

# Successful repair of an arch aneurysm with acute aortic dissection in a patient with Marfan syndrome using a hybrid surgical approach and the stent-assisted balloon-induced intimal disruption and relamination in aortic dissection repair technique

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## ABSTRACT

Endovascular management of aortic complications in patients with Marfan syndrome (MFS) is uncommon. We treated a patient with MFS with a diagnosis of a 75-mm aortic arch aneurysm and uncomplicated aortic type B dissection using single-stage hybrid surgery combining total arch replacement with elephant trunk and the STABILISE (stent-assisted balloon-induced intimal disruption and relamination in aortic dissection repair) technique for complete aortic remodeling. The repair was successful, and the aortic true lumen was completely expanded. At 6 months after surgery, clinical evaluation confirmed the early success of the intervention. This type of surgery must be studied further before it can become routine treatment for patients with MFS but it proved safe and feasible. (*J Vasc Surg Cases Innov Tech* 2021;7:392-5.)

**Keywords:** Aortic arch repair; Aortic dissection; Aortic remodeling; Marfan syndrome; STABILISE

For patients with Marfan syndrome (MFS), the current guidelines recommend endovascular treatment for emergencies or as a bridge to surgery,<sup>1</sup> because open repair yields excellent results in the proximal aorta. Recent reports of success and technical progress have resulted in renewed interest in endovascular treatment of patients with MFS, in particular, for type B aortic dissections (TBADs); however, a healthy proximal landing zone is crucial. Furthermore, the stent-assisted balloon-induced intimal disruption and relamination in aortic dissection repair (STABILISE) technique for TBAD appears promising, although its application for patients with MFS has been infrequent,<sup>2</sup> as has endovascular management of arch aneurysms associated with TBAD.<sup>3</sup> We report a case of arch aneurysm and subacute TBAD in a patient with MFS treated using a hybrid

approach and the STABILISE technique. The patient provided written informed consent for the report of her case.

## CASE REPORT

A 50-year-old woman with MFS and three previous cardiovascular operations (Bentall procedure in 1983, mitral valve replacement in 1991, and Bentall distal anastomosis replacement in 2001) was admitted to our emergency department for acute thoracic pain. The computed tomography angiography scan showed a 75-mm aortic arch aneurysm with an uncomplicated acute aortic dissection type B<sub>2-9</sub>.<sup>4</sup> Renal arteries originated from the true lumen, celiac trunk, and superior mesenteric artery (SMA) from the false lumen (Fig 1). The patient was admitted to the intensive care unit for monitoring and pain management and scheduled 3 days later for semi-elective hybrid surgery owing to the huge arch aneurysm size.

The open procedure involved total arch replacement using a quadrifurcated Dacron graft (Intergard woven aortic arch prosthesis 24/10/8/8/10-mm; Getinge AB, Stockholm, Sweden) prolonged by an elephant trunk (Gelweave plus aortic prosthesis 22-mm; Terumo Aortic, Vascutek Ltd, Scotland, United Kingdom), with the patient under systemic circulatory arrest, moderate hypothermia for 37 minutes at 23.1°C, and antegrade cerebral perfusion. Because of the fragile distal anastomosis, it was decided intraoperatively to switch to a frozen elephant trunk technique with TEVAR with the available on-stock endografts. Correct positioning of the stiff wire in the true lumen and ascending graft was assessed using transesophageal ultrasound. A cTAG thoracic stent-graft (26-mm × 10-cm; W.L. Gore & Associates, Flagstaff, Ariz) was deployed just distal to the left subclavian branch. Next, a Valiant thoracic stent-graft (31/31 × 97-mm;

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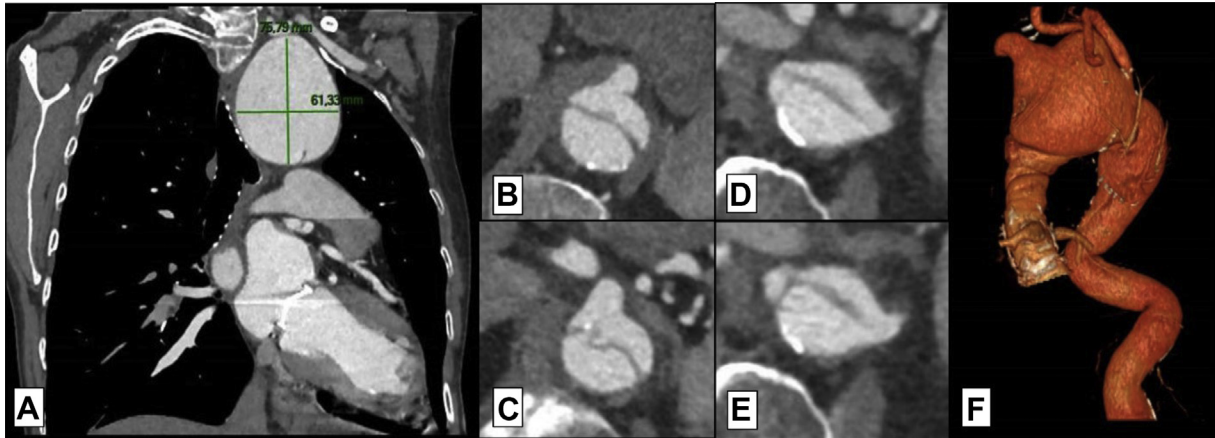
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**Fig 1.** **A**, Scan showing 75 × 61-mm aneurysm of the aortic arch. **B**, Celiac trunk arising from the false lumen. **C**, Superior mesenteric artery (SMA) arising from the false lumen. **D,E**, Renal arteries arising from the true lumen. **F**, Three-dimensional reconstruction.

Medtronic Vascular, Santa Rosa, Calif) was placed with a 5-cm overlap and a 36 × 180-mm dissection stent (Zenith dissection endovascular stent; Cook Medical, Bloomington, Ind) was implanted down to the infrarenal aorta. Perforation of the dissection membrane was achieved with a steerable sheath (Oscor, Palm Harbor, Fla) and a 0.0018-in. Astato wire (Asahi Intecc Medical, Tokyo, Japan) to allow for placement of a guide-wire in the SMA. The stent-graft overlap and dissection stents were remodeled using the STABILISE technique and a compliant aortic balloon.<sup>5</sup> A balloon-expandable stent (Omni-link Elite 8 × 19-mm; Abbott Vascular, Chicago, Ill) was finally placed in the SMA. Angiography showed complete obliteration of the false lumen without an endoleak and preserved perfusion of the visceral and renal arteries. The patient left the intensive care unit on day 1 and the hospital on day 15. Six months later, a computed tomography angiogram confirmed successful repair of the arch aneurysm and complete expansion of the true lumen with remodeling around the TEVAR and dissection stents and excluded any endoleak or false lumen perfusion, confirming the early success of the intervention (Fig 2).

## DISCUSSION

We have reported the successful hybrid treatment of an acute TBAD with an associated arch aneurysm in a patient with MFS, combining total arch replacement and the STABILISE technique. Thoracoabdominal aortic open repair was proposed to our patient with MFS, who denied it. To avoid a repeat sternotomy, endovascular arch repair using a branched endograft could have been considered. However, it requires experience, and because the patient required emergency surgery, such an endograft could not have been ordered. Because the patient had presented as an emergency case from another country, it was not possible to obtain any additional information on the MFS diagnosis.

Although TEVAR has now replaced open surgery for TBAD, it does not prevent further aneurysmal development owing to the absence of total thrombosis of the false lumen.<sup>6</sup> This is now considered the main risk factor for incomplete aortic remodeling.<sup>7</sup> Moreover, its use for patients with MFS is not the reference standard because it results in increased rates of false lumen persistent flow and late dilatation owing to stent-graft new entry in ≤33%.<sup>8</sup>

The STABILISE technique seems to induce false lumen obliteration, favoring long-term aortic remodeling and has been used in our center for cases of complicated acute and subacute type B aortic dissection. The high likelihood of aneurysmal degeneration of the thoracic aorta justified its use in our patient with MFS. We considered the safety of aortic ballooning in a patient with weakened aortic walls during the subacute phase. Neither Faure et al<sup>2</sup> nor Soler et al<sup>3</sup> reported any case of aortic rupture. This technique was used in seven patients with MFS, with complete aortic remodeling in all seven patients (100%) at 2 years after treatment.<sup>2</sup>

The endovascular approach remains under evaluation for patients with MFS or other heritable aortopathies, and these patients require close follow-up after TEVAR owing to the risk of further dilatation. When late reintervention, whether endovascular or open, is necessary, it could be complicated by the dissection stent. However, we, and others,<sup>9</sup> have successfully performed endovascular repair of chronic dissection using a fenestrated endograft after an aortic bare stent had been implanted, without any difficulty with catheterization through the stent.

The distal sealing zone in the thoracic aorta could have been achieved with an occlusion device such as a candy plug in the false lumen instead of the STABILISE technique. However, we were more confident with the



**Fig 2.** **A**, Repair of the arch with a quadrifurcated graft and elephant trunk, with the beginning of the thoracic endograft. **B**, Ostium of the celiac trunk. **C**, Stented superior mesenteric artery (SMA) through the dissection stent. **D**, Outcome at the level of the renal arteries. **E**, Three-dimensional reconstruction with arch repair and thoracic endograft, followed by two dissection stents.

STABILISE technique, and no candy plug was available at the time.

Soler et al<sup>3</sup> reported treatment of a type B<sub>1-10</sub> aortic dissection using a hybrid approach and the STABILISE technique. After an uneventful initial procedure, the patient experienced aneurysmal degeneration between the covered thoracic endograft and the dissection stent owing to stent dislocation. The investigators recommended long stent overlapping, especially when the anatomy is tortuous, which is frequent in patients with MFS.<sup>3</sup>

The presence of diseased aortic walls raise questions regarding the quality of the proximal landing zone. However, few data exist on hybrid aortic repair for TBAD in patients with MFS. Because the aortic arch will be involved in >25% of all patients with TBAD,<sup>10</sup> surgeons will need to combine arch procedures with thoracic stent grafting to achieve a better proximal sealing zone. Some centers have suggested that TEVAR for patients with MFS should be used only if deployed in a graft to avoid late complications.<sup>10</sup> Faure et al<sup>2</sup> reported only one type I endoleak (14%), a rate similar to the 12.5% reported by Botta et al<sup>11</sup> and the rates of type Ia endoleaks reported in patients without MFS (range, 17%-21%). Kouchoukos<sup>12</sup> considered that using a Dacron graft as a proximal landing zone for TEVAR instead of the native aorta was an ideal indication in patients with MFS, precluding any previous antegrade deployment of the stent-graft in the thoracic aorta.

Another option would be a frozen elephant trunk using a hybrid graft combining the Dacron graft and stent-graft, which was not available for our case. Moreover, implantation of the stent-graft from above could lead to perforation, a new entry tear in the acute phase of TBAD, or extensive aortic coverage in small patients, increasing the risk of spinal cord ischemia.

## CONCLUSIONS

The use of a hybrid intervention in a patient with MFS, combining the STABILISE technique with open arch repair and elephant trunk proved to be safe, feasible, and completely successful. Because the STABILISE technique remains under evaluation, further studies and longer follow-up are necessary to allow this approach to be used in routine practice for patients with MFS.

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