THE MARFAN FOUNDATION

Know the signs. Fight for victory.

THE MARFAN FOUNDATION
PROFESSIONAL ADVISORY BOARD STATEMENT

Thoracic Endovascular Aortic Repair (TEVAR) and Endovascular Aneurysm Repair (EVAR) are not the Optimal Treatments for Aortic Aneurysm and Dissection in Patients with Marfan Syndrome and Related Connective Tissue Conditions

The Marfan Foundation Professional Advisory Board does not recommend use of endovascular stent grafts for thoracic or abdominal aortic aneurysm and dissection in patients with Marfan syndrome and related connective tissue conditions because of high risk of complications and high rate of re-intervention. Conventional "open" surgical repair for aneurysms and dissections remain the gold standard and ideally should be performed at highly experienced surgical centers. This recommendation is based on data primarily from patients with Marfan syndrome, but also may apply to related connective tissue conditions, such as vascular Ehlers Danlos and Loeys-Dietz syndromes, as well as familial thoracic aortic aneurysm disorders.

In all clinical trials for TEVAR (Thoracic Endovascular Aortic Repair) and EVAR (Endovascular Aneurysm Repair) devices, patients with Marfan syndrome and other related connective tissue conditions have been excluded because of anticipated high failure rates due to the fragility of their aortic tissues. Consequently, these devices have not been approved for patients with Marfan and related connective tissue conditions and their use is considered off-label. There are numerous guidelines and expert consensus statements advising against endovascular repair for patients with Marfan syndrome and other related connective tissue conditions (1-3). Despite these guidelines, endovascular stent-grafting has steadily increased in this population. A recent review of 371 patients with suspected or confirmed syndromic or familial genetically triggered thoracic aortic conditions indicated that 8.5% underwent TEVAR procedures (4).

Recent long-term outcome studies indicate that the rate of re-intervention after TEVAR in the Marfan and related connective tissue conditions population is approximately 41.9% (4) compared to 15% in the general population (5). For Marfan and related disorder patients, early treatment failure rates were 25-43% for primarily type I endoleaks, 5-33% for early post-intervention repair, and 13-33% for secondary failure. Complications documented include endoleaks, loss of seal, device migration or erosion, induced new entry tears and retrograde ascending aortic dissection (6). These issues have led to subsequent need for reoperation, stent graft explantation, open repair of the aorta, and late death (4, 6). Additionally, there is limited, incomplete data regarding the impact of persistent radial forces on the abnormal aortic tissue at each end of stent-grafts in patients with Marfan syndrome. Post-intervention surveillance confirms that the aorta continues to expand despite successful stent-graft deployment, even when the false-lumen is thrombosed (1-2).

22 Manhasset Avenue, Port Washington, NY 11050
t 516 883 8712 f 516 883 8040 w Marfan.org

Evidence shows that patients with Marfan syndrome and related connective tissue conditions suffering from dissections have a high risk of re-interventions, approximately 52% in the distal aorta, ultimately leading to replacement of the entire thoracoabdominal aorta (7-9). Risk for re-intervention for patients with type B dissections can be as high as 86% (10). Late open repair of the descending aorta after stent-grafting in these patients can be very challenging. A surgical team experienced in open aneurysm repair in the setting of prior TEVAR or EVAR, is important to minimize catastrophic complications such as stroke, paraplegia, paraparesis, and death.

There are a few limited circumstances when TEVAR or EVAR should be considered in patients with Marfan syndrome and related connective tissue conditions. These include patients who have previous surgically placed grafts present both proximally and distally, which can anchor the endograft. Additionally, in some desperate, life-threatening emergencies, a stent-graft can serve as an emergent life-saving temporizing bridge to stabilize the patient prior to more definitive open surgical repair. Such situations include (1, 4):

- aortic rupture in a patient where the open intervention risk far exceeds that of the planned percutaneous procedure
- acute type B aortic dissection complicated by malperfusion
- rupture where extreme hemodynamic instability precludes safe transport to an experienced thoracic aortic surgical referral center for standard surgical intervention

The Marfan Foundation Professional Advisory Board suggests that patients with Marfan syndrome and related connective tissue conditions with complicated cases be referred to centers highly experienced in open surgical repair of complex aortic disease rather than pursue endovascular surgical solutions locally under the misconception that those procedures are safer and that endovascular specialists with experience in non-Marfan disease have translatable experience. Highly satisfactory outcomes with open repair of Marfan patients with distal aortic disease have been reported in the last decade from several experienced surgical centers (11-15). In the largest series of 127 patients with distal aortic dissection, 5-year survival was 87% and freedom from late repair failure was 86% at 8 years (4, 11). Because every patient is unique, the final recommendation should come from a genetic aorta team (vascular specialist in combination with a surgeon, cardiologist and geneticist) that has significant experience in Marfan syndrome and related connective tissue conditions.

References

1. Svensson LG, Kouchoukos NT, Miller DC, Bavaria JE, Coselli JS, Curi MA, Eggebrecht H, Elefteriades JA, Erbel, R Thomas, Gleason G, Lytle BW, Mitchell, RS, Nienaber CA, Roselli EE, MD, Safi HJ, MD, Shemin RJ, Sicard GA, Sundt TM, Szeto WY, and Wheatley GH. Expert Consensus Document on the Treatment of Descending Thoracic Aortic Disease Using Endo-Vascular Stent-Grafts. Ann Thorac Surg 2008; 85:S1-41.

2. Hiratzka LF, Bakris GL, Beckman JA, Bersin RM, Carr VF, Casey DE Jr, Eagle KA, Hermann LK, Isselbacher EM, Kazerooni EA, Kouchoukos NT, Lytle BW, Milewicz DM, Reich DL, Sen S, Shinn JA, Svensson LG, Williams DM; American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines; American Association for Thoracic Surgery; American College of Radiology; American Stroke Association; Society of Cardiovascular Anesthesiologists; Society for Cardiovascular Angiography and Interventions; Society of Interventional Radiology; Society of Thoracic Surgeons; Society for Vascular Medicine. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with Thoracic Aortic Disease: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. Circulation 2010; Apr 6;121(13):e266-369.

3. Erbel R, Aboyans V, Boileau C, et al. 2014 ESC Guidelines on the diagnosis and treatment of aortic diseases: Document covering acute and chronic aortic diseases of the thoracic and abdominal aorta of the adult. The Task Force for the Diagnosis and Treatment of Aortic Diseases of the European Society of Cardiology (ESC). Eur Heart J2014;35:2873-926.

4. Shalhub S, Eagle KA, Asch FM, LeMaire SA, Milewicz DM; GenTAC Investigators for the Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions (GenTAC) Consortium. Endovascular thoracic aortic repair in confirmed or suspected genetically triggered thoracic aortic dissection. J Vasc Surg. 2018 Aug;68(2):364-371. doi: 10.1016/j.jvs.2017.11.095. Epub 2018 Mar 19.

5. Hsu. Hung-Lung and Shih, Chun-Che. Re-intervention after thoracic endovascular aortic repair is high, but we should keep optimistic. J Thorac Dis. 2017;9(2):E162-E163.

6. Kouchoukos NT. Endovascular surgery in Marfan syndrome: CON. Ann Cardiothorac Surg. 2017 Nov;6(6):677-681. doi: 10.21037/acs.2017.10.05.

7. Engelfriet PM, Boersma E, Tijssen JG, et al. Beyond the root: dilatation of the distal aorta in Marfan's syndrome. Heart 2006;92:1238-43. 10.1136/hrt.2005.081638.

8. Finkbohner R, Johnston D, Crawford ES, et al.Marfan syndrome. Long-term survival and complications after aortic aneurysm repair. Circulation 1995;91:728-33. 10.1161/01.CIR.91.3.728.

9. Mimoum L, Detaint D, Hamroun D, et al. Dissection in Marfan syndrome; the importance of the descending aorta . Eur Heart J 2011;32:443-9. 10.1093/eurhertj/ehq434.

10. Schoenhoff FS, Jungi S, Czerny M, et al. Acute aortic dissection determines the fate of initially untreated aortic segments in Marfan syndrome. Circulation 2013;127:1569-75.

11. Coselli JS, Green SY, Price MD, et al. Results of Open Surgical Repair in Patients With Marfan Syndrome and Distal Aortic Dissection. Ann Thorac Surg 2016;101:2193-201.

12. Dardik A, Krosnick T, Perler BA, et al. Durability of thoracoabdominal aortic aneurysm repair in patients with connective tissue disorders. J Vasc Surg 2002;36:696-703.

13. Kalkat MS, Rahman I, Kotidis K, et al. Presentation and outcome of Marfan's syndrome patients with dissection and thoraco-abdominal aortic aneurysm. Eur J Cardiothorac Surg 2007;32:250-4.

14. Mommertz G, Sigala F, Langer S, et al. Thoracoabdominal aortic aneurysm repair in patients with marfan syndrome. Eur J Vasc Endovasc Surg 2008;35:181-6.

15. Omura A, Tanaka A, Miyahara S, et al. Early and late results of graft replacement for dissecting aneurysm of thoracoabdominal aorta in patients with Marfan syndrome. Ann Thorac Surg 2012;94:759-65.

December 2018