Chimneys and sandwiches for endovascular arch repair in patients with Marfan syndrome: Are we snorkeling in cloudy waters?

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Elective repair of the proximal aorta for aneurysmal disease is standard of care for patients with Marfan syndrome (MFS) and has greatly improved life expectancy.\(^1,2\) Late distal aortic events, including dissection, rupture, and aneurysmal degeneration, are common and are the leading cause of death in this population. Multiple operations, with increasing complexity, morbidity, and mortality are often required in those surviving acute dissection.

Zhang and colleagues\(^3\) report a case of a 33-year-old patient with MFS who previously underwent a Bentall procedure and mechanical aortic valve replacement who presented with an acute dissection with the entry tear proximal to the left subclavian artery. The patient refused a conventional redo operation. Although medical therapy may have been an acceptable option in this circumstance, the authors offered endovascular repair, with a chimney for the innominate artery, a sandwich for the left common carotid artery, a left carotid-subclavian bypass, and 2 thoracic stent-grafts across the arch. The entry tear was covered successfully, although a type III endoleak was observed and persisted at 30 days.

Total endovascular reconstruction of the arch may involve custom-made fenestrated or branched devices, in-situ or back-table physician modifications to off-the-shelf devices, or parallel grafts (eg, chimneys, periscopes, and snorkels [CHIMPS]). Several modifications of the latter technique have been developed since Criado applied it to the arch,\(^4\) but the common principle remains the preservation of flow to vital aortic branches. The benefit of a chimney graft is that there is no wait for a costly custom-made device; thus, it is suitable for patients in urgent need. The primary concern has been long-term durability, given the potential for so-called gutter type I endoleaks, which are reported in 10% of patients at 30 days.\(^5,6\)

Endovascular techniques have revolutionized the care of patients with descending aortic disease. Zhang and colleagues\(^3\) pushed the envelope by treating an acute arch dissection in a patient with MFS. They report early technical success owing to proximal fixation in the surgical graft and nondilated arch vessels and aorta. However, high rates of endoleaks, stentgraft migration, and unfortunate cases of sudden aortic rupture have been reported in patients with MFS treated with thoracic endovascular aortic repair (TEVAR).\(^7,11\) Endovascular stent-grafts have been used as a life-saving bridge to definitive repair,\(^12\) or in cases where the stent-graft is anchored in surgical grafts proximally and distally.\(^13\) In this case, the presence of multiple arch grafts in dissected connective tissue raises serious concerns for the stability of the repair. As appropriately suggested by the authors, close lifelong surveillance is mandatory. Although late endoleaks may be managed by an endovascular approach, complex open procedures with partial or complete removal of the stent-grafts may be required in this young patient with MFS.

Zhang and colleagues\(^3\) report procedural success of a complex arch stent-graft in a patient with MFS and an acutely dissected arch. In light of the poor outcomes of standard TEVAR procedures in patients with MFS, such complex procedures should be limited to truly nonoperative candidates who meet acceptable anatomic criteria. Developments in
technology may further refine the currently limited role of TEVAR in the management of complex arch pathologies in patients with inherited aortic disease.

References