Stent graft repair of descending aortic dissection in patients with Marfan syndrome: An effective alternative to open reoperation?

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Objective: Aneurysms or dissections can involve multiple aortic segments in patients with Marfan syndrome, requiring staged replacement of the entire aorta. The optimal treatment of descending aortic dissection in these patients is a major challenge. We investigated the feasibility and outcomes of endovascular repair of the descending aorta in patients already submitted to open aortic root/arch surgery.

Methods: From March 1998 to July 2008, 12 patients (10 male and 2 female patients; mean age, 37.8 ± 11.6 years) affected by Marfan syndrome underwent endovascular treatment for dissection of the descending aorta after previous open aortic root/arch surgery. Stent graft procedures were performed urgently in 5 patients and electively in 7 patients.

Results: Neither in-hospital deaths nor perioperative paraplegia or stroke occurred. Follow-up (median, 31 months; range, 3–57 months) was 100% complete. One patient needed surgical conversion for persistent type I endoleak, leading to false lumen expansion 3 months after endovascular repair. Extension of the dissection occurred in 2 patients 1 month and 2 years after the procedure, respectively. No late death or aortic rupture was observed.

Conclusions: Endovascular repair of the dissected descending thoracic aorta can be performed in patients with Marfan syndrome with a low risk of death or major complications. In case of staged procedures, stent graft treatment can be considered a possible alternative to open reoperation. Long-term durability remains to be determined.

Marfan syndrome is a relatively common heritable disorder of connective tissue that affects numerous organ systems, but the most severe complication is aortic aneurysm and dissection.^{1–3} Aneurysms or dissections can frequently involve multiple aortic segments, requiring staged replacement of the entire aorta.^{3–5} Although progress of surgical techniques for the ascending aorta improved life expectancy in patients with Marfan syndrome,^{5–7} results of conventional surgical intervention or medical treatment for descending aortic dissection are still disappointing.^{8–11} The feasibility of endovascular aortic repair provided new alternatives, but the potential risks are not fully determined. The aim of this study was to analyze procedural feasibility, early and midterm results, and clinical outcome after endovascular aortic repair of the descending aorta in 12 consecutive patients with Marfan syndrome already submitted to open aortic root/arch surgery.

MATERIALS AND METHODS

Patients

From March 1998 to July 2008, 115 patients underwent endovascular repair for dissection of the descending aorta. Of these, 12 (10.4%) patients were affected by Marfan syndrome according to the Ghent nosology. There were 10 male and 2 female patients (mean age, 37.8 ± 11.6 years). The mean American Society of Anesthesiologists class was 3.2 ± 0.7 . Patient characteristics are shown in Table 1.

Thoracic Aortic Involvement and Previous Aortic Surgery

All patients have been previously submitted to open aortic surgery for annuloaortic ectasia (4 Bentall procedures and 1 “David 1” operations) and acute (2 Bentall procedures and 3 supracoronary replacements) and chronic (1 Bentall procedure and 1 “David 1” operation) type A aortic dissection. Hemiarth or total arch replacement was performed in 5 and 4 patients, respectively. The elephant trunk technique was used in 3 patients. A mitrotricuspid valve plasty was associated with a Bentall procedure in 1 patient. In the same patient an orthotopic heart transplantation was performed for end-stage cardiomyopathy 10 years later. Three patients who underwent the Bentall procedure plus hemiarth replacement (1 patient) and supracoronary replacement plus hemiarth replacement (2 patients) for acute type A dissection underwent reoperations for aneurysmal degeneration of the aortic arch.

Presenting Signs, Symptoms, and Interventional Indications

A classical type B aortic dissection with an abrupt onset of pain was diagnosed in 5 (41.7%) patients, all previously submitted to aortic root surgery for aneurysm a median of 168 months (range, 24–264 months) previously. Chest pain was reported by all patients, whereas abdominal and leg pain was experienced by 2 and 1 patients, respectively. Preoperative acute malperfusion was detected in 3 patients (mesenteric ischemia,

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Abbreviations and Acronyms

- CT = computed tomography
- MRI = magnetic resonance imaging
- TEE = transesophageal echocardiography

acute renal failure, and limb ischemia in 1 patient each). Preoperative hypertension appeared at presentation in 4 of 5 patients. No patient was admitted with shock or tamponade, but left hemothorax was found in 4 patients. Endovascular repair was indicated in the acute phase for an impending aortic rupture in 2 (16.7%) patients and for visceral or limb ischemia in 2 and 1 patients, respectively. In chronic cases (7 patients) indications were the increasing diameter of the descending aorta during follow-up (all) and recurrent pain (4 patients).

Diagnostic Evaluation

Routine examination of heart, lung, liver and kidney functions, as well as contrast-enhanced computed tomographic (CT) analysis, magnetic resonance imaging (MRI), and/or angiographic analysis, were conducted both in emergency and elective cases. Hemodynamically compromised patients underwent only CT scanning just before emergency/urgent stent graft repair. Anatomic assessment of the descending aorta was made by using 1 or more imaging methods, including CT scanning in 83.3%, aortographic analysis in 75%, and MRI in 33.3%. Preoperative chest radiographic analysis was performed in all patients and showed a widened mediastinum in 100%. Tomographic imaging revealed pleural effusion in 33.3% of patients and a periaortic hematoma in 8.3% of patients, with signs of free rupture in 1 patient. Aortic measurements were performed in multiple planes perpendicular to the major vessel axis. At least 5 to 10 measurements were performed at the neck sites in patients undergoing elective operations. The mean aortic diameter of the pseudoaneurysm was 51.1 ± 3.8 mm.

Stent Graft Procedure

Patients underwent mechanical ventilation after achievement of general anesthesia. Blood pressure was monitored by means of right radial artery cannulation. Ceftriaxone (2 g administered intravenously) was administered before the procedure. The common femoral artery was used for access after surgical exposure in all patients. After exposition of the artery, a 6F sheath was inserted, and 2500 IU of heparin was administered. In patients with active bleeding into the pleural space, mediastinal space, or both, no systemic heparin was administered. Angiographic analysis and transesophageal echocardiographic (TEE) analysis were performed to identify the lesion and landing zones and the lesion's relation to side branches in all patients. Twenty-nine thoracic stent grafts (21 Talent and 8 Valiant; Medtronic, Santa Rosa, Calif) were loaded on an extrastiff guidewire and delivered under fluoroscopic and TEE control with induced hypotension (systolic pressure, <70 mm Hg) to prevent inadvertent downstream displacement of the stent graft during delivery. The proximal end of endografts was always an uncovered stent (free-flow end). On the basis of CT/MRI/TEE measurement, a maximum oversizing of 10% was applied in the choice of stent graft diameter. A shaping balloon was used when necessary under TEE control. An elephant trunk was used as the landing zone of the endografts in 3 patients. Postprocedural angiographic and TEE controls were performed to reveal the final result. There were no local complications related to the presence of the sheath, and no thrombectomies were performed in any case. No aortic fenestration was necessary in this series. Intraoperative details are reported in Table 2.

Statistical Analysis

All statistical analyses were performed with the SPSS software package (version 14.0; SPSS, Chicago, Ill). Incidence rates of events are reported as

TABLE 1. Patients' preoperative features

Age (y) ± SD	37.8 ± 11.6
Sex (male/female)	10/2 (83.3/16.7)
NYHA classification	
I–II	10 (83.3)
III–IV	2 (16.7)
ASA class ± SD	3.2 ± 0.7
Presentation of type B aortic dissection	
Acute	5 (41.7)
Chronic	7 (58.3)
Hemothorax	4 (33.3)
Hypertension	4 (33.3)
Smokers	4 (33.3)
COPD	1 (8.3)
Diabetes	1 (8.3)
Renal failure	3 (25)
Previous cardiac/aortic surgery	12 (100)
Aortic measures	
Aortic diameter (mm) ± SD	51.1 ± 3.8

Values are presented as the mean ± standard deviation or the number of patients experiencing the event followed by the corresponding percentage in parentheses, unless otherwise noted. SD, Standard deviation; NYHA, New York Heart Association functional class; ASA, American Society of Anesthesiologists; COPD, chronic obstructive pulmonary disease.

the number of patients experiencing the event followed by the corresponding percentage. Continuous data are reported as the mean ± standard deviation, median and range, or both of values observed. The Kaplan–Meier actuarial method was used to generate survival estimates and freedom from endoleaks and from reoperation reported with 95% confidence limits.

RESULTS

Technical success (insertion and deployment of the stent graft in the target aortic segment) was achieved in all patients. More than 1 stent was necessary in 11 (91.7%) of 12 patients. One stent graft was implanted in 1 patient, 2 stent grafts each were implanted in 5 patients, and 3 stent grafts each were implanted in 6 patients, according to the number and site of re-entries. The need for 2 or 3 stents is related to many factors, both anatomic and physiopathologic: the extent of the disease, the presence of multiple large re-entry sites (the clinical relevance of which is evident only after deployment of first grafts), the length of the thoracic aorta (related to body surface area), and the necessity to land in a safe proximal zone (usually more proximal than in standard procedures) or inside a previous vascular graft. Neither intraoperative mortality nor surgical conversion occurred. Postoperatively, 5 (41.7%) patients were transferred to the intensive care unit, and 7 were transferred directly to the ward after extubation in the operating room. The median intubation time was 2.5 hours (range, 1–13 hours). Median intensive care unit and in-hospital stays were 20 hours (range, 6–21 hours) and 9 days (range, 2–19 days), respectively. Only 1 patient received blood cell transfusion. In all patients blood pressure was tightly controlled (systolic blood pressure, <120 mm Hg). Fever, increased C-reactive

TABLE 2. Intraoperative details

Patient no.	PAO	Time between PAO and EVAR (mo)	Technical success	Surgical access	Left subclavian artery closure	Stent grafts per patient	Endograft diameters	Stent graft length	Fluoroscopic time	Contrast media	Surgical conversion	Transfusions	Mortality
1	Reimplantation	24	Y	RCFA	N	2	32/26 30/30	105 116	25	110	Y	N	N
2	Bentall	264	Y	RCFA	N	1	24/30	151	12	120	N	N	N
3	Ascending aortic replacement	41	Y	RCFA	N	3	30/30 34/30 32/32	115 113 115	32	120	N	N	N
4	Bentall procedure	108	Y	LCFA	N	3	32/32 34/30 36/32	115 113 112	18	100	N	Y	N
5	Reimplantation + arch + ET	2	Y	RCFA	N	3	32/32 36/36 38/38	100 150 114	25	100	N	N	N
6	Bentall procedure	168	Y	RCFA	N	2	36/32 34/30	112 113	12	90	N	N	N
7	Ascending aortic replacement	20	Y	RCFA	N	2	36/32 34/30	112 113	10	70	N	N	N
8	Bentall procedure + MVP + TVP	168	Y	RCFA	Y	3	32/32 36/32 34/34	200 150 48	25	210	N	N	N
9	Ascending aortic replacement + arch	58	Y	RCFA	Y*	2	30/30 34/30	200 150	14	100	N	N	N
10	Bentall procedure + arch + ET	66	Y	RCFA	N	3	28/28 32/28 32/28	150 150 150	100	90	N	N	N
11	Bentall procedure + Arch + ET + MVR	2	Y	RCFA	N	2	32/32 36/32	200 150	40	110	N	N	N
12	Bentall procedure	408	Y	RCFA	N	3	30/30 34/34 34/34	200 100 200	19	180	N	N	N

Values are presented as absolute numbers. PAO, Previous aortic operation; EVAR, endovascular aortic repair; Y, yes; RCFA, right common femoral artery; N, no; LCFA, left common femoral artery; ET, elephant trunk; MVP, mitral valve plasty; TVP, tricuspid valve plasty; Y*, yes with previous left subclavian artery–left common carotid artery bypass; MVR, mitral valve replacement.

protein levels, and leucocytosis were observed in all patients starting from the first postoperative day in the absence of any evident infectious agent. The systemic inflammatory response was mild and self-limiting in the majority of patients, lasting from 2 to 10 days and responding to nonsteroidal anti-inflammatory drug administration, but it was prolonged in 3 patients (from 15 days to >3 months) without leucocytosis, positive blood culture results, or both.

One patient required a femorofemoral bypass 24 days after endovascular repair for acute limb ischemia caused by distal extension of the dissection. Early type II endoleaks caused by

a backflow from an intercostal artery (1 patient) and from the left subclavian artery after intentional closure without previous revascularization (1 patient) were detected in 2 patients. Both endoleaks resolved spontaneously within 6 to 12 months. Type I endoleak, leading to aneurysmal expansion, was detected in 1 patient who was converted to surgical intervention 3 months later. A left thoracotomy was performed to remove the stent graft, and an interposition vascular graft was used in the thoracic aorta without serious complications.

Neither paraplegia nor other perioperative complications occurred. In-hospital results are summarized in Table 3.

TABLE 3. Early and midterm results

In-hospital results	
Intubation time (h)*	2.5 (1–13)
ICU stay (h)*	20 (6–21)
In-hospital stay (d)*	9 (2–19)
Paraplegia	0
Stroke	0
Acute renal failure	0
Respiratory insufficiency	0
Early endoleak	
Type I	1 (8.3)
Type II (R)	2 (16.7)
Late endoleak	
Type I	1 (8.3)
Follow-up	
Surgical conversion	1 (8.3)
Second endovascular procedure	1 (8.3)
New dissection during follow-up	2 (16.7)
Overall mortality	0

Values are presented as the number of patients experiencing the event followed by the corresponding percentage in parentheses, unless otherwise noted. *ICU*, Intensive care unit; *(R)*, spontaneous resolution during follow-up. *Median (range).

All patients were subjected to a strict follow-up protocol, which was 100% complete. The median follow-up time was 31 months (range, 3–57 months). Clinical examination and CT scan or MRI controls were performed at discharge; at 1, 3/6, and 12 months after treatment; and every 12 months thereafter (Figure 1). Retrograde blood flow distal to the implanted endograft and preventing the complete thrombosis of the false lumen in the distal descending aorta was observed in all patients at 1 to 3 months after the procedure. However, a reduction of mean diameters of the involved aortic segments and a progressive thrombosis of the false lumen involving the entire thoracic aorta was usually observed after almost 6 months. Persistent false lumen expansion of the distal descending thoracic aorta occurred in 1 patient and was treated with a second endovascular procedure (stent graft extension) 38 months after the first procedure. The second procedure was successful, and progressive thrombosis of the false lumen in the thoracic aorta was obtained. At last follow-up, the patient is in good general condition without complications. A new dissection of the native arch, interposed between a vascular graft proximally and the endograft distally, was detected in 1 patient 2 years after the procedure. The aortic diameter was less than 3 cm, and the patient is currently in follow-up. No late deaths occurred during follow-up. Actuarial freedom from endoleak and from any type of reoperation is shown in Figures 2 and 3.

DISCUSSION

Marfan syndrome is an autosomal dominant disorder of connective tissue characterized by highly variable clinical manifestations, primarily in the skeletal, ocular, and cardiovascular organ systems. Mutation in the fibrillin gene is

responsible for the alteration of extracellular matrix in the connective tissue, leading to aortic dilation and dissection.^{1,12,13}

Long-term surveillance of the aorta before and after surgical intervention and prophylactic resection of the dilated segment has become standard practice in patients with Marfan syndrome, prolonging survival similarly to that seen in the healthy population.^{2,3} Excellent results have been obtained in elective surgical intervention of the ascending aorta with composite grafts and valve-sparing operations,^{6,7,14–17} whereas relatively low morbidity and mortality rates are actually associated with urgent or emergency procedures.¹⁴ Given the benefits of prophylactic aortic surgery, there are a growing number of patients who might require secondary aortic operations as a result of new or residual dissections, increasing diameter, or new aneurysms.

All patients of our series underwent previous surgical intervention of the ascending aorta, aortic arch, or both. Retrospective analysis of our data showed that patients undergoing operations for aortic root aneurysm associated or not with aortic valve disease had a new-onset type B dissection requiring interventional treatment after a median of 14 years from the first operation. On the other hand, patients treated for type A aortic dissections underwent endovascular repair earlier (2.5 years after the first operation). A close follow-up of all patients with Marfan syndrome is necessary to detect asymptomatic changes requiring repair.^{4,18} After aortic root replacement, aortic operations in the descending aorta are required in 50% of patients over a mean period of 26 years.¹⁹ The risk of aortic dissection increases with increasing aortic diameter, but it can also occur in nondilated aortas. Carrel and colleagues⁴ reported a high incidence of reoperation in patients with acute type A aortic dissection, with unexpected fatal outcome even if the aorta is normal sized. In contrast to the aortic root, at present, no clear guidelines elucidate definite criteria for descending aortic operations either for prophylactic or urgent surgical repair.^{3,5,19} As a matter of fact, the optimal treatment for patients with acute or residual dissections of the descending aorta continues to be debated. The prognosis for patients with type B aortic dissection is bleak and determined primarily based on dissection-related and patient-specific risk factors.^{10,20–24} International Registry of Acute Aortic Dissection investigators⁹ reported an overall in-hospital mortality of 33.9% after conventional surgical intervention for acute type B dissection. The actuarial survival estimates for all patients were 71%, 60%, 35%, and 17% at 1, 5, 10, and 15 years, respectively, and were similar for the medical and surgical patients.²⁵ Reoperation and late aortic complications were predicted by the presence of patients with Marfan syndrome. Mortality is related either to retrograde progression of dissection with involvement of the proximal aorta with even higher mortality or to expansion of the false lumen and formation of a thoracic aneurysm with an inherent risk of rupture.

FIGURE 1. Volume-rendering (A) and maximum-intensity projection (MIP) images (B) of the acute aortic type B dissection with the entry tear at the level of the left subclavian artery ostium in a patient with Marfan syndrome initially operated on for an annuloaortic ectasia and proximal arch aneurysm. In panel A the white arrow shows the vascular graft used for ascending aortic and arch replacement. Dissection of the descending aorta in axial projection is shown in panel C. Volume-rendering (D) and MIP images (E) show a correct position of the stent graft and the absence of endoleak 3 months after the procedure; the white asterisk indicates patency of the false lumen (FL) distally to the implanted graft. Panel F shows the complete thrombosis of the FL in the descending thoracic aorta in axial projection after 3 months.

FIGURE 2. Freedom from any type of endoleak.

The feasibility of endovascular repair in patients with Marfan syndrome is widely debated because of the potential risk induced by radial forces of the stent graft on the fragile aortic wall. Stent grafting into synthetic graft “necks” proximally and distally might be a safer alternative than a repeat thoracotomy in selected cases. In their update on treatment of aortic disease in patients with Marfan syndrome, Milewicz and associates⁵ suggest that stent grafts should not be used in either the abdominal or the thoracic aorta in patients with this syndrome or other connective tissue diseases. Endovascular stent grafts have been successfully used to treat persistent distal aortic dissection after previous root replacement, as reported by Ince and coworkers.²⁶ In their series of 6 patients, elective conversion to surgical repair was necessary in 2 patients at 22 and 43 months after stent graft implantation, respectively, and was being considered at 74 months in a third patient. Our experience showed acceptable results with low morbidity rates and a low incidence of vascular complications and endoleaks. However, progression of dissection or a new dissection, which often characterizes the course of Marfan disease, might be accelerated by the effect of the stent graft. In our series 2 patients presented with a new dissection during follow-up: in 1 patient a new dissection of an interposed aortic arch between a Dacron graft in the ascending aorta and the stent graft in the descending aorta occurred several months after the procedure; the clinical course was uneventful, and the patient is still in follow-up without significant arch enlargement. The second case, a distal dissection of the descending aorta, occurred 24 days after the procedure. According to retrospective case review, the dissection, which was adjacent to the distal part of the stent graft, was probably related to an excessive oversizing with respect to the aortic diameter.

FIGURE 3. Freedom from any type of reoperation (surgical and endovascular).

Despite a minimal inflammatory cell infiltration described at the level of the aortic wall in patients with Marfan syndrome,²⁷ all patients of our series experienced a self-limiting postimplantation syndrome in the postoperative period characterized by fever and leucocytosis. The trigger of this systemic reaction is unclear, but it is possible that endograft materials and the remodeling of endovascular thrombus could play an important role in the stimulation of the immune response. Another peculiar finding in our experience has been the presence of a large retrograde blood flow in the false lumen distal to the implanted endograft, initially preventing the immediate thrombosis of the false lumen, probably because of larger re-entry sites than in patients without Marfan syndrome. However, complete and enduring thrombosis of the false lumen in the thoracic aorta was observed almost 6 months after the procedure in all patients without endoleak.²⁸

CONCLUSION

Endovascular repair of the dissected descending thoracic aorta can be performed in patients with Marfan syndrome, with low risk of death and major complications. In case of staged procedures, stent graft treatment can be considered a possible alternative to open reoperation. Because of the particular fragility of the aortic wall in these patients, careful case selection avoiding hostile anatomy (an excessive tortuosity of the aorta, an angulated aortic arch or extension of the lesion to the aortic arch, the presence of a graft in the abdominal aorta, the presence of large entry or re-entry sites close to the celiac trunk and a short proximal neck [<5 mm from left subclavian artery], landing zones with a diameter of ≥ 42 or ≤ 20 mm, and diameter of the femoral or iliac arteries of ≤ 8 mm),

accurate measurement and limited stent graft oversizing are mandatory to avoid potential complications. Long-term durability remains to be determined. A strict follow-up is necessary to detect possible failures.

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