

# Treatment of thoracoabdominal aortic disease in patients with connective tissue disorders



Natalia O. Glebova, MD, PhD, FACS,<sup>a</sup> Duke E. Cameron, MD, FACS,<sup>b</sup> and James H. Black III, MD, FACS,<sup>c</sup>  
*Rockville and Baltimore, Md; and Boston, Mass*

## ABSTRACT

**Objective:** Aneurysmal degeneration of the entire aorta is common in patients with connective tissue disorders (CTDs). Potential treatment options of these aneurysms include open repair and endovascular repair. Our objective herein was to review available evidence for different approaches to treatment of thoracoabdominal aortic disease in patients with CTDs.

**Methods:** We performed a systematic literature review using PubMed and referenced manuscripts on open and endovascular treatment of thoracoabdominal aortic aneurysms and dissections in patients with CTDs.

**Results:** A total of 28 studies were identified for inclusion in this review, 8 reporting on outcomes after open thoracoabdominal aortic aneurysm repair in patients with CTD, 8 on open branched graft use, and 12 on endovascular aortic repair in this population of patients. Reported outcomes were characterized by low perioperative morbidity and mortality, good branch patency, and low rate of reintervention for open repair and significant rates of endograft-related complications and substantial need for secondary endovascular interventions and open conversions for endovascular repair.

**Conclusions:** There is a lack of high-quality evidence to support any particular approach to aortic repair in patients with CTD and a dearth of comparative data between open repair and endovascular repair. There are distinct differences in the published lengths of follow-up between the two repair approaches as well as in the prevalence of their use in an acute vs elective setting. It is evident that endovascular interventions for aortic disease in patients with CTDs are associated with many device- and aorta-related complications both in the short term and in the long term. Despite the lack of level 1 evidence, open repair currently remains the standard approach to treatment of aortic disease due to CTDs. Open branched graft repair in particular is the preferred technique. Endovascular interventions may be cautiously used in patients with CTDs in selective circumstances. (*J Vasc Surg* 2018;68:1257-67.)

**Keywords:** Thoracoabdominal aortic aneurysm; Connective tissue disorder

Connective tissue disorders (CTDs) are a group of syndromes characterized by abnormalities in the integrity of connective tissues, such as the vascular wall, and leading to variably penetrant weakness of these tissues. Patients with CTDs such as Marfan, Loeys-Dietz, and vascular Ehlers-Danlos syndromes are prone to extensive cardiovascular anomalies, including aortic dissection and aneurysmal dilation of the entire thoracoabdominal aorta starting with the aortic root.<sup>1</sup> Aortic dissection is more often the underlying pathophysiologic insult

necessitating aortic repair in CTD patients, in contrast to non-CTD patients, in whom degenerative aneurysms are usually the cause for repair. The characteristic propensity to aortic dilation, especially after dissection, in patients with CTDs and the particular challenges with tissue integrity in these patients highlight the importance of determining the most appropriate aortic repair technique with the best short- and long-term outcomes.

Endovascular repair has become the dominant technique for treatment of aortic aneurysms, with open repair decreasing during the last two decades.<sup>2</sup> However, the use of endovascular techniques in patients with CTDs remains controversial as device trials uniformly excluded these patients; thus, level 1 data on the safety and efficacy of these devices in patients with CTDs are not available (similar to level 1 evidence for open thoracoabdominal aortic aneurysm [TAAA] repair). Furthermore, the long-term durability of endovascular aortic repair in patients with CTDs, who at the time of repair tend to be younger on average than patients with degenerative aneurysms, is unknown.

Nevertheless, several groups have described results of endovascular repair of aortic aneurysms in patients with CTDs, although the reported endovascular experience is not as extensive as the open aortic repair

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From the Mid-Atlantic Permanente Medical Group, Rockville<sup>a</sup>; the Division of Cardiac Surgery, Department of Surgery, Massachusetts General Hospital, Boston<sup>b</sup>; and the Division of Vascular Surgery and Endovascular Therapy, Department of Surgery, Johns Hopkins Hospital, Baltimore.<sup>c</sup>

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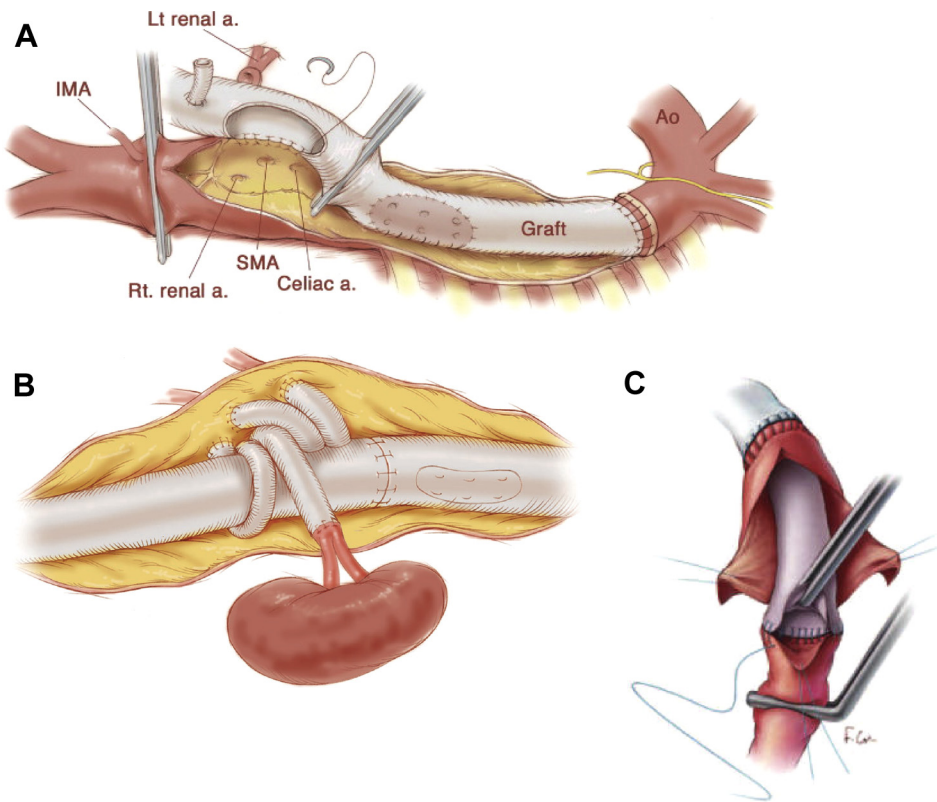
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Correspondence: James H. Black III, MD, Division of Vascular Surgery and Endovascular Therapy, Department of Surgery, Johns Hopkins Hospital, Halsted 668, 600 N Wolfe St, Baltimore, MD 21287 (e-mail: [jhblack@jhmi.edu](mailto:jhblack@jhmi.edu)).

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**Fig 1.** Technical aspects of aortic aneurysm repair. **A**, Carrell patch reconstruction during thoracoabdominal aortic aneurysm (TAAA) repair; visceral plate is reimplemented as a patch. Ao, Aorta; IMA, inferior mesenteric artery; SMA, superior mesenteric artery. (From Black JH 3rd. Technique for repair of suprarenal and thoracoabdominal aortic aneurysms. *J Vasc Surg* 2009;50:936-41.) **B**, Branched graft reconstruction during TAAA repair; visceral arteries are reimplemented individually. (From Glebova NO, Hicks CW, Alam R, Lue J, Propper BW, Black JH. Technical aspects of branched graft aortic reconstruction in patients with connective tissue disorders. *J Vasc Surg* 2016;64:520-5.) **C**, Elephant trunk reconstruction during aortic arch aneurysm repair. (Reprinted with permission from the American Journal of Roentgenology. From Johnson PT, Corl FM, Black JH, Fishman EK. The elephant trunk procedure for aortic aneurysm repair: an illustrated guide to surgical technique with CT correlation. *AJR Am J Roentgenol* 2011;197:W1052-9.)

experience in this population of patients. Overall, open TAAA repair in patients with CTDs is an effective operation, with a cumulative 5-year survival rate of 53% at mean follow-up of 27 months.<sup>3</sup> Whereas long-term outcomes after endovascular interventions in this population of patients are less clear, several studies describe short- and medium-term results and are reviewed here.

In this review, we have subdivided the analysis of results of open aortic repair into two separate groups because of an important detail of the open repair technique in patients with CTDs. Typically, two distinct techniques are used during open aortic repair: the Crawford inclusion technique, characterized by direct reimplantation of visceral and intercostal vessels in the aortic graft in the form of a Carrell patch; and the branched graft technique, with individual reconstructions of visceral vessels using branches of the prosthetic graft (Fig 1).<sup>4-6</sup> The branched graft technique has the advantage of obviating the development of visceral patch aneurysms that occur at a high frequency of 18% in CTD patients, but it is characterized by longer operative times and

potentially greater blood loss.<sup>7</sup> Thus, branched graft repair outcomes are summarized separately from open nonbranched repair to detect these differences.

Here, we review the published literature on open aortic repair, branched graft technique, and endovascular repair of thoracoabdominal aortic diseases in patients with CTDs. Our analysis surveys the possible approaches to aortic reconstruction in patients with CTDs and clarifies methods optimally suited to these patients.

## METHODS

**Systematic review of the literature.** We performed a systematic review of the literature by searching PubMed electronically using the following terms: "aortic reconstruction connective tissue disorder," "aortic reconstruction branched graft," "endovascular aorta connective tissue disorder," and "branched aortic graft connective tissue disorder." The reference list of each article was searched manually for relevant publications. Selection criteria included case series in English on different approaches to thoracoabdominal aortic reconstruction

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