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# Aortic endovascular stenting in patients with systemic connective tissue disorders: does the prohibitive dogma still stand tall?

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**Abstract**

Endovascular repair of thoracic aortic diseases can provide satisfactory outcomes in elective and certain emergency cases involving the descending thoracic and aortic arch. However, open repair remains the gold standard method of aortic root pathologies and certain aortic arch pathologies, such as extended dissection. Nevertheless, the use of endovascular stenting in patients with connective tissue disorders has not been fully explored because the aortic tissues are fragile and the likelihood of keeping the stent in place is low because of its progressive dilatation and subsequent requirement for open repair at a later stage when the stent graft fails. Our brief review focuses on current evidence of the use of stents in patients with connective tissue disorders and whether such practice can be expanded further.

**Keywords**

Aorta, endovascular, open repair, connective tissue disorders, stent graft, thoracic aorta

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## Current evidence

In September 1990, Parodi et al.[1] performed the first successful endovascular aneurysm repair procedure in the Western world in a patient with severe chronic obstructive pulmonary disease and an abdominal aortic aneurysm. Less than 2 years later, the first thoracic aortic stent graft repair (thoracic

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endovascular aortic repair) was performed in a patient with an enlarging descending thoracic aortic false aneurysm 30 years after aortic coarctation repair.[2] The frequency of endovascular stent grafting procedures has since dramatically increased, and such procedures have become the treatment of choice for descending thoracic aortic disease in many centers worldwide.[3,4] However, their use in patients with connective tissue disorders as an alternative to open surgery remains controversial and largely unsupported by current guidelines and expert consensus.[2,5,6]

Connective tissue disorders that have a known association with aortic disease include Marfan syndrome (MFS), Ehlers–Danlos syndrome, Loeys–Dietz syndrome (LDS), and familial thoracic aortic aneurysms and dissections (FTAAD). Several recent studies have focused on the use of endovascular stenting in patients with MFS.[7–15] Parisi et al.[16] noted the technical feasibility of endovascular repair in these patients, quoting success rates approaching 100% and associated low early in-hospital morbidity and mortality rates. However, an analysis of mid- to long-term follow-up data revealed a high reintervention rate secondary to primary and secondary endoleaks and a low rate of positive aortic remodeling.[15] This is attributed to the radial force exerted by the stent, resulting in circumferential stress at the site of the landing zones of an abnormally fragile aortic wall affected by connective tissue disease.

Importantly, most patients with MFS and other connective tissue disorders are young with minimal comorbidities mandating the need for a durable repair. According to the published findings from the International Registry on Acute Aortic Dissections, patients with MFS and type B aortic dissections experienced lower 5-year freedom-from-reintervention rates than patients without MFS, particularly when endovascular therapy (32.0% vs. 71.5%, respectively) rather than open surgery (54.4% vs. 88.0%, respectively) was pursued.[17] Similarly, in a systematic review of 54 patients with MFS who developed type B aortic dissection and underwent endovascular repair across 12 previous publications, Pacini et al.[18] noted low rates of in-hospital mortality (1.9%) and early complications [spinal ischemia (1.9%), stroke (1.9%), and conversion to open surgery (3.7%)] but a high rate of subsequent complications at follow-up. Higher rates of endoleaks were reported following repairs for chronic dissections (31%) than acute dissections (9%). Of these, 16% of patients required additional endovascular stent grafting and 18% required open surgery. Twelve percent of patients died during an average follow-up period of 2.5 years. Patients in whom the stent graft was deployed on landing zones within previous surgical grafts did not develop endoleaks. In contrast to this, Coselli et al.[19] reported a low operative mortality rate (4%) and an 86% rate of freedom from late repair failure at 8 years with open repair, exceeding the medium- and long-term outcomes from all endovascular series in such patients. Furthermore, in a retrospective study of patients undergoing endovascular repair for type B dissection, Dong et al.[20] reported a high incidence of retrograde aortic dissection with endovascular repair in patients with MFS. The association of this complication with connective tissue disease was also subsequently shown in a European registry study.[21] The incidence of stent graft–induced new entry tears was also shown to be significantly higher in patients with than without MFS (33% vs. 3%).[22]

Data on endovascular treatment of aortic aneurysms and dissections for patients with Ehlers–Danlos syndrome, LDS, and FTAAD are scant.[23] Outcomes similar to those in patients with MFS are expected because of the similar aortic fragility and predisposition to progressive aortic dilatation. Aortic disease is more aggressive in patients with LDS than MFS, with aortic

dissection described in patients with aortic diameters as small as 39 mm.[24] The risk of progressive arterial dilation predisposes to stent graft failure when landing zones lie within areas of the native aorta. A uniform management strategy is lacking for patients with FTAAD because of the variation in genetic penetrance and in genotype and phenotype correlations. Similar to other genetic aortic diseases, endovascular therapies for FTAAD remain unendorsed by the pioneers in aortic surgery. In a study of 255 familial abdominal aortic aneurysms, van de Luijtgaarden et al.[24] reported a higher complication rate (35.3% vs. 19.1%), reintervention rate (39.2% vs. 20.1%), and aneurysmal growth rate (20.8% vs. 9.5%) following endovascular aneurysm repair in patients with than without familial disease.

Although the general consensus is that endovascular repair is not recommended in patients with connective tissue disorders unless the risks of surgical repair are considered prohibitive, endovascular stenting is considered permissible in certain clinical scenarios assessed on a case-by-case basis.[25,26] First, endovascular repair can be considered if the stent graft is set to be deployed between two previously placed surgical grafts where the feared risk of subsequent landing zone dilation is already circumvented. Second, intercostal patch aneurysms that occur after surgical repair of thoracoabdominal aneurysms, which pose particular challenges when managed surgically and are associated with increased surgical morbidity, can be considered for endovascular treatment if the proposed landing zones also lie within the surrounding surgical graft(s). Finally, in selected patients with ruptured type B dissections, emergency endovascular bridging repairs can be pursued.[3,21,23]

## Conclusion

Despite major improvements in stent graft materials and implantation techniques, radial force and circumferential stress on the native aorta remains a significant source of concern because of the resultant stent graft failure in patients with aortic fragility secondary to connective tissue disorders. Open surgical repair remains the gold standard in most patients.

### Declaration of conflicting interest

The authors declare that there is no conflict of interest.

### Ethics

This study did not require ethical approval because it did not involve examination of human tissue or disclosure of patient information.

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