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Commentary: Endovascular repair in Marfan syndrome: Viable bailout but not ready for prime time

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Thoracic endovascular aortic repair (TEVAR) has evolved to become standard of care for patients with degenerative aortic disease of the descending thoracic aorta, blunt aortic injury, and complicated type B aortic dissection (TBAD).¹ Advantages include ease of implantation, less surgical trauma and physiological derangements, and lower early morbidity and mortality compared with open distal aortic repair.² Nonetheless, TEVAR has been associated with an increased risk of aortic-related reinterventions.³

Patients with hereditary thoracic aortic disorders (HTAD), including Marfan syndrome, are considered to have a relative contraindication for TEVAR.¹ Despite reasonable perioperative success, significant concerns arise in mid to late follow-up. Complications, including endoleak, aortic reintervention, stent migration, and retrograde type A dissection, all occur at substantially higher rates than in patients without HTAD.^{4,5} Although Jiang and colleagues⁶ reported good early success and late survival in 26 patients with TBAD treated with TEVAR over a period of 10 years, endoleaks occurred in 27% of patients and they observed a 50% reintervention rate at 10 years. Furthermore, of patients who died during follow-up, 50% had an aortic-related death. The authors' experience provides a

Stent-induced new entry tear in a Marfan patient ultimately requiring open distal repair.

CENTRAL MESSAGE

Endovascular repair for type B dissection is a viable bailout strategy, but demands lifelong surveillance in patients with hereditary thoracic aortic disorders.

cautionary tale about the malignant natural history of stent-induced new entry tears in patients with HTAD and highlights the importance of lifelong surveillance of these patients, particularly those with a patent false lumen after TEVAR.

The authors did not discuss what led them to select an endovascular approach for these young patients with Marfan syndrome. Although some studies have reported high rates of morbidity and mortality with open distal aortic repair, particularly when analyzing regional or national registries, centers of excellence provide excellent results for open repair for patients with HTAD.⁷ In the GenTAC registry, operative mortality following open thoracoabdominal aortic repair in patients with HTAD was 1.3%;⁷ Coselli and colleagues⁸ reported an operative mortality of 4% in patients with Marfan syndrome undergoing distal aortic repair for dissection.

We agree that TEVAR may be used as an emergency bailout procedure in patients with HTAD and life-threatening complications such as rupture, malperfusion, or aorto-enteric or bronchial fistulas. Furthermore, TEVAR is an excellent option when landing in graft proximally and distally, such as in patients with intercostal patch aneurysms. We do not think endovascular repair with fenestrated or branch grafts for the arch or thoracoabdominal aorta is a viable option given the fragile aortic tissue, and risk of branch vessel complications in these patients. If TEVAR is to be used in HTAD, the following principles should be

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applied: excluding stent grafts with bare stents and barbs; landing in a preexisting graft or a long segment of nondilated native aorta; minimizing oversizing; and avoiding balloon dilatation.

Despite their encouraging results, we agree with the authors that TEVAR for TBAD in patients with Marfan syndrome should only be used in those who are at high or prohibitive risk for open repair and with suitable anatomy for TEVAR. If TEVAR is performed in these patients, closer surveillance should occur, particularly if false lumen thrombosis is not achieved.

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