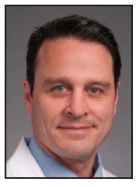
Chasing the Progressive Aortic Dissection

A case report illustrating the extreme complexity of aortopathy and our current knowledge in addressing this potentially devastating presentation.

BY BENJAMIN W. STARNES, MD



On July 5, 2007, Derek, a 42-year-old engineer, was traveling in Iceland with his wife when he experienced the acute onset of tearing back pain, like someone was stabbing him with a knife. It began in his upper middle back and traveled down to his lower back and then into his abdomen. He was taken by the Icelandic Coast Guard by helicopter to Landspitali Reykjavik University Hospital and was diagnosed with a Stanford type B aortic dissection, with an entry tear just beyond the left subclavian artery. He was managed medically with anti-impulse therapy for 18 days and returned to his home in Seattle on July 23, 2007.

Derek has a history of Marfan syndrome and had undergone aortic root replacement for an ascending aortic aneurysm in March 1999 at age 34. Despite this history, it was recommended that he undergo thoracic endovascular aneurysm repair (TEVAR) of his dissection 4 months later. He underwent this procedure with placement of an endoprosthesis, but 10 days later, his stent graft collapsed, and Derek became paraplegic. He underwent urgent placement of a giant Palmaz stent (Cordis Corporation, Bridgewater, NJ) but, unfortunately, never regained use of his lower extremities.

On May 22, 2008, Derek travelled to Cleveland to meet with Dr. Roy Greenberg at the Cleveland Clinic regarding enlargement and progression of his descending thoracic aorta, which had grown aneurysmally to > 6 cm, with persistent false lumen perfusion. Dr. Greenberg recommended endovascular repair instead of open repair because of the underlying severe aortic valvular insufficiency and the patient's inability to tolerate an aortic cross-clamp. I met Derek in July of that year and, after consultation with Dr. Greenberg, extended the repair from his existing stent graft to his diaphragm with a Zenith TX2 device* (Cook Medical, Bloomington, IN).[1,2] Amazingly, Derek's aorta was completely remodeled with false lumen thrombosis and an eventual normal appearance of the aorta 6 years later (Figure 1).

This case illustrates the extreme complexity of aortic dissection. Whether it be dissection in the setting of a connective tissue disorder, as in this case, or a result of illicit drug use, there is still much that we do not know about this aortopathy. With much of the current enthu-

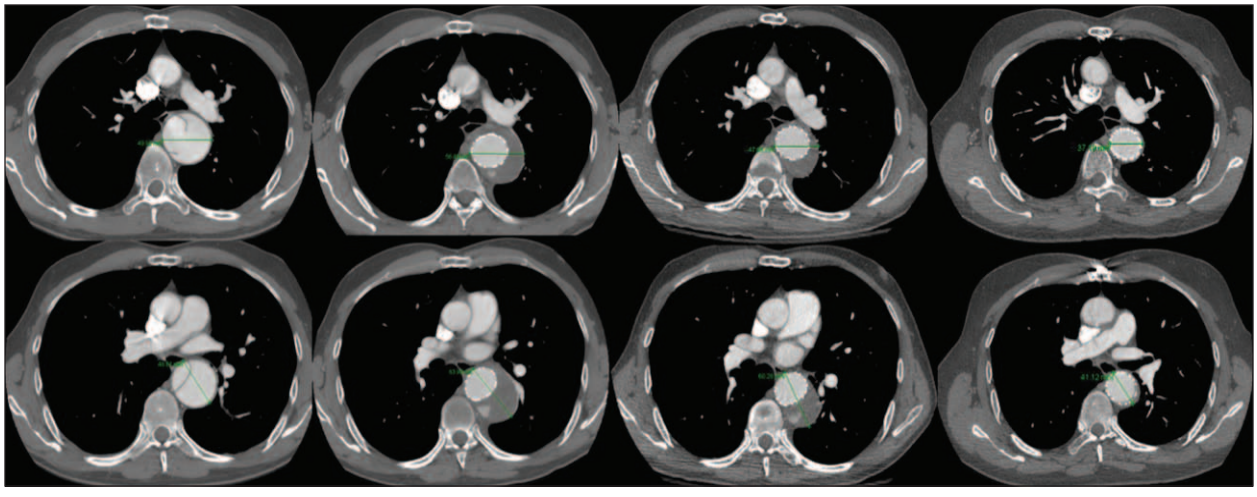


Figure 1. Six-year follow-up with representative axial CT images taken at the level of the carina (top panels) and the top of T-7 (lower panels) on October, 11, 2007; February 27, 2008; July 26, 2009; and December 4, 2013; respectively.

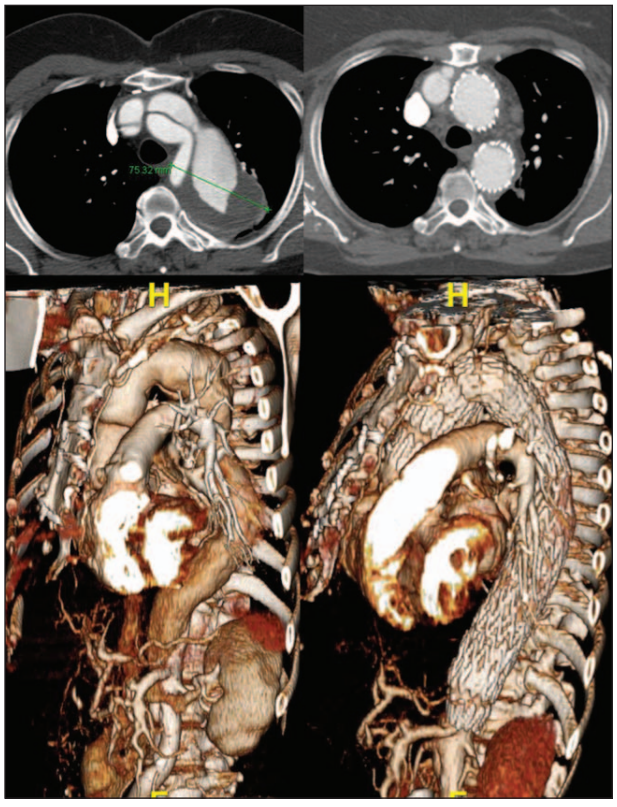


Figure 2. Hybrid repair of a chronic aortic dissection utilizing aortic root replacement, arch debranching, and successful TEVAR, with dramatic aortic remodeling during a 4-year period.

siasm about treating all patients presenting with aortic dissection using endovascular methods, this case example should cause some consternation. What have we learned about TEVAR and aortic dissection in the endovascular era? Whom should we treat? Whom should we not treat? Do the successes outnumber the failures? Is this simply a chronic disease with an eventual death sentence? Although many of these questions remain ill-defined and unanswered, we must understand that aortic dissection is progressive, and to successfully manage these patients, we must have the tenacity to continue to chase the disease until the aorta is stable.

WHAT WE KNOW NOW

The natural history of aortic dissection has been better defined as of late. The International Registry of Acute Aortic Dissection has provided data on outcomes relating to acute dissection of both the ascending and descending aorta. Acute dissection of the ascending aorta has a poor prognosis when treated medically, and roughly 60% of patients will die in the short-term. The outcome of acute dissection of the descending aorta when managed medically is much better, with an in-hospital mortality of 13%,[1] but one-third of these patients will eventually require surgical or endovascular intervention in their lifetime.

There are multiple case reports of the successful endovascular management of either acute or chronic aortic dissection with aortic stent grafts (apropos our first case example). Hybrid approaches have also been shown to be highly successful in certain situations involving the aortic arch either in combination with or without aortic root replacement (Figure 2).

BUT WHAT HAPPENS WHEN ENDOVASCULAR THERAPY DOESN'T WORK?

Do we have the devices and tools we need to adequately treat this disease in its chronic state, or should we focus our efforts on early management of the disease for all patients? Why do some patients do well with medical management and some patients don't? Figure 3 shows a patient with a Stanford type B chronic aortic dissection with rapid aneurysm enlargement who

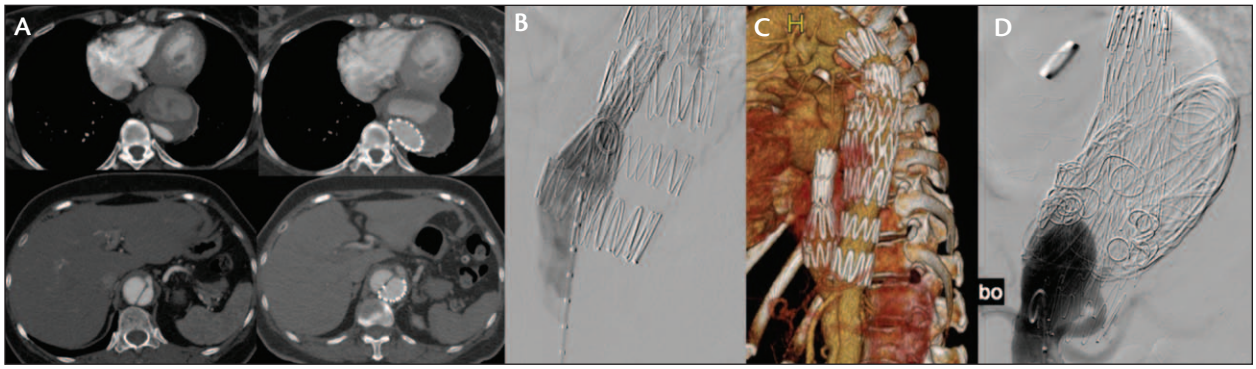


Figure 3. Imaging depicting the expanding aneurysm in association with chronic aortic dissection and subsequent TEVAR with persistent flow in the false aneurysm (A, B). Adjunctive measures were undertaken, resulting in successful false lumen embolization (C). A final coil embolization procedure successfully obliterated the false lumen and arrested expansion of the aneurysm 3 years later (D).

underwent TEVAR in July 2010. On follow-up CT angiography, the patient was noted to have a persistently patent false lumen and sac enlargement. This was confounded by the fact that she was chronically anticoagulated for a mechanical heart valve. She underwent multiple reinterventions, with subsequent complete false lumen thrombosis and complete obliteration of the aneurysm sac (Figure 3).

A NEW PERSPECTIVE

For nearly 50 years, we have classified aortic dissection based upon the anatomic location of the dissection in the aorta, with the implications affecting treatment choices, either surgical or medical. The widespread adoption of endovascular therapy to manage this disease has challenged the established classification systems to adequately account for the features that remain critical to making therapeutic decisions. In an attempt to modernize the classification of aortic dissection with relevance to endovascular therapies, the Working Group on Aortic Disease of the DEFINE Project recently proposed DISSECT, a new mnemonic-based approach to the categorization of aortic dissection (see the DISSECT Classification of Aortic Dissection sidebar).[2]

In the coming years, it will be crucial for us to appropriately classify patients so that we may compare apples to apples and oranges to oranges. We all know that a patient with a D:Ch, I:D, S:70 mm, SE:AI, C:C, T:P (previously known as simply a complicated Stanford type B or Debakey type 3) is entirely different from a D:Sa, I:D, S:30 mm, SE:D, C:UC, T:CT. In other words, not all type B dissections are the same, and our therapies should be directed at the natural history of the disease. I believe this new classification system is a step in the right direction toward classifying and managing patients presenting with a wide variety of anatomical and clinical manifestations.

LIFE GOES ON

I had coffee with Derek the other day at a local Starbucks. He is wheelchair bound, his legs taken from him in the prime of his life. Not a single day goes by that Derek doesn't think about his own aortic calamity. We owe our patients much better treatment methods, devices, and technology for aortic dissection in this new century.

We owe Derek. ■

**The Zenith TX2 is FDA approved with indications for use in the endovascular treatment of patients with aneurysms or ulcers of the descending thoracic aorta having vascular morphology suitable for endovascular repair.*

It is CE Mark approved with indications for use in the treatment of patients with atherosclerotic aneurysms, symptomatic acute or chronic dissections, contained ruptures, growing aneurysms and/or resulting in distal ischemia, in the descending thoracic aorta having vascular morphology suitable for endovascular repair.

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1. Suzuki T, Mehta RH, Ince H, et al. Clinical profiles and outcomes of acute type B aortic dissection in the current era: lessons from the International Registry of Aortic Dissection (IRAD). Circulation. 2003;108(suppl 1):II312-317.
2. Dake MD, Thompson M, van Sambeek M, et al. DISSECT: a mnemonic based approach to the categorization of aortic dissection. Eur J Vasc Endovasc Surg. 2013;46:175–190.

"DISSECT" CLASSIFICATION OF AORTIC DISSECTION

Duration (D) defined as time from onset of symptoms:
- Ac = acute: < 2 weeks from initial onset of symptoms
- Sa = subacute: 2 weeks to 3 months after symptom onset
- Ch = chronic: > 3 months from initial onset of symptoms

Intimal (I) tear (primary) location within the aorta:
- A = ascending aorta
- Ar = aortic arch
- D = descending aorta
- Ab = abdominal aorta
- Un = unknown

Size (S) of the aorta based on maximum transaortic diameter by centerline analysis at any level within the dissected segment of aorta

Segmental extent (SE) of aortic involvement from proximal to distal boundary:
- A = ascending aorta exclusively
- Ar = aortic arch exclusively
- D = descending exclusively
- Ab = abdomen exclusively
- AAr = ascending to arch
- AD = ascending to descending
- AAb = ascending to abdomen
- AI = ascending to iliac
- ArD = arch to descending
- ArAb = arch to abdomen
- ArI = arch to iliac
- DAb = descending to abdomen
- DI = descending to iliac

Clinical complications (C) related to dissection:
- C = complicated
  - Aortic valve involvement
  - Cardiac tamponade
  - Rupture
  - Branch vessel malperfusion: symptomatic branch vessel involvement defined as anatomic and clinical manifestations of branch vessel compromise (eg, static and/or dynamic branch involvement with accompanying stroke, paraplegia, coronary, mesenteric, visceral, renal, and/or extremity symptoms)
  - Progression of aortic involvement with proximal or distal extent of dissection
  - Other: uncontrollable hypertension, uncontrollable clinical symptoms, or rapid false lumen dilation and/or overall transaortic enlargement of > 10 mm within the first 2 weeks of initial diagnosis
- UC = uncomplicated (absence of complications listed above)

Thrombosis (T) of aortic false lumen:
- P = patent aortic false lumen: evidence of flow or contrast opacification within the false lumen throughout the length of dissected aorta
- CT = complete thrombosis of the aortic false lumen: no evidence of flow or contrast opacification within the following segments of the dissected aortic false lumen
  - A = ascending aorta
  - Ar = aortic arch
  - D = descending
  - Ab = abdomen
- PT = partial thrombosis of the aortic false lumen: longitudinal thrombosis of a portion of the aortic false lumen or circumferential thrombus that partially fills the false lumen constitute partial or incomplete thrombosis within the following segments of the dissected aorta:
  - A = ascending aorta
  - Ar = aortic arch
  - D = descending
  - Ab = abdomen

Data adapted from Dake MD, et al. Eur J Vasc Endovasc Surg. 2013;46:175–190.[2]