In this issue, Urbanski and colleagues\(^1\) report a single-center analysis of 42 patients fulfilling the original Marfan syndrome (MFS) Ghent criteria who received “patient-tailored aortic root repair.” Depending on the observed pathology and aortic root asymmetry, patients underwent a modified remodeling technique with replacement of 1, 2, or 3 sinuses of Valsalva. The majority of patients (64\%) in this small cohort (including all those younger than age 40 years) underwent replacement of all 3 sinuses with a patch technique: Each sinus was resected and replaced with a polyethylene terephthalate patch sewn directly onto the aortic annulus rather than onto a rim of remnant aortic tissue. This technical point is purported to be the important feature that distinguishes it from the original remodeling technique and may theoretically permit annular stabilization. Surgical results were excellent with no operative mortality, no late proximal aortic events, and an acceptable freedom from aortic valve replacement of 91\% at 8 years, although the mean follow-up was only 6.1 years. In our experience with root remodeling in patients with MFS, failure due to annular dilatation did not ensue until the second decade of follow-up.\(^2\)

Patients with MFS have an abnormality of the fibrous skeleton of the heart leading to annuloaortic ectasia and in some to mitral valve prolapse.\(^3\) Histologic examination of the aortic wall media in MFS demonstrates elastic fiber fragmentation and an increase in mucopolysaccharides that is more pronounced in aneurysmal tissue, but is still pathologic in nondilated aortic tissue.\(^4-6\) The authors\(^1\) state that in their cohort, “the cause of preoperative aortic insufficiency... was mostly cusp- rather than root-related.” In our experience, patients with MFS do not have a primary abnormality of their aortic cusps. Aortic insufficiency (AI) in these patients is often central and results from a dilated aortic annulus and/or sinotubular junction, which temporally precede and result in the development of AI. AI does not occur secondary to isolated aneurysms of the sinuses of Valsalva and primary cusp pathology most often results in eccentric AI. Long-standing AI may lead to secondary changes in the cuspal tissue such as stress fenestrations, significant thickening of the free margin of the cusps and apparent infolding or rolling over of the free margin toward the sinuses. Cusp prolapse may be induced during root repair, or less often may occur primarily, and should be corrected during surgery.

Urbanski and colleagues\(^1\) are to be commended for developing an individualized approach to aortic root repair. We believe the authors’ modified remodeling technique will prove to be durable in older patients with a degenerative aneurysm and a nondilated annulus. For those with MFS and other hereditary aortopathies, the best surgical option will almost certainly remain an aortic valve-sparing operation with reimplantation. The excellent very-long-term results observed in several centers following reimplantation\(^2,7-9\) result from the surgeons’ ability to address every component of the pathologic root, including correcting and stabilizing annuloaortic ectasia, excising all pathologic proximal aortic tissue, and correcting any associated cusp abnormality.

References


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Three-dimensional reconstruction of an aneurysmal aortic root in a patient with Marfan syndrome.


