EDITORIAL

LARS G. SVENSSON, MD, PhD
Director, Aortic Surgery, and Marfan Syndrome and Connective Tissue Disorder Clinic, Department of Thoracic and Cardiovascular Surgery, Cleveland Clinic

Acute aortic syndromes: Time to talk of many things

"The time has come," the Walrus said,
"To talk of many things:
Of shoes—and ships—and sealing-wax—
Of cabbages—and kings—
And why the sea is boiling hot
And whether pigs have wings."

—Lewis Carroll, The Walrus and the Carpenter (from *Through the Looking-Glass and What Alice Found There*, 1872).

LEWIS CARROLL'S POEM of 1872 is a useful starting point for identifying issues resulting from confusion over the variously described acute aortic syndromes—and, for oysters, the dangers of listening to walruses.

See related article, page 7

TALK OF MANY THINGS

In cases of aortic dissection (splitting or separation of the layers of the aortic wall), it is important to establish the type (ie, the location and extent) and class (ie, the structure) of the dissection, because these distinctions determine the treatment.[1] Similarly, in cases of painful or leaking degenerative aneurysms, we need to know the location of the aneurysm and whether the presenting pain is from compression of surrounding tissue, particularly of the vertebral bodies, or from leakage.

The location and extent of an aortic dissection can be classified in three ways (see **FIGURE 3** in Smith and Schoenhagen's excellent review of the use of computed tomography [CT] in acute aortic syndromes in this issue of the *Cleveland Clinic Journal of Medicine*[2]):

- **The DeBakey system** (type I, II, or III)
- **The Stanford system** (type A or B)
- **Distal or proximal** to the left subclavian artery.

Of note, the DeBakey system does not include tears in the arch that extend distally without ascending involvement. The original Stanford system included arch tears with distal extension in type B; hence, type B excluded all patients without ascending involvement.

The importance of the extent of dissection is that most patients with Stanford type A or DeBakey type I or II dissections should undergo immediate surgery, as most of them would die without it. Surgery is also indicated for arch tears (non-DeBakey, original Stanford type B).

Because these classifications are somewhat confusing, the simplest approach is to note whether the dissection extends proximal or distal to the left subclavian artery, because proximal dissections need surgery and distal ones are first managed medically.

The classes of dissection also have bearing on treatment.[1] These are:

- **Class I**—classic aortic dissection in the media with two lumens separated by a "flap" or septum
- **Class II**—intramural hematoma in the aortic wall from dissection in which the intimal tear cannot be imaged (these are nearly always found during surgery or autopsy)
- **Class III**—localized confined intimal tears without extensive undermining of the intima or flap formation. These are often seen with Marfan syndrome and can rupture or cause tamponade, as can any type of proximal dissection. The typical appearance is of a bulging bubble in the aortic wall.
- **Class IV**—penetrating atherosclerotic ulcers with localized dissections or wall hematomas, often with calcium at the base of a mushroom-shaped area of extraluminal contrast. Of note, the plane of dissection is often between the media and adventitia.
- **Class V**—iatrogenic or posttraumatic dissection.

All class I to class IV tears of the proximal aorta require surgery, whereas distal class IV and V tears may require either open or endovascular surgical intervention. Surgery is also indicated for patients with distal

dissections who have severe narrowing of the true lumen, distal ischemia, uncontrolled pain, severe hypertension, or evidence of leaking, particularly with class IV tears.

In distal dissections that are subacute (2–6 weeks old), the Investigation of Stentgrafts in Patients With Type B Aortic Dissection (INSTEAD) trial found that inserting a stent prophylactically provided no benefit. Further-more, there is no proof that stenting is beneficial if the aortic dissection is chronic, ie, more than 6 weeks old.[1,3–5]

WHICH SHOE FITS?

There is no ideal procedure to detect dissection, although the trend is towards CT angiography, as Smith and Schoenhagen report.[2] Although some investigators have optimistically estimated CT's sensitivity and specificity as 100%, cardiovascular surgeons are well aware of both false-positive and false-negative CT studies. Thus, for emergency repairs of proximal dissections, transesophageal echocardiography should be done after intubation and before opening a patient's chest if time allows. Magnetic resonance imaging, CT, and transesophageal echocardiography may all miss class III tears, but these are frequently evidenced by eccentric "bubbles" or "ballooning."[1]

SHIPS

Patients with either acute aortic dissection or severe pain associated with degenerative aneurysms need to be "shipped" promptly to a tertiary medical center after diagnosis, since larger volumes of procedures appear to be associated with better outcomes.

SEALING WAX

Using current surgical methods, the aortic valve can be preserved during aortic dissection repair unless the valve is bicuspid or the patient has Marfan syndrome.[1,3,4,6–8]

Sealing wax in the form of biological glues, rather than for letters, is a new innovation. A caveat remains, however: we have seen patients who have required reoperation for false aneurysms or infection. Hence, glues should be used with caution.

CABBAGES

A dilemma is whether patients should undergo coronary catheterization (or CT angiography—a separate question) and subsequent coronary artery bypass grafting (CABG), if needed, at the time of aortic dissection repair. The problem is that approximately one-third of patients have coronary artery disease that may require CABG, but the delay for catheterization increases the risk of rupture or tamponade before surgery.

Indeed, 40% of patients with proximal dissections die immediately, and 1% to 3% die in the hour before surgery. The short-term (in-hospital and 30-day) mortality rates range from 3.4% (Cleveland Clinic 2006 data) to 25%, and of the survivors only about 50% are alive 5 years after surgery.

Though dismal, the prognosis is improving. In 162 patients with aortic dissection and Marfan syndrome or connective tissue disorders who underwent surgery at Cleveland Clinic in the years 1978–2003, the 5-year survival rate in those with aortic dissection was 75% and the 10-year rate was 55%.[7] In those without dissection, the 10-year survival rate was approximately 90% ($P < .001$).

KINGS

Noted personalities who have had aortic dissection include King George II of England (who died in 1760), Lucille Ball, Conway Twitty, Jan Larson, and most recently John Ritter. None of these famous people survived their aortic dissections. Indeed, dissection and diseases of the aorta or its branches cause between 43,000 and 47,000 deaths annually,[9] more than from breast cancer, murders, or motor vehicle accidents. The main reason for these dismal statistics is that the disease is often misdiagnosed at the time of presentation.

BOILING SEA

Careful studies from Olmsted County, Minnesota,[10] have shown a tripling of the incidence of aortic disease, particularly in women, even though the rate of deaths from coronary artery disease has been decreasing. Furthermore, Olsson et al[11] report that the incidence of aortic dissection in men in Sweden increased to approximately 16 per 100,000 per year from 1987 to 2002, a 52% increase. The aging of the population must play a large role, but other factors may exist that are not well understood or defined and require further research.

PIGS HAVE WINGS

Will it be possible to overcome this rising problem? The answer is a definite yes. The results of aortic surgery have never been better. Many new innovations

are available, such as aortic root preservation and endovascular stenting procedures. It may be possible to slow the growth of or prevent some aneurysms and aortic dissections, particularly with beta-blockers and, potentially, with losartan (Cozaar) for Marfan syndrome patients.

One of the keys to preventing aortic catastrophes and aortic dissection is to repair aortic aneurysms. The threshold for surgery, however, depends on a surgeon's experience and results, the underlying pathology, and the aortic size.

We observed that 12.5% of dissections in patients with bicuspid valves and 15% of those in patients with Marfan syndrome were in aortas smaller than 5.0 cm in diameter, that aortic dissection occurred at smaller diameters in shorter patients, and that the risk of dissection increased exponentially with the size of the aorta. Subsequently, we found that a better measure of risk is the maximal aortic cross-sectional area in cm^2 divided by the patient's height in meters; if this ratio exceeds 10, then surgery is recommended.[12]

Results of surgery are good in experienced hands. In patients who undergo surgical repair of bicuspid aortic valves with or without concurrent repair of the ascending aorta (mostly in patients with an aortic cross-section-to-height ratio > 10), the perioperative mortality rate is about 1.0% for both groups, and at 10 years about 98% of patients are free from re-operation on the aorta and more than 90% are free from re-operation on the aortic valve.[8] This is important because these are typically young patients who would do better without biological replacement valves (which are not very durable) or mechanical valves (which necessitate lifelong anticoagulation). Results are also good in surgery of the aortic arch and even better in patients with tricuspid aortic valves.[4,6,8]

Increasingly, in patients at high risk, we are inserting thoracic, abdominal, and thoracoabdominal stent grafts, with excellent early results. An even newer innovation is to replace the aortic valve in high-risk patients via a transcatheter balloon-expandable valve stent inserted through the groin or left ventricular apex.

These treatment innovations have been big strides, but aortic disease continues to increase. Indeed, our volume of thoracic aortic surgery at Cleveland Clinic increased from 190 procedures in 1999 to 717 in 2006. Early detection—before acute emergency surgery is required, with its concomitant high risk of death—is the key to successful surgical outcome and long-term survival. ■

REFERENCES

1. **Svensson LG, Labib SB, Eisenhauer AC, Butterly JR.** Intimal tear without hematoma: an important variant of aortic dissection that can elude current imaging techniques. Circulation 1999; 99:1331–1336.
2. **Smith AD, Schoenhagen P.** CT imaging for acute aortic syndrome. Cleve Clin J Med 2008; 75:7–24.
3. **Svensson LG, Nadolny EM, Kimmel WA.** Multimodal protocol influence on stroke and neurocognitive deficit prevention after ascending/arch aortic operations. Ann Thorac Surg 2002; 74:2040–2046.
4. **Svensson LG, Kim KH, Blackstone EH, et al.** Elephant trunk procedure: newer indications and uses. Ann Thorac Surg 2004; 78:109–116.
5. **Greenberg RK, Haddad F, Svensson L, et al.** Hybrid approaches to thoracic aortic aneurysms: the role of endovascular elephant trunk completion. Circulation 2005; 112:2619–2626.
6. **Svensson LG.** Sizing for modified David's reimplantation procedure. Ann Thorac Surg 2003; 76:1751–1753.
7. **Svensson LG, Blackstone EH, Feng J, et al.** Are Marfan syndrome and marfanoid patients distinguishable on long-term follow-up? Ann Thorac Surg 2007; 83:1067–1074.
8. **Svensson LG, Blackstone EH, Cosgrove DM 3rd.** Surgical options in young adults with aortic valve disease. Curr Probl Cardiol 2003; 28:417–480.
9. **Svensson LG, Rodriguez ER.** Aortic organ disease epidemic, and why do balloons pop? Circulation 2005; 112:1082–1084.
10. **Clouse WD, Hallett JW Jr, Schaff HV, Gayari MM, Ilstrup DM, Melton LJ 3rd.** Improved prognosis of thoracic aortic aneurysms: a population-based study. JAMA 1998; 280:1926–1929.
11. **Olsson C, Thelin S, Ståhle E, Ekbom A, Granath F.** Thoracic aortic aneurysm and dissection: increasing prevalence and improved outcomes reported in a nationwide population-based study of more than 14,000 cases from 1987 to 2002. Circulation 2006; 114:2611–2618.
12. **Svensson LG, Kim KH, Lytle BW, Cosgrove DM.** Relationship of aortic cross-sectional area to height ratio and the risk of aortic dissection in patients with bicuspid aortic valves. J Thorac Cardiovasc Surg 2003; 126:892–893.

ADDRESS: *Lars Svensson, MD, PhD, Department of Thoracic and Cardiovascular Surgery, F24, Cleveland Clinic, 9500 Euclid Avenue, Cleveland, OH 44195.*