapy. Concerns over the development of aortic valve insufficiency or a pseudoaneurysm necessitating further surgery have been the primary arguments against these operations.¹

In this issue of the *Journal*, Fraser and colleagues,² present single-institution data from a 20-year period describing 100 consecutive cases of valve-sparing aortic root replacement (VSRR) in patients aged less than 18 years (median, 13.6 years). As expected, the majority of the patients had underlying connective tissue disease, such as Marfan syndrome (51%) or Loeys–Dietz syndrome (39%). Although 4 different procedures were used to achieve a VSRR over the past 20 years, the majority (84%) of patients underwent the reimplantation technique.³ Concomitant aortic valve repair was performed in 9 patients undergoing VSRR. Perioperative mortality was only 2%, with both patients having connective tissue disease. With a median follow-up of 4.7 years, 7 additional patients died, 4 of whom died of complications related to connective tissue disease. Reimplantation VSRR was found to be durable with a 5-year freedom from reintervention or 94%. Six patients required late reintervention for the development of a pseudoaneurysm.⁴ A total of 4 patients (4.8%) who underwent the reimplantation VSRR required late aortic valve replacement because of the development of annular dilatation and significant aortic insufficiency. Of note, none of the 12 patients with a bicuspid aortic valve required subsequent reoperation for aortic valve insufficiency.

The significance of the article is understandable in that Fraser and colleagues² present the largest experience with VSRR in the pediatric population. The group at Johns Hopkins have a well-deserved reputation as being the experts in VSRR surgery. Their reimplantation technique appears reproducible, even in pediatric patients with aortic valve insufficiency or a bicuspid aortic valve. The possibility of aortic valve replacement after the reimplantation VSRR is low. In addition, the authors provide valuable surgical technical tips when performing the reimplantation VSRR. Furthermore, the authors have gained an incredible amount of experience in the management of patients with connective tissue aortopathies. Through this understanding and to our benefit, they have been able to better define their guidelines for timing of aortic root replacement.

There are a few limitations to the study. David and colleagues⁵ demonstrated that patients with Marfan syndrome undergoing aortic valve-sparing root replacement have progressive aortic insufficiency over time. Given that the median follow-up in the current study is 4.7 years, one must acknowledge that the risk for surgical intervention may be higher with time. In addition, these are the results of a high-volume center performing a technically demanding procedure, and lower-volume centers may not be able to duplicate the results.⁶ Last, more detail on the failures of the patients undergoing reimplantation VSRR may have been helpful. One has to wonder about the mechanism of annular dilatation and the selective need for better aortic root stabilization.

Fraser and colleagues² show that the reimplantation VSRR is an effective, reproducible technique in children. With midterm follow-up, the risk for surgical reintervention is low. Modifications in annular stabilization in select patients may further improve the excellent outcomes. Longer follow-up on aortopathy in pediatric patients with connective tissue disorder will undoubtedly aid in defining surgical timing and the need for any adjustments.

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