Type A dissections in patients with Marfan syndrome: When less is not more

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Advantages of alternative TAAD management strategies in patients with Marfan syndrome.

Central Message

An aggressive initial approach combining total arch replacement and treatment of the descending aorta in patients with Marfan syndrome and type A aortic dissection reduces late reoperation rates.

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Ideal management of patients with Marfan syndrome (MFS) and type A aortic dissection (TAAD) remains an unresolved dilemma between mitigating the potentially devastating consequences of untreated aortic disease and minimizing morbidity and mortality associated with surgical intervention. MFS, a connective tissue disorder resulting from a mutation in the fibrillin-1 gene, is associated with a number of aortic pathologies,^{1,2} and approximately 20% to 25% of affected individuals will have TAAD develop in their lifetimes.³ Optimal treatment of TAAD in these patients requires consideration of the unique challenges associated with MFS, including high rates of recurrent aneurysm in the distal arch as well as thoracic and thoracoabdominal aortic involvement, requiring subsequent reoperations.^{4,5}

Acute TAAD in the general population is often treated with a minimalist approach. The life-threatening arch dissection is addressed in a limited and urgent manner, followed by subsequent elective repair of remaining distal aortic disease as needed. This strategy is suboptimal in the long term in patients with MFS, who have greater than 12-fold increased odds of requiring distal aortic reintervention than do patients without MFS.⁴ In this issue of the *Journal*, Ma and colleagues⁶ report on an alternative approach. Their group had previously published a consecutive series of patients with MFS and TAAD treated with an aggressive early approach combining total arch replacement and open endovascular stent placement into the descending aorta (“frozen elephant trunk” technique).⁷ The approach involves the use of a specially designed 4-branched arch graft in conjunction with the Cronus stent graft (MicroPort Scientific Corporation, Shanghai, China) to treat the descending aorta.⁶ The current study is one of the largest published cohorts of patients with MFS treated with this extensive approach, and the long-term follow-up by Ma and colleagues⁶ in this issue of the *Journal* provides much needed insight into the progression of disease and late causes of morbidity and mortality in this high risk population.

The study of Ma and colleagues⁶ challenges the “expert consensus” to avoid stent grafts in patients with MFS because of concerns regarding the chronic effects of radial forces on an intrinsically weak aorta and the durability of stent grafts.⁸ Of the 106 subjects, 11 required additional interventions for thoracoabdominal aortic aneurysm (n = 8) or thoracic endovascular aortic repair for distal new entry (n = 3); however, 8 years from initial operation, freedom from reoperation was 84.2%, with a 74.1% survival. In contrast, Schoenhoff and coworkers³ reported a 48% reoperation rate in the distal aorta for patients with MFS who presented with acute aortic dissection and were treated primarily with proximal aortic interventions. The experience of Ma and colleagues⁶ demonstrates that superior outcomes can be achieved in patients with MFS by using their total arch replacement plus frozen elephant trunk approach. Furthermore, the protective effect of youth in this cohort (age 34.5 ± 9.7 years) should not be underestimated, and further supports more extensive intervention at the initial encounter. Many lessons can be gleaned from the experience of Ma and colleagues⁶: the use of standardized conduits, emphasis on early cerebral and distal aortic reperfusion, a low threshold for concomitant aortic root repair, and close radiographic and clinical follow-up have the potential to improve long-term outcomes in high-risk patients with MFS who present with TAAD. We commend Ma and colleagues⁶ on their

impressive results in this complex patient population. Further multi-institutional analysis is warranted to investigate widespread generalizability.

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