

Open repair of an aortic aneurysm in a patient with Loeys-Dietz syndrome using Gore hybrid vascular branch grafts

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A 44-year-old woman with Loeys-Dietz syndrome (transforming growth factor- β [*TGFBR2*] gene mutation) presented with a retrograde type B dissection. She developed rapid expansion of the thoracoabdominal aortic false lumen aneurysm. The patient was successfully treated with open thoracoabdominal repair using Gore Hybrid Vascular Grafts (W. L. Gore and Assoc, Flagstaff, Ariz) for revascularization of the celiac trunk, the superior mesenteric artery, and both renal arteries. Follow-up imaging documented patency for all visceral branches. The described off-label use for sutureless revascularization might be a fast, simple, and reliable solution for revascularization during open thoracoabdominal repair. Furthermore, anastomotic aneurysm in patients with connective tissue disease might be prevented by sutureless anastomosis. (*J Vasc Surg Cases* 2015;1:69-72.)

Surgical treatment of aneurysms in patients with genetic aortic syndromes (GAS) is challenging. We present a patient with GAS who was successfully treated with open thoracoabdominal repair using Gore Hybrid Vascular Grafts (GHVGs; W. L. Gore and Assoc, Flagstaff, Ariz) for revascularization of the celiac trunk, the superior mesenteric artery, and both renal arteries.

CASE REPORT

A 44-year-old woman with suspected GAS and retrograde type B aortic dissection after a ruptured infrarenal aortic aneurysm developed enlarged false lumen aneurysm of the thoracoabdominal aorta. Furthermore, a penetrating aortic ulcer (PAU) had developed at the junction of aortic arch and descending aorta on the minor curvature as well as additional aneurysms of two intercostal arteries and both internal mammary arteries, with a maximal diameter of 19 mm (Fig 1, A).

In the referring hospital, the ruptured infrarenal aortic aneurysm had been replaced by a 16-mm Dacron (DuPont, Wilmington, Del) tube graft in an emergency operation under intermittent manual and medicamentous cardiopulmonary resuscitation due to hemodynamic instability. A periprocedural retrograde type B dissection occurred postoperatively.

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The patient was then transferred to our institution and was initially administered conservative antihypertensive medication. Because of the complicated vascular phenotype, we ordered testing for GAS.

During the following 6 weeks, the patient experienced recurrent pain and progressing false lumen expansion (from 47 mm to 53 mm) and finally required immediate intervention (Fig 1, B). Owing to the high rupture risk, treatment could not await test results for GAS.

Our interdisciplinary vascular board, consisting of specialists for open and endovascular surgery, cardiac surgery, angiology, cardiology, and connective tissue disease, including GAS, decided to proceed immediately by open thoracoabdominal aortic repair with reimplantation of the visceral branches. The time required to produce and deliver an individualized four-fenestrated endograft would have taken about 2 months, which was considered too long to wait for the repair of this rapidly progressing aneurysm. Moreover, because the patient was young and otherwise healthy, no significant concern against open repair was raised. In a second procedure, the previously diagnosed PAU and the mammary artery aneurysms would be repaired by thoracic endovascular aortic repair and coil embolization, respectively.

Patients with GAS are known for their high risk to develop postoperative anastomosis aneurysms. We therefore favored the implantation of GHVGs over a traditional suture-mediated anastomosis to the visceral and renal arteries. The treatment options were discussed by our interdisciplinary vascular board, and the patient gave informed consent for the off-label use of GHVG implantation and for the publication of her data.

Surgical technique. At present, the GHVG is generally used to create arteriovenous access but is also used for lower extremity revascularization and for debranching purposes during cerebral, renal, and mesenteric revascularization.¹⁻⁴ We recently expanded its use to vertebral revascularization in a patient with GAS.⁵

GHVG placement does not require sutures or cross-clamping, thereby minimizing trauma and avoiding a time-consuming anastomosis.¹⁻⁵ It is composed of a tubular, heparin-coated, expanded

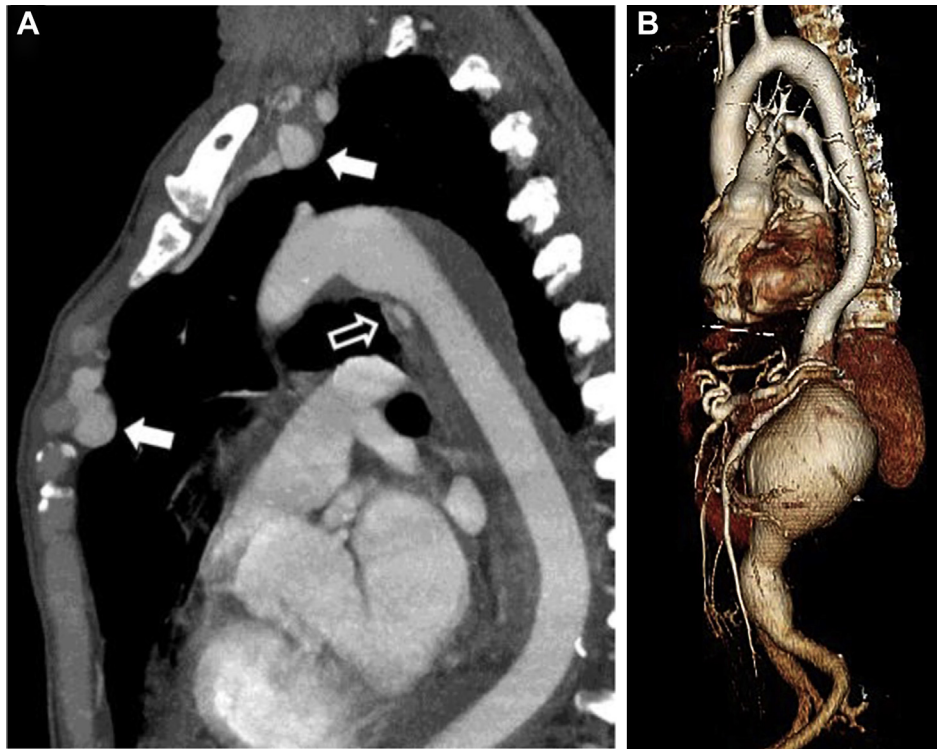


Fig 1. **A**, Preoperative multiplane reconstruction in the computed tomography scan shows a penetrating aortic ulcer (PAU) at the junction of aortic arch and the descending aorta on the minor curvature (*blank arrow*) as well as additional aneurysms of two intercostal arteries and both internal mammary arteries, with a maximal diameter of 19 mm (*filled arrows*). **B**, Preoperative three-dimensional volume-rendered computed tomography scan of the supraceliac aorta shows rapidly progressing false lumen expansion with aneurysmatic dilatation. The PAU and the intercostal and mammary artery aneurysms are not visible in this reconstructed image.

polytetrafluoroethylene vascular graft with a nitinol stent reinforcement at one end that allows an easy and atraumatic insertion and deployment of the graft into the target vessel. Here, we used GHVGs in a patient with suspected GAS, and therefore with increased risk for aneurysmal formations, for sutureless revascularization of the celiac trunk, the inferior mesenteric artery, and both renal arteries during open thoracoabdominal aortic repair to avoid clamping of the visceral and renal arteries.

After placement of a lumbar drain, induction of general anesthesia, and installation of perioperative monitoring for somatosensory evoked potentials (SEPs), we exposed the descending and retroperitoneal juxtarenal aorta by thoracolaparotomy via Crawford access through the sixth intercostal space. We dissected the proximal part of the infrarenal Dacron tube graft and controlled the celiac trunk, superior mesenteric artery, and both renal arteries with elastic vessel loops.

The left femoral artery and vein were cannulated, and we started a partial extracorporeal bypass with moderate cooling to 32°C. We connected a four-branched tubing system, including catheters with balloon-inflatable tips, to the extracorporeal bypass for selective perfusion of the celiac trunk, superior mesenteric artery, and both renal arteries.

After proximal and distal cross-clamping and transection of the aorta, we maintained distal infrarenal aortic perfusion at a mean pressure of at least 70 mm Hg. Flow-controlled and

pressure-controlled selective perfusion of the visceral arteries was performed under continuous SEP monitoring.

We completed the proximal anastomosis of the descending aorta using a 16-mm tube graft in an end-to-end fashion. Because of the fragile consistency of the aortic wall, the suture was stabilized by Teflon (DuPont) felt. After limited surgical dissection of the visceral arteries, we placed the reinforced section of the GHVG sequentially about 4 cm into each of the four visceral target vessels via the native aortic entry. Balloon dilatation was not performed due to suspected GAS and adequate oversizing of the GHVG. We cannulated each of the GHVGs sequentially with the perfusion catheter, inflated the balloon for fixation, and started flow-controlled selective perfusion. We cut the Dacron tube graft to length and performed a distal anastomosis. Thereafter, we removed the clamps of the Dacron graft.

Under continuous selective perfusion, we cut the GHVGs to length and anastomosed them subsequently terminolateral to the tube graft under partial cross-clamping with 6-0 Prolene (Ethicon, Somerville, NJ; Fig 2). SEP readings were stable throughout the entire procedure. After rewarming and decannulation, intraoperative angiography confirmed graft patency of all grafts.

Postoperative follow-up. Postoperatively, we monitored the patient for 24 hours in our intensive care unit. She recovered well, no postoperative complications occurred, and she was discharged



Fig 2. Terminolateral anastomosis of the four Gore Hybrid Vascular Grafts (GHVGs; W. L. Gore and Assoc, Flagstaff, Ariz) to the tube graft under partial cross-clamping (6-0 Prolene; Ethicon, Somerville, NJ).



Fig 3. A 3-month follow-up three-dimensional volume-rendered computed tomography scan confirms patent visceral grafts without any deterioration of renal function and successful exclusion of the aneurysm.

on postoperative day 9. Antiplatelet therapy was administered using acetylsalicylic acid (100 mg) without additional anticoagulation.

Three months after testing for GAS was ordered, our clinic for Marfan and GAS patients confirmed a mutation of the transforming growth factor- β receptor 2 gene (*TGFBR2*) indicative for Loeys-Dietz syndrome.

Follow-up ultrasound examinations at 1 month and computed tomography at 3 months revealed patent visceral grafts, without any deterioration in renal function, and successful exclusion of the aneurysm (Fig 3). Coiling of the internal mammary artery

aneurysms will complete the treatment. At present, the previously described PAU at the junction of aortic arch and descending aorta on the minor curvature is not perfused and will therefore be monitored only.

DISCUSSION

Loeys-Dietz syndrome is an autosomal-dominant aortic aneurysm syndrome caused by mutations in the *TGFBI* and *TGFBI2* genes.⁶ It is characterized by facial dysmorphism, cleft palate, and aggressive vascular pathologies, including dilatation of the aorta, blood vessel tortuosity, and high risk for aortic dissection and early rupture of aneurysms.⁶

Treatment of patients with connective tissue disease is often challenging because multifocal aneurysmal formations require multiple surgical interventions.^{7,8} In addition, during open surgery, the risk of anastomotic aneurysms frequently requires reoperations.⁴ Minimal operative exposure and trauma is essential to minimize perioperative risk in these patients.

The use of complex endovascular aortic repair in post-dissection aneurysms with fenestrated and branched endografts is increasingly in development but currently implemented by a few centers only.^{9,10} Therefore, establishing novel, less-invasive therapies for these patients seems mandatory.¹¹ The technique for sutureless revascularization of the visceral and renal arteries as an off-label use of otherwise approved hybrid vascular grafts seems a promising treatment option for these high-risk patients.

The distinct advantages of the GHVG requiring only one central suture anastomosis over conventional open surgery approaches include decreased ischemia and surgery time, avoidance of cross-clamping, less invasiveness and trauma, and relatively easy access to surgically challenging regions. Particularly during mesenteric and renal revascularization, a short ischemic time is of major importance to minimize renal dysfunction and prevent visceral reperfusion injury.² Furthermore, the sutureless outflow anastomosis might prevent future formation of anastomotic aneurysms in patients with GAS.⁵

CONCLUSIONS

Use of the GVHG for visceral debranching is a fast, simple, and reliable technique that may be useful especially in challenging patients with GAS.

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