Masters of Cardiothoracic Surgery

Hybrid repair of a thoracoabdominal aortic aneurysm in female patient with Loeys-Dietz syndrome

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Open thoracoabdominal aortic repair has reportedly good mid- and long-term results, can be performed with reasonable morbidity and mortality rates in high-volume centers and, therefore, represents a durable treatment for patients with connective tissue disease (CTD) (1,2).

Despite the use of contemporary strategies for organ protection and an experienced surgical and anesthetic team, these procedures remain challenging. To reduce surgical trauma and the risk for spinal cord ischemia (SCI), an individualized approach with staged procedures, including endovascular strategies, can benefit patients and should be considered, depending on the anatomical features and the operative risk (1,3). Particularly when safe landing zones in prosthetic material are available and the operative risk is deemed high, an individualized, combined endovascular/open approach can also be considered in patients with CTD (1,3). Patients pretreated with thoracic endovascular aortic repair (TEVAR) and/or frozen elephant trunk (FET) might be good candidates for thoracoabdominal aortic aneurysm (TAAA) hybrid repair using the Thoracoflo® device, as the stent grafts serve as stable landing areas (4,5).

Clinical vignette

A 46-year-old female with Loeys-Dietz syndrome type 2, mutation *TGFBR2*-gene [c.1319A>C (p.Glu440Ala)], who developed acute type B dissection with rapid progressive (within 4 months) TAAA type II (8.5 cm) and was selected for staged repair. Due to the ascending aneurysm (5.0 cm), she was selected for supracoronary ascending and arch repair using the Thoraflex Hybrid® graft. Due to previous radiation treatment for breast cancer, including mastectomy, and concern for poor wound healing, thoracotomy was avoided, and the patient was selected for TEVAR extension, followed by hybrid repair using the Thoracoflo® hybrid graft.

Surgical technique

First stage

The first step was to create a safe proximal landing zone by aortic arch repair using the FET technique. Treatment of the descending thoracic aorta was completed 5 months later by TEVAR extension (placed about 3 cm above the celiac trunk ostium) to create a landing zone for the Thoracoflo® hybrid device, which was implanted 6 weeks thereafter, due to manufacturing of a customized Thoracoflo® graft. The device was deployed via transfemoral access. During the same procedure, coil embolization of the intercostal arteries at the level of T10 was performed to avoid backbleeding during the procedure and to reduce the risk of SCI by staged repair.

Second stage: device implantation

The patient is placed in the supine position and median laparotomy is performed. The abdominal aorta, including the celiac trunk, superior mesenteric artery, left renal and both iliac arteries, are exposed via right visceral rotation in a secondary retroperitoneal approach. The Thoracoflo® graft consists of a proximal stent graft with ring stents connected to a distal multibranched, gelatin-sealed polyester prosthesis via a collar (4,5). Before implantation, all side branches, except the left iliac side branch, are ligated. The left iliac branch is temporarily anastomosed end-to-side to one common iliac artery to be used for retrograde visceral perfusion (SPIDER technique) and clamped until the stent graft is deployed and the graft is deaired.

After direct puncture of the access site (aorta or visceral artery ostium), the stent-grafted section is introduced over the guidewire, which is externalized through the sidehole in the nose cone of the introducer. Then the guidewire is extracted, the peel-away sheath retracted, and the proximal fixation is released. To avoid bleeding and collapse of the stent-grafted section during deployment, cardiac output reduction is recommended by in-flow occlusion or rapid pacing.

The delivery system is carefully retracted until the nose cone is in the access branch, the access branch is clamped, the splitter is removed, and the handle is completely removed. After de-airing, cardiac output reduction is released, and the clamp on the iliac side branch is removed to establish retrograde visceral and antegrade iliac pulsatile blood-flow (SPIDER technique). Visceral arteries and the left renal artery are sequentially attached to the corresponding branches of the graft.

The infrarenal aorta is cross-clamped and suprarenal aorta is opened up to the collar. The right renal artery can be temporarily perfused via a perfusion catheter attached to the right iliac side branch while the collar is attached to the native aorta to prevent backbleeding and distal graft migration. The right renal artery is then attached from inside the aorta and both iliac arteries are reattached.

Postoperative course

The patient tolerated the operative procedure well and was extubated the following day. The postoperative course was uneventful, and the patient was able to leave the intensive care unit on postoperative day 5. At 1-year follow-up, she showed good clinical recovery with patent graft branches and no residual aneurysm or signs of infection, as well as uncomplicated wound healing without herniation.

Comments

Surgical and endovascular treatment for extensive aortic disease in patients with CTD remains a challenge, especially in female patients with small access vessels. Despite this, open repair remains the gold standard, with reasonable outcomes when performed in experienced, high-volume centers.

Staged repair has become increasingly important, depending on previous aortic repair, for patients with suitable anatomy and comorbidities (1-3). It is mandatory to create safe proximal and distal landing zones to avoid the risk of reintervention. The Thoracoflo® hybrid device can serve as a new treatment option in this specific patient cohort. Thoracotomy and extracorporeal circulation can be avoided, blood loss minimized, and ischemia times and the risk of SCI can be reduced by allowing pulsatile blood flow to the collateral network. To date, three female patients with CTD have been successfully treated with this staged repair strategy.

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Footnote

Conflicts of Interest: ALE is a consultant for WL Gore, CryoLife, Edwards Lifesciences, and Terumo Aortic. The other authors have no conflicts of interest to declare.

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