Acute aortic syndromes encompass a number of life-threatening aortic emergencies. These include aortic dissection, penetrating atherosclerotic ulcer, intramural hematoma, and aortic aneurysmal leakage or ruptured abdominal aortic aneurysm (see chapter 60, Aneurysmal Disease).

Acute aortic syndromes are uncommon but frequently fatal. The incidence ranges from 2.9 to 4.7 cases per 100,000 people per year. The 1-year, 5-year, and 10-year actuarial survival rates are 92%, 77%, and 57%, respectively, for operative patients. Twenty-two percent of cases are undiagnosed prior to death. The most common cardiovascular complication of Marfan's syndrome is aortic root disease and type A dissection (ascending aorta). The identification of gene mutations associated with Marfan's syndrome, such as \(TGFBR2\) and \(FBN1\), combined with regular follow-up can reduce fatal outcomes.

**PATHOPHYSIOLOGY**

Acute aortic syndromes occur in the setting of chronic hypertension and other factors that lead to degeneration of the media of the aortic wall. Bicuspid aortic valve, Marfan's syndrome, Ehlers-Danlos syndrome, and familial history of aortic dissection all predispose to aortic syndromes. Chronic cocaine or amphetamine use accelerates atherosclerosis, increasing the risk for dissection. Prior cardiac surgery is another risk factor for aortic dissection. All mechanisms involve weakening of the medial layer and intimal wall stress. Response to stress may include aortic dilation, aneurysm formation, development of a penetrating ulcer, intramural hemorrhage, aortic dissection, and aortic rupture.

Aortic dissection occurs after a violation of the intima allows blood to enter the media and dissect between the intimal and adventitial layers. The dissecting column of blood forms a false lumen and may extend proximally, distally, or in both directions. Blood may dissect and reenter the intima, and this may clinically suggest a spontaneous cure. Alternatively, the blood may dissect through the adventitia, which nearly always proves rapidly fatal.

Aortic dissection has a bimodal age distribution. The first peak involves younger patients with specific predisposing conditions such as connective tissue disorders. The second peak includes those aged >50 years.
with chronic hypertension. Other atherosclerotic risk factors appear to be only minor contributors to pathogenesis of acute aortic syndromes.

Aortic dissections are classified using two separate systems, the Stanford and DeBakey systems. The Stanford classification considers any involvement of the ascending aorta a type A dissection. Stanford type B dissections are restricted to only the descending aorta. DeBakey type 1 dissections simultaneously involve the ascending aorta, the arch, and the descending aorta. DeBakey type 2 dissections involve only the ascending aorta, and type 3 involve only the descending aorta.

An aortic intramural hematoma results from infarction of the aortic media, usually from injury to the vasa vasorum. An intramural hematoma may resolve spontaneously or may lead to dissection. Penetrating atherosclerotic ulcer can lead to intramural hematoma, aortic dissection, or perforation of the aorta.

**CLINICAL FEATURES**

**HISTORY**

The site of initial intimal disruption predicts the initial symptoms. Symptoms may change as the dissection extends along the aorta or involves other arteries or organs. Classically, dissection presents with abrupt and severe pain in the chest that radiates to an area between the scapulae and may be accompanied by a feeling of impending doom. In a case series of 464 dissections, 60% of patients had anterior chest pain (more common in Stanford type A); abdominal pain is more common in Stanford type B. Most patients describe the pain as severe or the worst they have ever experienced. Sixty-four percent describe it as sharp pain and 50% as tearing or ripping pain. Syncope occurs almost 10% of the time (more common in Stanford type A).

Twenty-two percent of dissections occur in patients with prior cardiac surgery.

Dissection in or near a carotid artery may present as a classic stroke, and 20% of patients with type A dissection display neurologic findings, which predicts a poorer prognosis. Interruption of blood supply to the spinal cord may lead to paraplegia. Further distal dissection may present as back, flank, or abdominal pain. A proximal dissection to the aortic root may lead to cardiac tamponade and is generally fatal.

**PHYSICAL EXAMINATION**

For most patients with aortic dissection, examination findings are relatively normal. An aortic insufficiency murmur may occur (32%), and a pulse deficit in radial arteries or femoral arteries can be found (15%). Hypertension is common (49%), but hypotension occurs in 18% to 25% and worsens prognosis. Aneurysmal dilation of the aorta may compress regional structures such as the esophagus, the recurrent laryngeal nerve, or the superior cervical sympathetic ganglion, causing dysphagia, hoarseness, or Horner syndrome.
Using retrospective data from the International Registry of Acute Aortic Dissection,\textsuperscript{15,16} three clinical categories (underlying condition; pain quality and location; examination findings) were parsed into 12 features\textsuperscript{15,16} associated with acute aortic dissection (Table 59-1).

\begin{table}
\centering
\caption{Acute Aortic Dissection: Features From the International Registry of Acute Aortic Dissection}
\begin{tabular}{|l|l|l|}
\hline
\textbf{Category 1: Underlying Condition} & \textbf{Category 2: Pain in Chest, Back, or Abdomen} & \textbf{Category 3: Abnormal Examination} \\
\hline
Marfan’s syndrome & Abrupt onset & Systolic blood pressure differential in extremities or pulse amplitude difference \\
Family history of aortic disease & Severe in intensity & Focal neurologic deficit and chest, back, or abdominal pain \\
Aortic valvular disease & Ripping or tearing & New murmur of aortic insufficiency and chest, back, or abdominal pain \\
Recent aortic manipulation & & Shock or hypotension \\
Thoracic aortic aneurysm & & \\
\hline
\end{tabular}
\end{table}

\section*{DIAGNOSIS}

The large differential diagnosis for chest pain plus the many end-organ ischemic manifestations associated with aortic dissections make the diagnosis challenging. The most important differential diagnoses are listed in Table 59-2.

\begin{table}
\centering
\caption{Differential Diagnosis of Aortic Dissection}
\begin{tabular}{|l|}
\hline
Myocardial infarction or acute coronary syndromes \\
Pericardial disease \\
Stroke \\
Musculoskeletal disease of the extremity \\
Spinal cord injuries and disorders \\
Intra-abdominal disorders \\
Pulmonary disorders, including pulmonary embolus, pneumonia, pleurisy, pneumothorax \\
\hline
\end{tabular}
\end{table}
Ischemic manifestations may change with time (as the dissection progresses), and this may distract the physician from making the correct diagnosis. Rupture of the dissection into the true aortic lumen may cause a cessation of symptoms, and the correct diagnosis may then be inappropriately dismissed. History, physical examination, and chest radiography can suggest the diagnosis, but only if one is alert to aortic dissection as one of the diagnostic possibilities in a patient with acute chest pain, syncope, or acute focal neurologic signs. Factors associated with misdiagnosis include walk-in mode of admission, normal mediastinal width/aortic contour on chest radiograph, absent extremity pulse amplitude differences, and nonspecific symptoms. 17,18,19

**ECG**

It may be difficult to differentiate aortic dissection from acute coronary syndromes on ECG, because both conditions are associated with ECG changes; dissection may limit or obstruct coronary artery blood flow. Abnormal ECG findings include new Q waves or ST-segment elevation in 3% to 4%, ST depression in 15% to 22%, and nonspecific ST and T-wave changes in 41% to 62%. 5,9,20 The ECG is normal in only 19% to 31% of patients. 5,20

**BIOMARKERS**

Several potential biomarkers have been investigated for their utility to identify or exclude aortic dissection. 21 D-Dimer is the marker most thoroughly investigated. A meta-analysis of seven studies involving 298 subjects with acute aortic dissection and 436 without found a sensitivity of 97% (95% confidence interval, 94% to 99%) and negative predictive value of 96% (95% confidence interval, 93% to 98%) using a D-dimer cut point of 500 ng/mL (1620 nmol/L). 22 The specificity was low at 56% (95% confidence interval, 51% to 60%). Guidelines do not endorse the use of D-dimer as the sole means of excluding aortic dissection, 15 and several authors have cautioned against this practice. 23,24,25,26 One report found that young adult patients with short dissection length and thrombosed false lumen were likely to have a false-negative D-dimer. 24 The false-negative rate using D-dimer is as high at 18%. 25

**IMAGING**

A plain chest radiograph may provide important clues for the diagnosis. However, from 12% to 37% of patients have no abnormality, and this study should not be used to exclude dissection. 5,27 The most common radiographic abnormality is a widened mediastinum or abnormal aortic contour. Other possible findings include pleural effusion, displacement of aortic intimal calcification, and deviation of the trachea, mainstream bronchi, or esophagus (Figure 59-1).

**Figure 59-1.** Abnormal aortic contour on chest radiography. Frontal and lateral radiographs of the chest in a patient with type B aortic dissection reveal an abnormal aortic contour (arrow). A right pleural effusion is present, and multiple postoperative clips and wires are also seen.
CT (especially multidetector-row CT) is the imaging modality of choice for diagnosis of dissection. CT can reliably identify a false lumen (Figure 59-2) and can provide additional details such as the anatomy of the dissection, the location of the dissection flap, extension of the flap into great vessels (Figure 59-3), signs of aortic rupture, and signs of end-organ damage. CT protocols should be both with and without IV contrast. Invasive catheter angiography is rarely necessary.

**FIGURE 59-2.**
CT image of a type A aortic dissection. True and false lumens are present in the ascending aorta and descending aorta (descending false lumen at arrow) on noncontrast (left) and contrast (right) images. AF = ascending false lumen; AT = ascending true lumen; DT = descending true lumen.
Type B dissection into the iliac arteries. Contrast CT image of dissection extending into the iliac arteries (anterior to vertebral body). True and false lumens are visible in both arteries (arrows).

CT may also diagnose intramural hematoma and penetrating atherosclerotic ulcer.\textsuperscript{28} Penetrating atherosclerotic ulcer can be difficult to distinguish from large atheromatous plaques (Figure 59-4). CT diagnosis of penetrating atherosclerotic ulcer depends on extension of the ulcer past the intima. Ulcers often have overhanging edges and focal outpouchings of the aorta itself. Intramural hematoma is often identified by a high-signal mass in the aorta on CT (Figure 59-5). This often appears as a crescent and is best seen on noncontrast images.

FIGURE 59-4.
Noncontrast CT image of a penetrating aortic ulcer in the descending aorta (arrows), demonstrating an outpouched, abnormal contour of the aorta in three sections. [Image used with permission of Dr. Ernest]
FIGURE 59-5. Contrast CT image of an intramural hematoma (arrows point to the crescent-shaped lesion along the posterior lateral aortic wall) in the descending aorta. [Image used with permission of Dr. Ernest Scalzetti, MD.]
In experienced hands, transesophageal echocardiography may be as sensitive and specific as CT. The procedure generally has to be performed under moderate sedation or even general anesthesia. Known esophageal disease is a relative contraindication. Sound transmission is disrupted by air in the trachea or left bronchia, which may make evaluation of the ascending aorta difficult. The accuracy and precision of transesophageal echocardiography are highly operator dependent. MRI has been used to evaluate stable patients with suspected aortic disease.\(^2\)

**Coronary CT angiography**, or the "triple rule-out," can diagnose and differentiate coronary artery disease, pulmonary embolism, and acute aortic dissection.\(^29\),\(^30\) However, it requires a specialized contrast infusion protocol to image the three vascular beds of interest and an increased radiation dosage.\(^31\) Furthermore, the "triple rule-out" study has not been shown to improve diagnostic yield, reduce clinical events, or diminish downstream resource use.\(^32\)

**TREATMENT**

**ANTIHYPERTENSIVES: NEGATIVE INOTROPIC AGENTS**

While aortic dissections may cause hypotension that requires fluid or blood product resuscitation, suspected aortic dissection commonly requires antihypertensive treatment. Initial treatment is with a negative inotropic agent in order to lower blood pressure without increasing the shear force on the intimal flap of the aorta. β-Blockade is ideal, and short-acting β-blockers such as propranolol, labetalol, or esmolol are preferred over long-acting β-blockers. The ideal target blood pressure is undefined by controlled trials and must be tailored to each patient (see [chapters 57](#), "Systemic Hypertension" and [58](#), "Pulmonary Hypertension"). However, a **systolic pressure of 120 to 130 mm Hg is a reasonable starting point; guidelines suggest a goal of 100 to 120 mm Hg.**\(^1\)

Esmolol may be given as an initial bolus of 0.1 to 0.5 milligram/kg IV over 1 minute followed by an infusion of 0.025 to 0.2 milligram/kg/min. Labetalol (a β-blocker with limited α-blocking characteristics in a 7:1 ratio) also may be used at an initial dose of 10 to 20 milligrams IV with repeat doses of 20 to 40 milligrams every 10 minutes to desired effect or a maximum dose of 300 milligrams. Calcium channel blockers may be used in the event of a contraindication to β-blockade, but experience with their use in the setting of aortic dissection is limited. β-Blocker use has been associated with improved survival in the International Registry of Acute Aortic Dissection (IRAD) database.\(^33\),\(^34\)

**VASODILATORS**

Vasodilators such as nitroprusside may be added for further antihypertensive treatment after successful administration of a negative inotrope. Adequate β-receptor or calcium channel blockade should be achieved prior to starting a vasodilator (see [chapters 57](#) and [58](#)). Nicardipine, a parenteral dihydropyridine calcium channel blocker, has been used with success as a replacement for nitroprusside.\(^35\)
DEFINITIVE REPAIR

Rapid referral to a surgeon is mandatory. Dissection with involvement of the ascending aorta requires prompt surgical repair.

Endovascular repair treats some aortic type A and complicated type B dissections (patients with malperfusion, persistent severe pain, persistent false lumen, resistant hypertension, or expanding aortic diameter), penetrating ulcers, and intramural hematomas. Stenting has been combined with fenestration for patients with malperfusion. Endovascular therapy has uncertain long-term effects but has shown short-term benefit. Endovascular treatment is minimally invasive and avoids sternotomy and circulatory arrest. In treating dissection, goals of therapy include expansion and stabilization of the true lumen and passive resorption of thrombosis of the false lumen. In addition, visceral artery blood flow can be restored passively or by fenestration of the initial flap. Refractory pain in patients treated medically with type B dissection may be an indication for invasive intervention.

DISPOSITION AND FOLLOW-UP

Patients with acute aortic syndromes are likely to require admission to an intensive care unit for hemodynamic therapy and careful monitoring. Acute intermural hematomas and penetrating ulcers have an unclear clinical course and natural history. Therefore, the management of patients with these disorders remains controversial. Clearly no patient with an acute aortic syndrome should be discharged without consultation with a cardiovascular or vascular surgeon.

SPECIAL CONSIDERATIONS

AORTIC DISSECTION COMPPLICATING PREGNANCY

Aortic dissection in pregnancy is rare and usually occurs in the third trimester and postpartum period. Risk factors are bicuspid aortic valve, connective tissue disorders, hypertension, and a family history. Pregnancy increases the risk of dissection in patients with Marfan's syndrome, complicating 4.4% of pregnancies in women with the syndrome. Depending on the gestational age of the fetus, cesarean section with concomitant aortic repair is recommended for type A dissection. Simultaneous consultation with obstetrics and cardiovascular surgery is needed if the diagnosis is considered.

PRACTICE GUIDELINES

Guidelines have been published on the management of thoracic aortic disease, including type A dissection, by the American College of Cardiology/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions,
Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine. Because of the relative infrequency of this condition, most of the recommendations are consensus based.

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