Endovascular fenestration and stenting for renovisceral malperfusion in a pediatric patient with type II Loeys-Dietz syndrome

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ABSTRACT

A 16-year-old girl with Loeys-Dietz syndrome presented with an acute, complicated type B aortic dissection (AD) with mesenteric and right renal malperfusion owing to a dynamic obstruction. The anatomy of her AD and her genetic aortography were suboptimal for thoracic endovascular aortic repair. Given the concern for anticipated late aortic degeneration and the need for open aortic repair, she underwent successful transfemoral endovascular septal fenestration with stenting of the fenestration into the superior mesenteric artery and additional stenting of the right renal artery. Her renal failure and mesenteric angina resolved, and she was discharged home. Endovascular fenestration provides an elegant solution for AD-associated dynamic malperfusion of aortic branch vessels without compromising future open aortic repairs. (J Vasc Surg Cases Innov Tech 2024;10:101514.)

Keywords: Endovascular fenestration; Aortic dissection; Renovisceral malperfusion; Connective tissue disorder; Loeys-Dietz syndrome

CASE REPORT

A 16-year-old girl with type II Loeys-Dietz syndrome (LDS; pathogenic variant in TGFβR2, exon 7, c.1583 G>A, p.Arg528His) presented with an acute type B aortic dissection (TBAD) and right renal malperfusion. She had a history of vascular and skeletal manifestations of LDS including aortic root dilation (3.6 cm) and spondyloptosis of the lumbar spine requiring fusion. Vitals were notable for a heart rate in the 70s and relative hypertension with a systolic blood pressure (SBP) of 120 to 130 mm Hg, compared with her baseline of 90 to 100 mm Hg. Her weight and height were 52.6 kg and 160 cm, respectively, for a body mass index of 20.5 kg/m^2. Her vascular examination was consistent with symmetric upper extremity pulses and diminished femoral pulses with pedal signals. On examination, there was no abdominal tenderness or distension, and she had robust bilateral hip flexion. She had a white blood cell count of 6000/mm^3, hemoglobin 12.1 g/dL, hematocrit 38.5%, creatinine 0.7 mg/dL (baseline 0.4 mg/dL), and lactate 1.3 mmol/L. Her TBAD involved an entry tear at the level of the left subclavian artery in the setting of a bovine arch variant (Fig 1, *A*); the dissection extended through the descending thoracic aorta (DTA) into the abdominal aorta and terminated in the proximal common iliac arteries. The true lumen (TL) gave rise to the celiac artery, superior mesenteric artery (SMA), and right renal artery (RRA) and was significantly compressed in the abdominal aorta (Fig 1, *B*) despite a fenestration at the level of the SMA and RRA. Furthermore, there was a dynamic obstruction of the SMA (Fig 1, *B*) and RRA (Fig 1, *C*). The left renal artery arose from the false lumen. The DTA total aortic diameter was 33 mm proximally and 22 to 23 mm distally.

The TBAD was initially managed with anti-impulse control, and her back pain resolved. However, she developed transient, postprandial mesenteric angina with liquid intake, consistent with intestinal malperfusion. Additionally, her acute kidney injury worsened, evidenced by a creatinine of 2.0 mg/dL, resulting in hypervolemia and acute hypoxic respiratory failure requiring nasal cannula. As a result, operative intervention was warranted. Given a proximal landing zone requiring cervical debranching and her connective tissue disorder (CTD), including a particularly virulent subtype of LDS, with an increased risk of a retrograde type A AD, she was not an ideal candidate for thoracic endovascular aortic repair (TEVAR), and open aortic replacement was high risk given the acute nature of her dissection. Endovascular septal fenestration was chosen to relieve the dissection-associated dynamic malperfusion of the SMA and RRA as it would not compromise anticipated open aortic repair (OAR).

After a diagnostic aortogram confirmed severe TL compression (Fig 2), she underwent transfemoral endovascular fenestration of the paravisceral AD septum just opposite the SMA with the Rösch-Uchida transseptal needle (Cook Medical, Bloomington, IN) in the TL under intravascular ultrasound (IVUS) guidance in the FL (Fig 3, *A* and *B*). Over a transfemoral wire crossing from the TL to the FL, septal angioplasty was serially performed with a noncompliant balloon (Fig 4, *A*), up to 18 mm. The SMA was cannulated through this targeted but large fenestration, and

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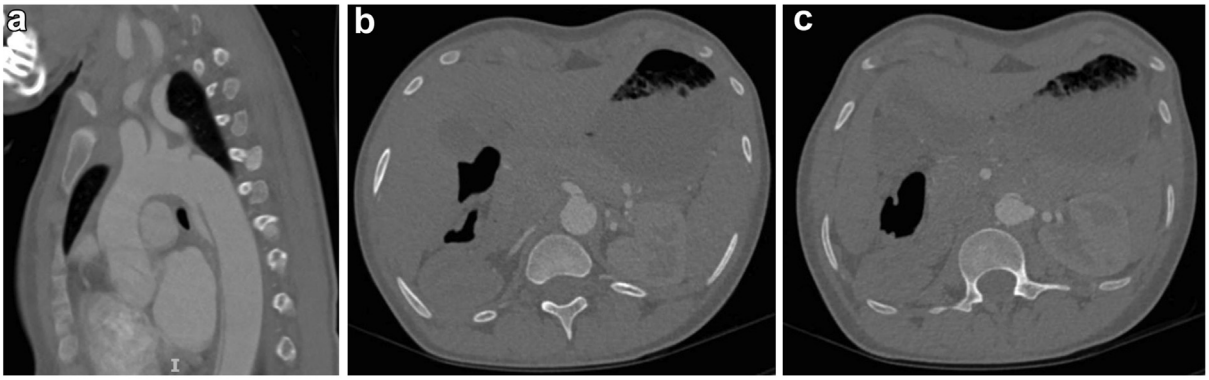

Fig 1. Sagittal **(A)** and axial **(B, C)** images of a computed tomography (*CT*) angiogram of the chest, abdomen and pelvis demonstrating an acute type B aortic dissection (*TBAD*) with an entry tear at the level of the left subclavian artery in the setting of a bovine arch variant **(A)**. The dissection extends through the thoracoabdominal aorta with an obliterated true lumen (*TL*) and a dynamic obstruction of the superior mesenteric artery (SMA) **(B)** and RRA **(C)**, which both arise from the TL.

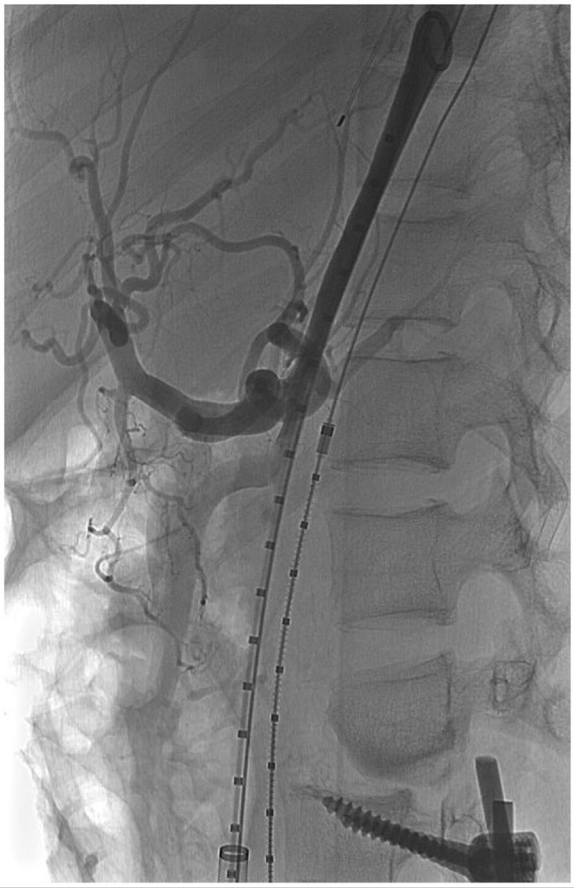

Fig 2. Lateral diagnostic aortogram demonstrating true lumen (*TL*) collapse.

an 8 mm × 40 mm self-expandable bare metal stent (BMS) from the FL into the SMA was deployed, stabilizing this fenestration (Fig 4, *B* and *C*) and equalizing the previously measured pressure gradient between the TL (SBP 50 mm Hg) and FL (SBP 90 mm Hg). The RRA was stented from the TL with a 7 mm × 40 mm self-expandable BMS. Completion aortogram (Fig 4, *C*) demonstrated resolution of dynamic obstruction of the SMA and RRA with significantly improved flow in these vessels. Her postoperative course was complicated by delirium and ileus. Her renal failure and mesenteric angina resolved. She was discharged home on postoperative day 23 on oral antihypertensives.

On surveillance imaging, her proximal DTA degenerated to 56 mm (Fig 5, *A*) over the following 5 months owing to continued FL flow, and the stenting of the septal fenestration (Fig 5, *B*) remained patent. She underwent open type 2 thoracoabdominal aortic repair and aortorenomesenteric bypasses with cardiopulmonary bypass, deep hypothermic circulatory arrest, and cerebrospinal fluid drainage. Cardiopulmonary bypass avoided aortic arch cross-clamping in a fragile aorta at risk of retrograde AD. The stented fenestration (Fig 5, *C*) was patent at the time of surgery, and the SMA and RRA stents were partially explanted after septal excision. Her postoperative course was complicated by an upper extremity deep venous thrombosis requiring anticoagulation and pneumonia. The patient's legal guardian provided written informed consent for patient information and images to be published.

DISCUSSION

AD-related malperfusion syndrome can manifest as renovisceral, extremity, and/or spinal cord ischemia.[1] Therapeutic options historically were limited to OAR, including replacement[1] and fenestration.[2,3] The high morbidity and mortality associated with emergent OAR led to the development of endovascular means to relieve AD-related distal vessel ischemia.[4,5] Endovascular techniques focus on TL expansion by coverage of the entry tear with a stent graft[6,7] to depressurize the FL or by fenestration of the intimal flap[8] to equalize the pressure gradient between the TL and FL. More than a decade ago, the INvestigation of STEnt grafts in Aortic Dissection (INSTEAD) trial demonstrated the long-term survival benefit of TEVAR in uncomplicated TBAD.[7] TEVAR has

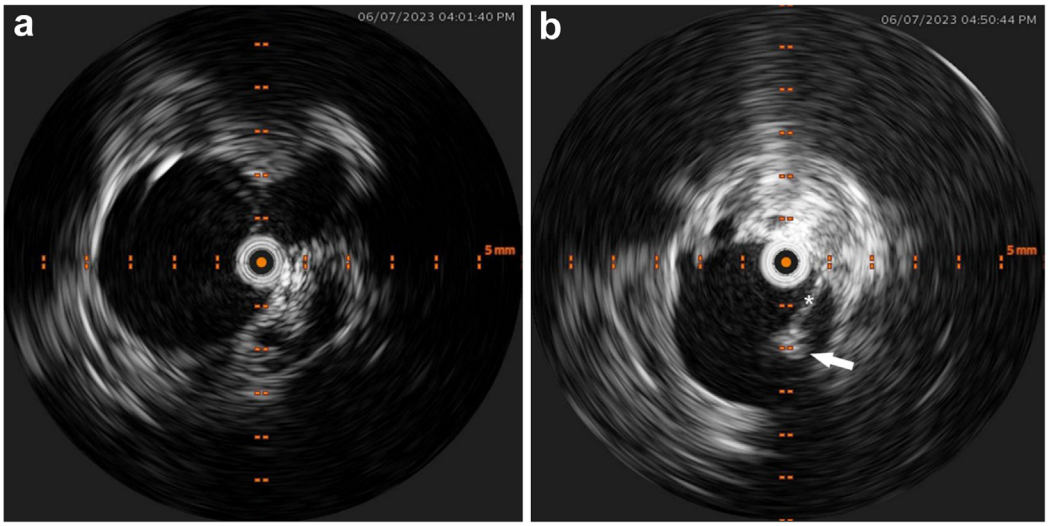

Fig 3. Intravascular ultrasound (*IVUS*) images confirming compressed true lumen (*TL*) at the level of the superior mesenteric artery (SMA) **(A)** followed by a tented dissection flap (*) by Rösch-Uchida needle (*arrow*) that is rotated and angled posteriorly **(B)**.

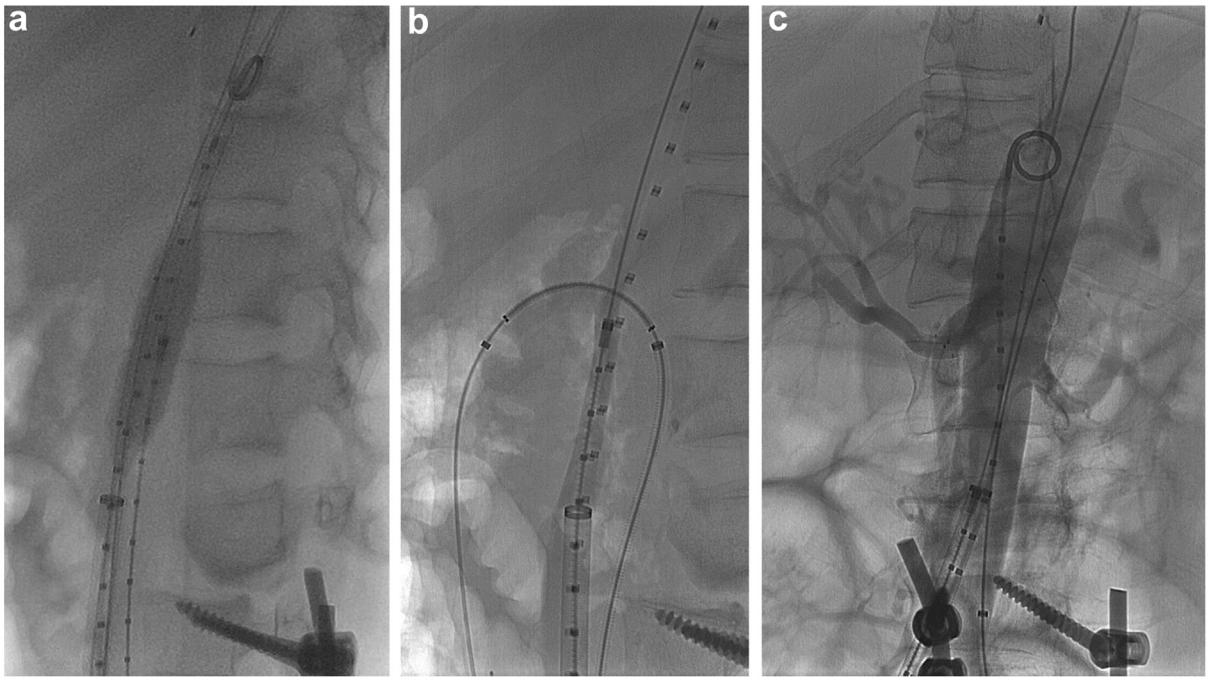

Fig 4. Septal angioplasty with an 18-mm, noncompliant balloon **(A)**. The superior mesenteric artery (*SMA*) was cannulated from the FL and stented with an 8 mm × 40 mm self-expandable bare metal stent (*BMS*), stabilizing this fenestration **(B, C)**. Completion aortogram **(C)** demonstrated resolution of dynamic obstruction of the SMA and right renal artery (*RRA*) with significantly improved flow in these vessels.

also become the mainstay of therapy for acute aortic syndromes, including complicated, acute TBAD.[9]

Endovascular septal fenestration can be used as a primary therapy when aortic stent grafting is not feasible[8,10] or as an adjunct to TEVAR[9,10] in the setting of persistent malperfusion. It is typically performed with a transseptal needle[8] under IVUS guidance[11] or a bare wire,[12,13] followed by longitudinal septotomy[12,13] or balloon angioplasty with or without adjunctive stenting.[8,10] Septal fenestrations were classically described in the DTA and infrarenal abdominal aorta,[8] but later expanded to the abdominal aorta.[10] Adjunctive stenting of the aortic TL,[10] the branch vessel,[8,10] and, less commonly, the septal fenestration[8] optimizes TL flow.

Device-associated complications raise concerns about the usefulness of TEVAR in patients with genetic

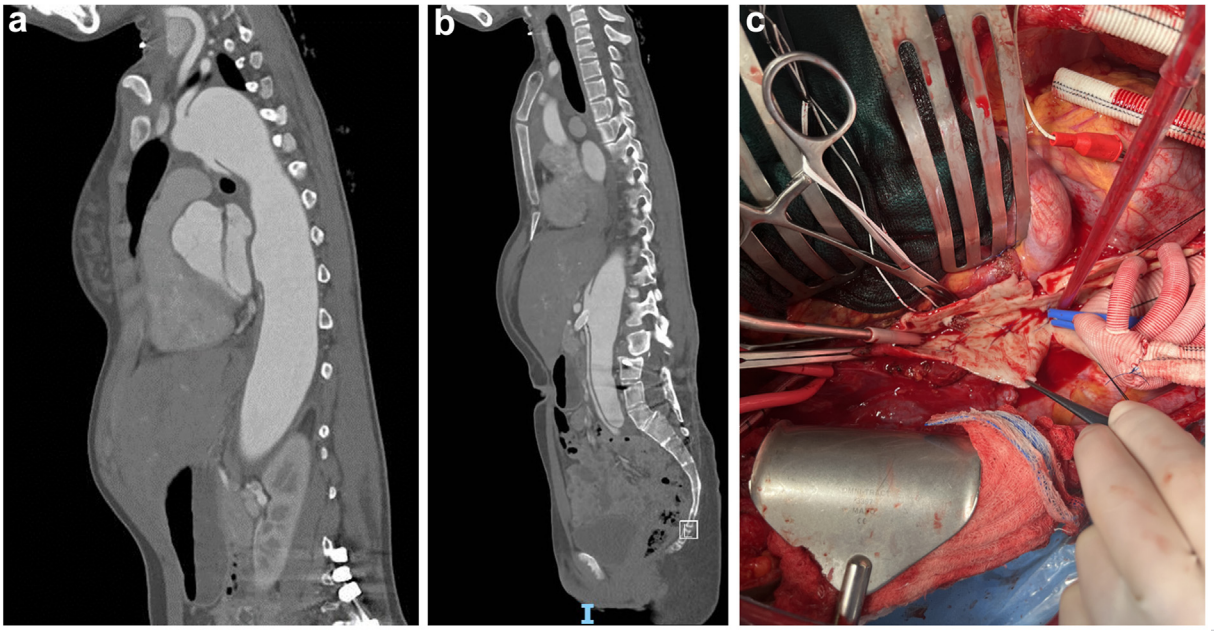

Fig 5. Sagittal **(A**, **B)** images of a computed tomography angiogram of the chest, abdomen, and pelvis demonstrating aneurysmal degeneration of the thoracoabdominal aorta, most pronounced in the proximal descending thoracic aorta (*DTA*) **(A)**. The true lumen (*TL*) remains obliterated in the DTA but equalizes in the abdominal aorta owing to the stented fenestration from the false lumen (*FL*) into the superior mesenteric artery (*SMA*) **(B)**. After completion of the proximal aortic anastomosis and sequential aortic replacement of the DTA, a longitudinal aortotomy of the visceral aorta is performed, revealing a dominant FL with a patent stented fenestration **(C)**.

aortopathies. This concern is largely based on the chronic radial force from the stent graft on the fragile aorta.[14] In one study evaluating long-term outcomes of TEVAR for TBAD, stent graft-induced new entry (SINE) occurred in one-third of Marfan patients compared with 3% of non-Marfan patients with a mean follow-up of 11 ± 16 months.[15] As a result, consensus guidelines[16] continue to recommend open repair for genetic aortopathies. OAR for acute TBAD continues to be associated with high in-hospital mortality of ≤30%, even in the contemporary literature.[4] To mitigate the perioperative morbidity of open surgery, TEVAR has become increasingly popular for acute TBAD.[17] The EVICTUS study challenged the gold standard of OAR in CTD with endovascular aortic repair, demonstrating high rates of procedural success and 5-year survival coupled with low perioperative mortality.[18] More than 40% of this cohort underwent emergent TEVAR with an indication of malperfusion syndrome in nearly 10% of patients.[18]

Pediatric patients with CTD represent a unique cohort in which TEVAR for AD-related malperfusion syndrome is often suboptimal. As discussed previously, OAR in acute TBAD results in significant risk,[4] and TEVAR is not feasible at times owing to the proximity of the entry tear to the arch vessels and risk of SINE in patients with CTD. Endovascular septal fenestration and stenting have a particularly beneficial role in this population, which ultimately requires future OAR. It mitigates the risks inherent to emergent OAR, namely, tissue friability and ensuing coagulopathy. Furthermore, it avoids the risks associated with TEVAR, including SINE, stent graft infection in setting of sepsis, and cervical debranching when adequate proximal seal requires partial great vessel coverage.

The present case had several salient points, highlighting the challenges of decision-making in this clinical scenario. First, this patient was not a candidate for TEVAR given her virulent subtype of LDS and entry tear at the level of the left subclavian artery in the setting of a bovine arch variant. Second, the Rösch-Uchida transseptal needle created a fenestration from the TL to the FL. IVUS guidance in the FL[11] was essential to observe rotation of the needle to the center of the septum and at the level of malperfused target vessel. Intestinal malperfusion dictated fenestration at the level of the SMA. Third, after septal angioplasty, adjunctive self-expandable BMS stenting of the fenestration from FL into the TL of the SMA was essential to maintain patency of the fenestration and resolve the dynamic SMA obstruction. Stenting through the septal fenestration into a target vessel is rare, and Slonim et al[8] described this adjunct in the iliac system for lower extremity malperfusion. In addition, the resolution of RRA malperfusion after branch vessel stenting represented the more typical adjunctive stenting

described in the literature.[8,10] Endovascular septal fenestration provides an elegant solution for AD-associated static and/or dynamic malperfusion of aortic branch vessels when endografting of the primary entry tear is not feasible.

CONCLUSIONS

Endovascular septal fenestration with stenting is a useful technique for AD-associated malperfusion syndromes that does not compromise options for future repair. It is particularly useful as a temporizing solution in pediatric patients with CTD-associated AD, where open repair for aneurysmal degeneration remains the standard of care.

DISCLOSURES

None.

REFERENCES

1. Conrad MF. Aortic dissection: epidemiology, pathophysiology, clinical presentation, and medical and surgical management. In: Sidawy AN, Perler BA, eds. *Rutherford's vascular surgery and endovascular therapy*. 10th ed. Philadelphia: Elsevier; 2023: 1098–1111.
2. Webb TH, Williams GM. Abdominal aortic tailoring for renal, visceral, and lower extremity malperfusion resulting from acute aortic dissection. *J Vasc Surg*. 1997;26:474–480.
3. Cambria RP. Surgical treatment of complicated distal aortic dissection. *Semin Vasc Surg*. 2002;15:97–107.
4. Trimarchi S, Nienaber CA, Rampoldi V, et al. Role and results of surgery in acute type B aortic dissection: insights from the International Registry of Acute Aortic Dissection (IRAD). *Circulation*. 2006;114:I357–I364.
5. Pruitt EY, Scali ST, Arnaoutakis DJ, et al. Complicated acute type B aortic dissection: update on management and results. *J Cardiovasc Surg*. 2020;61:697–707.
6. Tjaden BL, Sandhu H, Miller C, et al. Outcomes from the Gore Global Registry for Endovascular Aortic Treatment in patients undergoing thoracic endovascular aortic repair for type B dissection. *J Vasc Surg*. 2018;68:1314–1323.
7. Nienaber CA, Kische S, Rousseau H, et al. Endovascular repair of type B aortic dissection: long-term results of the randomized investigation of stent grafts in aortic dissection trial. *Circ Cardiovasc Interv*. 2013;6:407–416.
8. Slonim SM, Miller DC, Mitchell RS, Semba CP, Razavi MK, Dake MD. Percutaneous balloon fenestration and stenting for life-threatening ischemic complications in patients with acute aortic dissection. *J Thorac Cardiovasc Surg*. 1999;117:1118–1126.
9. Lombardi JV, Hughes GC, Appoo JJ, et al. Society for vascular surgery (SVS) and society of thoracic surgeons (STS) reporting standards for type B aortic dissections. *J Vasc Surg*. 2020;71: 723–747.
10. Norton EL, Wiliams DM, Kim KM, et al. Management of acute type B aortic dissection with malperfusion via endovascular fenestration/ stenting. *J Thorac Cardiovasc Surg*. 2020;160:1151–1161.
11. Chavan A, Hausmann D, Dresler C, et al. Intravascular ultrasound–guided percutaneous fenestration of the intimal flap in the dissected aorta. *Circulation*. 1997;96:2124–2127.
12. Barshes NR, Gravereaux EC, Semel M, Bolman RM 3rd, Belkin M. Endovascular longitudinal fenestration and stent graft placement for treatment of aneurysms developing after chronic type B aortic dissection. *J Vasc Surg*. 2015;61:1366–1369.
13. Midulla M, Renaud A, Martinelli T, et al. Endovascular fenestration in aortic dissection with acute malperfusion syndrome: immediate and late follow-up. *J Thorac Cardiovasc Surg*. 2011;142:66–72.
14. Sorber RA, Black JH. Aneurysms caused by connective tissue abnormalities. In: Sidawy AN, Perler BA, eds. *Rutherford's vascular surgery and endovascular therapy*. 10th ed. Philadelphia: Elsevier; 2023:1098–1111.
15. Dong Z, Fu W, Wang Y, et al. Stent graft-induced new entry after endovascular repair for Stanford type B aortic dissection. *J Vasc Surg*. 2010;52:1450–1458.
16. Isselbacher EM, Preventza O, Black JH, et al. 2022 ACC/AHA guideline for the diagnosis and management of aortic disease: a report of the American heart association/American college of cardiology joint committee on clinical practice guidelines. *Circulation*. 2022;146: e334–e482.
17. Botta L, Russo V, La Palombara C, Rosati M, Di Bartolomeo R, Fattori R. Stent graft repair of descending aortic dissection in patients with Marfan syndrome: an effective alternative to open reoperation? *J Thorac Cardiovasc Surg*. 2009;138:1108–1114.
18. Olsson KW, Mani K, Burdess A, et al. Outcomes after endovascular aortic intervention in patients with connective tissue disease. *JAMA Surg*. 2023;158:832–839.

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