Stent grafting in Marfan syndrome? We are not convinced

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In this issue of the *Journal*, Faure and colleagues¹ present a well-executed, novel approach to stent grafting in patients with Marfan syndrome. To summarize, they describe a strategy for treatment of type B dissection in which a stent graft is used to cover the entry tear of a type B dissection, and bare-metal, uncovered stents are used to expand the intimal flap against the adventitially bound false lumen. They term this technique *STABILISE*. As a bit of background, this is an evolution of a technique in which a covered stent was used to cover the entry tear and bare-metal, uncovered stents were used to line the true lumen of the distal aorta, but without expanding the bare-metal stent to appose the intimal flap with the adventitially bound false lumen. This was termed the *STABLE* strategy.² The putative advantage of the *STABILISE* strategy is that the false lumen is obliterated at the index procedure by balloon expansion of the bare-metal stent. The requirement for future reverse remodeling in the stented segment is thus eliminated, because the false lumen is obliterated at the index procedure within the stented segment, although the false lumen remains patent distally beyond the stents, as was demonstrated experimentally with this approach in 1991.³

Faure and colleagues¹ present data showing fewer peri-procedural complications than have been reported by other experienced centers with stent grafting in connective tissue disease. One can only conclude either that device technology has improved—they used the covered stent-graft C-TAG device proximally (Conformable Gore TAG stent graft; W.L. Gore & Associates, Flagstaff, Ariz) and the uncovered nitinol Zenith Dissection Endovascular Stent device distally (Cook Medical, Bloomington, Ind)—or that their procedural technique is particularly delicate in these dissected, abnormal aortas. Either way, however, it is an overstatement to call the reported results midterm outcomes, because the median follow-up is 15 months, which is short given the long-term outlook in patients with Marfan syndrome.

One also must emphasize the principles and literature regarding stent grafting in patients with Marfan syndrome and other connective tissue disorders. In an expert

Open surgery remains the criterion standard for patients with Marfan syndrome.

Central Message

Stent grafting in Marfan syndrome remains questionable in light of numerous studies demonstrating high morbidity and mortality. Open operation carries low risk and remains the treatment of choice.

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consensus document from the Society of Thoracic Surgeons, stent grafting is not recommended in patients with Marfan syndrome and connective tissue disorders unless the risk of open surgical repair is prohibitive. Further, because these patients are young and the durability of stent grafts is unknown, and because the radial force of stent grafts on abnormal aortas is considered risky, stent graft placement is deemed not prudent and to be avoided.⁴ The 2010 American Heart Association guidelines similarly recommend open operation for treatment of aneurysms resulting from descending thoracic aortic dissection, particularly in the setting of connective tissue disease, in a document coauthored with the American Association for Thoracic Surgery and the Society for Thoracic Surgery.⁵ Similarly, the 2014 European Society of Cardiology guidelines recommend consideration of endografting in patients with Marfan syndrome only in emergencies or as a bridge to definitive surgical therapy.⁶

The complications associated with stenting or stent grafting in patients with connective tissue disorders have been widely described. First, there is a remarkably high rate of endoleak necessitating operative intervention, owing to continued dilation of the abnormal aorta, which is left in situ and used as a landing zone. In a systematic review from the University of Bologna, the overall postprocedural endoleak rate was 29% when stent grafts were landed in native aorta in patients with Marfan syndrome.⁷ The vast

majority of endoleaks were type I, occurring at the junction of the stent graft with the aorta. Conversion to open surgery was necessary in 18% of patients. Mortality was 12.8% at 2.5-year follow-up, and most deaths were aortic related. Similarly, in a series of 16 patients with Marfan syndrome who underwent 19 stent-grafting procedures for descending thoracic and abdominal aortic pathology, 44% had primary treatment failure, with most of these failures necessitating conversion to an open operation, and there was a 25% mortality at 9.3 months.⁸ Overall, at 6.8-month follow-up, only 38% had a successful endovascular procedure and required no subsequent interventions, and 1 of these patients died of multiorgan failure. Treatment failures in this series included cases of type I endoleak, as well as that of a patient who had a retrograde type A dissection, a life-threatening complication. Retrograde type A dissection is a known complication of stent grafting in type B dissection, and its risk is greatly elevated in patients with Marfan syndrome.⁹ In fact, despite the tendency to exclude patients with Marfan syndrome from stent grafting, between 13.5% and 27% of retrograde type A dissections reported in the literature occur in patients with Marfan syndrome.⁹

Consistent with the data on endoleaks in patients with Marfan syndrome, Faure and colleagues¹ report that 1 in 7 patients at 15 months required an arch replacement to manage a type Ia endoleak. This endoleak resulted from the continued enlargement—which one would expect—of the native Marfan aorta. Arch replacement in this case would likely have been avoided by open repair of the type B dissection as the index operation, ideally after a 4- to 6-week period to stabilize the dissection before elective replacement. The finding of Faure and colleagues¹ represents a failure rate of 15% at 15 months, consistent with the expected failure rate derived from reports of stent grafting in the literature.

Long-term risk of rupture is not reduced, and may be increased, by the STABILISE treatment. In this technique, when balloons are used to expand the uncovered metal stent against the intimal flap, this flap, because of fenestrations, will not entirely reappose with the adventitia. Therefore, relamination—a word in the STABILISE acronym—is misleading because the 3 layers of the aorta are not rejoined along its entire length. There are areas of bare adventitia not lined with intima that are up against an uncovered metal stent that has been expanded against this adventitia, which is in contact with pressurized blood in the aorta. Such expansion of an uncovered stent against weak adventitia in a connective tissue disorder may pose a high risk for rupture in the long run.

When is stent grafting appropriate for patients with Marfan syndrome and other connective tissue disorders? We suggest 3 circumstances.¹⁰ First, if the stent graft can be placed between 2 previously placed surgical grafts, this is acceptable because the landing zones for the stent graft

will not dilate with time. Second, intercostal patch aneurysms after open thoracoabdominal aneurysm repair are difficult to expose surgically and represent an important area for stent grafting. In these cases, the stent graft will also lie within a surgical graft, consistent with the previously stated principle. More liberal use of multibranched grafts to reconstruct the visceral segment should decrease this challenging long-term sequela of side-to-side visceral patch anastomoses. Third, emergency cases of rupture can arguably be addressed with stent grafting if the perceived surgical risk is too high for open repair or if a later open surgical revision is planned. Otherwise, the literature confirms that elective repair of type B dissection in patients with Marfan syndrome should be done by open operation. Although the new technology and complex acronyms make STABILISE appear attractive, one must recall that patients with connective tissue disorders are young, and they tend to be excellent operative candidates who seek restoration of a nearly normal life expectancy and reintervention-free long-term survival. In the largest series of open surgical repair of descending thoracoabdominal aortic aneurysms in the setting of dissection in patients with Marfan syndrome, Coselli and colleagues¹¹ describe an operative mortality of 4%, lower than many endovascular series, and a freedom from late repair failure of 86% at 8 years, well above that of any endovascular series. We continue to advocate for open operations for these patients.

In summary, stent grafting in patients with Marfan syndrome remains questionable in light of numerous studies demonstrating high rates of subsequent open operation for aneurysm expansion and type Ia endoleak, stent graft–induced dissection including retrograde type A dissection, and risk of rupture in segments treated with uncovered metal stents. Open operation has low rates of morbidity and mortality in the young population with Marfan syndrome, and it remains the treatment of choice for type B dissection and thoracoabdominal aneurysm in this population.

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