

One ring* to rule them all.

*lat. anulus

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When valve-sparing root replacement was introduced, it soon became an attractive option for children and adolescents presenting with aortic root aneurysm due to disorders of connective tissue. In this young patient population, a Bentall procedure with a mechanical valve and life-long anticoagulation seemed a particularly uninviting option. At that time, many surgeons chose to perform a remodeling procedure as it was thought to be simpler in execution. While the initial results were very promising, patients soon started to come back with aortic valve regurgitation due to annular dilatation. This prompted many surgeons to adopt the reimplantation technique as the method of choice for young patients with disorders of connective tissue.

In the current edition of the *Journal*, Chauvette and colleagues report results from the AVIATOR registry that encompasses a large number of patients that underwent valve-sparing root replacement using different techniques¹.

This article conveys several important messages: (1) Valve-sparing root replacement is safe. There was only 1 perioperative death resulting in a 0.4% mortality. (2) Supporting the aortic anulus is key. As long as the aortic anulus is supported, it does not matter whether a reimplantation or a remodeling technique is performed. (3) Valve-sparing root replacement in young patients with disorders of connective tissue is a durable procedure – but not as durable as we had hoped for. The authors report 22% of AR \geq 2 after 10 years. While this in line with previous reported results in patients with Marfan syndrome, it is underwhelming to note that the median time to develop AR \geq 2 was less than 3 years. Given the vast experience of participating centers that have contributed to this study, mistakes in strategy and surgical technique seem unlikely to explain this rather high incidence of aortic valve regurgitation after such a short period of time. This raises the important question how we can identify those patients that will develop aortic regurgitation over time? Is it just bad luck or are there anatomical factors that predispose patients for failing valve-sparing root replacements that we have not yet identified?

Dr. Cameron and colleagues from Johns Hopkins have always stressed the fact that the cusps of patients with Marfan syndrome are less resistant than those of patients without connective tissue disorders. Patients often present with fenestrations earlier in life than we usually see^{2,3}. Are we sometimes overestimating the durability of these thin cusps? Of course, one can argue

that previous research has shown that the annual risk associated with a reoperation is less than the risk for a major neurological event with a mechanical valve⁴. Nevertheless, I hope that the AVIATOR investigators will help us to tease out additional factors that will make outcomes after valve-sparing root replacement more predictable.

The authors have to be commended for including the number of patients with disorders of connective tissue that underwent AVR or a Bentall procedure during the same time frame (n=134) and those that underwent VSSR without having a (known) connective tissue disorder (n=1004). These numbers provide important epidemiological data and allow readers to set things in perspective.

For several decades, Marfan syndrome has been the only seriously considered differential diagnosis in young patients presenting with aortic root aneurysm. Over the past decade, the community has slowly accepted the idea that patients presenting with aortic aneurysms and dissections are part of a wide spectrum of genetically mediated diseases that present in syndromic as well as non-syndromic forms. This study comes at a time where we rapidly expand our knowledge on the molecular basis of thoracic aortic disease and discover new mutations associated with aortic aneurysms each year. It becomes evident that we have to apply the knowledge gained from patients with disorders of connective tissue to all young patients that we operate on.

Recent years have seen new surgical methods to treat aortic root aneurysm in young patients with disorders of connective tissue such as the PEARS procedure in which a custom-made synthetic mesh is wrapped around the aortic root. Proponents of this technique suggest interventions at diameters that would not be considered appropriate thresholds for surgery according to current guidelines⁵.

While this was not the primary aim, the current study by the AVIATOR investigators provides a much-needed landmark analysis regarding the morbidity and mortality associated with VSRR in a contemporary cohort of patients with disorders of connective tissue.

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