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

The most common connective tissue disorders (CTDs) associated with aortic diseases include Marfan syndrome (MFS), Ehlers-Danlos syndrome type IV, and Loeys-Dietz syndrome; all can cause aortic dissection and aneurysmal dilatation of the thoracic aorta. The estimated prevalence of MFS is approximately 1 in 5,000 and is even lower for Ehlers-Danlos syndrome at 1 in 10,000 to 15,000. The exact prevalence of Loeys-Dietz syndrome is not yet known. The most common cardiovascular complications in patients with MFS are aortic root dilatation, aortic valve insufficiency, and aortic dissection. The life expectancy of patients with MFS has improved significantly, from 45 years before the era of open heart surgery to approximately 70 years with current medical and surgical treatment.[1]

The thoracic aorta comprises the ascending aorta, aortic arch, and descending and thoracoabdominal aorta. This article focuses on the descending and thoracoabdominal aorta in patients with CTDs. The main pathologies considered for open or endovascular interventions include aortic aneurysm, dissection, intramural hematoma, and penetrating ulcers. In general, aneurysms are caused by degenerative processes or by previous aortic dissection. However, in patients with MFS, aneurysms form as a result of mutations in the *FBN1* gene encoding fibrillin in the extracellular matrix, which is a major component of extracellular microfibrils.

In patients with CTDs, the predominant indications for intervention are (complicated) acute aortic dissection, aortic aneurysm, and postdissection aortic aneurysm. Patients with MFS are significantly younger than patients with degenerative aneurysms,[2,3] and therefore their physical condition is much better as compared with the other patients.[4] Despite this, there is a higher incidence of dissection and rupture in MFS patients with descending thoracic aneurysms (DTAAs) and thoracoabdominal aortic aneurysms (TAAAs). The recommended threshold for DTAA and TAAA repair is an aneurysm diameter of 5 to 5.5 cm.[5]

OPEN SURGICAL REPAIR FOR DTAAS AND TAAAS

During the last few decades, open DTAA and TAAA repair has changed from the "clamp-and-go" technique to a controlled procedure with extracorporeal support, selective organ perfusion, and neuromonitoring of the spinal cord. The largest experience of TAAA repair in MFS patients (137 confirmed, 163 suspected) has been described by LeMaire et al.[6] The 30-day mortality rate was 4.3%, and freedom of repair failure was significantly better in patients with confirmed MFS compared with patients with suspected MFS (90% vs 82% at 10 years; $P = .001$). Of these 300 patients, 31 had DTAAs and 178 had TAAAs. Surgical mortality was < 6% and major complications included renal failure in 6% and neurologic deficit in 4%. The authors concluded that operative treatment of aortic pathology in MFS patients provides excellent results and long-term survival.

In our aortic center, we have comparable results in patients with MFS undergoing open TAAA repair. In a subseries of 22 patients with MFS and TAAAs, all patients survived, and major complications such as paraplegia, renal failure, stroke, and myocardial infarction were not encountered. At 38-month follow-up, all patients were alive and had returned to work.[7] Kalkat et al[3] found a similar outcome, which further justifies that surgical repair of DTAAs and TAAAs provides excellent results in patients with MFS. Obviously, these complex operations should be performed in high-volume centers with adequate infrastructure and multidisciplinary expertise.[7]

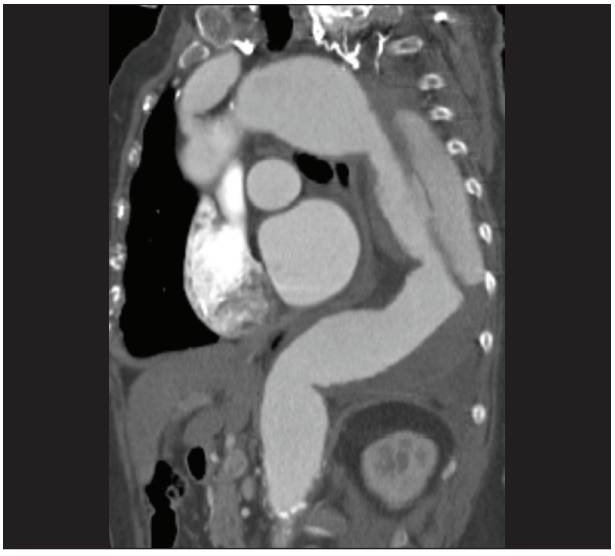

Figure 1. Aneurysm affecting the aortic arch and entire thoracoabdominal aorta (with dissection in the descending aorta), not offering adequate landing zones for stent grafts.

Figure 2. CT scan of a 28-year-old man with MFS. He had a post–type A (DeBakey type I) aortic aneurysm of the ascending aorta, arch, and thoracoabdominal aorta. After ascending aorta and arch replacement, he developed malperfusion caused by extreme narrowing of the true lumen.

SHOULD ENDOVASCULAR TECHNIQUES BE USED IN PATIENTS WITH CTDs?

It has been previously stated that, "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."[1] Based on the vulnerable tissue and progressive expansion of the aortic tissue in these patients, we would still agree with this statement; however, several small series report that endovascular repair is feasible and safe.

There are no randomized trials comparing endovascular and open surgical techniques for TAAAs; however, meta-analyses of data from nonrandomized trials and registries clearly demonstrate reduced early mortality, paraplegia, renal failure, cardiac complications, pneumonia, and length of stay after endovascular repair for DTAAs as compared to open surgery.[8] With regard to paraplegia, it should be emphasized that endovascular treatment for TAAAs is associated with a significantly higher incidence of neurologic complications.[9] However, these outcomes are reported for DTAAs and TAAAs in general, not specifically for patients with CTDs.

Experience With Aortic Endovascular Techniques in CTDs

After the first case reports approximately 12 years ago, the first small series showed the technical feasibility of endografting in subacute and chronic expanding aortic dissection.[10,11] Several of these patients already underwent ascending or TAAA repair. The authors proposed that this endovascular strategy might be used to bridge or even avoid repeat aortic surgery in patients with CTDs.

Endovascular strategies in nondissected DTAAs and TAAAs in patients with MFD are scarcely reported. The published results of endovascular aortic treatment in patients with MFS mainly address post–type B dissection pathologies.[12-14] In these series, patients were relatively young and almost all had already undergone surgery of the aortic root or arch. Nordan et al[12] described seven patients with aneurysmal degeneration of the thoracic aorta secondary to chronic dissection. They successfully implanted the endograft, but postintervention surveillance confirmed continuous dilatation of the aorta despite graft deployment and false lumen thrombosis. Botta et al[13] treated 12 patients for descending aortic dissection. They experienced that endovascular treatment is technically feasible in patients with MFS and can be considered a valuable alternative to open reoperation. In the experience of Eid-Lidt et al,[14] secondary endoleak was encountered in 44% of patients with a late reintervention rate of 33%.

In a study of 15 patients with MFS treated with thoracic endovascular repair, Waterman et al[15] reported 44% primary treatment failure and a mortality rate of 43%, mainly induced by the necessary conversion to complex open surgical repair. They concluded that open repair remains the gold standard therapy but that endovascular treatment can be offered to patients unfit for open surgery or, in selected cases, as a bridge to open repair.

Pacini et al[16] performed a systematic review of studies addressing the results of endovascular treatment in MFS patients with type B aortic dissection. The primary endpoints included perioperative and late death, major

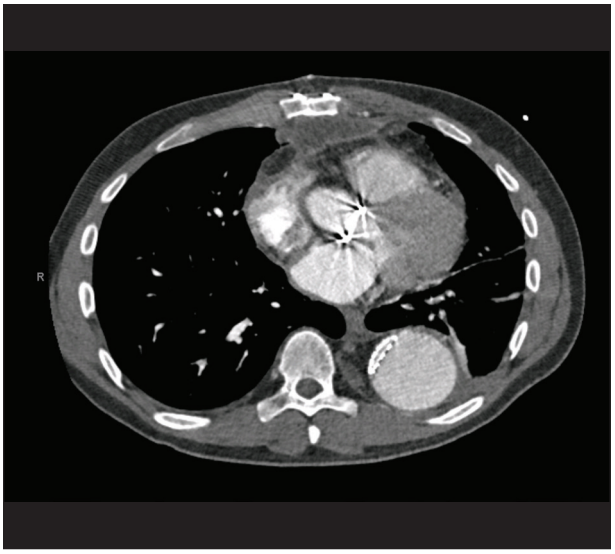

Figure 3. The same patient with MFS as shown in Figure 2 after an attempt to open the true lumen with a long bare-metal stent. This image shows a collapsed stent and inability to open the true lumen.

Figure 4. Scan showing postdissection aneurysm affecting the ascending aorta, arch, and thoracoabdominal aorta technically not amenable for endovascular repair.

complications, endoleak, surgical conversions, and need of additional endovascular procedures. Twelve studies were identified involving 54 patients: 11 (20.4%) underwent endovascular treatment for acute dissection and 43 (79.6%) for chronic dissection. Periprocedural mortality was only 1.9%; however, the incidence of endoleaks was very high (overall 22%). The latter was of great importance given that the majority of patients had already undergone aortic surgical procedures, offering adequate landing areas in aortic grafts, either proximal or distal. In patients in whom the stent graft landed in native aortic tissue, the rate of endoleak approached 30%. During follow-up, high endoleak rates persisted, requiring many reinterventions. At follow-up, mortality increased to 12%, which is obviously high in a relatively young population. The authors concluded that the complications after endovascular repair are too high to consider this a safe approach for acute and chronic dissections in MFS patients and cautioned against the routine use of the approach in this patient population.

Considerations

After reviewing the literature, it becomes clear that there is no evidence at present to change the approach from open to endovascular treatment for descending aortic dissection and aneurysms in patients with CTDs. This statement specifically applies for patients with CTDs who did not undergo complex aortic operations previously and who are fit for surgery. The current data on endovascular treatment of aortic pathologies in CTD patients are very limited—there are fewer than 100 patients reported in the literature. The reported experience is confined to small series in heterogeneous groups of patients and do not include patients with TAAAs.

In almost all published studies, patients had MFS; studies evaluating other CTDs with associated aortic disease are sparse. In general, the patients with MFS were young and had already undergone aortic root, ascending, and/or arch repair. When considering endovascular strategies, adequate proximal and distal landing zones are required. In case of preexisting aortic grafts, deployment and fixation will be safer than in native aortic tissue affected by CTDs. However, in both circumstances, early and late complications frequently occur and remain of great concern.[16]

TECHNICAL ISSUES INVOLVING THE DESCENDING THORACIC AORTA

The two main indications for treatment of the descending thoracic aorta are dissection and aneurysm. The aneurysm can be secondary to a type A or type B dissection or does not involve dissection at all. As discussed previously, the literature mainly describes postdissection pathologies despite the fact that a substantial number of patients with CTDs had nondissected aneurysms. In the latter patients, aneurysms most often involve the entire thoracoabdominal aorta, frequently affecting the ascending aorta and arch as well (Figure 1), not allowing for adequate endovascular repair because of absent landing zones. Collapsed or narrow true lumen can also be a burden for endovascular stent grafting, especially in chronic dissection, in which the rigid septum can hamper full apposition of the proximal stent. Figure 2 shows a postdissection TAAA in a 28-year-old patient with MFS. As a first procedure, the aneurysmatic

ascending aorta and arch were replaced via sternotomy. Postoperatively, the patient developed malperfusion of the liver, and CT showed an extremely narrow true lumen (Figure 2). An attempt to open this true lumen with a long bare-metal stent did not succeed (Figure 3). The stent did not open the small true lumen, and open repair of the type II TAAA was performed.

Other challenges include postdissection descending thoracic aneurysms secondary to extensive type A (DeBakey type I) dissection (Figure 4). Normally, the ascending aorta would be replaced, leaving a dissected arch in place. This common situation prohibits adequate proximal fixation, as endograft deployment in a dissected aortic segment is hazardous. Future developments should emphasize more extensive ascending and arch repair, including frozen elephant trunk repair, serving as a safe proximal docking area for secondary open and endovascular procedures of the descending and thoracoabdominal aorta.

TECHNICAL ISSUES INVOLVING THE THORACOABDOMINAL AORTA

Fantastic progress has been achieved in the endovascular treatment of TAAAs, both in degenerative and postdissection aneurysms.[17] However, endovascular treatment of TAAAs in patients with CTDs has only been performed in a few individual cases, mostly because of previous surgery, "frozen" chest or abdomen, or candidates unfit for open repair.

Several technical issues remain challenging in open repair. Cross-clamping a fragile aorta in patients with CTDs can be catastrophic, especially if more proximal clamping is not feasible. Retrograde dissection, like in endografting, can easily occur after cross-clamping distal to the left subclavian artery, even when performed with utmost care and caution. Rapid conversion is required in these cases, illustrating the need for adequate infrastructure and multidisciplinary teams.

There are, however, several indications for an endovascular approach or hybrid strategies. As mentioned previously, patients with CTDs can already have had several thoracotomies, making open surgical access rather menacing. Also, anatomic deformities such as severe pectus excavatum, extreme scoliosis, and rib disfigurements can hinder appropriate surgical access to the distal aortic arch and descending aorta. In these circumstances, proximal implantation of an endograft and open repair of the abdominal aorta with selective grafting of the visceral and renal arteries might constitute an acceptable hybrid alternative.

SUMMARY

There are fewer than 100 endovascular cases of descending or TAAA repair in CTD patients reported in the literature. 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 options can be included in the open surgical management of dissected or aneurysmal ascending aorta and aortic arch by implanting a (frozen) elephant trunk, allowing secondary open or endovascular treatment of descending and abdominal aortic pathology. 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. ■

Prof. Michael Jacobs, MD, PhD, is Professor and Chief of Surgery, Department of Surgery, Maastricht University Medical Center in Maastricht, The Netherlands. He has stated that he has no financial interests related to this article. Prof. Jacobs may be reached at m.jacobs@mumc.nl.

Dr. Barend Mees, MD, PhD, is from Maastricht University Medical Center in Maastricht, The Netherlands. He has stated that he has no financial interests related to this article.

Prof. Geert Willem Schurink, MD, PhD, is Professor of Surgery, Maastricht University Medical Center in Maastricht, The Netherlands. He has stated that he has no financial interests related to this article.

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