

# XII DISEASES OF THE AORTA

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## The Normal Aorta

The normal aorta is composed of three distinct layers: the inner intima, the elastic media, and the thin outer adventitia. In the media, layers of elastic elements intertwine with collagen and smooth muscle cells, providing the elastic strength that enables the aorta to withstand the pulsatile stress produced by the ejection of blood during ventricular systole. In diastole, the potential energy stored in the aortic wall in systole is transformed into kinetic energy as it propels the blood forward. With aging, the normal elastic elements of the aorta degenerate, reducing its elasticity and distensibility.

The aorta consists of three anatomic segments: the ascending aorta, the aortic arch, and the descending aorta. The ascending aorta consists of the aortic annulus, the aortic valve cusps, the sinuses of Valsalva, the sinotubular ridge, and the tubular portion of the ascending aorta. The ascending aorta connects the left ventricular outflow tract at the aortic valve to the aortic arch, which begins at the brachiocephalic artery. The arch provides branches to the head and neck vessels, coursing just in front of the trachea and then proceeding to the left of the esophagus and the trachea. The descending aorta begins in the posterior mediastinum at the ligamentum arteriosum and courses in front of the spine as it descends to the bifurcation of the leg vessels.

## Aortic Aneurysms

Aortic aneurysm is a potentially life-threatening entity for which both effective screening and curative therapy are available. An aorta is considered aneurysmal when its diameter exceeds 1.5 times the expected normal diameter at any location along its length; the normal diameter is approximately 2 cm.<sup>1</sup> Alternatively, an aorta may be considered aneurysmal when its maximum diameter is greater than 3 cm; this definition is often applied to the abdominal aorta.<sup>2,3</sup> Aneurysms are divided into those that affect the abdominal aorta and those that affect the thoracic aorta. More extensive aneurysms (termed thoracoabdominal) involve both aortic areas. In addition, aneurysms are defined as either fusiform (i.e., spindle shaped) or saccular (i.e., sack shaped).

### ABDOMINAL AORTIC ANEURYSMS

Aneurysms of the abdominal aorta are more common than thoracic aortic aneurysms. Among the risk factors for aneurysms, perhaps the most important are age and a history of tobacco use. The incidence of aneurysms increases in men older than 55 years and in women older than 70 years. Depending on age, men are two to five times more likely to develop aortic aneurysms. Additional risk factors are hypertension, elevated cholesterol, and a family history, suggesting a genetic predisposition to aneurysms.<sup>4,5</sup> Several reports show that aneurysms develop in as many as 25% of first-degree relatives of patients with abdominal aortic aneurysms.<sup>3,6</sup> The infrarenal aorta is the most commonly affected region.

### SCREENING FOR ABDOMINAL AORTIC ANEURYSMS

Current recommendations are for noninvasive screening of

male patients older than 60 years who have a family history of aneurysm. Screening of men 65 to 75 years of age who have a history of smoking is also recommended, but there is less evidence (class IIa) in support of such screening.<sup>3</sup> Screening with abdominal ultrasound may be particularly effective for obese patients, in whom abdominal palpation is of limited value. A large-scale trial of ultrasound screening in 67,900 men 65 to 74 years of age demonstrated a reduction of 43% in aneurysm-related death in those patients who underwent routine testing.<sup>7</sup> The actual cost-effectiveness of screening strategies has yet to be demonstrated.<sup>8</sup>

### Clinical Presentation

Most abdominal aortic aneurysms produce no symptoms and are discovered during a routine physical examination, as a result of noninvasive screening, or are incidentally detected during imaging for an unrelated reason. The most common symptom is pain, often described as a steady, gnawing discomfort in the lower back or hypogastrium. Generally, the pain is not affected by movement.

In some patients, the abdominal aortic aneurysm is first discovered during a period of rapid expansion or an impending rupture, which is often marked by severe discomfort in the lower abdomen or back, radiating to the buttocks, groin, or legs. Rupture is accompanied by the abrupt onset of back and abdominal pain, abdominal tenderness, the presence of a palpable pulsatile mass, hypotension, and shock. Of note, a ruptured aneurysm may mimic other conditions, including abdominal colic, renal colic, diverticulitis, and gastrointestinal hemorrhage. Not surprisingly, more than 25% of patients presenting with rupture or expansion of an aortic aneurysm are initially misdiagnosed.

Patients with impending or actual rupture must be managed as a surgical emergency in a manner similar to that used for patients with major trauma. Such patients rapidly develop hemorrhagic shock, manifested by peripheral vasoconstriction, hypotension, mottled skin, diaphoresis, oliguria, disorientation, and cardiac arrest. Patients with retroperitoneal rupture may show evidence of hematomas on the flank and in the groin. Although rare, rupture with erosion into the duodenum may present as massive gastrointestinal hemorrhage.

### Diagnostic Evaluation

**Physical examination** The abdominal aorta is usually detectable on deep palpation, particularly in thin persons. In obese patients, the normal aortic impulse may not be palpable. Obese patients may harbor a large aneurysm without any symptoms or findings on physical examination, unless the aneurysm is exerting pressure on an adjacent structure. Thin patients, in contrast, may feel a pulsatile mass in the abdomen when an abdominal aneurysm has developed.

When palpable, an aneurysm will be identified as a pulsatile mass extending from as high as the xiphoid process to the supra-pubic area. Because of the variable amount of tissue between the examiner's fingers and the aneurysm, the transverse diameter of the aneurysm is typically overestimated on physical examination. It is also difficult to differentiate an ectatic aorta from an aneurysm. Some aneurysms are sensitive to palpation and may be tender if they have recently expanded or if rupture is imminent. Patients with aneurysms often have evidence of other peripheral vascular disease.

**Imaging studies** Several diagnostic tools can help identify and measure the size of abdominal aortic aneurysms. Previously, aortography was considered the gold standard for evaluating aortic aneurysms. One advantage of aortography is that it can be used to evaluate associated iliofemoral disease and involvement of the renal and mesenteric branches of the aorta. Aortography is invasive and requires intravascular contrast, which carries a risk of nephrotoxicity. Its use has declined with the development of abdominal ultrasonography, multidetector computed tomography angiography (MDCTA), and magnetic resonance angiography (MRA).

Abdominal ultrasonography is the most practical and most frequently used screening method.<sup>9</sup> Ultrasonography has a sensitivity of nearly 100% for diagnosing aneurysms of significant size and can discriminate size to within  $\pm 3$  mm. Ultrasonography is inexpensive and noninvasive but may be inadequate for evaluating the most superior or inferior extent of an aneurysm and is generally considered inadequate as a sole diagnostic technique for planning surgical resection.

MDCTA can determine aneurysm size to within  $\pm 2$  mm. Because MDCTA can determine the inferior and superior extent of the aneurysm and its shape, it is more useful for planning surgical repair. However, the need for radiographic contrast is a relative disadvantage. When computed tomography is compared with an image derived from abdominal ultrasonography, the size of the aneurysm as determined by CT is larger by approximately 2.7 mm.<sup>10</sup> MRA can be successfully used for both screening and the planning of surgical or endovascular repair. MRA identifies the size and extent of an aneurysm with a high degree of accuracy. Anatomic landmarks are easily distinguished in the three-dimensional images created with MRA, which correctly defines the distal and proximal extent of an aneurysm in more than 75% of the cases examined.<sup>11</sup>

#### *Management to Reduce Risk of Aneurysm Rupture*

Current management of abdominal aortic aneurysm is directed at reducing the risk of rupture by intervening with timely surgical resection or endovascular stent grafting. Studies of the natural history of aneurysms show that the likelihood of rupture is greatest in patients with symptomatic, large, or rapidly expanding aneurysms. Aneurysms smaller than 4 cm in diameter have a low (<2%) risk of rupture.<sup>3,12,13</sup> Aneurysms exceeding 10 cm in diameter have a greater than 25% risk of rupture over 2 years. Because aneurysms tend to expand with time, current strategies call for identifying and observing aneurysms that are asymptomatic and are small enough to have a low risk of rupture. The median rate of expansion is slightly less than 0.5 cm a year.<sup>3,12-14</sup> However, the tendency for expansion is variable and may not be linear. More rapidly expanding aneurysms are more likely to rupture than are stable aneurysms. Aneurysms larger than 5.5 cm in diameter should be referred for repair, whereas aneurysms less than 4 cm in diameter are generally watched.<sup>3,12-17</sup> Evidence of expansion, particularly if the diameter of the aneurysm has exceeded 5.0 to 5.5 cm, is often taken as an indication to operate.<sup>18</sup>

#### *Surgical Treatment*

Surgical treatment consists of the resection of the aneurysm and the insertion of a synthetic (Dacron) graft. Additional distal surgery is often necessary; frequently, such surgery requires the resection and interposition of grafts into one or both iliac arteries. For most large aneurysms, the aneurysm wall is left intact, and the Dacron graft is placed inside the aneurysm. The surgical

treatment of abdominal aneurysms carries an average operative mortality of 4% to 6%.<sup>3</sup> Surgical mortality is 2% in low-risk patients but may be as high as 20% in patients with impending rupture. Patients in shock caused by aneurysm rupture require emergency surgery; operative and perioperative mortality in these patients may be as high as 50%.

#### *Endovascular Therapy*

Endovascular stent grafting is a relatively recent development in the therapy of thoracic and abdominal aortic aneurysms.<sup>19,20</sup> These stents can be deployed in any relatively straight section of the thoracic or abdominal aorta; they can be used to bridge a focal aneurysm or to exclude areas of severe atherosclerotic disease or penetrating ulcer from the aortic lumen. When properly deployed, the stent graft will increase the structural integrity of the aorta in that location. Planning for endovascular therapy requires precise determination of aortic dimensions. The placement of a stent graft into the aorta necessitates a femoral artery cutdown, which is an open surgical approach. Endovascular therapy for both thoracic and abdominal aortic aneurysms may also entail a simultaneous procedure into a branch vessel, most commonly an iliac artery.

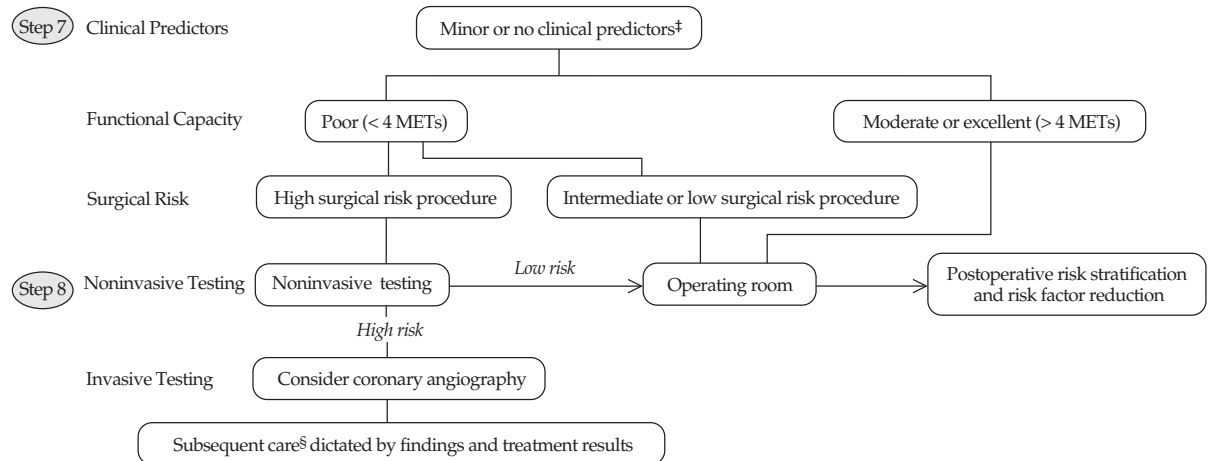
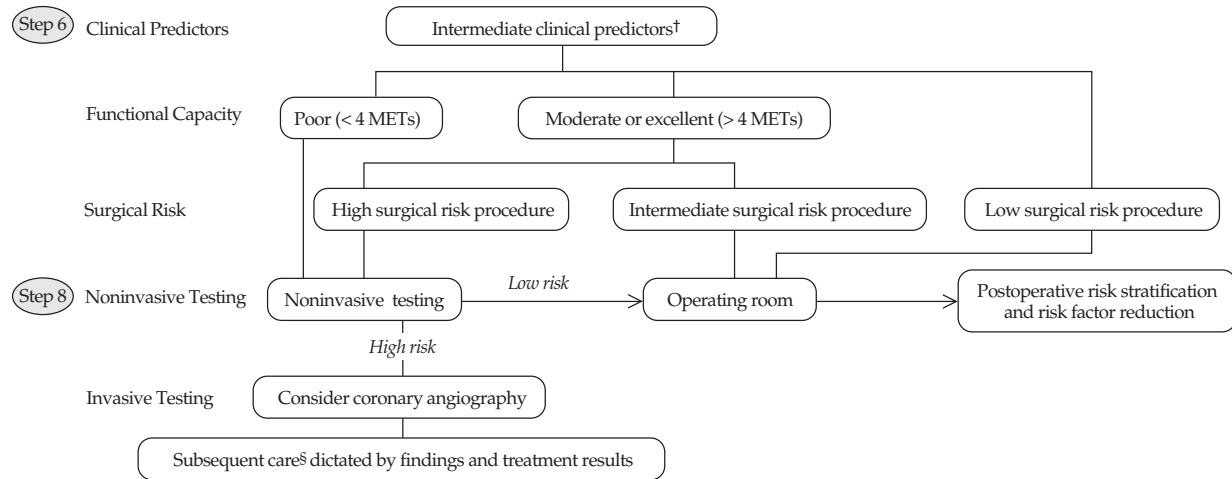
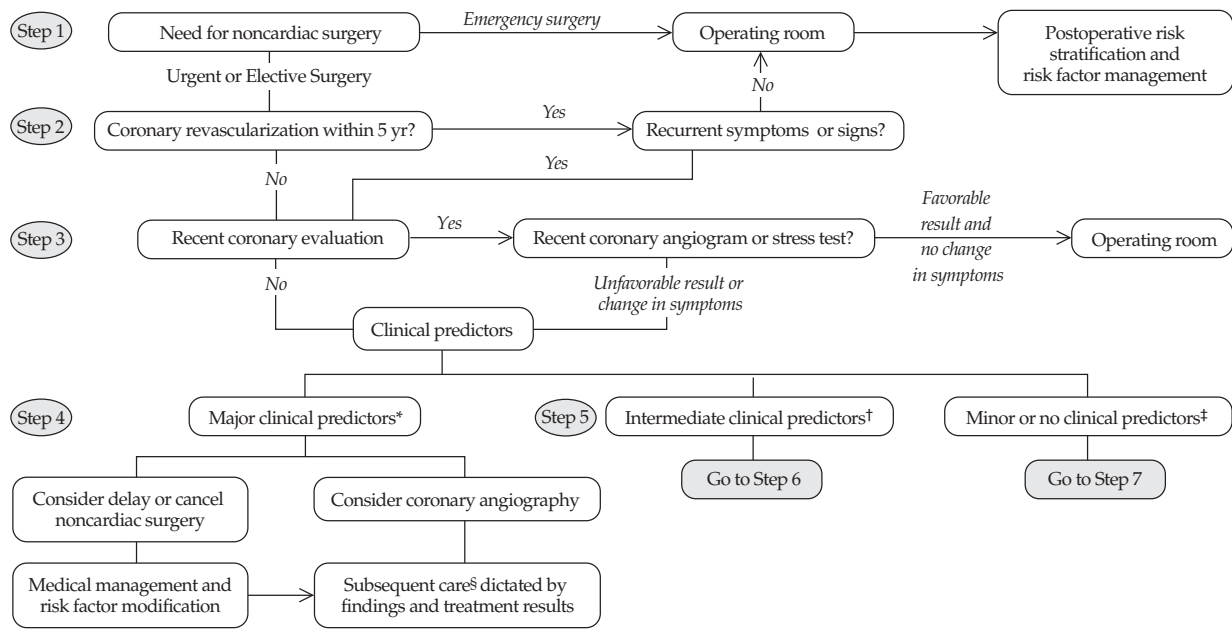
Endovascular therapy, although still evolving, is being used in as many as 50% of patients in high-volume centers. A decision to proceed with endovascular therapy rather than traditional open aneurysm resection is made on the basis of a patient's operative risk. Survival rates of patients who underwent endovascular repair of an infrarenal abdominal aneurysm appear to be equivalent to survival rates of patients who underwent open repair; however, this finding may be heavily dependent on patient selection.<sup>3</sup>

A complication of endovascular therapy is the development of a leak around or through the stent graft (referred to as an endoleak).<sup>21</sup> Four types of endoleaks can occur. A type 1 endoleak occurs at the margin of a stented cuff and can result in high pressure flow into the residual aneurysm, thus increasing the risk of rupture; it can be corrected by the use of a longer stent or by conversion to an open repair. A type 2 endoleak represents retrograde blood flow from branch vessels that have been excluded from the lumen of the aorta by the endograft. Many type 2 endoleaks seal spontaneously. If the leak persists, branch vessels can be embolized in an angiographic procedure. A type 3 endoleak is present when blood leaks directly through the graft material; because a type 3 leak can result in aortic rupture, it is an indication of further repair. A type 4 endoleak represents diffuse leakage through the relatively porous graft material; it often resolves spontaneously.

#### *Preoperative Evaluation and Management*

Appropriate preoperative evaluation and management are critical in a patient undergoing elective aortic aneurysm resection. Reports suggest that one third to two thirds of perioperative deaths can be attributed to coronary artery disease. A guideline published by the American College of Cardiology (ACC) and the American Heart Association (AHA) reviewed the literature regarding preoperative assessment; the ACC/AHA provides a simple algorithm to help determine which patients should be considered for preoperative noninvasive testing for coronary disease [see Figure 1].<sup>22</sup>

The first consideration is whether the vascular surgery is urgent or emergent. By definition, emergent surgery cannot be delayed, and in emergency cases, the risk is higher. In both urgent



\* Major clinical predictors: unstable coronary syndrome, decompensated CHF, significant arrhythmias, severe valvular disease.  
 † Intermediate clinical predictors: mild angina pectoris, prior MI, compensated or prior CHF, diabetes mellitus.  
 ‡ Minor clinical predictors: advanced age, abnormal ECG, rhythm other than sinus, low functional capacity, history of stroke, uncontrolled systemic hypertension.  
 § Subsequent care of patient may include cancellation or delay of surgery, coronary revascularization followed by surgery, or intensified care.

Figure 1 Stepwise approach to preoperative cardiac assessment. (CHF—congestive heart failure; METs—metabolic equivalents; MI—myocardial infarction)

and emergent cases, the usual medical approach is to assume the patient may have preexisting coronary disease. Unless contraindicated, beta blockers should be used to treat such patients. Ideally, beta blockers should be started days to several weeks before surgery; the dose should be titrated to achieve a target heart rate of 50 to 60 beats a minute.<sup>23</sup> The clinical status, electrocardiographic findings, and hemodynamics of these patients should be monitored carefully after surgery.

For determination of perioperative risk, the first issue to address is whether the patient has recently undergone coronary revascularization. If the patient has undergone coronary bypass surgery within the past 6 years and has had no subsequent coronary symptoms, the risk of perioperative events is relatively low. A second issue is whether the patient has recently undergone coronary evaluation. Further preoperative testing is not usually required for a patient who underwent a stress test or a coronary angiogram within the past 2 years that indicated minimal or no coronary disease, particularly if the patient is not diabetic and has had no change in clinical or functional status.

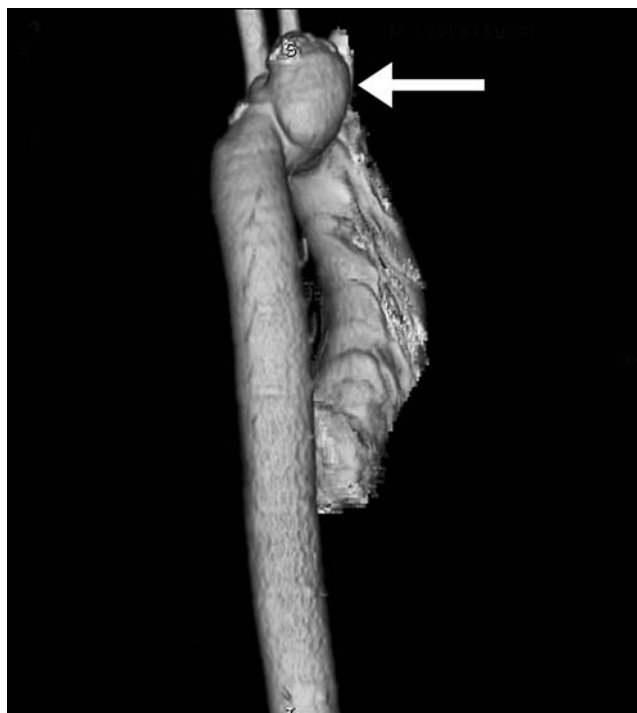
Patients with a history of myocardial infarction (MI), angina, diabetes, or congestive heart failure should be more thoroughly evaluated. If such patients have poor functional capacity and have not undergone recent coronary evaluation, they should undergo a preoperative stress test to evaluate the severity of coronary disease and to determine the status of left ventricular function. When possible, exercise is generally the preferred method of stress testing and appears to be safe in most patients.<sup>24</sup> For patients who are unable to exercise, pharmacologic stress testing with either dobutamine echocardiography or adenosine sestamibi or thallium imaging is appropriate. The risk of perioperative cardiac events is directly related to the presence and extent of left ventricular dysfunction and ischemia.

The relative risk of perioperative cardiac morbidity or mortality is low (1% to 5%) in patients with no inducible ischemia and who are without evidence of fixed perfusion defects or wall motion abnormalities. In patients with extensive areas of ischemia or prior infarction, as detected during preoperative testing, perioperative event rates (e.g., death and MI) may be as high as 20% to 40%. Such patients should probably undergo coronary angiography and possibly coronary revascularization before undergoing major operative procedures.

Although the indications for coronary bypass surgery or percutaneous coronary interventions are generally the same for the preoperative patient and the general population, the evaluation for potential heart disease that a patient with an aneurysm undergoes before aneurysm resection may be the patient's first such evaluation. Coronary artery disease must be treated to the fullest extent before undertaking a potentially stressful operation on the aorta. The same preoperative risk assessment is advisable for a patient undergoing an endovascular procedure, because of the potential that the endovascular procedure may need to be converted to an open surgical procedure.

#### *Postoperative Modification of Risk Factors*

Long-term modification of cardiovascular risk factors is a frequently overlooked issue in the management of patients undergoing abdominal aortic aneurysm resection. The preoperative period represents an excellent opportunity to identify and treat hypertension, diabetes, hypercholesterolemia, smoking, obesity, and poor functional status. All patients identified as having vascular disease should take aspirin daily to reduce the risk of cardiovascular events. Often, beta blockers are prescribed for pa-



**Figure 2** A three-dimensional echocardiogram rendered from a MDCTA scan of a patient with a discrete aortic arch aneurysm. Note the normal caliber of the descending thoracic aorta and mild dilation of the ascending thoracic aorta. A prominent sacular aneurysm is seen arising from the posterior superior aspect of the aortic arch (white arrow).

tients with coronary artery disease; the cholesterol profiles of such patients should be routinely assessed and treatment initiated as appropriate. Studies suggest that secondary prevention of vascular disease is enhanced by aggressive treatment of hypercholesterolemia, particularly in persons with a low-density lipoprotein cholesterol level exceeding 100 mg/dl. Currently, the best evidence suggests that the broad class of statin drugs is effective therapy. Beta blockers have been championed both for their ability to reduce the risk of MI and for their potential to reduce the risk of aneurysm (i.e., the development of new aneurysms or the expansion of existing yet undetected aneurysms) in patients who previously had significant aneurysms.

#### THORACIC AORTIC ANEURYSMS

Thoracic aortic aneurysms are less common than abdominal aneurysms; they are classified according to the involvement of the ascending aorta, the descending aorta, or a combination of the two. Aneurysms of the descending aorta are the most common. The etiology of thoracic aneurysms correlates with their location. Aneurysms of the ascending aorta are often associated with cystic medial necrosis. This association is particularly common in patients with Marfan syndrome, Ehlers-Danlos syndrome, and annuloaortic ectasia, which is the result of loss of elastic tensile strength in the aorta. Descending thoracic aortic aneurysms are often seen in hypertensive patients with extensive atherosclerosis. They usually originate beyond the left subclavian artery and may be either fusiform or sacular. Aneurysms of the aortic arch are often contiguous with aneurysms of the ascending or descending thoracic aorta.

### *Clinical Presentation*

More than half of thoracic aortic aneurysms are symptomatic; the rest are discovered only incidentally, often after a routine chest x-ray. Symptoms usually reflect pressure on a contiguous structure or consequences such as concomitant aortic insufficiency. Local mass effects may include a superior vena cava syndrome, caused by obstruction of the superior vena cava; pressure on the trachea, leading to cough or wheezing; and, occasionally, dramatic hemoptysis, resulting from fistula formation between the aneurysm and a major airway. Pressure on the esophagus may produce dysphagia. Pressure on the recurrent laryngeal nerve may result in hoarseness from vocal chord paralysis. Chest pain is usually caused by direct pressure of the aneurysm on an intrathoracic structure or by erosion of a bony structure.

A leaking or ruptured aneurysm usually presents with dramatic symptoms. Most such aneurysms leak or rupture into the left pleural space or pericardial sac, resulting in hypotension and the sudden onset of severe pain. Aortoesophageal fistulas are a very rare presentation that may produce life-threatening gastrointestinal bleeding.

### *Diagnostic Evaluation*

The diagnosis of thoracic aortic aneurysms is rarely suspected on physical examination. The thoracic aorta is generally not palpable. Rarely in the presence of a large aneurysm of the ascending aortic arch, the aortic impulse can be palpated just above the sternum or at the right upper sternal border.

The diagnosis is often initially suspected on chest x-ray and then confirmed with noninvasive or invasive imaging. On chest x-ray, most aneurysms appear as a widening of the mediastinal silhouette. Small aneurysms may not be detected. MRI and MDCTA are the most commonly used methods for delineating the size and extent of thoracic aneurysms. Transthoracic echocardiography (TTE) can be used to diagnose, measure, and monitor ascending aortic aneurysms, but it can evaluate only the proximal 3 to 5 cm of the ascending aorta and the arch. Neither TTE nor transesophageal echocardiography (TEE) is useful for evaluating aneurysms below the diaphragm. By using MDCTA and magnetic resonance imaging, it is possible to create a three-dimensional reconstruction of the aorta, which may provide unique information pertinent to diagnosis and management [see Figure 2].

### *Management to Reduce Risk of Aneurysm Rupture*

The natural history of a thoracic aneurysm can shed light on the disease process that has led to the aneurysm, on the risk factors that may affect the rate of aneurysm expansion, and on the concomitant presence of other vascular disease, including peripheral and coronary disease, that might affect long-term survival. Because size is a critical issue in terms of the risk of rupture, the initial size and potential growth of an aneurysm are important factors in the decision whether to operate on asymptomatic aneurysms. Aneurysms that are invading local structures or creating a marked vascular effect should usually be resected. Careful control of blood pressure is crucial for all patients and may require medical therapy, particularly with beta blockers, which may also slow the rate of aneurysm growth, particularly in the patient with Marfan syndrome.<sup>25</sup>

The initial size of a thoracic aneurysm is an important predictor of subsequent growth. In general, small aneurysms tend to grow slowly, whereas large aneurysms have a higher probability of expansion and rupture. On average, thoracic aneurysms grow at 0.4 cm/yr, but the growth rate varies greatly.<sup>26</sup> Small

aneurysms (i.e., < 5 cm in diameter) grow at about 0.1 cm/yr. Large aneurysms (i.e., > 5 cm) grow at 0.5 to 1.0 cm/yr. Although these average growth rates are reassuring, it should be emphasized that rapid expansion can occur and can dramatically affect the natural history and management. In general, thoracic aneurysms smaller than 5 cm in diameter are unlikely to rupture, whereas those larger than 7 cm are at high risk for rupture. Currently, most thoracic centers recommend surgery for aneurysms that exceed 5.5 cm in a patient who is otherwise an acceptable surgical candidate.<sup>27,28</sup> Because of their relatively young age, absence of associated disease, and low risk of elective repair, patients with Marfan syndrome should undergo surgery when aneurysms reach 5 cm in diameter, particularly if the aneurysm is expanding. At some high-volume centers, the threshold for elective repair of an ascending aortic aneurysm in a Marfan patient may be even lower.<sup>29</sup> As in the treatment of patients with abdominal aneurysms, the use of percutaneously placed aortic stent grafts may emerge as an attractive option in some patients with thoracic aneurysms.<sup>30</sup>

### *Surgical Treatment*

The choice of surgical procedure used to correct a thoracic aortic aneurysm depends on the location of the aneurysm. Ascending aortic aneurysms are typically treated by prophylactic surgical repair. Surgery is usually recommended when the aorta has a diameter of 5.5 cm; patients with Marfan syndrome are frequently recommended for surgery when the diameter of the aorta is 5 cm. The procedure usually involves the use of a composite graft that incorporates a bioprosthetic or mechanical valve. In skilled centers, a valve-sparing procedure may be used; in these procedures, the native aortic valve is left in place, and extensive reconstruction of the aorta is undertaken at the level of the sinotubular junction and, often, into the sinuses of Valsalva. The valve-sparing procedure obviates the need for an aortic valve replacement and leaves in place the native, reconstituted, and newly functional aortic valve.<sup>31,32</sup> A patient with an aneurysm of the arch or the descending thoracic aorta is typically recommended for surgical repair when the aneurysm reaches a diameter of 5.5 cm, when the aneurysm is rapidly expanding, or when the patient is symptomatic. As for the descending thoracic aorta, the surgical approach that is used is a resection of the aneurysm with an end-to-end anastomosis of a prosthetic graft. In the arch, the vasculature to the upper extremities and cerebral vasculature obviously will need to be accommodated. Most of the issues regarding endovascular therapy for abdominal aortic aneurysms (see above) also pertain to aneurysms of the descending thoracic aorta.

### *Postoperative Complications*

Neurologic sequelae are the most serious of potential postoperative complications. Currently, the risk of stroke after thoracic aneurysm resection ranges from 3% to 7%.<sup>33,34</sup> Efforts to reduce diffuse brain injury include hypothermic arrest and the use of either antegrade or retrograde cerebral perfusion.<sup>33,34</sup> Efforts to reduce central nervous system embolic events focus on meticulous surgical technique to avoid dislodging atheroemboli present in the aortic margins and to avoid air embolism during surgery. These issues are especially pertinent in aneurysms of the ascending aorta and arch. Surgery on the descending thoracic aorta carries a different neurologic risk—namely, postoperative paraplegia as a result of interruption of the supply of arterial blood to the spinal cord; postoperative paraplegia has been reported in up to 5% of patients. The incidence of spinal cord complications

is related to the extent of thoracic aortic replacement. Several methods have been devised to deal with this risk, but no definitive solution has yet emerged. Some centers have suggested that reattaching critical intercostal arteries leads to improved outcome,<sup>35</sup> whether or not epidural cooling of the spinal cord is employed during the surgery.<sup>36</sup>

### Aortic Dissection

The incidence of recognized aortic dissection in the United States is estimated to be 10 to 20 per million population, or about 5,000 cases a year. We stress, however, that the incidence of MI is greater than 500,000 cases annually; that is, MI is at least 100 times more common than aortic dissection. For most patients, dissection entails a tear in the intima, with the subsequent development of a propagating hematoma between the intima and the adventitia. Approximately two thirds of aortic dissections are initiated by a tear in the intima just above the aortic valve. Most of the remaining cases develop in the descending aorta at the attachment of the ligamentum arteriosum. Often, multiple communication sites are present between the true lumen and the false lumen, and the site of the initial tear cannot be determined with certainty. The dissection often spirals as it courses retrograde or antegrade along the aorta. Approximately 5% of aortic dissections are caused by intramural hematoma, which is spontaneous rupture of the vaso vasorum within the media, creating a hematoma in the media without a communication into the lumen.<sup>37</sup> This hematoma may extend a variable distance and may eventually rupture into the lumen, resulting in a more typical dissection.

#### CLASSIFICATION

Aortic dissections are classified as acute or chronic and according to their location. Dissections are termed acute when they are diagnosed within 2 weeks after the onset of symptoms; dissections diagnosed after 2 weeks of symptom onset are termed chronic. A key feature for classification is involvement of the ascending aorta, regardless of where the dissection began. Ascending aortic dissections are also called type A dissections. Dissections not involving the ascending aorta are typically classified as distal, or type B, dissections. Ascending aortic involvement identifies a patient population with high mortality if not treated surgically. A subset of patients with isolated aortic arch dissection has also been described.

The predisposing factors for type A and type B dissections differ somewhat. Disorders of the media that result in cystic medial necrosis are a precursor of type A dissection. Affected patients may include those with Marfan syndrome or other heritable connective tissue disorders. Another risk factor for ascending aortic dissection is aortic valve disease, such as a bicuspid valve. Although these conditions are classically associated with aortic dissection, over 90% of patients with acute aortic dissection do not have any recognized substrate for dissection.<sup>38,39</sup> Distal, or type B, aortic dissection is most often seen in patients with long-standing hypertension. Patients with type B dissection are older on average than patients with type A dissection. An unexplained relation between aortic dissection and pregnancy also exists, perhaps because of changes in cardiac output, blood pressure, or blood volume or the effects of pregnancy on the aortic wall itself.<sup>40</sup> Aortic dissection after inhalation of crack cocaine has also been reported.

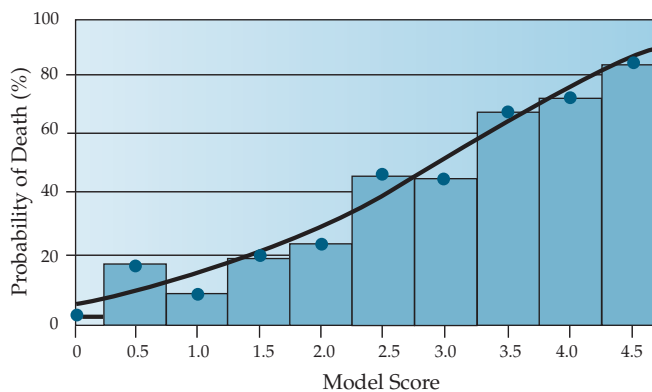
#### CLINICAL PRESENTATION

The most common distinguishing clinical feature of aortic dis-

section is the abrupt onset of pain.<sup>38</sup> The abruptness of onset is one of the clinical features reliably distinguishing the pain of aortic dissection from that accompanying other cardiovascular pathology (e.g., myocardial ischemia). This instantaneous pain may begin in the chest or back and may migrate to involve the neck, head, back, and legs as the dissection propagates. All three components of the classic triad—abrupt tearing pain, pulse deficits, and aortic insufficiency—are seldom observed in a single patient.<sup>38,41</sup> Other presentations of type A dissection are syncope,<sup>42</sup> abdominal pain,<sup>43</sup> or hypotension,<sup>44</sup> resulting from dissection into the pericardial space; stroke, resulting from interruption of the blood supply to one or both internal carotid arteries; and, in rare instances, isolated congestive heart failure, when the dissection involves the ascending aorta and interrupts aortic valve function. Approximately 6% of patients with acute dissection may present without pain.<sup>45</sup>

The typical presentation of type B dissection is onset of severe interscapular pain, which may radiate down the back toward the legs. Type B dissection is frequently accompanied by hypertension, whereas type A dissection more often occurs in the presence of normal or low blood pressure.<sup>38</sup> Ischemia of the spinal cord, limbs, and mesentery is most frequently encountered in type A dissection that has extended to involve the descending aorta. Aortic insufficiency is noted on auscultation in 35% to 50% of the cases of ascending aortic dissection; however, it is rather unusual in cases of type B dissection. Pulse deficits are seen in about 25% of patients with type A dissection and in perhaps 5% to 10% of patients with type B dissection.<sup>38</sup>

Acute dissection remains highly lethal. Mortality is commonly quoted as 1% per hour for the first 24 hours. Advanced age, hypotension, and limb and visceral ischemia are all predictors of greater mortality.<sup>43-49</sup> A published review of 500 patients with acute type A dissection identified a number of clinical factors that are predictors of death [see Figure 3].<sup>48</sup> Mortality in the study cohort ranged from 10% to 80%, depending on the number of adverse risk factors.<sup>48</sup> Specific subsets of patients may have a markedly worse prognosis, including those with periaortic hematoma<sup>50</sup> and those presenting with painless dissection.<sup>45</sup>



**Figure 3** Graphic demonstration of the increasing mortality in type A aortic dissection when multiple risk factors are present. There is an observed increase in mortality in type A dissection that parallels that of the predictive model. The total risk score was the sum of individual risk factors that were determined from regression analysis to be significantly linked to outcome. The individual factors and their individual scores (in parentheses) were as follows: renal failure (1.6), hypotension/shock/tamponade (1.1), abrupt onset of pain (1.0), pulse deficit (0.7), abnormal ECG (0.6), age  $\geq 70$  (0.5), female (0.3). (bars—observed findings; line—model probabilities)

## DIAGNOSTIC EVALUATION

Because acute aortic dissection is a life-threatening emergency, rapid and accurate diagnosis is crucial to patient survival. Therefore, sophisticated imaging modalities may be required. Although type A dissection may affect one of the coronary arteries and lead to a transmural MI in 1% to 2% of patients, more common findings include nonspecific ST-T wave changes or left ventricular hypertrophy related to long-standing hypertension.

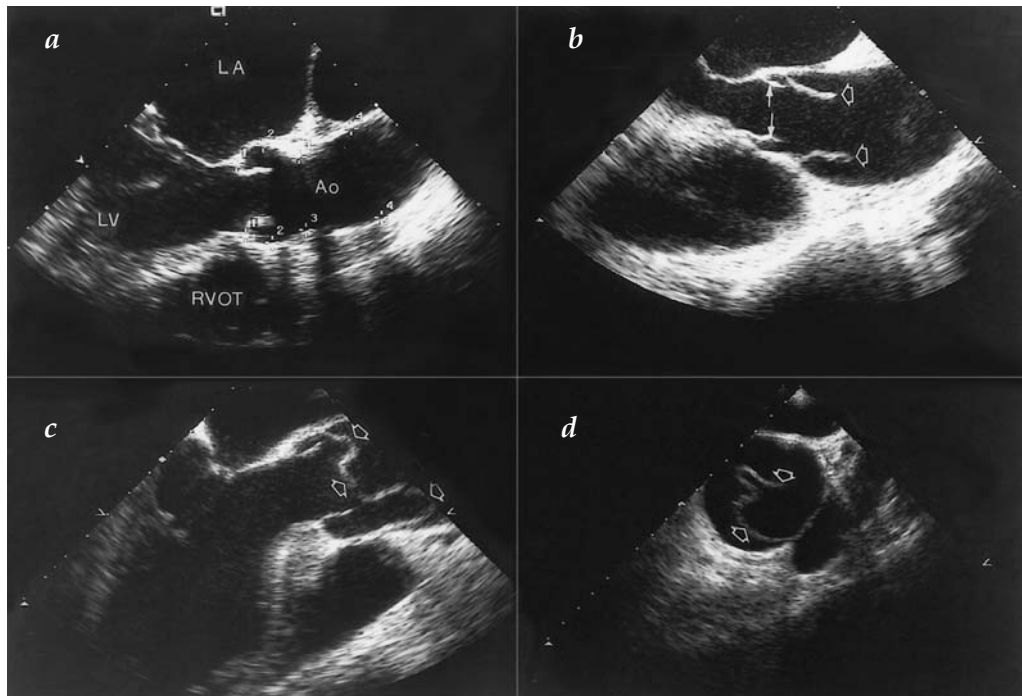
The typical chest x-ray reveals widening of the mediastinal silhouette; it may also demonstrate evidence of a pleural effusion, cardiomegaly, or congestive failure if severe aortic regurgitation is present. A normal-appearing chest x-ray is seen in more than 10% of documented cases of acute aortic dissections.<sup>38</sup> Other laboratory abnormalities are generally nonspecific. An increase of smooth muscle myosin is present in more than 85% of patients presenting within 3 hours after onset of acute aortic dissection.<sup>51</sup> Other biomarkers, including D-dimer,<sup>52,53</sup> and the presence of soluble elastin fragments in the serum<sup>54</sup> have shown promise for both diagnosis and prognosis in acute aortic dissection.

After a careful history and physical examination, the key to diagnosis is rapid identification of the aortic dissection, ascertainment of whether the ascending aorta is involved, and urgent

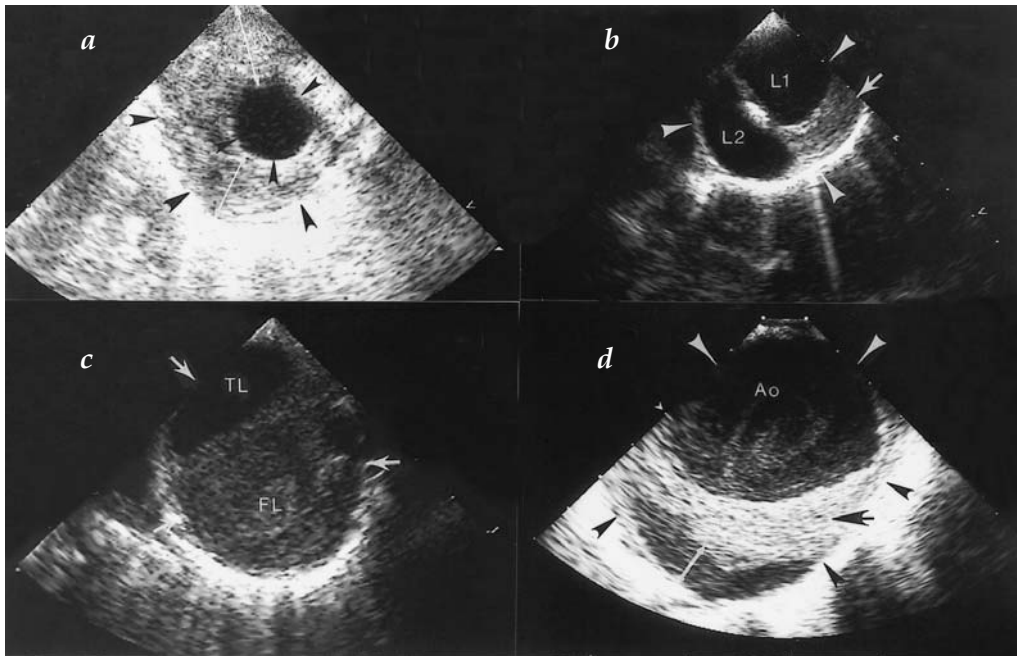
cardiac surgery if proximal aortic dissection is diagnosed. The importance of rapid diagnosis and institution of definitive therapy for aortic dissection cannot be overemphasized. Given the 1% to 2% mortality per hour in the first 24 hours after presentation, even brief delays in diagnostic imaging are unacceptable.

Currently, four diagnostic tools are used to evaluate patients with suspected dissection: MDCTA, echocardiography, MRI, and aortography.<sup>55-61</sup> In general, the choice of which imaging modality to initially employ will depend on local expertise and availability. In most hospitals, the choice is either MDCTA or TEE, and the majority of patients undergo more than one imaging study.<sup>61</sup>

MDCTA is widely available in most community and tertiary-care hospitals. MDCTA scanning gives greater resolution than is available using the older scanners, and its reported sensitivity and specificity for aortic dissection exceeds 95%.<sup>57</sup> TEE offers significant advantages in diagnosis [see Figures 4 and 5]. The primary attraction of TEE is its portability, making it suitable for performance in the emergency department, intensive care unit, or operating room. In addition to being highly sensitive for the identification of type A dissection, TEE is also useful when involvement of the aortic valve and the status of the left ventricle, pericardial space, and right and left coronary artery ostia are unknown.



**Figure 4** Transesophageal echocardiograms from a patient with a normal ascending aorta (panel A) and three different ascending aortic dissections (panels B through D). In the normal ascending aorta, the cardiac chambers are noted. The ascending aorta is well visualized, including its annulus (point 1), the coronary sinuses (point 2), the sinotubular junction (point 3), and the true ascending aorta (point 4). Note that the aorta dilates at the level of the sinuses, narrows at the sinotubular junction to a dimension equivalent to that of the annulus, and then slightly dilates further in the ascending aorta. Shown is a normal aortic valve in its open position. Panel B was recorded in a patient with a proximal aortic dissection. The orientation is identical to that in panel A. The solid arrows denote the position of an open aortic valve leaflet. The open arrows represent the margins of a dissection that originated at the sinotubular junction and extended distally. Panel C was recorded in a patient with an ascending aortic dissection (orientation identical to that in panels A and B), and the aortic valve is open. In this instance, a convoluted intimal flap (open arrows) is clearly visualized in the proximal ascending aorta. Panel D was recorded in the short axis of the aorta in a patient with an aortic dissection. In the circular ascending aorta, multiple convolutions of an intimal flap are clearly visualized (open arrows). Note that a communication point (between the downward pointing arrow and the wall of the aorta) allows free communication of flow between the two lumens. (Ao—ascending aorta; LA—left atrium; LV—left ventricle; RVOT—right ventricular outflow tract)



**Figure 5** Panels A through D represent four transesophageal echocardiograms recorded in a short-axis view of the descending thoracic aorta in patients with aortic pathology. Panel A was recorded in a patient with an ascending aortic aneurysm and a large periaortic (adventitial) hematoma extending distally along the thoracic aorta. The smaller black arrows denote the boundaries of the normal-diameter descending thoracic aorta. The larger black arrows pointing inward mark the full dimension of the periaortic hematoma; the full dimension is also noted by the double-headed white arrows. In this instance, the intima of the descending thoracic aorta was not involved in the dissection process. However, a large periadventitial hematoma ruptured along the course of the descending thoracic aorta. Panel B was recorded in a patient with an aortic dissection localized to the descending thoracic aorta. The maximum external dimensions of the aorta are noted by the large white arrowheads. The white arrow notes an area of atherosclerosis and thrombus within the aorta. Two distinct lumens (L1 and L2) can be seen at this level. Panel C was recorded in a patient with an aortic dissection extending from the aortic valve to the bifurcation of the aorta. The large white arrows denote the outer dimension of the aorta. There is an echo-free lumen, or true lumen (TL), and a false lumen (FL) with early thrombus formation. Note the vague echo densities within the false lumen. Panel D was recorded in a patient with a large descending thoracic aortic aneurysm and intramural hematoma. The large arrowheads (black and white) denote the outer dimensions of the aorta. The dilated aortic lumen (Ao) is also noted. The black arrow denotes an area of marked atherosclerosis within the aorta, and the double-headed white arrow denotes an area of intramural hemorrhage, characterized by a lower echo density than the atherosclerotic components. Note also the low-density echoes, which represent stagnant blood flow within the aorta.

TEE can be very useful in detecting the mechanism of aortic insufficiency and in determining the feasibility of repair.<sup>62,63</sup> Valves in which aortic insufficiency is the result of sinotubular dilatation or extension of the dissection into the sinus are often candidates for repair. Patients with intrinsic disease of the aortic valve leaflets are less optimal candidates for repair.

MRI is less commonly used unless the MRI scanner is part of the emergency department. For most hospitals, however, the delay required in getting a patient into the MRI suite and completing the study makes this technology less efficient than TEE or MDCTA.

Finally, although aortography is still used in some hospitals, it is seldom the initial test for aortic dissection. The reported false negative rate for aortography ranges from 5% to 15%.<sup>64</sup> Aortography frequently misses lesions such as an intramural hematoma. In addition, the time required to get a patient to an angiography suite and complete the study is generally considerably longer than that for TEE or MDCTA. Our medical center and many others follow an algorithmic approach to the evaluation and treatment of a suspected aortic dissection [see Figure 6].

#### TREATMENT

The treatment of aortic dissection includes aggressive medical therapy for all patients and definitive surgical therapy in selected patients. The decision to perform surgery depends first and foremost on the site of the aortic dissection [see Figure 6].

#### *Surgical Repair*

**Type A aortic dissection** Any involvement of the ascending aorta carries with it a much greater risk of rupture into the pericardial space; development of coronary or cerebral ischemia, aortic regurgitation, and congestive heart failure; or free rupture of the aorta into the thorax. Thus, definitive surgical repair is carried out as quickly as possible for patients with proximal or type A aortic dissection who are appropriate candidates for the procedure.<sup>65,66</sup>

For patients with type A dissection complicated by malperfusion, medical therapy plus percutaneous reperfusion utilizing aortic stenting or fenestration, or both, and selective branch stenting may allow stabilization and reduce risk associated with the operation. After a period of recovery, repair of the patient's ascending aorta may be undertaken.<sup>67,68</sup>

Definitive aortic repair includes resection of the dissected aorta and insertion of an aortic graft. In high-volume centers, valve replacement is required in only 25% of cases.<sup>65</sup> For most patients, the aortic repair includes reimplantation of the coronary arteries. In some patients, this repair includes resection and placement of a graft to the aortic arch. Even in the best of centers, surgical mortality ranges from 10% to 35%, depending on comorbidity.<sup>48,65,69,70</sup> Major contributors to surgical mortality include hypotension and shock, pulse deficits, and the presence of cardiac tamponade.<sup>70</sup>

**Type B aortic dissection** Surgery for type B dissection is indicated for patients with life-threatening complications that require a surgical approach.<sup>71,72</sup> Examples include patients who experience ischemia of both kidneys, leading to reversible renal failure; development of bowel ischemia; ischemia involving one of the legs or arms; development of a progressive aneurysm; impending rupture; and recurrent extension of the dissection. In some centers, percutaneous insertion of aortic stents or endovascular stent grafts has been used to stabilize dissections of the descending aorta.<sup>73-77</sup> This strategy may be preferable to surgery in some candidates. In particular, stenting may promote thrombosis of the false channel and thereby reduce the long-term risk of aneurysm formation and aortic rupture.

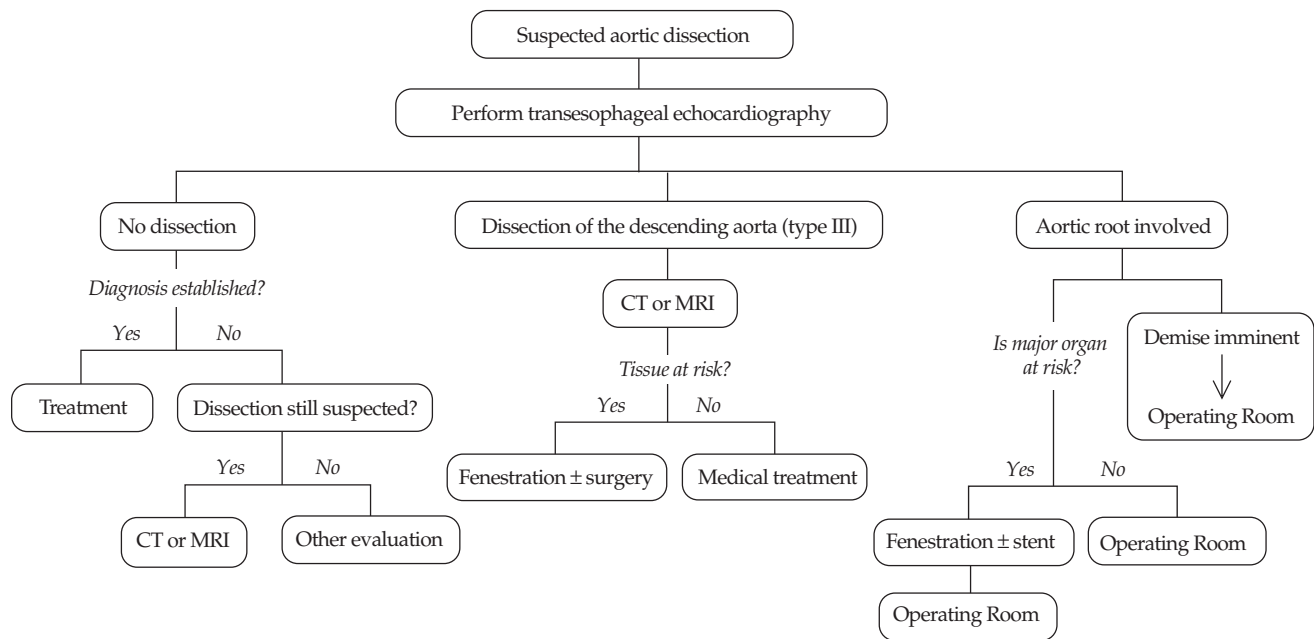
#### Postoperative Complications

The same issues regarding operative complications for ascending and descending aortic aneurysms apply to dissection,

including the need to preserve cerebral perfusion. Because the operation to repair acute dissection occurs in an emergency setting, complication rates may be higher.

#### Medical Therapy

All patients with aortic dissection receive aggressive medical therapy. This treatment is first directed at controlling the blood pressure.<sup>78</sup> For hypertensive patients, administration of intravenous beta blockers followed by oral beta blockers, along with the concomitant administration of intravenous or oral vasodilators, is imperative. Heart rates should be maintained at or below 60 beats/min, and the blood pressure should be kept as low as possible while allowing perfusion of the brain, kidneys, and other vital organs. For normotensive patients, the heart rate should be low (at or below 60 beats/min), and blood pressure should be maintained in the low-normal range. The likelihood of the propagation of dissection is believed to be in part related to acceleration of flow in the aorta—that is, the force of the aortic jet per unit time (i.e.,  $dP/dt$ ). Accordingly, beta blockers have been the most important therapy for the medical treatment of aortic dissection. Long-term management of aortic dissection requires aggressive medical therapy and careful surveillance. Patients who retain patency in the false channel of the aorta after either medical treatment or surgical repair have a significant risk of aneurysm formation and rupture of the false channel, especially in the first 6 months after initial therapy. Expansion, rupture, or both are more common in patients who are older and have poorly con-



#### Indications for Surgical Intervention in Aortic Dissection

Ascending Aorta Only	Ascending and Descending Aorta	Descending Aorta Only
Emergent repair in appropriate candidates	Emergent repair if any of the following are present: Aortic insufficiency with CHF Hypotension Pericardial effusion with compromise If ischemia in kidney, CNS, bowel, or major limb, attempt stabilization with stent or fenestration before surgery	Emergent repair if any of the following are present: Rupture Impending rupture Uncontrolled pain Major organ at risk (stabilization with stent or fenestration not feasible)

Figure 6 Decision algorithm for evaluation and treatment of a suspected aortic dissection. Type 3 dissection originates in the descending aorta and extends distally down the aorta or, in rare instances, retrograde into the aortic arch and ascending aorta. (CHF—congestive heart failure)

trolled hypertension and chronic obstructive pulmonary disease.<sup>79</sup> Aggressive treatment of high blood pressure and heart rate and careful monitoring of the patient's status with physical examination and noninvasive imaging are essential. At many centers, either MDCTA or MRI is performed on a regular basis after initial treatment of the dissection. Imaging of the aorta is repeated 3 to 6 months after surgery to screen for the development of aneurysm in the false channel or at the margins of a surgical repair. Persistence of blood flow in the residual false lumen may indicate that a patient is likely to experience continued expansion of the aneurysmal aorta.<sup>80</sup> After the 6-month screening, patients undergo aortic imaging annually, and scrupulous attention is directed to antihypertensive therapy and modification of risk factors. Following successful surgical repair of type A dissection, long-term survival is most influenced by the preexisting underlying comorbidities, including age, prior cardiac surgery, and atherosclerosis.<sup>81</sup>

### Atypical Aortic Dissection

Several forms of acute aortic pathology present in a manner similar to that of classic acute aortic dissection. Because of the similar clinical manifestations, the term acute aortic syndrome has been applied to these presentations.<sup>82</sup>

#### AORTIC DISSECTION WITHOUT INTIMAL TEAR

About 5% to 10% of patients presenting with symptoms suggestive of aortic dissection actually have aortic dissection without an intimal tear (intramural hematoma).<sup>37,83-90</sup> This hemorrhage into the medial layer of the aorta may produce a localized or discrete hematoma or may extend for a various distance by dissecting along the plane of the aortic media. Clinically, this hemorrhage mirrors aortic dissection in terms of both its risk factors and its presentation. Intramural hematoma is generally not identified on aortography. It is most easily diagnosed with MDCTA. With noncontrast imaging, the hematoma appears as a crescent-shaped high-attenuation area along the aortic wall; moreover, this region cannot be enhanced with contrast imaging. MRI reveals the same crescent-shaped high-intensity area, whereas on TEE, an intramural hematoma may appear as a circular or crescentic thickening.

Studies of the natural history of the intramural hematoma suggest that the outcome is similar to that of classic aortic dissection,<sup>84,85,88</sup> although some recent studies have suggested a more benign prognosis.<sup>86,89</sup> By 30 days, the rate of aortic expansion or death in patients with medically treated ascending aortic intramural hematoma approaches 50%. Patients with aneurysmal aortas are at particular risk.<sup>86</sup> By contrast, the mortality for intramural hematoma in the descending aorta appears to be between 10% and 15%, a rate similar to that for type B aortic dissection. Generally, therapy for acute intramural hematoma is the same as for classic aortic dissection.

#### PENETRATING ATHEROSCLEROTIC ULCER

A second form of aortic disease that may have an acute presentation is a penetrating atherosclerotic ulcer<sup>90,91</sup> [see Figure 7]. Penetrating ulcers result from erosion of the intima of the aorta, usually because of extensive atherosclerosis. Ulcer formation may produce a hematoma in the media that extends several centimeters from its origin up or down the aorta. Occasionally, pseudoaneurysms are created that may extend into the adventitia and, in rare instances, may rupture. This aortic process devel-



**Figure 7** Contrast-enhanced MDCTA of the arch of the aorta in a patient presenting with acute chest pain. In the proximal arch, note the three penetrating ulcers (arrows) diagnosed by the radiographic contrast protruding out of the anticipated border of the aortic arch.

ops gradually in elderly patients with extensive atherosclerosis and often is heralded by chest pain or back pain and hypertension. Because it usually presents as a localized process, it is seldom associated with other symptoms of aortic dissection, such as pulse deficit, aortic valve regurgitation, or neurologic defects. The management of penetrating ulcers remains controversial. Asymptomatic patients who experience progressive enlargement or recurrent atheroemboli may require surgical therapy. Endovascular therapy can be used to exclude the ulcerated area from the lumen and stabilize the aortic wall. For most patients, however, medical therapy suffices; management entails aggressive treatment of atherosclerotic risk factors, including cessation of smoking, control of hypertension, aggressive lipid-lowering therapy, and careful surveillance. The role of antiplatelet or anticoagulant therapy for this condition is not clear.

### Aortic Atheromatous Emboli

Atherosclerosis of the aorta may be so extensive that it leads to overlying thrombosis and subsequent dislodgment of thrombi, cholesterol particles, or fibrinous material into the cerebrovascular or peripheral circulation. Risk factors are hypertension, diabetes, hyperlipidemia, advanced age, and other vascular diseases. Atheromatous disease is most common in the distal aorta but may also occur in the ascending aorta and arch. Evidence of ulceration of atherosclerotic plaques is an independent risk factor for stroke, as is the identification of a mobile, large, protruding aortic atheroma detected with TEE. Plaques more than 4 mm in dimension (dimension refers to the maximum size of the atheroma that protrudes into the lumen of the aorta) in the ascending aorta are particularly associated with an increased risk of ischemic stroke.<sup>92</sup> Atheroemboli or cholesterol-particle emboli may also involve the peripheral extremities, leading to ischemic lesions on the feet or toes (so-called blue-toe syndrome). These emboli may present as abdominal pain as a result of bowel ischemia. Acute nonoliguric renal failure is another occasional manifestation, as is gastrointestinal bleeding or pancreatitis. Cutaneous involvement may produce a characteristic skin lesion called livedo reticularis.

Cholesterol embolism syndrome is most common in patients following cardiac catheterization or other angiographic procedures in which catheters or wires are manipulated within the aorta. Because the occurrence of atheroemboli may be delayed after aortic manipulation, the relation between the two may not be apparent when the patient is first examined. If cutaneous manifestations are present, a biopsy of the lesions will often identify needle-shaped clefts in the arteriolar lumen characteristic of cholesterol embolization.

Treatment of cholesterol embolism syndrome begins with avoidance of further aortic manipulation (e.g., cardiac catheterization), if this has been a precipitant. Aggressive treatment of hypercholesterolemia is warranted. A search for an aortic aneurysm or protruding mobile atheromas is appropriate in patients for whom the syndrome develops without a concomitant iatrogenic source. Occasionally, recurrent emboli warrant the resection of an aneurysm or of a severely diseased segment of atheromatous aorta.<sup>93</sup> The role of anticoagulant and antiplatelet drugs in this syndrome is uncertain.

## Takayasu Arteritis and Giant Cell Arteritis

### TAKAYASU ARTERITIS

Takayasu arteritis is a rare inflammatory condition that affects the aorta and its major branches. Other names include aortic arch syndrome, pulseless disease, and young female arteritis.<sup>94,95</sup> Although Takayasu arteritis is seen throughout the world, most cases occur in Asia and Africa. A specific etiologic agent has yet to be identified, but current evidence favors an autoimmune mechanism. Some studies suggest it may be linked to rheumatic fever, streptococcal infections, certain HLA subtypes, rheumatoid arthritis, and other collagen vascular diseases. Takayasu arteritis is substantially more prevalent in women than in men. Patients are younger than 40 years; the average age is 29 years. Takayasu arteritis has been divided into three types.<sup>95</sup> Type I involves the aortic arch and its branches, type II involves the distal aorta and spares the arch, and type III may affect both the ascending aorta and the descending aorta. A suggested fourth category involves the pulmonary arteries.

#### *Pathophysiology*

Takayasu arteritis is a granulomatous arteritis of the aorta and its branches, with subsequent involvement of the media and adventitia. Later, the disease may progress to a sclerotic stage in which the intima is hyperplastic, the media degenerates, and the adventitia develops fibrosis. This late fibrotic process may encroach on the lumen of the aorta or its branches. Common areas of involvement are the main aorta and branch points of its major branch vessels. The pulmonary artery may also be involved. The coronary arteries are affected in fewer than 10% of patients. In some patients, involvement of the ascending aorta may lead to aortic valve regurgitation.

#### *Clinical Presentation*

The initial symptoms are often typical of an acute or systemic inflammatory process, including fever, loss of appetite, weight loss, night sweats, and arthralgias. Involved vessels may have accompanying localized tenderness over them. By the time the diagnosis is established, most patients have reached a sclerotic phase, in which vascular insufficiency is causing the predominant symptoms. It may involve the upper or lower extremities.

Hypertension occurs in more than half of patients. Congestive heart failure occurs in 25% of patients because of hypertension, aortic valve insufficiency, or involvement of the coronary arteries. Aneurysms may develop in 15% to 25% of patients.

#### *Diagnostic Evaluation*

Laboratory findings in patients with Takayasu arteritis generally include an elevated erythrocyte sedimentation rate, mild anemia, and a slightly increased white blood cell count. The chest x-ray may demonstrate a rim of calcification around the involved vessels. Aortography often shows an irregular intimal surface with stenoses of the aorta or its branch arteries. Poststenotic dilatation or frank arterial aneurysms may be visible. Similar diagnostic features can also be detected by TEE and MRI,<sup>96</sup> which may also detect arterial wall thickening and edema.

#### *Treatment*

The management of Takayasu arteritis begins with high-dose glucocorticoid therapy, which usually leads to abatement of constitutional symptoms and the laboratory signs of inflammation. Serial sedimentation rates are useful for monitoring the benefits of treatment. For patients who fail to respond to steroid therapy, cyclophosphamide has been used. Alternatively, low-dose methotrexate may enhance the efficacy of steroids or allow steroid tapering. Surgery may be necessary to treat unremitting peripheral ischemia or aortic valve disease or to treat renal artery stenosis that causes severe hypertension. For patients with involvement of the coronary ostia, bypass surgery may be indicated as well. Percutaneously placed arterial stents have been successfully used to treat segmental disease in a variety of vessels in patients with this syndrome.

### GIANT CELL ARTERITIS

Giant cell arteritis is another form of aortoarteritis, which pathologically is similar to Takayasu arteritis. In contrast to Takayasu arteritis, this illness is more commonly seen in Europe and the United States and in patients older than 50 years (the mean age at onset of disease is 67 years).

#### *Pathophysiology*

This form of arteritis often affects the branches of the proximal aorta, particularly the branches supplying the head and neck, the extracranial structures (including the temporal arteries), and the upper extremities. Aortic involvement often coexists with temporal arteritis and polymyalgia rheumatica. Unlike Takayasu arteritis, giant cell arteritis less often has a sclerotic phase that progresses to occlusion of vessels. However, giant cell arteritis may lead to aneurysm formation, aortic regurgitation, or aortic dissection.<sup>97,98</sup>

#### *Clinical Presentation*

The classic presentation of giant cell arteritis consists of headache, tenderness over involved arteries in the scalp or the temporal region, jaw claudication, difficulty combing one's hair, and constitutional symptoms. Fever is common, and the blood vessels involved are thick and tender. Pulses may be diminished, and bruits may be present. Occasionally, signs of aortic valve regurgitation are present.

A serious complication of this syndrome is blindness, which results when arteritis affects the ophthalmic artery. The progression to total blindness may be rapid. Visual symptoms of some type occur in as many as 50% of patients. An initial high dose followed by prolonged therapy with corticosteroids remains the

treatment of choice.<sup>99</sup> In rare instances, giant cell arteritis may lead to diminished upper-extremity pulses and reductions in blood pressure, as well as arm or leg claudication. It also may cause coronary ischemia or abdominal angina in rare cases. Unlike Takayasu arteritis, giant cell arteritis virtually never affects the kidneys. Aortic aneurysms occur in 15% of patients with giant cell arteritis, most commonly involving the ascending aorta. Such aneurysms may develop late in the disease, leading to rupture, aortic dissection, or severe aortic valve regurgitation.

### Diagnostic Evaluation

A higher-than-normal erythrocyte sedimentation rate is characteristic of this disease. The diagnosis is confirmed by biopsy of an involved artery, usually the temporal artery. Clinicians need to be aware, however, that the results of temporal artery biopsy may be negative in as many as 15% of patients with confirmed disease; therefore, a second biopsy may be necessary in patients with a high likelihood of temporal arteritis. Temporal artery ultrasound may detect evidence of arterial inflammation and thickening and be used for diagnosis or to direct a targeted biopsy.<sup>100,101</sup>

### Treatment

Standard therapy for giant cell arteritis is high-dose glucocorticoid therapy. Methotrexate may be used to reduce the need for steroids or to treat patients who respond inadequately to steroids. Cyclophosphamide may also be useful for reducing the need for glucocorticoids. Surgery is typically reserved for patients who experience progressive ischemic symptoms or aortic aneurysms.

### Traumatic Disease of the Aorta

Partial or complete transection of the aorta occurs as a result of major blunt thoracic trauma, most commonly as a result of a high-speed motor vehicle accident. Most patients with complete aortic transection do not survive long enough for hospital evaluation. If rapidly diagnosed, patients with partial transection may survive long enough to undergo surgical correction. Evidence of aortic trauma is often obscured by other major organ trauma. Patients with aortic transection are typically in shock related to the major trauma and may have diminished lower-extremity pulses. The transection is usually located at the distal arch, immediately after the origin of the left subclavian artery.

Rapid diagnosis is the key to the survival of patients with aortic transection. The routine chest x-ray typically reveals a widened mediastinum, often with pleural effusions. The gold standard for diagnosis of this disorder remains aortography, but TEE,<sup>102</sup> MDCTA,<sup>103</sup> and MRI have also been used successfully.<sup>104</sup> Successful treatment requires vigorous fluid and blood resuscitation and surgical repair of the aortic transection.

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