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Commentary: Thoracic endovascular aortic repair in Marfan syndrome—dancing with dogma

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Thoracic endovascular aortic repair (TEVAR) in patients with Marfan syndrome (MFS) is controversial, and current clinical practice guidelines recommend against the routine use of endovascular stent-grafts in this subgroup of patients due to unpredictable outcomes.^{1,2} Specifically, issues surrounding aortic fragility with subsequent development of retrograde type A dissection (RTAD) and overall high rates of reintervention have raised concerns about safety, efficacy, and durability. Notably, in 2012, our center published an initial TEVAR experience in 16 patients with MFS, who were deemed to be poor open surgical candidates, and we reported 6% (N = 1) 30-day mortality but a significant reintervention rate (50%), mandating close follow-up but suggesting safety and feasibility in selected patients.³

More contemporaneously, Pellenc and colleagues⁴ documented successful TEVAR deployment in 18 patients with MFS with zero mortality, stroke, spinal cord injury, and/or RTADs. The authors attributed their favorable outcomes to a treatment protocol that included several technical and anatomical features, including avoidance of bare stents; favoring landing into prosthetic graft material (especially in the proximal landing zone); minimizing oversizing in the native aorta and distal landing zone under-sizing in cases performed for dissection. Nevertheless, one nonelective open arch repair was required, and there was a 55% reintervention rate at

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TEVAR has been avoided in Marfan syndrome, but novel reports are challenging dogma.

mean follow-up of 21 ± 14 months (although 5 were planned).

In the current report, Jiang and colleagues⁵ demonstrate outcomes of TEVAR in 26 patients with MFS who predominantly presented with uncomplicated type B aortic dissection (TBAD). No perioperative deaths were reported, and acceptable survival (82.9%) at 10 years was documented. Of note, this experience represents a small (1.48%) subset of the 1573 patients with TBAD treated at their center during the study interval.

One major concern with TEVAR in acute TBAD is precipitation of RTAD, but that did not occur acutely in this series (although 1 patient experienced RTAD 5 years postoperatively). When possible, waiting 14 days for aortic stabilization before TEVAR of TBAD has been advocated and was largely performed in this series except in 3 cases of malperfusion, intractable pain, and hemodynamic instability. Interestingly, the authors were liberal with left subclavian artery (LSCA) coverage, and spinal drains were not routinely employed with stent lengths of 150 to 200 mm. Despite early postoperative endoleaks being documented in 4 patients (type Ib, N = 2; type II, N = 2 from the LSCA), all resolved spontaneously. Late endoleaks were identified in 3 patients, which were managed successfully without conversion to open repair (type Ia, N = 1 treated with extension; type II, N = 2 also from the LSCA treated expectantly; type III, N = 1 treated expectantly).

Due to various failure modes and elevated rates of long-term reintervention, all subjects undergoing TEVAR, and especially patients with MFS, require close follow-up.⁶ Moreover, 4 of the 26 patients in the current series had documented stent graft–induced new entry tears during surveillance, and corresponding freedom from reintervention

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was 83.4% and 50.3% at 5 and 10 years, respectively.⁵ Importantly, false lumen (FL) thrombosis was achieved around the stent-graft in 65% (N = 17), which was associated with better overall survival when compared with patients without FL thrombosis. As the authors suggest, patients with persistent FL perfusion may require even greater surveillance intensity.

So how does this series inform current clinical practice? This experience adds to a growing literature that supports selective use of TEVAR in patients with MFS. It is well-described that patients with MFS are at high risk for aneurysmal degeneration, but if TEVAR can be performed safely in acute TBAD presentations, as shown in this series and others—perhaps it is time to revisit dogma and take a fresh look at TEVAR in this complex group of patients.

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