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1 **Optimising Aortic Endovascular Repair in Patients with Marfan Syndrome**

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14 **Keywords:** Aortic dissection, Endovascular aortic repair, Marfan syndrome, thoracic
15 endovascular aortic repair,

16 **Glossary of abbreviations:**

17 TEVAR: thoracic endovascular aortic repair; MFS: Marfan syndrome; aTBAD: acute type B
18 aortic dissection; cTBAD: chronic type B aortic dissection; rcTAAD: residual chronic type A
19 aortic dissection; CT: computed tomography; CTA: computed tomography angiography; FL:
20 false lumen

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Objectives: In Marfan syndrome (MFS) patients, endovascular repair carries a risk of aortic wall injury, which may result in retrograde aortic dissection, dilatation, or false aneurysms at the landing zones. It was hypothesised that these complications may be potentially avoided using modified practices. This study aimed to describe our experience with the use of a specific protocol of endovascular aortic repair in patients with MFS.

Methods: All MFS patients treated by aortic endovascular repair between February 2015 and August 2018 were prospectively included. The following rules were applied: (i) excluding stent grafts with bare stents and barbs, (ii) proximal landing in a pre-existing graft, or (iii) minimisation of proximal oversizing in the case of proximal landing in native aorta (<10%), and (iv) distal undersizing in case of chronic dissection.

Results: In eighteen patients (55% men, mean age: 47 ± 17 years), the index procedures were initial endovascular aortic repair ($n = 10$), elephant trunk completion ($n = 6$), and anastomotic pseudo-aneurysm after thoracic open repair ($n = 2$). The technical success rate was 100%. Proximal landing was in the native aorta in 11 patients (61%), with a mean proximal oversizing of 2.4 mm (8% oversized). Distal landing in the native aorta was performed in 16 cases (89%), with a mean distal undersizing of 8.9 mm (–23%). No mortality, spinal cord ischaemia, stroke, and retrograde aortic dissection occurred during the post-operative course. One type 1b endoleak was observed. The mean follow-up was 21.4 months. Aortic aneurysm related mortality was 5% ($n = 1$) and occurred after distal thoracoabdominal surgery planned from the outset (prior to endovascular repair). Another patient presented a proximal landing

zone complication with aortic enlargement. The mean maximal aortic diameter significantly decreased from 59 mm to 45 mm ($p = 0005$) after treatment.

Conclusion: The specific protocol described in this study seems to optimise the results of endovascular aortic repair in MFS patients with significant aortic remodelling.

INTRODUCTION

Marfan Syndrome (MFS) is the most common connective tissue disorder with multisystem features involving the aorta. It is caused by mutations in the fibrillin-1 gene (*FBN-1*) in most cases. Multimodal treatment is based on early diagnosis and preventive measures, which include avoidance of violent sports (brisk increase in blood pressure), beta-blocker therapy, and preventive surgery of ascending aorta.¹ Recently, owing to the decreased prevalence of aortic dissection related to preventive aortic surgery of the ascending aorta, the occurrence of dissection/aneurysm of the descending aorta is becoming a main issue in this population.²

Open descending thoracic aortic surgery remains the gold standard treatment for aneurysms and chronic dilatations after aortic dissection in MFS patients.³ Acute type A dissections are obviously treated with open repair, but endovascular repair is an emergent option to treat complicated acute type B dissections or chronic dissection not involving the ascending aorta in the general population without genetic diseases. However, the current guidelines do not recommend thoracic endovascular aortic repair (TEVAR) in patients with MFS.

Indeed, TEVAR of aortic dissections in patients with Marfan disease is not indicated as a first line treatment but can be considered in the event of a life threatening emergency in both acute and chronic complicated dissections.⁴ In this specific population, the risk of aortic wall aggression associated with the use of a thoracic stent is consider important, as it may result in retrograde aortic dissection, dilatation, or false aneurysms at the landing zone, which leads to endoleaks and rupture during follow up.

It was hypothesised that these complications may be potentially avoided by specific choices of the type and size of the stent graft to be used. Therefore, a specific protocol for stent graft placement is followed in patients with MFS who are referred to the department by the French National Reference Centre for Marfan Syndrome and Related Disorders.

METHODS

The local institutional review board approved this study, and all the patients provided their full consent for participation. This study was a retrospective analysis of prospectively collected database.

Patients

All patients endovascularly treated for aortic pathology between February 2015 and August 2018 who fulfilled the MFS criteria according to the Ghent nosology⁵ and were *FBNI* gene mutation carriers were included. In case of a life threatening aortic condition in patients not previously treated in the centre who were clinically suspected as having MFS, genetic tests were performed post-operatively.

A multidisciplinary team of vascular surgeons, cardiac surgeon, cardiologists, and anaesthesiologists discussed all the cases. Patients who were declared as poor candidates for open surgical intervention or at high risk of peri-operative complications on the basis of medical status or anatomic considerations were considered for endovascular therapy.

In our institution, management of Marfan patients with aortic dissection is as follows:

- i. Type A dissection requires Bentall or Aortoplasty (Tirone David in our institution) in emergency. The procedure is completed by arch repair with frozen elephant trunk when the entry tear is located on the arch.
- ii. Acute type B dissection are managed according to standard rules:

a. Non-complicated type B aortic dissections in Marfan patients are treated medically.

b. Complicated acute dissections (rupture, mesenteric or renal perfusion, refractory pain) are treated with TEVAR. Rapid aortic dilatation (i.e. aortic diameter above 40 mm during the acute phase) is considered a complication, and also considered for TEVAR.

iii. Chronic dissection of the descending aorta (either type B or residual type A) is considered for surgery according to guidelines (i.e. maximal aortic diameter ≥ 55 mm or diameter increase above 10 mm/year). TEVAR is chosen depending on the proximal landing zone: either the aortic arch has been replaced ($n = 7$), or the patient was considered to be too high risk for surgery

Rules followed for all the patients included (since January 2015):

- Excluding bare stents and barbs in the choice of a stent graft
- Proximal landing in a pre-existing graft (graft-in-graft technique)
- If impossible, minimisation of proximal oversizing in the case of proximal landing in the native aorta ($< 10\%$)
- Use of distal undersizing (adjusted diameter to the main diameter of the true lumen)

Technical aspect

All the procedures were performed under general anaesthesia by using a mobile C-arm with a flat-panel detector (Cios-Alpha, Siemens Medical Solutions, Forchheim, Germany), in an operating room, except the last two cases, in which the procedures were performed in a hybrid room (Infini-X Sky+, Canon Medical, Tochigi, Japan).

All TEVAR implantations were performed through the femoral artery. The choice of a percutaneous or surgical approach for TEVAR was left to surgeon preference. The insertion

side depended on the dissection morphology and extension. In case of aortic dissection, the guide wire was positioned in the true lumen under angiography guidance, with a catheter placed through the guide wire at the levels of the visceral and thoracic aortas. Intravascular ultrasonography was not used.

When total exclusion of the dissection was mandatory (urgent cases and large aneurysms), embolisation of the false lumen was performed. The technique that we used was previously reported.⁶

Preventive use of Cerebrospinal fluid drainage before TEVAR for spinal cord ischemia prevention was routinely used. Indications for drainage were:

- Long aortic coverage ≥ 200 cm
- TEVAR in level T8-L1
- False lumen embolisation
- Previous abdominal aortic repair

Computed tomography and imaging follow up

Computed tomography (CT) angiography (CTA) was performed using varied scanners, obtaining axial images with contiguous 1 mm thick sections from the beginning of the supra-aortic vessels to the femoral arteries. All CTA scans were performed with contrast material enhancement (100–120 mL of contrast). Acquisitions were performed at the arterial phase and late (delayed) phase (240 – 300 s) to highlight low flow endoleaks and false lumen (FL) patency in particular.⁷ The main aortic diameter was measured in arterial phase acquisition and perpendicularly to the centreline of the aorta. FL patency was defined in the late (delayed) phase acquisition. Post-operative CTA was systematically performed early after surgery and after 1, 3, 6, and 12 months and yearly thereafter.

Clinical follow up

Data were obtained during patient visits to the hospital, hospital stay, or telephone interviews. All follow up examinations were conducted by board certified vascular surgeons experienced in treating aortic dissections.

Outcome definitions

Initial technical success was defined as the achievement of thoracic stent graft delivery without type 1a proximal endoleak and surgical conversion. Early outcomes are defined as 30 day outcomes. The primary outcome was thoracic aortic shrinkage of ≥ 5 mm on the last follow- up CTA. Stroke was defined as a neurological deficit lasting for > 24 hours and confirmed by imaging. Spinal cord ischaemia was defined as postoperative paraparesis or paraplegia. Renal failure was defined as a decrease in glomerular filtration rate of $> 20\%$ or the need for haemodialysis. Respiratory failure was defined as the need for post-operative reintubation. Mid-term technical success was defined as aneurysmal stability or shrinkage on the last follow up CTA, with no landing zone dilatation/pseudo-aneurysm.

Statistical analyses

Statistical analysis was performed using Prism 7.00 (Graphpad Software, San Diego, CA). Numerical values are expressed as mean \pm standard deviation. Categorical variables are presented as percentages. Variables were compared between the initial and follow up CT scans using the Wilcoxon test. A p value $< .05$ was considered statistically significant.

RESULTS

Patient characteristics

During the study period (3.5 years), 69 patients underwent open thoracic or thoracoabdominal repair for dissection, aneurysms, graft infections. Among them, 26 were MFS patients

Eighteen patients were included (55% men, $n = 10$), with a mean age of 47 ± 17 years at the index procedure. The underlying pathology was an aortic dissection in all the cases.

The indications for endovascular repair were as follows:

- Acute type B aortic dissection (aTBAD) according to the following guidelines:⁴ aortic rupture, mesenteric or renal malperfusion, poor prognosis factor such as acute dilatation with a maximal thoracic aortic diameter of > 40 mm, and refractory pain ($n = 5/18$)
- Chronic aortic dilatation of a type B dissection (cTBAD) or residual chronic type A dissection (rcTAAD) after a previous arch repair, with a maximal thoracic aortic diameter of > 55 mm ($n = 10/18$)
- Acute abdominal aortic dissection ($n = 1/18$) treated with thoracic stent graft placement
- Proximal or distal aortic pseudo-aneurysm after thoraco-abdominal open repair (one proximal and one distal) for chronic type B dissection ($n = 2/18$)

The patients' demographics and comorbidities are shown in **Table 1**. Ten patients (55%) had American Society of Anesthesiologists scores of ≥ 3 . Sixty-six per cent of the patients had already undergone a previous aortic open repair ($n = 12$). Among the patients, seven had a previous aortic arch open repair ($n = 6$) and an open type 1 open thoraco-abdominal repair ($n = 1$), which allowed for a graft in graft proximal landing. Therefore 11 patients underwent TEVAR without previous aortic arch repair. Six patients (33%) underwent TEVAR in acute settings.

Procedures

Cerebrospinal fluid drainage was performed in nine patients (50%) before TEVAR (**Table 2**).

The index procedures were as follows:

- Initial endovascular aortic repair in 10 patients (nine TEVAR and one EVAR)
- Elephant trunk completion in six patients
- Late complication of open surgery: anastomotic pseudo-aneurysm after thoracic open repair (one proximal and one distal) for chronic dissection

TEVAR was performed through percutaneous access of the common femoral artery in eight patients (44%) and through surgical exposure in 10 patients (56%). Proximal landing was in the native aorta in 11 patients (61%) and in the previous surgical graft (graft in graft) in seven patients (39%). In case of native aorta proximal landing, the mean stent graft proximal oversizing was 2.4 ± 1.1 mm (8% oversized). Distal landing in the native aorta was performed in 16 patients (89%), and the mean distal stent graft downsizing was -8.9 mm \pm 12.1 (-23%).

In case of proximal landing in the native aorta, the devices used were mainly the Cook TX2 Dissection stent graft (Cook Medical, Bloomington, IN, USA) or Cook Custom made device based on the TX2 stent graft (with no barbs) in 44% ($n = 8$) of the patients. The other stent graft without barbs used was the Conformable-TAG (WL Gore and Associates, Flagstaff, AZ, USA) in 16% ($n = 3$) of the patients. In patients with proximal landing in the previous surgical graft (graft in graft), the device used was the Cook Zenith alpha thoracic stent graft (Cook Medical). No distal component was used. The mean covered length of the thoracic aorta was 185 ± 33 mm (range 145 – 261 mm). The technical success rate was 100% (Fig. 1).

Planned supra-aortic trunk debranching was necessary in five cases (zone 2, $n = 4$; zone 1, $n = 1$), and debranching of both renal arteries was performed in one case and performed in one procedure just before TEVAR. The patient with a ruptured acute abdominal aortic dissection required left renal artery revascularisation using the parallel graft technique (Fig. 2).

30 day outcomes

The post-operative data are summarised in Table 3. During the post-operative course, no mortality, spinal cord ischaemia, stroke, and retrograde aortic dissection were observed.

One type 1b endoleak through the FL was observed in a patient with a large thoracic aneurysm (93 mm) complicating a chronic type B dissection. This patient was operated by endovascular elephant trunk completion associated with thoracic FL embolisation. Despite the embolisation, the FL flow persisted on the late phase CTA at discharge.

Mid-term outcomes

The mean CT-scan follow-up period was 21.4 ± 14 months (range 3.5 – 46.2 months). Clinical follow up was 23.5 ± 14.6 months (range 6.1 – 47.4 months). One patient (5%) died from multiple organ failure after open thoraco-abdominal surgery. The 29 year old patient underwent an aortic arch open repair completed with TEVAR and FL embolisation to achieve exclusion of a 98 mm aneurysm in the posterior arch and aortic isthmus, complicating a chronic type B aortic dissection. A distal open thoracoabdominal repair was planned in the initial strategy because of a 60 mm type 2 thoracoabdominal aneurysms. The overall survival was 82% (95% CI 0.61 – 1.00).

No type 1a endoleak was observed (Table 4). The type 1b endoleak observed early persisted after two additional FL embolisation sessions. The 69 year old patient was unfit for open repair, so a branched thoraco-abdominal endovascular repair was planned. However, she died of a spontaneous subdural haematoma during follow up. One patient had a type 2 endoleak through the intercostal arteries that spontaneously resolved 6 months after surgery. No stent graft migration occurred.

Dilatation of the proximal landing zone without false aneurysm was observed in a 27 year old woman 11 months after TEVAR. This procedure was performed for a post-partum complicated acute type B dissection with mesenteric dynamic malperfusion. TEVAR allowed

complete salvage. The oversizing was larger in this study (+5 mm, 19.5% oversized). This case remains the only exception of oversizing of > 10%. Owing to symptomatic aortic enlargement, the patient underwent a total open arch repair with distal anastomosis on the stent graft with a simple postoperative course. No late surgical conversion with stent graft explantation was needed. The mid-term clinical success rate was 83% ($n = 15/18$).

A new aortic procedure was required in 10 patients (55%; Table 4):

- Planned strategy: In five patients (28%), an open thoraco-abdominal repair distal to the stent graft was planned prior to the TEVAR, in the pre-operative strategy of the repair. Mean interval of treatment between TEVAR and distal open repair was 11.5 ± 8.5 weeks.
- Enlargement of the untreated dissected native aorta (aneurysmal degeneration): It required a new procedure in four patients (22%), including open abdominal repair ($n = 1$), new TEVAR ($n = 1$), EVAR ($n = 1$), and FL embolisation ($n = 1$).
- TEVAR complications: One open arch repair was required and was considered as a TEVAR complication (5%).

Over the 18 patients, the maximal aortic diameter at the level of the treated aortic segment decreased from 59 mm to 45 ± 14 mm (Table 4). The Wilcoxon test revealed a significant reduction in thoracic diameter ($p = .0005$) after treatment. And 13 patients (72%) showed a favourable aortic remodelling with an aneurysmal shrinkage of ≥ 5 mm during follow-up.

DISCUSSION

This study represents a larger published series of patients with genetically authenticated MFS treated with TEVAR. No deaths related to the TEVAR procedure were observed. Dilation of the landing zone occurred in only one patient (5%) and is a complication of the largest

oversizing in this series. Furthermore, the protocol for using TEVAR in this population led to a significant favourable aortic remodelling in 72% of the cases at midterm follow up in this particularly challenging pathology.

Genetic diagnosis

In surgical series, MFS is theoretically diagnosed clinically in accordance with the revised Ghent criteria,⁵ although the multidisciplinary approach necessary to certify the diagnosis is difficult to obtain in this severely ill population. Without genetic testing, many “Marfan-like” other related diseases may be considered Marfan (FBN 1 mutation),⁸ and this leads to the heterogeneity of the population in different studies.^{9 – 11} Indeed, many genetic mutations in many signalisation pathways between the smooth muscle cells (SMCs) and the extracellular matrix can occur^{12 – 14} and lead to aortic diseases. Furthermore, many of these mutations may be associated with skeletal features evocative of MFS. The association of some mutations with a more severe phenotype, which may impact the result of TEVAR, has now been well established.

TEVAR complications

Mitigated outcomes after TEVAR in MFS are also explained by the heterogeneity of TEVAR practices. Devices used and sizing techniques are usually not detailed in the literature. Landing zone complications are the first cause of TEVAR failure in patients with MFS reported in the literature. In addition, the aortic reinterventions rate ranges from 14% to 60%, and the rate of conversion to open surgical technique ranges from 0 to 33% mainly owing to late failure of TEVAR.^{10,11,15 – 21}

Excessive oversizing worsens the aortic wall injury and increases the risk of retrograde type A dissection.²² Furthermore, patients with MFS present a higher risk (×10) of stent graft induced new -entry (SINE) tears, with a reported incidence rate of 33%.²³ SINE tears after

TEVAR are more frequent when TEVAR is performed for chronic rather than for acute dissection. They may cause retrograde false lumen flow and pressurisation, which result in potential aneurysmal degeneration. Distal oversizing appears to be a significant risk factor of SINE tears after TEVAR of aortic dissection.²⁴ Traditional teaching recommends oversizing seal zones by 10% – 25%, but this should probably be adapted according to the disease process and clinical circumstance. Oversizing should be lower in aortic dissections than in aortic aneurysm. In a same way, it should be lower in patients with MFS than in patients with no connective tissues disorders. Thus, it was decided to minimise the proximal oversizing in our practice in such patients. Furthermore, the distal diameter of the stent graft is now sized according the true lumen main diameter. This kind of practice requires the use of several stent grafts with different diameters or the use of tapered stent grafts that are available off the shelf if the tapering is ≤ 8 mm. When the tapering is larger, a custom made device has to be used. The use of stent grafts with a proximal bare stent is less clearly related to retrograde type A aortic dissection after TEVAR.²² However, it appears to be a significant risk factor in some papers.²⁵ As a precautionary principle, this kind of stent graft was not used.

Staged strategy and extensive aortic repair in MFS

The TEVAR complication rate may be reduced when included in a multistage strategy for extensive aortic repair. After an acute type B or acute operated type A dissection, aortic aneurysmal evolution of the descending aorta is observed in $> 40\%$ of patients²⁶ and probably even more often in patients with MFS. The main expansion zone is the proximal descending aorta. In a large cohort study, Mimoun *et al.*²⁷ observed that aortic event free survival directly correlated with the remaining dissected native aorta. These findings advocate an extensive aortic repair from the aortic root to the iliac arteries in Marfan disease with dissection of the entire aorta and aneurysmal degeneration.

Open surgical repair remains the gold standard treatment for the descending thoracic aorta. This technique appears to be more durable, with acceptable morbidity and mortality. The spinal cord ischemia rate ranges from 4% to 7%, and the mortality rates range from 5% to 14% in patients with connective tissue disorders.^{28,29} Longer follow up is available after open surgical repair in the literature, indicating fewer additional new aortic procedures.²⁹

Extensive endovascular aortic repair is an emergent strategy. TEVAR associated with the use of a distal aortic bare stent in Marfan patients has been recently reported.¹¹ The technique using stent assisted balloon induced intimal disruption and relamination concept (STABILISE) was first described in 2014 by Peter Mossop and collaborators.³⁰ In a study of seven patients with suspected MFS who presented complicated acute type B aortic dissections, the STABILISE technique showed favourable early outcomes. Aortic complications were observed in only 14% of the patients (one case of dilatation of the proximal aortic landing zone dilatation with type 1a endoleak). A favourable aortic remodelling was observed in all the cases.

Furthermore, Lille's team published the first study of fenestrated or branched stent grafts to treat a thoraco-abdominal aorta in chronic aortic dissections associated with connective tissue disorders.⁹ They reported 17 patients unfit for surgery, including four with MFS and known *FBNI* gene mutation. The early outcomes were favourable with no spinal cord ischemia or mortality. Aneurysmal shrinkage was observed in > 35% during midterm follow up.

Finally, extensive aortic repair can also be performed as a staged repair. A leading European team in open surgery recently presented outcomes of a two stage open strategy for thoracoabdominal repair.³¹ In two not entirely comparable cohorts, a two stage type 2 open thoraco-abdominal repair compared favourably with one stage repair in terms of in hospital

mortality (0% vs. 22%) and long term survival. Indeed a multi-stage repair is an attractive strategy in such complex pathologies.

In Bichat Hospital, staged hybrid repair for type 2 thoraco-abdominal aneurysmal dissections is, as much as possible, performed as described by Jain *et al.*³² TEVAR simplifies the second stage elephant trunk³³ and probably lowers the risk of spinal cord ischemia in hybrid stage aortic repair³⁴ when the distal thoracoabdominal aorta needs to be repaired.

Limitations

This study has several limitations. The depth of the statistical analysis was limited because of the relatively small number of patients included in this study. Comparison with open repair is limited due to patients' conditions (they were declared as poor candidates for open surgical intervention). This study included two types of devices and they could have different performance in such population. Finally, the follow up in this study is intermediate and a longer follow up is mandatory to confirm this promising results.

CONCLUSION

In this study of MFS patients with aortic dissection, endovascular aortic repair appears to be an efficient therapeutic alternative provided specific rules are observed. A delicate approach, using atraumatic stent grafts, limited oversizing, and graft in graft technique, seems to avoid acute complications and allows positive remodelling of the aorta. Aortic stent grafting, which nowadays plays an important role in the treatment of life threatening conditions, may become part of a multistage aortic repair, allowing complete treatment of the dissected aorta. Multicentre prospective enrolment of patients and longer follow up are required to confirm these encouraging results with aortic endovascular repair in patients with MFS.

CONFLICT OF INTEREST

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Figure 1. A 64 year old patient with Marfan syndrome with a 59 mm thoraco-abdominal aneurysm complicating a type B chronic aortic dissection. Volume rendering technique pre-operative view (A), axial views showing patency of the false lumen at thoracic and abdominal level (B). He underwent thoracic endovascular aortic repair with Cook dissection thoracic stent graft (proximal stent graft: ZDEG 30 × 142, distal stent graft: ZDEG 32-28 × 162, Cook Medical, Bloomington, IN, USA) with left subclavian to left common carotid artery transposition. Follow-up computed tomography angiography at 26 months shows favourable expansion of the stent graft, with two vascular plugs inside the false lumen leading to complete thrombosis of it (C and D).

Figure 2. A 59 year old patient with Marfan syndrome with a medical history of mechanic mitral and aortic replacement and chronic heart failure, presented acute abdominal pain with hypotension during hospitalisation for flutter ablation. Computed tomography angiography (CTA) revealed a ruptured acute abdominal aortic dissection with renal arteries involvement (A). Axial view shows the infrarenal entry tear and the rupture (B). He was considered unfit for an open suprarenal repair. He underwent a life-saving procedure with abdominal aorta coverage with two thoracic devices (distal stent graft: C-TAG 37 × 150 and proximal stent graft: 37 × 100, WL Gore and Associates, Flagstaff, AZ, USA) and a left renal

479 revascularisation with the parallel graft technique (“sandwich technique”) using 2 Gore
480 Viabahn stents (WL Gore and Associates) re-inforce with a nitinol bare stent (Absolut pro,
481 Abbott Vascular, Redwood City, CA, USA). Proximal oversizing was 3 mm and distal
482 oversizing was 5 mm. Six month follow up CTA shows good exclusion of the false lumen and
483 patency of the left renal artery (C) with no landing zones complications.

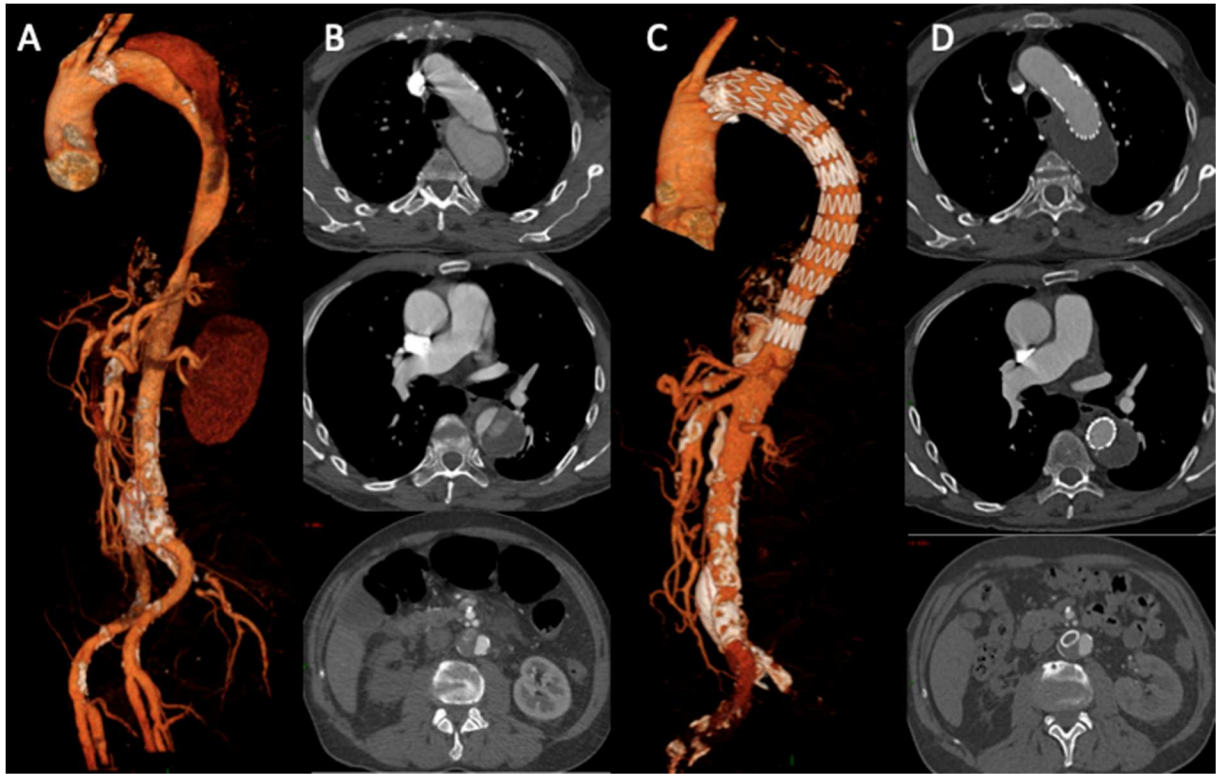


Figure 1: a 64 year-old Marfan patient with a 59 mm thoracoabdominal aneurysm complicating a type B chronic aortic dissection. Volume rendering technique preoperative view (A), axial views showing patency of the False lumen (FL), at thoracic and abdominal level (B). He underwent TEVAR with Cook Dissection thoracic stentgraft (proximal stentgraft; ZDEG 30x142, distal stentgraft: ZDEG 32-28x162, Cook Medical, Bloomington, IN) with left subclavian to left common carotid artery transposition. Follow-up CTA at 26 months shows favorable expansion of the stentgraft, with 2 vascular plugs inside the FL leading to complete thrombosis of it (C and D).

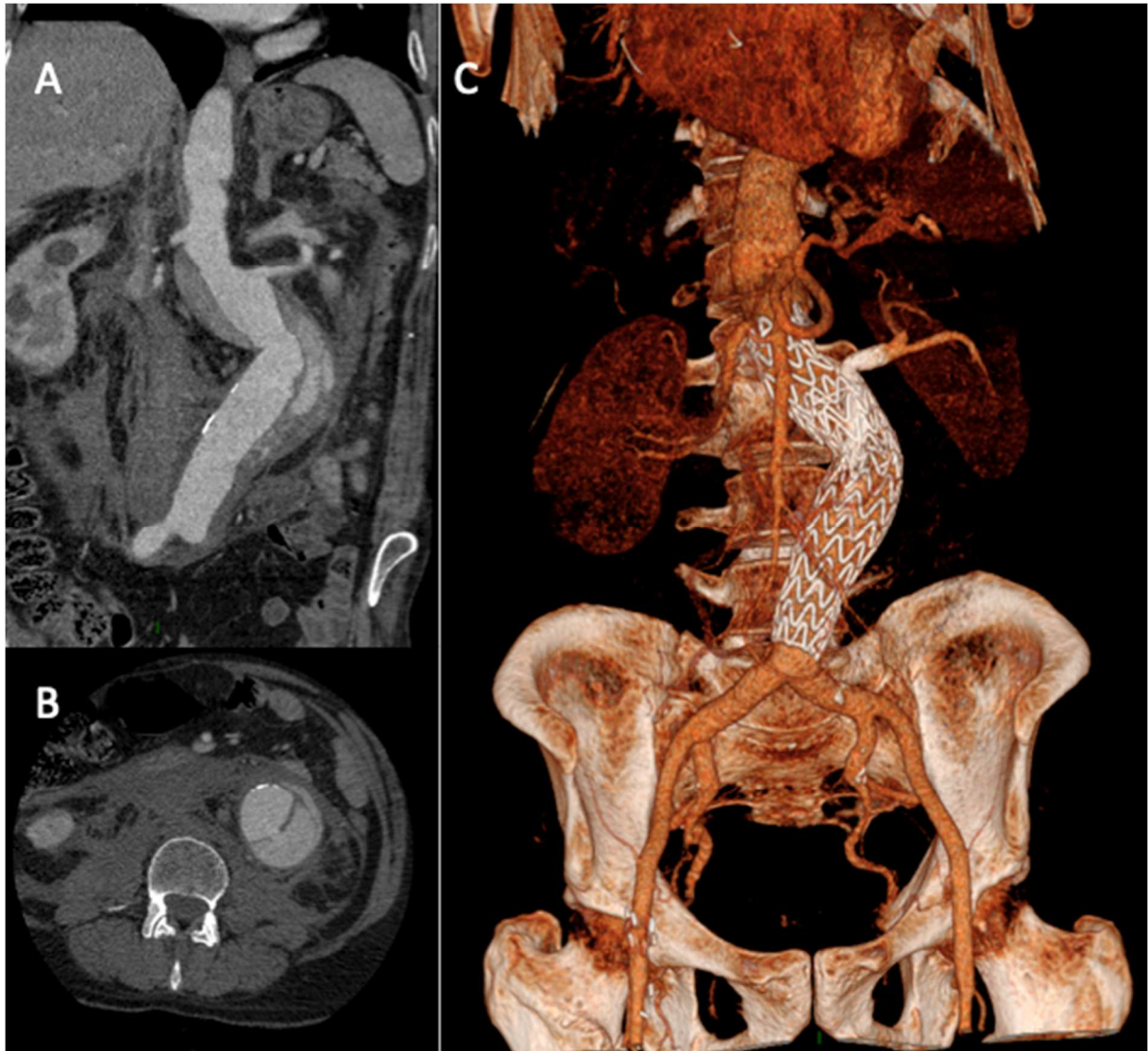


Figure 2: A 59 year-old Marfan patient, with a medical history of mechanic mitral and aortic replacement and chronic heart failure, presented acute abdominal pain with hypotension during hospitalization for flutter ablation. CTA revealed a ruptured acute abdominal aortic dissection with renal arteries involvement (A). Axial view shows the infrarenal entry tear and the rupture (B). He was considered unfit for an open suprarenal repair. He underwent a life-saving procedure with abdominal aorta coverage with 2 thoracic devices (distal stentgraft: C-TAG 37x150 and proximal stentgraft: 37x100, WL Gore and Associates, Flagstaff, Arizona) and a left renal revascularization with the parallel graft technique (“sandwich technique”) using 2 Gore Viabahn stents (WL Gore and Associates, Flagstaff, Arizona) reinforce with a nitinol bare stent (Absolut pro, Abbott Vascular, Redwood City, CA). Proximal oversizing

was 3mm and distal oversizing was 5 mm. Six-months follow-up CTA shows good exclusion of the FL and patency of the left renal artery (C) with no landing zones complications.

Table 1. Baseline characteristics of Marfan syndrome patients treated with optimised protocol for endovascular aortic repair.

Characteristics	<i>n</i> = 18
Age – y	47±17
Male	10 (55)
Marfan syndrome	18 (100)
<i>FBN1</i> mutation	18 (100)
ASA score ≥ 3	10 (55)
Hypertension	5 (28)
Diabetes mellitus	2 (11)
Smoker	6 (33)
Dyslipidaemia	4 (22)
Chronic heart failure	3 (17)
Chronic obstructive pulmonary disease	3 (17)
Acute aortic dissection	6 (33,3)
<i>Underlying type of dissection</i>	
Chronic type A (residual)	3 (17)
Type B	14 (78)
Acute	5 (28)
Chronic	9(50)
Acute abdominal	1 (5)
<i>Last previous aortic procedure</i>	12 (67)
Ascending aorta repair	2 (11)
Aortic arch repair	6 (33)
Thoracic or thoraco-abdominal repair	3 (17)

Abdominal repair	1 (5)
Mean maximal aortic diameter – mm	59±15

Continuous data are presented as mean±standard deviation and categorical data as number (%). ASA = American society of Anaesthesiology; y = years.

Table 2. Technical aspects of Marfan syndrome patients treated with optimised protocol for endovascular aortic repair.

Technical aspects	n=18
<i>Index procedure</i>	
Initial TEVAR	10 (55)
Elephant Trunk completion	6 (33)
Anastomotic false aneurysm	2 (11)
<i>Stents grafts</i>	
Cook TX2 dissection or CMD	8 (44)
Gore C-TAG	3 (17)
Cook alpha thoracic (graft in graft proximal landing)	7 (39)
Proximal landing in native aorta	11 (61)
Proximal oversizing (in native aorta) – mm	2.4 (8)
Distal landing in native aorta	16 (89)
Distal undersizing (in native aorta) – mm	-8.9 (-23)
Coverage length – mm	185±33
<i>Associated procedures</i>	
FL embolization	8 (44)
Supra aortic trunks debranching	5 (28)
Zone 1	1 (5)
Zone 2	4 (22)
Visceral debranching	1 (5)
Renal artery covered stenting (parallel graft)	1 (5)
CSF drainage	9(50)
Technical Success	18 (100)

Continuous data are presented as mean \pm standard deviation and categorical data as number (%). CMD = custom made device, CSF = cerebrospinal fluid; FL, false lumen; TEVAR = thoracic endovascular aortic repair;.

Table 3. 30 day outcomes of Marfan syndrome patients treated with optimised protocol for endovascular aortic repair.

30 day outcomes	n=18
In hospital mortality	0
Aortic related death	0
<i>Spinal cord ischemia</i>	
Transient paraparesia	0
Permanent paraparesia	0
Stroke	0
Renal failure	1 (5)
Respiratory failure	0
Heart failure/myocardial infarction	0
<i>Endoleak</i>	
Type 1a	0
Type 1 b	1 (5)
Type 2	1 (5)
Retrograde aortic dissection	0

Continuous data are presented as mean±standard deviation and categorical data as number (%).

Table 4. Mid-term outcomes of Marfan syndrome patients treated with optimised protocol for endovascular aortic repair.

Mid-term outcomes	n=18
Follow up – mo	21.4±14
<i>Endoleaks</i>	
Type 1a	0
Type 1b	1(5)
Type 2	0
<i>Landing zone dilatation/pseudo-aneurysm</i>	
Proximal	1 (5)
Distal	0
Stent graft induced new entry tear	0
<i>New aortic procedures</i>	10 (55)
Open aortic arch repair	1(5)
Distal open thoraco-abdominal repair	5 (28)
Open abdominal aortic repair	1 (5)
TEVAR	1(5)
EVAR	1 (5)
False lumen embolization	1 (5)
Aortic related death	1 (5)
Overall mortality	2 (11)
<i>Causes of death</i>	
Multiple organ failure (after distal open thoraco-abdominal repair)	1 (5)
Subdural hematoma	1 (5)
Mean maximal aortic diameter – mm	45±14
Aneurysmal shrinkage ≥5 mm	13 (72)
Mid-term clinical success	17 (95)

Continuous data are presented as mean±standard deviation and categorical data as number (%). TEVAR = thoracic endovascular aortic repair; EVAR = endovascular aortic repair; mo = months.