

# Hybrid approach to achieve secure distal seal zones during endovascular aortic repair in a patient with Marfan syndrome

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## ABSTRACT

This report details a hybrid approach for treatment of abdominal aortic aneurysm in a patient with Marfan syndrome (MFS). A 34-year-old patient with MFS and prior open thoracoabdominal aortic aneurysm repair underwent bilateral common iliac artery interposition graft repair and endovascular aortic repair. The bifurcated stent graft was implanted into the previous thoracoabdominal graft proximally and iliac interposition grafts distally. Postoperatively, the patient recovered uneventfully with sac regression to 4.5 cm through 2-year follow-up, without seal zone degeneration. This hybrid approach aimed to eliminate landing zone degeneration in patients with MFS undergoing endovascular repair. (J Vasc Surg Cases Innov Tech 2024;10:101595.)

**Keywords:** Marfan syndrome; Endovascular aortic repair; Abdominal aortic aneurysm; Aortic dissection; Iliac artery aneurysm

Marfan syndrome (MFS) is a congenital connective tissue disease resulting from pathogenic variants in *FBNI*, which causes systemic abnormalities.<sup>1</sup> The primary causes of major morbidity and mortality in MFS are thoracic aortic dissections and progressive aortic root enlargement.<sup>2,3</sup> Open surgical repair is recommended for definitive repair of aortic complications given its long-term durability.<sup>4,5</sup>

After dissection, patients with MFS experience aneurysmal degeneration affecting the entire aorta, necessitating multiple operations.<sup>6</sup> Reports indicate reintervention rates for aortic disease in patients with MFS ranging from 14% to 27%.<sup>7,8</sup> Furthermore, increasing numbers of patients have a history of open aortic surgery, with rates varying from 7.3% to 37.4%.<sup>9,10</sup> Staged repair can mitigate perioperative complications like spinal cord ischemia. However, redo open aortic surgery carries risk of intraoperative complications, including injury to adjacent structures and major bleeding.<sup>11,12</sup>

Endovascular aortic repair (EVAR) has become the dominant treatment modality for degenerative abdominal aortic aneurysm (AAA) owing to its lower perioperative risks and immediate recovery.<sup>13</sup> Similarly, thoracic

EVAR has become the preferred treatment for thoracic aortic pathologies.<sup>14,15</sup> However, endovascular repair in MFS remains controversial owing to limited data on durability.<sup>16</sup> In particular, concerns linger regarding seal zone integrity in patients with MFS experiencing progressive aneurysmal dilatation of their native aorta. However, early results of EVAR have been promising in a select subgroup of patients with MFS deemed high risk for open repair.<sup>17-19</sup>

Here, we report a case of a patient with MFS with a history of open thoracoabdominal aortic repair who was treated successfully with EVAR with concurrent bilateral common iliac artery interposition grafts to create secure EVAR seal zones. The patient provided written informed consent for the report of his case details and imaging studies.

## CASE REPORT

A 34-year-old man with MFS and a history of multiple open aortic replacements presented to our clinic for an enlarging infrarenal AAA. Twelve years earlier, the patient underwent elective valve-sparing aortic root replacement. Four years later, he developed a type B<sub>2-11</sub> dissection extending into the left common iliac artery, complicated by right lower extremity acute limb ischemia, requiring fasciotomies, skin grafts, and left to right femoral-femoral bypass. Three years thereafter, surveillance imaging revealed increased aneurysmal dilatation of a Crawford extent I post-dissection thoracoabdominal aortic aneurysm (TAAA) to 5.8 cm with occlusion of the previous femoral-femoral bypass (Fig 1). The patient underwent a successful open TAAA repair with a multibranch graft to the celiac, superior mesenteric, and bilateral renal arteries. Aortic replacement was confined to the juxtarenal aorta to decrease risk of spinal cord ischemia.

Five years later, computed tomography angiography revealed an enlarging 5.9-cm infrarenal AAA distal to the previous open repair, with false lumen extension to left common iliac artery. Genetic counseling was offered, but the patient deferred, having

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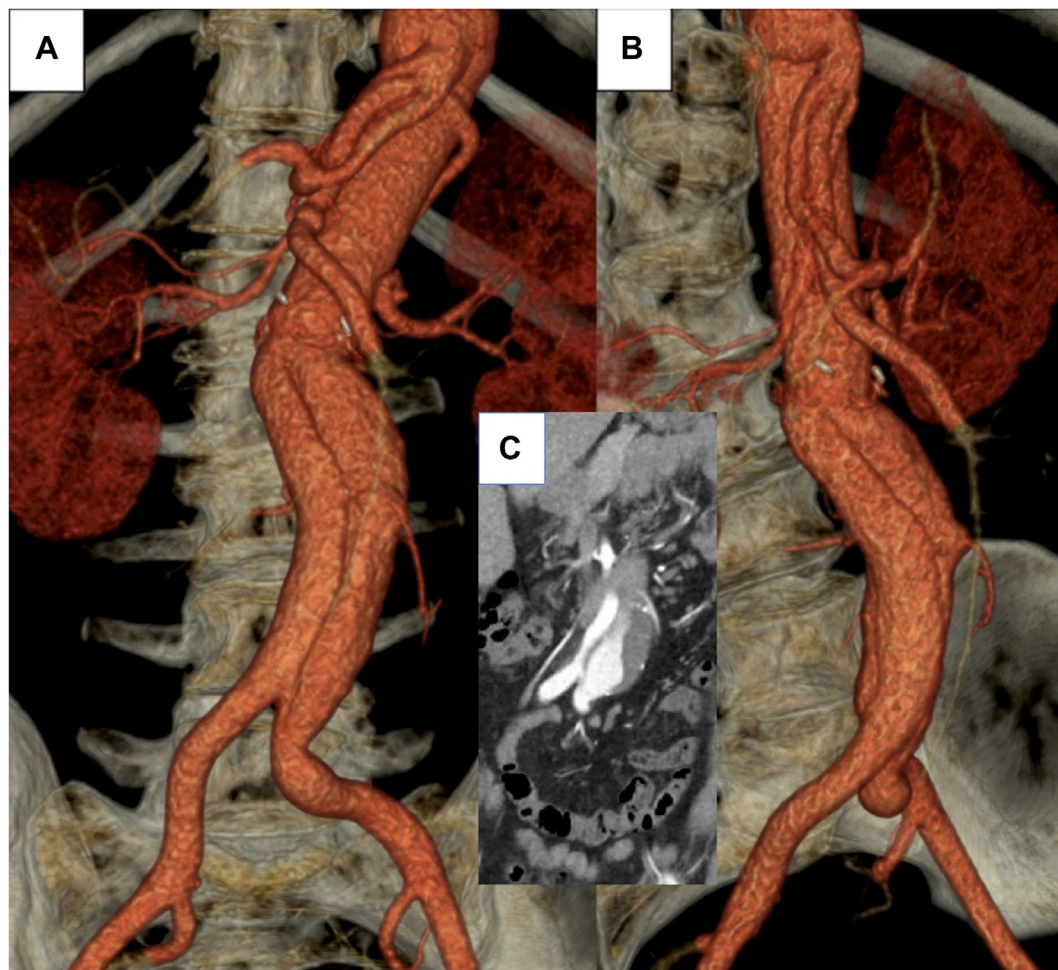
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**Fig 1.** Patient's thoracoabdominal aortic aneurysm (TAAA) seen on follow-up imaging after valve-sparing root repair in coronal view (A) and sagittal view (B). Proximal left common iliac artery with false lumen extension from the infrarenal aorta seen on computed tomography angiography in the coronal view (C).

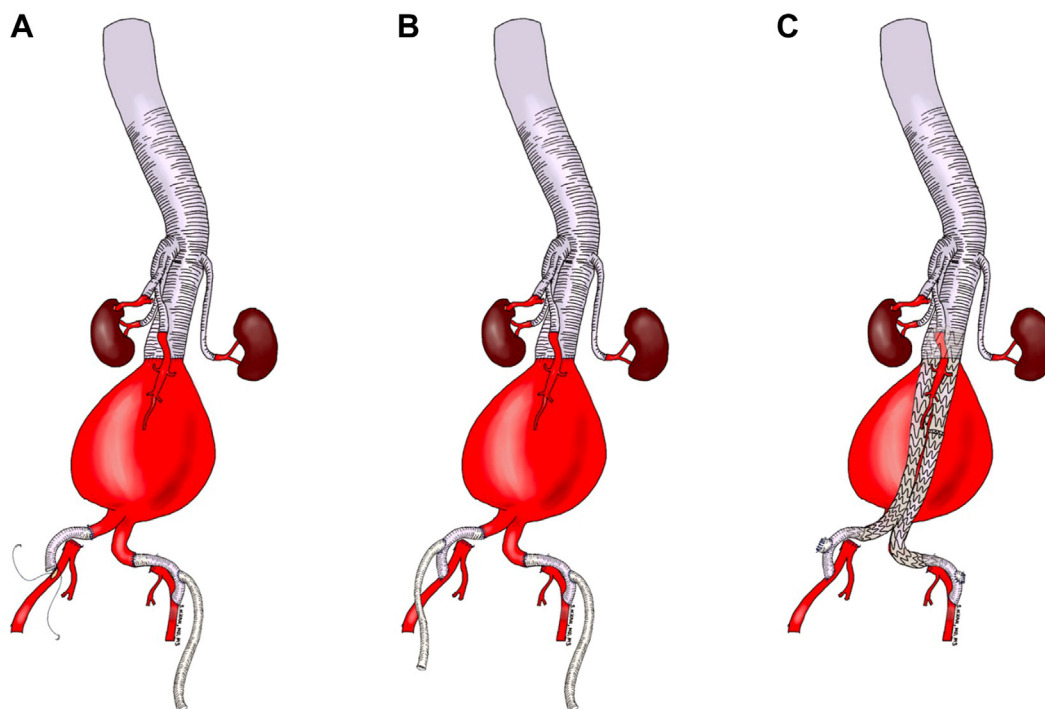
already received genetic confirmation of his MFS diagnosis. Given the previous operation involving the juxtarenal aorta, we elected a hybrid repair with open bilateral iliac artery interposition grafting, followed by concomitant EVAR landing proximally into the previous thoracoabdominal graft and iliac grafts distally.

Under general anesthesia, we performed a midline laparotomy from the umbilicus to the pubis, reflecting the mesentery to expose the left iliac bifurcation. The distal left common iliac artery was divided, and its distal end was oversewn. A 10-mm Dacron graft was anastomosed to the proximal stump in end-to-end fashion. The distal end of the bypass was anastomosed to the external iliac artery in end-to-side fashion, preserving flow to the internal iliac artery. A temporary iliac conduit was sewn onto this bypass graft in end-to-side fashion (Fig 2). The right iliac artery was replaced in similar fashion. Each conduit was accessed and upsized to 18F and a 12F sheath, through which a 35 × 14 × 14 cm Gore Excluder (W. L. Gore & Associates, Flagstaff, AZ) was positioned. The Excluder was deployed into the prior open TAAA Dacron and sealed distally into the iliac grafts. This technique ensured both proximal and distal landing

zones for the EVAR were within prosthetic material, eliminating potential native tissue dilation. The patient recovered uneventfully and was discharged on postoperative day 4. Two years later, computed tomography angiography demonstrated sac regression of the infrarenal AAA without further aneurysmal degeneration (Fig 3).

## DISCUSSION

This case demonstrates a hybrid approach to managing progressive aneurysmal degeneration of a dissected aorta in a patient with MFS. Although studies have demonstrated the safety and effectiveness of EVAR in specific patients with MFS, such as high-risk patients for open repair or patients with rupture, concerns persist regarding durability and seal zone integrity.<sup>20</sup> Current guidelines recommend an elective staged open surgical approach to maintain durability in patients with MFS. However, in staged open aortic repairs, transitional aortic segments can be challenging, because the distal end of the previous



**Fig 2.** Open repair of bilateral common iliac arteries with interposition grafting to repair aneurysmal false lumen of the infrarenal aorta extending into the proximal left common iliac artery (**A**) and placement of temporary iliac conduits for endovascular access (**B**). Endovascular aortic repair (EVAR) with implantation proximally into previous thoracoabdominal graft and distally into iliac grafts (**C**).

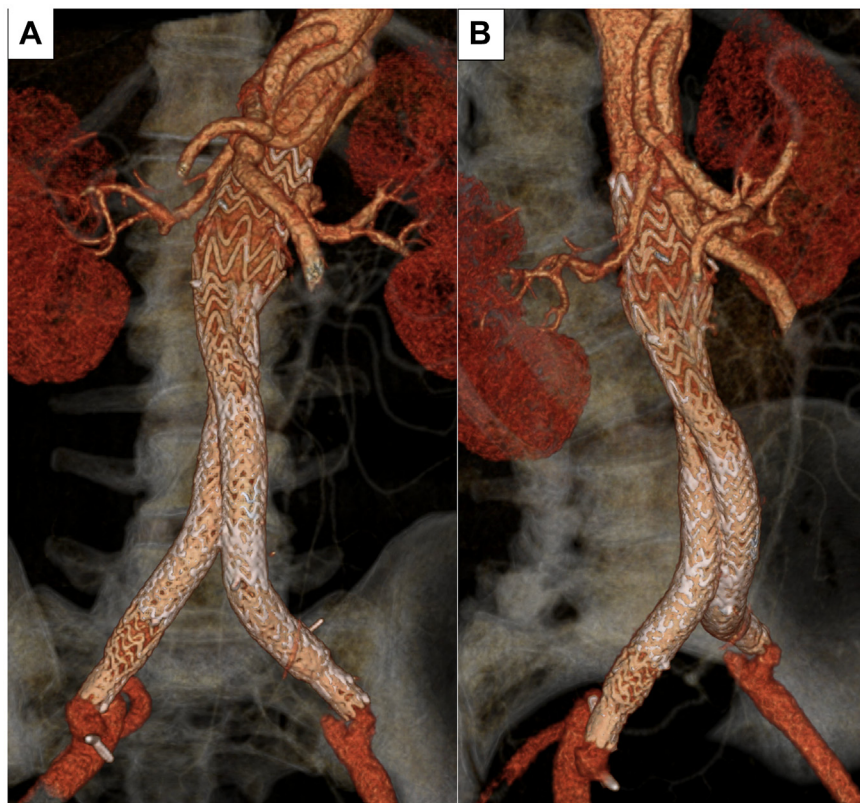
aortic repair needs to be dissected and controlled for proximal control during subsequent open repair. In the aortic arch and descending thoracic aorta, this challenge can be addressed by applying a hybrid frozen elephant trunk (FET) technique. This technique involves deploying a stent graft into the replaced aortic arch, which then serves as the site for proximal anastomosis during subsequent thoracoabdominal aortic reconstructions distally. In this case, our configuration applied the FET principle by landing the stent graft within prosthetic material both proximally and distally, effectively eliminating interaction between the stent grafts and native vessels along the EVAR seal zones. Additionally, this application circumvented the need for surgical dissection in redo fields for the proximal anastomosis, which would have been difficult in a patient with multiple aortic reoperations.

Previous studies report reintervention rates after EVAR in patients with MFS ranging from 11% to 55%.<sup>3,17,21</sup> Patients with MFS have higher rates of reintervention owing to continued degeneration of the aorta at landing zones and friable aortic tissue vulnerable to stent graft-induced new entry tears.<sup>19</sup> The incidence of these complications and patients at risk for reintervention after endovascular repair are not well-established. However, several studies

report that patients with chronic dissection are more likely to require reintervention, which may be important to note when considering EVAR.<sup>3</sup> Despite controversy surrounding the use of stent grafting in patients with MFS, endovascular therapies may be well-suited for a subset of patients, particularly those with prior open repairs where proximal and distal landing zones are within prosthetic grafts.<sup>20,22</sup> This hybrid approach has shown favorable results, including a decreased risk of retrograde dissection or endoleaks at the involved seal zones when the stent graft was landed proximally into a preexisting graft.<sup>16,23</sup> Our patient's 2-year follow-up has shown promising results with sac regression and no evidence of further aneurysmal degeneration. Continued surveillance is planned to assess long-term durability.

This case emphasizes the need for patient-specific strategies in managing MFS-related aortic complications. Moreover, comparative data with traditional open and total endovascular repair are needed to draw conclusions on optimal technique for treating patients with MFS. In this case, the patient's previous open thoracoabdominal replacement left adequate space below the renal branch graft, enabling a secure proximal seal zone for EVAR. This allowed us to avoid surgical dissection around the juxtarenal aorta.





**Fig 3.** Computed tomography imaging study at the 2-year follow-up demonstrating successful aneurysm exclusion with the Gore Excluder endoprosthesis in coronal view (A) and sagittal view (B).

## CONCLUSIONS

The principles of the FET hybrid approach through EVAR with concurrent bilateral common iliac artery interposition grafts can be used in patients with MFS with continued AAA degeneration. This technique with limited open replacement of EVAR seal zones may be a viable option in patients with genetic aortopathies and complex prior open aortic repairs.

## DISCLOSURES

S.M.H. is a consultant for W. L.Gore & Associates, Cook Medical, and Terumo Aortic, and a member of the scientific advisory board for W. L. Gore & Associates, Cook Medical, Terumo Aortic, VizAI, and Vestek.

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