TBAD Decision-Making for Patients With Connective Tissue Disorders

Dr. J. Westley Ohman discusses screening protocols, how connective tissue disorders inform decision-making, advice for counseling patients, and multidisciplinary collaboration.

What are your protocols and processes for screening type B aortic dissection (TBAD) patients for connective tissue disorders (CTDs)?

We use age, physical examination or radiographic features (such as the modified Ghent nosology), and family history to gauge the underlying risk of an inherited genetic aortopathy. In the absence of patient drug use history (eg, cocaine, methamphetamines), we have a high index of suspicion for aortopathies in patients aged < 40 years.

How does a patient having a CTD affect your decision-making with regard to TBAD treatment?

Even in patients with CTDs, medical therapy is still our first-line therapy for uncomplicated TBAD, and we continue our standard surveillance protocol of repeat CTA at discharge, 1 month, 6 months, and then annually thereafter.

For more elective interventions—whether at the time of initial TBAD or chronically for aneurysmal degeneration—we recommend consideration for open surgical repair given our experience and volume.

For urgent and emergent interventions, we still favor endovascular therapy as a bridge therapy. Our previously published experience showed a 40% reintervention rate at 1 year for endovascular therapy landing in native aortic tissue, and these patients are extensively counseled on the necessity of follow-up and close surveillance imaging.[1]

How do different CTDs (Vascular Ehlers-Danlos syndrome [VEDS], Marfan syndrome [MFS], Loeys-Dietz syndrome [LDS]) each uniquely affect or inform treatment options?

In general, there is a spectrum of fragility of the aortic wall in patients with CTDs—with VEDS being the most fragile, followed by LDS, and then MFS; however, we are starting to see individual genotype variants informing our understanding of the vessel fragility. I think this an incredibly exciting avenue for individualized treatment of patients based on their specific phenotype risk factors. We tend to be more aggressive with medical therapy in patients with VEDS and LDS given this risk, also recognizing that they are better treated via open surgery with anastomotic adjuncts.

When do you feel compelled to deviate from guidelines regarding TBAD management in the presence of a CTD?

I think the acceptance among our team of endovascular therapy as a bridge therapy has allowed us to be slightly more liberal with this technology, particularly in patients where the goal is alleviation of malperfusion rather than definitive aortic treatment.

Considering both a patient's CTD and the nature and extent of their dissection, what combination of factors most influence your decisions between medical therapy, endovascular approaches, or open repair?

We still can't lose sight of the centrality of medical therapy in all patients with TBAD, and the presence or absence of a CTD should not change that fact. However, the underlying pathology, extent of aortic repair necessary, and, if possible, our ability to stage the extent of aortic repair all influence our decision-making regarding the types of repairs offered to the patients. For example, it is not uncommon for proximal ascending and/or arch repairs to be performed with open surgery but an endovascular extension through the thoracic aorta as part of a planned strategy, for staging their spinal cord risk and making the eventual thoracoabdominal repair less extensive. There is a paucity of data for total endovascular repair of the thoracoabdominal aortic segment, particularly around the branch vessel behaviors, such that open surgery of this segment still remains our recommended approach.

What are you looking for on initial imaging or other pretherapeutic testing that specifically suggests CTD, as well as to guide your decisions?

In addition to the clinical manifestations of these diseases that most clinicians are familiar with, there are other radiographic and echocardiographic manifestations depending on the disease process. The most pronounced manifestations in MFS tend to be skeletal or neurologic,

such as kyphoscoliosis, pectus deformity, dural ectasia, or meningoceles, which are among the most common.

However, arterial-specific imaging findings that should prompt concern for a CTD include near/full circumferential delamination of the intima, multiple flaps, and tortuosity/redundancy of branch vessels (particularly cerebrovascular tortuosity).

More subtle findings of pulmonary artery dilation or paraseptal emphysema may be present, and mitral valve prolapse may be present on echocardiography. For patients with LDS, there is overlap, but the classic triad of arterial tortuosity (the vertebral and carotids are usually the best examples), hypertelorism (wide-set eyes), and nearly pathognomonic bifid uvula should prompt concern.

If thoracic endovascular aortic repair is selected as the optimal therapy, does your technical approach change?

Our experience (mirrored by University of Washington) is that percutaneous closure in the preclose technique is safe and effective in patients with CTD, and that is our preferred approach.[1,2] I favor the use of devices with staged deployment, as I believe that reduces the risk of intraoperative stent graft–induced new entry tears.

The overall approach depends on the goal of therapy. If the goal is resolution of renovisceral malperfusion, then limited aortic coverage is planned, and the devices are sized with a 5% to 15% oversizing based on intraoperative intravascular ultrasound measurements for native aortic tissue at the proximal landing zones. The total aortic diameter distally is measured to assess the need for tapered devices.

For landing zones, landing in surgical grafts and in areas of the aorta where the wall is as straight as possible is the goal, but the latter is rarely possible in landing zone 2 or 3.

How do you evaluate and stratify the potential risk for significant progression or rupture—and ultimately tailor your plan for follow-up and the potential need for reintervention in these populations?

The entire spectrum comes into consideration: acuity of presentation, indication for treatment, and presumed CTD based on clinical and radiographic features.

For patients with acute TBAD with rupture, we tend to favor aggressive serial imaging and have a low threshold for surgical conversion as their clinical picture evolves. This may involve CTA scans at 1- and 7-day intervals and 1-, 3-, and 6-month intervals unless their clinical picture warrants otherwise. For malperfusion, we tend to favor a more traditional protocol, with predischarge, 1-, 6-, and 12-month scans. For chronic TBAD, we often forgo the predischarge CTA if the completion angiogram is reassuring, even if the goal is to seal or isolate a degenerative segment. For patients with presumed VEDS based on clinical suspicion, we typically favor a more aggressive screening protocol, given their clinical observed increased vessel wall fragility and higher rates of degeneration.

What advice can you share about counseling patients regarding their options and involving them in informed decisions about their care?

I think most patients are fairly well versed in the natural history and pathology of the disease processes due to the work of The Marfan Foundation and its collaboration with the Loeys-Dietz Syndrome Foundation and The VEDS Movement. There is a wealth of information and patient experience out there that I actively encourage my patients to engage and involve themselves with.

Ultimately, we discuss a personalized approach for the patient based on their anatomy, our expectations of the repair, our local experience, and their stated goals and expected future aortic or arterial reconstructions. Having a true collaborative, multidisciplinary team with cardiac surgery and cardiology and presenting a transparent and unified message to patients and their families (particularly when the patient is a minor) is absolutely essential.

How and when do you communicate and collaborate with other physicians in the patient's care? Who comprises your multidisciplinary team?

We have a biweekly multidisciplinary aortic team consisting of cardiac surgery, vascular surgery, cardiology, chest radiology, and a (soon to be) medical geneticist. When patients on the outpatient side are referred to us, we discuss and then prioritize same-day visits with the relevant parties after a preliminary discussion has taken place. Afterwards, we discuss in person and if there's a change, update the patient. ■

1. Solomon JR, Braverman AC, Ohman JW. Endovascular and hybrid repair in patients with heritable thoracic aortic disease. Ann Vasc Surg. 2022;87:124-139. doi: 10.1016/j.avsg.2022.05.026
2. Sorber R, Smerekanych S, Pang HJ, et al. Utilization of percutaneous closure devices for large bore arterial access in patients with genetic aortopathy does not result in increased rates of access site complications. J Vasc Surg. 2025;81:582-589. doi: 10.1016/j.jvs.2024.11.001

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