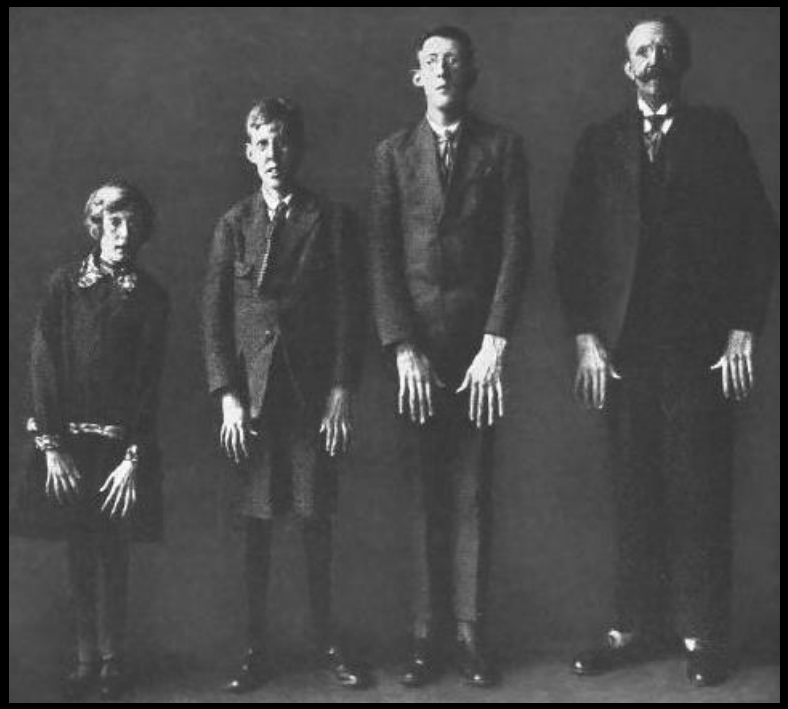
Is endografting an option in patients with connective tissue disorders?



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 diseases. 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

American Heart Association.

Endovascular management of chronic aortic dissection in patients with Marfan syndrome

Ian M. Nordon, MSc, Robert J. Hinchliffe, MD, Peter J. Holt, MD, Rob Morgan, FRCR, Marjan Jahangiri, MS, Ian M. Loftus, MD, and Matt M. Thompson, MD, *London, UK*

Conclusion: TEVR in Marfan syndrome patients with chronic aortic dissection is technically feasible. However, post intervention surveillance confirms that the aorta continues to dilate despite graft deployment and false lumen thrombosis. Endovascular repair may offer a viable option in patients who have contraindications to open surgery, but longer follow up of more patients is required to define the place of this therapy. (J Vasc Surg 2009;50:987-91.)

Journal of Vascular Surgery®

J ENDOVASC THER
2009;16:60–70

◆ ENDOVASCULAR THERAPY REVIEWS ◆

Treating the Thoracic Aorta in Marfan Syndrome: Surgery or TEVAR?

David G. Cooper, MS, FRCS; Stewart R. Walsh, MSc, MRCSEd; Umar Sadat, MBBS, MRCS; Paul D. Hayes, MD, FRCS; and Jonathan R. Boyle, MD, FRCS

Cambridge Vascular Unit, Cambridge University Hospitals NHS Trust, Addenbrooke's Hospital, Cambridge, UK.

Outcomes of endovascular repair are mixed, with questionable longevity. Reoperation is, however, common in MFS, and minimally invasive techniques may provide a bridging role or alternative solution when revisiting the hostile surgical field.

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

Alyson Lee Waterman, MD, MPH,[a] Robert Joseph Feezor, MD,[a] W. Anthony Lee, MD,[c] Philip J. Hess, MD,[b] Thomas M. Beaver, MD, MPH,[b] Tomas D. Martin, MD,[b] Thomas Stuart Huber, MD, PhD,[a] and Adam Wayne Beck, MD,[a] *Gainesville and Boca Raton, Fla*

Results: Between 2000 and June 2010, 16 patients were identified as having undergone 19 TEVAR/EVAR procedures. These included three emergent operations (two for acute dissection/malperfusion and one for anastomotic disruption early after open repair). All 16 patients had previously undergone at least one (range, 1-5) open operation of the ascending aorta or arch at a time interval from 33 years to 1 week prior to the index endovascular repair. During a median follow-up of 9.3 months (range, 0-46 months), there were four deaths (25%). Six patients (38%) had successful endovascular interventions. Despite early success, there was one death in this group at 1 month postintervention. Seven patients (44%) experienced primary treatment failure with five undergoing open conversion and one undergoing left subclavian coil embolization (the seventh was lost to follow-up and presented 4 months later in cardiac arrest and expired without repair). There were three deaths in the primary treatment failure group. Two patients experienced secondary treatment failure. One underwent the index TEVAR for acute dissection with malperfusion and required a subsequent TEVAR for more distal aortic pathology. He is stable without disease progression. The other patient underwent open conversion after a second EVAR with four-vessel "chimney" stent grafts and is stable with his entire native aorta having been replaced.

Conclusions: Aortic disease associated with Marfan syndrome is a complex clinical problem and many patients require remedial procedures. Endovascular therapy can provide a useful adjunct or bridge to open surgical treatment in selected patients. However, failure of endovascular therapy is common, and its use should be judicious with close follow-up to avoid delay if open surgical repair is required. (J Vasc Surg 2012;55:1234-41.)

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

Guering Eid-Lidt,[1*] MD, FSCAI, Jorge Gaspar,[1] MD, FSCAI, FACC, Gabriela Meléndez-Ramírez,[2] MD, MSC, Jorge Cervantes S.,[3] MD, Hector González-Pacheco,[4] MD, Félix Dámas de Los Santos,[4] MD, Aloha Meave-González,[2] MD, and Samuel Ramírez Marroquín,[3] MD

In-hospital mortality was 10%. At a mean follow-up of 59.6 ± 38.9 months, the cumulated mortality was of 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 8 years was 80.0%

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

Davide Pacini, MD,* Alessandro Parolari, MD, PhD,* Paolo Berretta, MD, Roberto Di Bartolomeo, MD, Francesco Alamanni, 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 est caution, and pros and cons [illegible]ated patient-by-patient. How[illegible]gical repair has been demon[illegible]ve in the treatment of chr[illegible]the distal thoracic aorta with more [illegible]s, even if associated with higher early mor-[illegible]ity rate of 9.6%. 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].

Published world expierence: 54 patients .

Ann Thorac Surg 2013;95:737–49

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

Scott A. LeMaire, MD, Stacey A. Carter, BA, Irina V. Volguina, PhD, Anne T. Laux, BS, Dianna M. Milewicz, MD, PhD, Garry W. Borsato, MD, Catherine K. Cheung, Hon BS, John Bozinovski, MD, Jennifer M. Markesino, 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%

THE ANNALS OF THORACIC SURGERY

Thoracoabdominal Aortic Aneurysm Repair in Patients with Marfan Syndrome☆

G. Mommertz,[1] F. Sigala,[1] S. Langer,[1] T.A. Koeppel,[1] W.H. Mess,[3]
G.W.H. Schurink[2] and M.J. Jacobs[1,2*]

European Vascular Center Aachen-Maastricht, [1]Department of Vascular Surgery, University Hospital Aachen and Maastricht, Germany, Departments of [2]Vascular Surgery, and [3]Neurophysiology, University Hospital Maastricht, The Netherlands

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

A. GREINER [1], J. GROMMES [1], S. LANGER [1], R. AUTSCHBACH [2], M. J. JACOBS [1]

Masters of Cardiothoracic Surgery

Open repair in chronic type B dissection with connective tissue disorders

Michael J. Jacobs, Geert Willem Schurink

Management of acute aortic dissection

Christoph A Nienaber, Rachel E Clough

In patients with connective tissue disease, remodelling is less successful and endovascular strategies are discouraged.[103–105]

Lancet 2015; 385: 800–11

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TEVAR

Endovascular Options in the Descending Thoracic Aorta in CTDs

A review of the data on thoracoabdominal aortic aneurysm repair in patients with connective tissue disorders.

BY PROF. MICHAEL JACOBS, MD, PHD; DR. BAREND MEES, MD, PHD; AND PROF. GEERT WILLEM SCHURINK, MD, PHD

Maastricht UMC+
Heart+Vascular Center

At present, clinical outcome is questionable, and there is not enough experience and evidence to consider endovascular repair as the first therapeutic option for descending aortic dissection and aneurysms in patients with CTDs. Open surgery remains the treatment of choice but only when performed in centers with high volume and adjunctive protective measures.

Endovascular repair of descending aortic dissection and aneurysms in CTD patients should also be considered in surgical revision cases, hostile chests, unfit patients, and severe anatomic deformities. ■