Stent grafts and endovascular repair are usually a poor option for treating aneurysm patients with TCD.

Prevalence connective tissue diseases
- Marfan 1/5000
- Ehlers-Danlos 1/10,000-15,000
- Loeys-Dietz unknown

Cardiovascular complications
- Aortic root dilatation
- Aortic valve insufficiency
- Aortic dissection-aneurysm, type A > type B
- TAAA – post dissection TAAA

Treatment of Aortic Disease in Patients With Marfan Syndrome
Dianna M. Milewicz, MD, PhD; Harry C. Dietz, MD; D. Craig Miller, MD
Endovascular Stent Grafting of the Aorta
In general, stent grafts should not be used in either the abdominal or the thoracic aorta in patients with MFS or other connective tissue disease. An exception is previous aortic replacement operations that have been complicated by a late localized false aneurysm. Stent grafting into old synthetic graft “necks” proximally and distally may be a safer alternative than a repeat thoracotomy in selected cases.

Circulation 2005
Endovascular management of chronic aortic dissection in patients with Marfan syndrome

Jan M. Norlin, MSc, Robert J. Hinsdale, MD, Peter J. Holt, MD, Rob Morgan, FRCSI, Marfan Johannesburg, MS, Ian M. Lofthouse, MD, and Matt M. Thompson, MD, London, UK

Conclusions. TEVAR in Marfan syndrome patients with chronic aortic dissection is technically feasible. However, poor intervention outcomes confirm that the aorta continues to dilate despite graft deployment and false lumen thrombosis. Endovascular repair may offer a viable alternative to surgical repair if complete endovascular exclusion of the aortic dissection is achieved. Further research and long-term follow-up of more patients is required to define the place of this therapy. (J Thorac Surg 2009;130:197-201)

Endovascular treatment of acute and chronic aortic pathology in patients with Marfan syndrome

Alyson Lee Wainerman, MD, MPH, Robert Joseph Foster, MD, W. Anthony Lee, MD, Philip J. Hess, MD, Thomas M. Barret, MD, MPH, Tomás D. Martín, MD, Thomas Smart Emhs, MD, PhD, and Adam Wynne Bash, MD, Guernsey and New Haven, RI

Results. Between 2009 and June 2015, 13 patients were identified as having undergone 17 TEVAR/TEVAR procedures. Three included thoracic aortic aneurysms (TAA) for acute dissection, deployment, and leak for chronic dissection, and repair of rupture. Endovascular treatment of TAA was associated with an overall 30-day mortality rate of 0%. No patients presented renal or neurological complications following TEVAR. In our experience, TAA for chronic dissection was associated with a 6-month mortality rate of 15%. Overall, endovascular treatment was associated with a 6-month mortality rate of 10% following TAA repair. (J Thorac Surg 2015;149:222-227)

Conclusion. Aortic disease associated with Marfan syndrome is a complex clinical problem and many patients require multiple procedures. Endovascular therapy can provide a valuable adjunct to bridge to open surgical resection in selected patients. The benefits of endovascular therapy, however, need to be weighed against the high-risk nature of the disease and the need for lifelong medical follow-up. (J Thorac Surg 2015;149:222-227)

Endovascular Treatment of Type B Dissection in Patients with Marfan Syndrome: Mid-Term Outcomes and Aortic Remodeling

Guering End-Lit, MD, PhD, Jorge Gaspar, MD, PhD, Alon Yavor, MD, Gabriela Meléndez-Ramírez, MD, Jorge Cervantes-Silva, MD, Hector González-Pacheco, MD, Félix Dámaso de los Santos, MD, Alvaro Munez-Gonzalez, MD, and Samuel Ramírez Marroquín, MD

In-hospital mortality was 10%. At a mean follow-up of 39.6 ± 30.9 months, the cumulative mortality was 20% and late mortality 11.1%. The rate of secondary endoleak was 44.4% and late reintervention of 33.3%. Survival freedom from cardiovascular death at 5 years was 80.9%.

Endovascular Treatment for Type B Dissection in Marfan Syndrome: Is It Worthwhile?

Davide Pacini, MD, *Alessandro Pardari, MD, PhD, Paolo Berretta, MD, Roberto Di Bartolomeo, MD, Francesco Alimanni, MD, and Joseph Bavaria, MD

Taken together, the data reported in this systematic review of the results of endovascular stent grafting for the treatment of type B dissection in MFS, especially in chronic presentations, are the proofs of concept that this type of approach needs to be considered with the greatest caution, and pros and cons need to be evaluated patient-by-patient. However, open surgical repair has been demonstrated as effective in the treatment of chronic dissection of the distal thoracic aorta with more stable results, even if associated with higher early mortality rate of 8.6%. In fact, the need of further aortic repairs is significantly lower after open surgical repair, with a freedom from aortic reoperation of 93% at 5 years and 83% at 10 years.[23]

Spectrum of Aortic Operations in 300 Patients With Confirmed or Suspected Marfan Syndrome

Scott A. LeMaire, MD, Stacey A. Carter, BA, Jennifer Volgaina, PhD, Anne T. Lau, BS, Diana M. Millozzi, MD, PhD, Garry W. Besette, MD, Catherine K. Chang, RN, BS, John Bizziowski, MD, Jennifer M. Marksino, BS, William K. Vaughn, PhD, and Joseph S. Coselli, MD

- 30 day mortality: 4.3%
- 10 year “freedom repair failure” 90%
- Renal failure: 6%
- Neurological complications: 4.3%
Thoracoabdominal Aortic Aneurysm Repair in Patients with Marfan Syndrome


Marfan Syndrome: when to operate TAA(A)s?

Open repair in chronic type B dissection with connective tissue disorders

Surgical correction of failed thoracic endovascular aortic repair

Stephen Langen, MD; Gerfried Hornemaker, MD; Thomas A. Keogel, MD; Gert W.H. Schurink, MD; Rodger Amstald, MD; and Michael J. Jacobs, MD.

Open surgery gold standard for aneurysms
Acute uncomplicated type B: conservative treatment
Acute complicated type B: TEVAR
Subacute/chronic type B with aneurysm: open surgery
Surgical protocol incl. ECC and neuromonitoring
Centralization is a must