Acute Aortic Dissection in the Emergency Department: Diagnostic Challenges and Evidence-Based Management

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KEYWORDS
• Aortic dissection • Emergency department • Evidence-based medicine

ACUTE AORTIC DISSECTION

Acute aortic dissection (AAD) is a rare but potentially catastrophic disease that remains difficult to diagnose in the emergency department (ED). It carries a significant in-hospital mortality of 27%, even when properly diagnosed.1 In a recently published guideline from the American College of Cardiology (ACC)/American Heart Association (AHA), mortality estimates for AAD were placed at 40% for immediate death, 1% per hour for incremental death thereafter if initially surviving the acute dissection event, and 20% for perioperative death.2 There is a 50% to 70% reported survival rate after initial surgery depending on patient age and cause. This article focuses on the epidemiology, clinical assessments, diagnostic challenges, and management strategies for AAD in the ED.

AAD: EPIDEMIOLOGY

The International Registry of Acute Aortic Dissection (IRAD) was created in 2000 to follow patterns and outcomes in AAD.3 This registry of 464 cases (mean age 63 years, men 65%), found an overall mortality of 27.4%, with a surgical versus nonsurgical
mortality of 26% versus 58% respectively for proximal (type A) dissections. Mortalities for distal (type B) dissections were 10.7% for medical management and 31% for surgical management (performed in 20% of type B candidates).

The true incidence of aortic dissection is difficult to define because acute AAD can be instantly fatal in the prehospital setting (and death may be attributed to other causes). In addition, AAD may be missed on initial presentation, leading to early mortality as a result of misclassification. Population-based prevalence studies suggest that the incidence of AAD may range from 2.5 to 3 cases per 100,000 person years in the United States (6000–10,000 new cases annually), to 16 cases per 100,000 annually in Sweden. There is a higher incidence of AAD in men (65%) and with increasing age.

Left untreated, patients with a proximal (type A) acute dissection have a mortality of approximately 75% within 2 weeks. With successful initial therapy, the 5-year survival rate is 75%. The 10-year survival rate for surgically repaired dissections is 40% to 60%.

The medicolegal issues surrounding missed diagnosis of AAD remain significant. There is no central repository of pooled medicolegal cases/outcomes in the United States available to physicians for quality assurance purposes. However, the Risk Factor Study conducted by the Sullivan Group in the United States in 2006 examined the documentation of risk factors for acute diagnoses in 91,286 ED records analyzed, and found that risk factors for AAD were poorly documented (26%) compared with 86% documentation for coronary artery disease risk factors. In this study, although medical physicians performed most patient assessments (83.6%), it was noted that nurse practitioners (48.9%) and template users (50.2%) were the best at documenting the relevant risk factors. In Canada, the Canadian Medical Protection Agency (CMPA) has issued 2 recent bulletins concerning AAD cases and outcomes. In the first bulletin, 2 cases of atypical chest pain with nonclassic features were reviewed. In the first case, the findings of experts suggested that a detected large blood pressure differential between patient arms should have alerted the treating physicians to the possible diagnosis of AAD. In the second, there was a complete lack of typical and atypical findings. The second bulletin reviewed 32 cases that generated 34 medico-legal cases; 56% of these were either dismissed or judged in favor of the physician. Common misdiagnoses included acute coronary syndrome (19%), musculoskeletal pain (20%), pneumonia/pulmonary embolism (20%), pericarditis (12%), gastrointestinal (GI) pain (9%), and other causes (20%). The bulletin cautioned physicians to consider the diagnosis of AAD in situations of sudden severe chest pain, accompanying visceral symptoms (nausea, vomiting, pallor, diaphoretic), normal/minimally abnormal electrocardiography (ECG) findings, and inappropriate reliance on classic features such as tearing chest pain, blood pressure/pulse discrepancies, new cardiac murmurs, and chest radiograph mediastinal widening as potentially misleading.

**AAD: PATHOPHYSIOLOGY**

Normal cardiac contractions involve swinging movements of the heart in the pericardium resulting in small flexions of the ascending and descending aorta; the latter is tethered just distal to the left subclavian artery. This repetitive swinging (37 million beats per year) creates repetitive stresses on the layers of the aortic wall. The aortic wall is composed of 3 layers: the innermost intima, the media (composed of elastic connective tissue and smooth muscle), and the outermost adventitia.

Hemodynamic stressors to the aortic inner walls can be the result of prolonged hypertension, inherently weakened connective tissue walls (eg, Ehlers-Danlos syndrome).
syndrome), or a bicuspid aortic valve that alters the laminar flow of ejected blood toward the aortic wall rather than the central vascular lumen. 

Dissection occurs when the medial layers have degenerated through normal aging or other pathologic processes, and pulsatile blood flow tears through the intimal layer into the media. The resulting false lumen can then extend distally or proximally. This distention can lead to obstruction of other arterial origins from the aortic trunk, rupture back into the true vascular lumen, or into the pericardial sac or pleural cavity. External rupture can be more common because the adventitial layers are thin walled. The most important predictors of continuing dissection are the degree of sustained hypertension, and the upstroke surge pressure (slope) of the pulse wave during contraction (upstroke pattern on apex cardiogram; change in pressure \(\frac{dP}{dt}\)). False lumens can also be the result of spontaneous hemorrhage of the vasa vasorum into the aortic wall; this occurs in 8% to 15% of cases and explains the absence of an intimal tear in certain cases. Intramural hematomas seem to be more common in the descending aortas of elderly hypertensive patients.

**CLASSIFICATION OF AAD**

The anatomic distribution as well as the acuity of the dissection have important prognostic and therapeutic implications. Consequently, dissections are classified according to anatomic location as well as acuity.

There are 2 different anatomic classification systems for aortic dissection that are commonly used, the Stanford and DeBakey classifications. The Stanford classification divides dissections into types A and B. Specifically, any dissection involving the ascending aorta (proximal to the brachiocephalic artery) is classified as type A, whereas type B dissections involve only the descending aorta (distal to the subclavian artery). The DeBakey system describes dissection according to the origin of the intimal tear and the extent of the dissection using type I, II, and III classifications. In DeBakey type I dissections, the intimal tear originates in the ascending aorta and extends to the arch and often beyond to the descending aorta. DeBakey type II dissections originate in, and are confined to, only the ascending aorta. DeBakey type III dissections originate in and propagate to the descending aorta. DeBakey type III dissections were then further subdivided into IIIa and IIIb classifications by Larson and Edwards, in which the former involves only the thoracic descending aorta and the latter extends below the diaphragm (Fig. 1, Table 1). Although vascular surgeons may debate the relative usefulness of either classification system for surgical management planning, for emergency physicians, it is not as important which classification system is used, so long as an accurate assessment of ascending versus descending aorta involvement can be made to differentiate management strategies. The Stanford system, with an A versus B dichotomy, is easier to use than the 3-level DeBakey system (with subdivisions).

For temporal classification purposes, AAD can be categorized as acute if diagnosis occurs within 2 weeks of pain onset, subacute if within 2 to 6 weeks, and chronic if more than 6 weeks after onset of pain.

According to the IRAD, 62% of dissections are type A, whereas the remaining 38% are type B. Although more common, type A dissections are also associated with a higher mortality. The overall mortality for patients with type A dissection in IRAD was 27.4% (26.6% mortality for those who underwent surgery compared with 58% for those who were treated medically). Conversely, with appropriate medical antihypertensive management, 90% of patients with uncomplicated type B dissections survived to hospital discharge.
Aortic dissection occurs most frequently in men and with advancing age.\textsuperscript{1} However, in the IRAD database, female patients with AAD were older than their male counterparts (mean age onset, 63 years), less likely to present within 6 hours of onset, less likely to endorse abrupt onset or have pulse deficits, and more likely to have mental status changes or congestive heart failure.\textsuperscript{2} Women were also less likely to be properly diagnosed within 24 hours of symptom onset, and subsequently had higher in-hospital mortality (30\% vs 21\%, $P = .001$) than men.

### Table 1

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<tr>
<th>Classification of Aortic Dissection</th>
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<tr>
<td><strong>Stanford</strong></td>
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<td>Type A</td>
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<tr>
<td>Type B</td>
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<tr>
<td><strong>DeBakey</strong></td>
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<tr>
<td>Type I</td>
</tr>
<tr>
<td>Type II</td>
</tr>
<tr>
<td>Type III</td>
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<td>Type IIIa</td>
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<td>Type IIIb</td>
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There are several known/reported risk factors for AAD (Table 2). Common medical risk factors include a history of hypertension (70%–90%), prior cardiac surgery/catheterization (18%), a bicuspid aortic valve (14%), or a history of connective tissue disorders (eg, Marfan syndrome, Ehler-Danlos syndrome). Inflammatory vascular disorders also hold a higher risk of AAD. IRAD data show that for patients less than 40 years old with AAD, 50% had a history of Marfan syndrome. Patients may also be at risk of acquired AAD with significant exertion or trauma. A positive family history in first-degree relatives has been associated in 13% to 19% of patients with AAD without an identified genetic syndrome.

There are specific populations that deserve special mention when considering the diagnosis of AAD in the ED. Cocaine, methylenedioxymethamphetamine (MDMA; known as ecstasy), and other stimulant use have been associated with an increased risk of AAD. Acute β-blocker withdrawal and fluctuations in circadian cortisol levels have also been implicated in AAD. There are case reports of spontaneous AAD in recreational weight lifters. Certain infectious diseases have also been associated with AAD, including tuberculous aortitis, and insidious *Salmonella* aortitis causing type B dissection. An increasing incidence of AAD in pregnancy has been reported, including cases associated with cocaine use, inflammatory arteritis, and otherwise previously healthy women with no prior aortic disease. Half of dissections of women younger than 40 years occur in pregnancy. Pregnancy has been associated with altered hemodynamic states (increased heart rate and stroke volume), and increased estrogen and progesterone changes, which may induce

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<th>Table 2</th>
<th>Risk factors for aortic dissection</th>
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<td><strong>Risk Category</strong></td>
<td><strong>Associated Disorders</strong></td>
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<tr>
<td>Increased aortic wall stress</td>
<td>Hypertension (especially uncontrolled)</td>
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<tr>
<td></td>
<td>Pheochromocytoma</td>
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<td></td>
<td>Cocaine/stimulant use</td>
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<td></td>
<td>Weight lifting/other Valsalva situations</td>
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<td></td>
<td>Deceleration injury/blunt trauma</td>
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<td></td>
<td>Aortic coarctation</td>
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<tr>
<td>Medical conditions affecting aortic medial layers</td>
<td>Genetic disorders/syndromes: Ehler-Danlos, Marfan, Turner, Loeys-Dietz, Noonan, congenital bicuspid aortic valve, familial dissections/aneurysms</td>
</tr>
<tr>
<td></td>
<td>Inflammatory vasculitides: syphilis, granulomatous arteritis, tuberculous, salmonella, Takayasu, giant cell, systemic lupus erythematosus, Behçet</td>
</tr>
<tr>
<td>Iatrogenic wall injury</td>
<td>Cardiac/valvular surgery, intra-aortic balloon pump use, aortic cannulation, cross-clamping sites, catheterization</td>
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<tr>
<td>Other</td>
<td>Male sex</td>
</tr>
<tr>
<td></td>
<td>Age &gt;50 y</td>
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<tr>
<td></td>
<td>Pregnancy</td>
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<td></td>
<td>Polycystic kidney disease</td>
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<td></td>
<td>Chronic corticosteroid use or immunocompromised states</td>
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histologic changes in arterial walls that predispose to dissection. In addition, it is possible to present to ED with AAD with no known risk factors; there is no clear reported evidence on the incidence of risk factor–free AAD.

**Presentations of Acute Thoracic Aortic Dissection**

**Historical features**

The diagnosis of acute thoracic aortic dissection (TAD) in the ED is complicated by it being a rare clinical condition with potentially catastrophic outcomes, and does not necessarily present with classic findings. Typical features include sudden acute chest pain (90%) that is excruciating, severe at onset, and of a sharp/ripping/tearing quality. It may be possible to localize the dissection origin based on location of pain: anterior (ascending aorta), neck/jaw (arch), interscapular (descending aorta), and lumbar/abdominal (subdiaphragmatic). Migratory pain may be rare (17%). Of concern, pain may ease or abate over time, or change as the dissection extends and begins to involve other organ systems. For example, type A dissections typically present with chest pain (71% anterior), and, less commonly, with back pain (47%) or abdominal pain (21%). Type B dissections are more likely to present as back pain (64%), followed by chest or abdominal pain (63% and 43% respectively). Visceral symptoms including nausea, vomiting, diaphoresis, apprehension, and lightheadedness may accompany the onset of dissection.

In the CMPA case review series of missed AAD (n = 32 patients), the symptom distribution was as follows: sudden-onset severe chest pain (91%), visceral symptoms (pallor, vomiting, diaphoresis 78%), intermittent pain (75%), radiation to back/neck/arms/jaw (69%), pleuritic/positional pain (44%), pyrexia (22%), syncope (9%), and tearing quality (3%). The author concluded that, although there may be some classic findings of TAD that should prompt the inclusion of TAD in a differential diagnosis of acute chest pain in the ED, the reliance on the presence of these features, or the absence thereof, may not be sufficient to include or exclude the diagnosis.

The most comprehensive systematic review to date by Klompas, and the subsequent abstract, summarize the difficulty of relying on certain clinical features for the diagnosis of TAD. The likelihood ratios (LRs) for various clinical findings are summarized in **Table 3**.

The studies suggest that there is poor reliability in the symptomatic descriptions of pain quality when determining the likelihood of TAD in a patient with chest pain in the

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<th>Symptom</th>
<th>+ LR (95% CI)</th>
<th>− LR (95% CI)</th>
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<tr>
<td>Sudden onset of pain</td>
<td>1.6 (1.0–2.4)</td>
<td>0.3 (0.2–0.5)</td>
</tr>
<tr>
<td>Tearing/ripping pain*</td>
<td>1.2 (0.2–8.1)</td>
<td>0.99 (0.9–1.1)</td>
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<td></td>
<td>10.8 (5.2–22.0)</td>
<td>0.4 (0.3–0.5)</td>
</tr>
<tr>
<td>Migrating pain*</td>
<td>1.1 (0.5–2.4)</td>
<td>0.97 (0.6–1.6)</td>
</tr>
<tr>
<td></td>
<td>7.6 (3.6–16.0)</td>
<td>0.6 (0.5–0.7)</td>
</tr>
<tr>
<td>History of hypertension</td>
<td>1.6 (1.2–2.0)</td>
<td>0.5 (0.3–0.7)</td>
</tr>
<tr>
<td>Focal neurologic deficit</td>
<td>6.6–33.0</td>
<td>0.71–0.87</td>
</tr>
<tr>
<td>Diastolic murmur</td>
<td>0.9–1.7</td>
<td>0.79–1.1</td>
</tr>
<tr>
<td>Pulse deficits</td>
<td>2.4–47.0</td>
<td>0.62–0.93</td>
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* Unpooled data from 2 studies.

Data from Klompas M. Does this patient have an acute thoracic aortic dissection? JAMA 2002;287:2262–72.
ED. However, the 2000 study by von Koloditsch and colleagues\(^{32}\) merits some attention. This work was a prospective study of patients in the ED with chest pain (n = 250, 128 confirmed TAD cases) presenting to a German university hospital center, with a mean age of 53 years, 78% men, and 61% type A dissection. The outcomes of this specific study suggest that the positive and negative LRs for tearing/ripping pain (10.8 and 0.4 respectively), and for migrating pain (7.6 and 0.6 respectively) approach clinically significant levels for ED physician decision making.\(^{33}\)

A final precaution remains, in that up to 12% of TAD cases may be painless, and present only with complications involving other body systems.\(^{4}\) These patients tended to be older, possible steroid users or patients with Marfan, or presenting with syncope, stroke, or congestive heart failure.\(^{2}\)

**Physical findings of AAD**

The physical examination findings associated with AAD are unreliable and frequently absent in patients with AAD, based on the location of the dissection and the extent of involvement of surrounding structures or organ systems. An examination of the peripheral pulses in the upper extremities or assessment of blood pressure differentials can reveal some important clues for the diagnosis. Historically, a blood pressure differential of 20 to 30 mm Hg was reported to be significant for TAD,\(^{18}\) and a differential greater than 20 mm Hg was confirmed in a prospective study by von Koloditsch and colleagues\(^{32}\) as a significant predictor of AAD. However, other studies have shown that up to 20% of normal patients may have a pulse differential of 20 mm Hg without AAD,\(^{18}\) and 53% of normal patients have difference of 10 mm Hg.\(^{21}\)

The Klompas\(^{30}\) meta-analysis of clinical findings with AAD suggested that the pooled sensitivity of pulse deficits is 31% (95% confidence interval [CI] 24–39) but a positive LR of 5.7 (95% CI 1.4–23.0). New aortic regurgitation murmurs arise in 32% to 76% of patients with AAD,\(^{1,2}\) but have a pooled sensitivity of 28% (95% CI 21–36) and a disappointing positive LR of 1.4 (95% CI 1.0–2.0).

Complications of acute aortic regurgitation may range from nonexistent to life-threatening congestive heart failure or cardiogenic shock, depending on the extent and magnitude of dissection into the aortic valve root. Other clues to significant aortic regurgitation complicated by pericardial tamponade may include jugular venous distension, muffled heart sounds, tachycardia, and hypotension.\(^{1}\) Acute aortic dissection into the pericardium with subsequent tamponade is reported to be the second most common cause of death in AAD.\(^{18}\)

AAD can be complicated by mass compression effects on adjacent structures, leading to superior vena cava syndrome, Horner syndrome (sympathetic chain), hoarseness (recurrent laryngeal nerve), dyspnea (tracheobronchial tree), and dysphagia (esophagus).\(^{4}\) A pulsatile sternoclavicular joint, although rare, may provide a diagnostic clue to the diagnosis with compression manifestations of upper chest/ lower neck phenomena.\(^{4}\)

**End-organ presentations of AAD**

**Cardiovascular complications** These can include aortic regurgitation and related disorders (discussed earlier), pulse deficits, blood pressure differentials, syncope, myocardial infarction, congestive heart failure, and cardiogenic shock.\(^{1,2,4,8}\) Low sensitivity findings from the Klompas\(^{30}\) review include pericardial rubs (sensitivity 6%, 95% CI 3%–13%), congestive heart failure (sensitivity 15%, 95% CI 4%–33%), shock (sensitivity 19%, 95% CI 15%–26%), and new myocardial infarction on ECG (sensitivity 7%, 95% CI 4%–14%). It is important to recognize an ST-elevation myocardial infarction (STEMI) in the setting of potential proximal AAD (incidence
3%), because thrombolytic therapy is contraindicated in these situations; this reportedly occurs in 0.1% to 0.2% of STEMI cases. STEMI is most common in the right coronary circulation, leading to posteroinferior infarctions caused by dissection into the right coronary ostium. Evidence of myocardial ischemia on ECG is reported in up to 19% of AAD cases. There are also reports of proximal dissections extending into the atrial septa leading to conduction abnormalities. Both atrial fibrillation and intractable supraventricular tachycardia cases have been reported. Painless acute congestive heart failure has been reported in the setting of a type B dissection. In addition, AAD as a cause of cardiac arrest should be suspected in patients of older age, known aortic aneurysms, male gender, and initial pulseless electrical activity rhythm, as well as those with polycystic kidney disease.

**Syncope** Syncope complicates approximately 13% of cases of AAD. This syncope can be the result of acute cardiac dysfunction (described earlier), vascular outflow obstruction in the arch/carotid arteries, neurologic vasovagal pain responses, or from acute hypovolemia caused by hemorrhage into third spaces. Syncope inappropriately attributed to heat-related illness in a young healthy male patient with a painless AAD has been reported. Painless syncope is also reported elsewhere in the emergency literature.

**Neurologic complications** Focal neurologic deficits can complicate AAD with a pooled sensitivity of 17% (95% CI 12%–23%). Neurologic deficits can result from hypotension, malperfusion, distal thromboembolism, or nerve compression from mass effects. Proximal arch dissections are more likely to cause intracranial and brainstem deficits, whereas distal arch dissections may involve the spinal cord and lower extremities. Cerebral ischemia and stroke syndromes are the most common central nervous system effects of proximal AAD, occurring 5% to 15% of the time; chest pain in the setting of new focal neurologic deficit is highly predictive of proximal AAD involving the cerebral circulation. It is critical for ED physicians to consider AAD in the setting of acute stroke syndromes, because contraindicated thrombolytic therapy in this setting can lead to catastrophic outcomes. Sudden coma from basilar artery occlusion and transient locked-in syndrome have both been described in the setting of painless AAD, as has acute vertigo. Focal central findings may be localized from vasococlusive or thromboembolic causes, or diffuse if resulting from systemic hypotension. The spinal cord may be susceptible to injury if AAD involves the origins of the intercostal spinal arteries, the artery of Adamkiewicz, or the thoracic radicular artery, resulting in clinical syndromes of transverse myelitis, anterior cord syndromes, and paraplegia or quadriplegia. This may be particularly true in watershed areas of the cord. Multiple cases exist of painless neurologic deficits of the legs of variable onset and duration (incidence 10%); the common theme is that the index of suspicion for ED physicians should be high for AAD in the setting of sudden paralysis of the lower extremities even in the absence of chest symptoms. Of interest, 50% of neurologic symptoms may be transient, and one-third may not present with chest pain, complicating the potential AAD diagnosis.

**Ears/nose/throat complications** As stated previously, several structures in the throat and upper thorax can be compressed by mass effect of proximal/arch dissection, including the trachea (dyspnea, stridor), esophagus (dysphagia), recurrent laryngeal nerve (hoarseness), and sympathetic chain (ipsilateral Horner syndrome). Serious comorbidities have resulted from initial benign presentations including sore throat, hoarseness, and hoarseness with collapse and neck bruising. The recurrent theme is of benign and potentially misleading complications in the face of painless AAD.
Respiratory complications  Respiratory effects of AAD can include mass effects on the tracheobronchial tree (dyspnea), hemorrhage into the lung tissues (hemoptysis 3%) and pleural space, pleural effusions, and death.\textsuperscript{2,53}

GI complications  Mesenteric ischemia is the most common GI complication of AAD, and the most common cause of death in type B dissection.\textsuperscript{2} By the time unreliable serum markers become positive, it is often too late to salvage the dead bowel. Rare but catastrophic GI bleeding can also result from mesenteric ischemia or from aortoenteric fistula.\textsuperscript{54,55}

Other atypical presentations  Chronic aortic dissection has been reported in the literature as presenting with fever of unknown origin,\textsuperscript{56} renal colic, and mesenteric ischemia.

Other diagnoses mimicking AAD  There are sparse case reports of thymic diseases presenting as an AAD, including a 49-year-old hypertensive woman with marfanoid features presenting with tearing chest pain and a para-aortic bulge on chest radiograph but normal aortogram, ultimately diagnosed as an invasive thymoma.\textsuperscript{57} Another case involved an 80-year-old hypertensive man presenting with sudden severe chest and back pain, but investigations ultimately revealed an thymic carcinoid tumor.\textsuperscript{58} A case of Takayasu arteritis presenting with AAD to the ED has also been described.\textsuperscript{59}

The value of combined clinical findings in arriving at a diagnosis of AAD has been examined.\textsuperscript{32} The findings examined in combination included severe sudden-onset tearing pain, blood pressure or pulse differentials between the arms, and/or mediastinal widening on chest radiograph. The positive LRs for diagnosing AAD with 0, 1, 2, or 3 combined findings increased exponentially through 0.1 (95% CI 0.0–0.2), 0.5 (95% CI 0.3–0.8), 5.3 (95% CI 3.0–9.4), and 66.0 (95% CI 4.1–1062.0) respectively. However, these 3 findings were only present in 27% of the 128 patients included in this study.

The overarching themes in the diverse literature regarding AAD assessment in the ED become readily apparent. The classic features of AAD are rarely present in combination to suggest an obvious diagnosis in the ED. The clinical manifestations of AAD may be a result of the dissection process itself, the location and extension of the dissection, and the subsequent end organs affected by the compromised blood flow or mass effects caused by the dissection. Seemingly unrelated clinical features more consistent with other, more common diagnostic entities may delay or confound the ultimate diagnosis of AAD, with potentially serious sequelae. In addition, there are many AAD mimics that may initially suggest AAD, but ultimately are diagnosed as something else. The lesson for all ED physicians is one of awareness and vigilance of common and atypical presentations of this dangerous condition, and of investigating them appropriately to make a proper and safe diagnosis.

AAD: DIAGNOSTIC STRATEGIES IN THE ED

Once the possibility of AAD has been considered, the emergency physician must make the proper diagnosis. Routine initial tests readily available in the ED (ECG, chest radiograph, laboratory markers) all have variable reliability in making a diagnosis of AAD.

Electrocardiography

The ECG is often normal or may show nonspecific changes in the setting of aortic dissection. Data collected from the IRAD revealed that the ECG was normal in 31% of cases, whereas 26% showed left ventricular hypertrophy reflecting long-standing hypertension.\textsuperscript{3} The Klompas\textsuperscript{30} review also noted a poor LR+ (0.2–3.2) and LR− (0.84–1.2) for left ventricular hypertrophy findings on ECG in the setting of AAD. The primary usefulness of the ECG in this clinical setting is to consider or exclude alternative diagnoses.
It is imperative to consider the possibility of aortic dissection in the setting of ischemia, especially inferior ischemia, because proximal dissections may extend to involve the right coronary artery. In the review by Klompas, 7% of patients with aortic dissection showed evidence of acute ischemia (either ST elevation or new Q waves) on the ECG. In this setting, the AHA 2010 guidelines on thoracic aortic disease recommended that, because of the uncommon event of dissection-associated coronary disorder, ST elevation should be treated as a primary cardiac event unless the patient is considered to be at high risk for aortic dissection.

Laboratory Markers: d-Dimer and Other Serum Biomarkers

The use of a screening D-dimer in the diagnosis of TAD has been shown to be a highly useful marker, with reported sensitivities ranging from 94% to 99%. Despite this high sensitivity, a 2008 review by Sutherland and colleagues commented that, because of the wide CIs quoted in the studies, the poorly defined eligibility criteria for study inclusion, and the possibility of false-negatives in patients with thrombosed false lumens, D-dimers should not be used as the sole screening tool in this clinical setting. This conclusion was echoed in the 2010 AHA guidelines for diagnosis and management of patients with thoracic aortic disease in which D-dimer screening was not recommended at this time because of the lack of large prospective evaluation, limitations in accurately assessing posttest probability of a negative D-dimer, and the potential for a negative D-dimer result in patients with a thrombosed false lumen or ascending aortic intramural hematoma. A recently published meta-analysis of 7 studies (298 patients with AAD, 436 without) suggests that using a D-dimer cutoff of 500 ng/mL had an excellent negative LR (LR− 0.06, 95% CI 0.03–0.12) to exclude AAD, but not a good positive LR to include an AAD diagnosis (LR+ 2.43, 95% CI 1.89–3.12). These investigators concluded that using a cutoff of less than 500 ng/mL may be useful to exclude the diagnosis of AAD and avoid the need for advanced imaging. However, there was significant heterogeneity in the included studies, and this information has not yet been prospectively validated in the ED. With further investigation and large prospective evaluation, D-dimer assays may show diagnostic promise and become part of a useful screening strategy or clinical decision rule for diagnosis of AAD.

Serum biomarkers reflecting smooth muscle damage have also been investigated in the setting of AAD. Specifically, serum smooth muscle myosin heavy chain (released with arterial wall smooth muscle damage) and calponin (a smooth muscle troponin-like protein) have been evaluated by Suzuki and colleagues. Although further investigation is required, both biomarkers show promise and, in the future, may assist in the diagnosis of aortic dissection.

Although not helpful in the screening or diagnosis of aortic dissection, C-reactive protein levels have been shown to have prognostic value and predict adverse long-term outcomes. In a small cohort study of 255 Austrian patients with symptomatic aortic disease, cumulative mortality from 1 to 6 months was 32% to 40%, and there was a near-linear increase in hazard ratio from 0.7 to 2.6 through increasing C-reactive protein level quartiles.

Chest Radiography

Approximately 90% of patients with aortic dissection have abnormalities on the chest radiograph, and therefore the presence of a normal chest radiograph may help to decrease the likelihood of aortic dissection. In the review by Klompas, a pooled analysis of 1337 chest radiographs also reported abnormalities in 90% of patients with aortic dissection. Furthermore, Klompas showed that, in the absence of an abnormal aortic contour or mediastinal widening, the likelihood of AAD is significantly decreased.
The most common radiographic changes associated with dissection were abnormal aortic contour (pooled sensitivity 71%) and widening of the mediastinum (pooled sensitivity 64%). Other radiographic findings may include pleural effusion, displacement of intimal calcification, abnormalities of the aortic knob, and displacement of trachea or nasogastric tube deviation to the right. Because of the lack of sensitivity and nonspecific chest radiography findings present in aortic dissection, it is imperative to proceed with additional imaging techniques.

Advanced Imaging Modalities

In the clinical setting, where a rapid diagnosis is crucial because of the critical nature of AAD, several factors are considered when deciding how to proceed with advanced diagnostic imaging. Specifically, variables such as testing risks and benefits, access and availability to imaging modalities, accuracy of technique, and individual patient variables are carefully considered. Historically, aortic dissection was evaluated with aortography, a modality that has now largely been replaced with noninvasive diagnostic strategies, including helical computed tomography (CT), magnetic resonance imaging (MRI), and transesophageal echocardiography (TEE). Although transthoracic echocardiography (TTE) may provide useful bedside information in the setting of aortic dissection, it does not have sufficient sensitivity or specificity to be the solitary diagnostic modality used. Specifically, TTE has shown a sensitivity of only 59.3% in the detection of AAD. Further limiting the use of TTE is the inability to visualize the entire aorta. As bedside ED ultrasound continues to grow and develop, findings consistent with aortic dissection may be observed at the bedside by the ED physician. A case report study recently described 5 cases in which ED physicians used bedside ED ultrasound to aid in the diagnosis of TAD.

Transesophageal echocardiography

Transesophageal echocardiography (TEE) may play an important role in the diagnosis of aortic dissection. This imaging modality may be of particular usefulness in the hemodynamically unstable patient, in which a timely diagnosis is imperative and transfer out of an acute care setting to the radiology department is not possible. TEE has been shown to have a high sensitivity with reported values of 98%. In the systematic review of different imaging modalities, TEE was shown to have comparable sensitivity (98%) and sensitivity (95%) with CT and MRI. TEE also offers the advantages that it is able to show aortic regurgitation and pericardial effusion. Although visualization of the branches of the aortic arch and distal ascending aorta historically limited the usefulness of TEE, probe technologic advancements have improved the visualization of these anatomic regions.

CT

According to the IRAD, the most common diagnostic modality initially used is CT, with 61% of patients undergoing CT. With technological advancements, helical CT offers many advantages and shows in a pooled analysis a high sensitivity and specificity of 100% and 98%, respectively. In this study, Shiga and colleagues concluded that helical CT was the optimal imaging modality for ruling out aortic dissection in patients with a low clinical pretest probability for aortic dissection. The limitations of CT include use of ionizing radiation and contrast media, need for patient transfer out of the acute care setting, and limited ability to assess the aortic valve. Conversely, CT is generally readily available, quick to complete, delineates anatomy of the entire aorta well, and may show alternative disorders considered in the differential diagnosis for aortic dissection.

In the meta-analysis of 16 studies conducted by Shiga and colleagues, CT, MRI, and TEE were found to be equally effective in ruling out or confirming the diagnosis
of TAD (Table 4). Accordingly, in the recent 2010 AHA guidelines for the diagnosis and management of patients with thoracic aortic disease, a class I recommendation was made that either urgent TEE, MRI, or CT imaging be used to determine a definitive diagnosis in patients with high clinical suspicion for aortic dissection. Furthermore, the AHA recommends that, if the initial diagnostic test is negative in a patient with high clinical suspicion for aortic dissection, a second imaging modality should be completed. In summary, TEE, CT, and MRI all show acceptable diagnostic abilities and the initial imaging modality should be dictated by patient characteristics and availability of resources. If the initial imaging test is negative in the context of a high clinical suspicion, the clinician should proceed with a second diagnostic imaging modality. The algorithm (Fig. 2) from the 2010 ACC/AHA guidelines summarizes a diagnostic approach to diagnostic decision making for AAD in the ED.

AAD: MANAGEMENT ISSUES

Once the diagnosis of AAD has been confirmed, the management decisions in the ED are straightforward. Patients with evidence of hypotension need to be urgently resuscitated with intravenous (IV) fluids, blood products, and immediately transported to the operating room to optimize survival chances. Although there may be some advocacy for permissive hypotension in the management of ruptured abdominal aortic aneurysms to limit the use of blood products and to suppress exsanguination, there is no literature to support this strategy in the hypertensive patient with AAD. The only contemporary report of minimal benefit with permissive hypotension in AAD management was in a small case series of endovascular patients who had endovascular stent grafts placed and observed for leaks; no comments were provided regarding permissive hypotension in medical management alone, and this is not addressed in the ACC/AHA 2010 guidelines. Conversely, in the hypotensive patient, use of vasopressors to support blood pressure must be cautious given the risk of propagating the dissection. Similarly, inotropic agents may increase ventricular contraction rate and force, which will increase shear forces on the aortic wall. Blood pressures in 4 limbs should be constantly monitored to follow potential evolution of intimal flaps obstructing flow into an extremity and causing pseudohypotension. In cases of severe hypotension/shock or pulseless electrical activity with presumed pericardial tamponade, emergency pericardiocentesis may be warranted. For patients being transported to a regional vascular center for definitive care, all appropriate measures to optimize hemodynamic stability and safe transfer should be achieved before transportation of the patient.

Medical Management of AAD

In patients with hemodynamically stable dissections, the goals of ED management include pain control, heart rate and blood pressure control to avoid excessive shear forces on the intimal layers of the arterial walls (dP/dt = speed at which blood is ejected into the aorta with each ventricular contraction). Analgesia can usually be achieved with titratable opioids, which relieve pain, decrease sympathetic tone, and augment the effects of rate control and vasodilation. Target systolic blood pressures of 100 to 120 mm Hg and heart rates less than 60 beats/min can be achieved using β-adrenergic blockers (esmolol, labetalol) or vasodilators such as sodium nitroprusside, nitroglycerin, or fenoldopam. However, the use of vasodilators as a single agent is discouraged because of the associated reflex tachycardia and resulting increased shear forces across arterial intimal walls (dP/dt). As a result, a concomitant β-blocker (eg, esmolol) should be used (assuming no contraindications such as...
<table>
<thead>
<tr>
<th>Imaging Technique</th>
<th>Including Studies No.</th>
<th>Sensitivity</th>
<th>Specificity</th>
<th>Positive LR</th>
<th>Negative LR</th>
<th>Diagnostic Odds Ratio</th>
</tr>
</thead>
<tbody>
<tr>
<td>TEE</td>
<td>10</td>
<td>98 (95–99)</td>
<td>95 (92–97)</td>
<td>14.1 (6.0–33.2)</td>
<td>0.04 (0.02–0.08)</td>
<td>6.1 (5.0–7.2)</td>
</tr>
<tr>
<td>Helical CT</td>
<td>3</td>
<td>100 (96–100)</td>
<td>98 (87–99)</td>
<td>13.9 (4.2–46.0)</td>
<td>0.02 (0.01–0.11)</td>
<td>6.5 (4.4–8.7)</td>
</tr>
<tr>
<td>MRI</td>
<td>7</td>
<td>98 (95–99)</td>
<td>98 (95–100)</td>
<td>25.3 (11.1–57.1)</td>
<td>0.05 (0.03–0.10)</td>
<td>6.8 (5.5–8.0)</td>
</tr>
</tbody>
</table>

Fig. 2. AHA/ACC 2010 evaluation algorithm for AAD. ACS, acute coronary syndrome; AoD, aortic dissection; BP, blood pressure; CNS, central nervous system; CXR, chest radiograph; MR, magnetic resonance. (Reproduced from Hiratzka LF, Bakris GL, Beckman JA, et al. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the diagnosis and management of patients with thoracic aortic disease. Circulation 2010;121:e266–369. doi:10.1161/CIR.0b013e33181d4739e; with permission.)
chronic obstructive pulmonary disease (COPD) or high-risk bronchospastic disease). Labetolol may be an attractive single-agent choice because it has both $\alpha_1$-specific and nonspecific $\beta$-adrenergic properties (7:1 ratio of $\beta$ vs $\alpha$ blockade for IV labetalol). Another advantage of these titratable IV agents is rapid onset and short duration of action, which can be turned off quickly as needed. Trimethaphan (a ganglionic blocker and vasodilator) has both $dP/dt$ and systolic blood pressure reduction properties, making it an acceptable alternative to esmolol/nitroprusside combination or labetalol, especially in the context of contraindications to either agent; side effects include tachyphylaxis, significant hypotension, respiratory depression, urinary retention, and ileus. These blood pressure targets should be cautiously maintained without causing end-organ ischemia. Calcium channel blockers are less desirable agents for blood pressure and heart rate control, particularly nifedipine (and all dihydropyridines), which has negligible inotropic and chronotropic activities and may increase reflex sympathetic tone and arterial wall stress.

The starting doses of various agents are listed in Table 5.

**Surgical Management of AAD**

Type A acute dissections require prompt surgical treatment in a qualified vascular or cardiac surgery center. Expeditious surgery can reduce in-hospital mortality to 27% compared with medically treated type A dissections (mortality 56%). Autopsy studies have suggested that untreated type A dissections can increase mortality by 1% per hour up to a cumulative 50% mortality within the first 48 hours of diagnosis.

The definitive management of type B dissections is less clear. Appropriate medical management can limit mortality to 10%. Surgery may be reserved for patients with ongoing pain, refractory hypertension, occlusion of major arterial trunk origins, frank leaking or rupture, or development of local aneurysms. Patients with local aneurysms have been reported to have in-hospital 30-day mortalities of 32%; the triad of hypertension, branch vessel involvement, and absence of chest pain is an independent predictor of in-hospital death.

**Longer Term Follow-Up**

After the acute phase of management, long-term $\beta$-blockade is warranted regardless of medical versus surgical management. Repeat surgery is required in 10% to 20% of cases because of redissection, compression of mediastinal structures, blood leakage, or aneurysm formation. Type B dissections initially managed medically often go on to elective surgery because of aneurismal dilatation or limb ischemia.

Routine follow-up examinations are recommended on a schedule of 3 to 6 months.

<table>
<thead>
<tr>
<th>Table 5</th>
<th>Initial medications for blood pressure control in aortic dissection</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Medication</strong></td>
<td><strong>Class (Agent)</strong></td>
</tr>
<tr>
<td>$\beta$-Blockers</td>
<td>1. Esmolol</td>
</tr>
<tr>
<td></td>
<td>2. Labetalol</td>
</tr>
<tr>
<td></td>
<td>Sodium Nitroprusside</td>
</tr>
</tbody>
</table>
Pitfalls in AAD Management

Because the incidence of AAD is rare compared with other important causes of acute chest pain in the ED (eg, acute coronary syndrome [ACS], pulmonary embolism, pneumothorax, esophageal rupture), a rapid and reliable approach to ruling out AAD is important, especially in the context of a time-dependent emergency such as STEMI, when medical and reperfusion treatments must be implemented as soon as possible.\(^\text{21}\) The incidence of STEMI is nearly 800 times that of AAD, but proximal dissection rarely can lead to STEMI.\(^\text{21}\) Use of antiplatelet agents, anticoagulants, and fibrinolytics can be disastrous in the patient with STEMI who has a proximal AAD with coronary involvement as opposed to the more common atherosclerotic plaque rupture causes. Fatality rates of 71% have been reported in patients misdiagnosed with AAD-STEMI due to catastrophic hemorrhage.\(^\text{21}\) Recognition of key risk factors, clinical features, or lack of response to conventional ACS treatments should prompt ED physicians to at least consider an AAD diagnosis before embarking on potentially irreversible fibrinolysis decisions.

Other barriers to expeditious diagnosis of AAD in the context of ED chest pain may include lack of awareness of AAD incidence/risk factors, lack of readily available screening tests for AAD, and institutional care pathways for more common chest pain presentations (eg, ACS, pulmonary embolism), which steer clinicians away from the rare, but equally dangerous, diagnosis of AAD.\(^\text{2}\)

Prognosis for AAD

There has been little progress in developing prognostic methods to accurately predict outcomes for patients with AAD. The most common determinant of outcomes seems to be the maximal aortic diameter measurement,\(^\text{75}\) but this is not universally reliable depending on different dissection types (A vs B). For type A dissections, observational data from the IRAD data registry (n = 591 patients) suggest that a diameter cutoff of 5.5 cm may be useful to guide elective surgery decisions, but there was no difference in mortality.\(^\text{76}\) For type B dissections, an incremental increase of 5 mm in maximal aortic diameter on CT scan had an odds ratio of 1.41 (95% CI 1.04–1.92) of in-hospital death, rupture, or organ malperfusion in one retrospective series of 220 patients,\(^\text{77}\) and a diameter of 40 mm or more was predictive of needing elective surgery in another series of 180 patients.\(^\text{78}\) In a recent review of the IRAD database (n = 1480 type B patients), increasing ascending aortic dimensions were associated with higher surgical incidence, but no difference in mortality or cause of death at different width cutoffs (<40 mm, 41–45 mm, >46 mm).\(^\text{79}\) However, it was noted that higher ascending aorta widths were associated with more open procedures involving root/aortic valve/ascending aorta/arch repairs, and those with widths less than 40 mm were more likely to undergo conservative endovascular repairs.

Recent ACC/AHA Guideline 2010 Recommendations

The recently published ACC/AHA guideline describes the following recommendations for management of AAD\(^\text{2}\):

1. Initial management (level of evidence)
   a. Class I recommendations
      i. IV \(\beta\)-blockade titrated to heart rate of 60 beats per minute or less in the absence of contraindications (C).
      ii. If \(\beta\)-blockade is contraindicated, consider nondihydropyridine calcium channel blockers for same goal as described earlier (C).
iii. After target heart rate is achieved, if persistent systolic blood pressure still is greater than 120 mm Hg then consider vasodilators (nitroprusside, angiotensin-converting enzyme inhibitors) administered intravenously to further reduce blood pressure without compromising end-organ perfusion (C).

iv. Beware β-blockade or calcium channel blockade if there is evidence of acute aortic regurgitation caused by loss of compensatory tachycardia (C).

b. Class III recommendations
   i. Initiate rate control before using vasodilators to avoid reflex tachycardia causing increased aortic wall stresses and propagation of dissection (C).

2. Definitive management (level of evidence)
   a. Urgent surgical consultation should be obtained for all patients diagnosed with AAD, regardless of anatomic location, once the diagnosis is made or highly suspected (C).
   b. Ascending AAD should be urgently evaluated for immediate surgery to avoid life-threatening complications (B).
   c. Descending AAD should be managed medically unless life-threatening complications arise (eg, malperfusion of end organs, dissection progression, enlarging aneurysm, worsening symptoms, or inability to control blood pressure [level B]).

Most of these recommendations are in keeping with other evidence sources reviewed in this article. These suggestions are manageable in the ED, provided that there is adequate access to resources for advanced diagnosis and critical care.

Acute Dissection in Pregnancy

Pregnant women with potential AAD present unique diagnostic problems for emergency physicians. Pregnancy is associated with several important hemodynamic and physiologic stresses for women.28 These stresses include large changes in intravascular volume, compensatory cardiac output demands (heart rate, inotropism), and intimal wall connective tissue changes due to hormonal influences. Patients are most at risk of AAD expansion and rupture in the third trimester, especially during the peripartum and postpartum periods (up to 3 months after birth). The unique circumstances of preeclampsia can raise the risk of missed type A dissection due to overlap with type B symptoms.28 Urgent surgical repair by specialized vascular and obstetric teams may be warranted because of unique considerations of aortic repair coupled to maintaining fetal viability. If the timing is appropriate for fetal development, urgent AAD repair may coincide with emergency delivery by cesarean section, with strict monitoring of maternal and fetal hemodynamics.26,28,80 Placental malperfusion is the leading cause of fetal death if the advancing dissection involves occlusion of the internal iliac artery and uterine arterial insufficiency. There are case reports of AAD in pregnancy managed surgically with concomitant operative delivery, with the presence of previously discussed risk factors including cocaine use,25 Ehlers-Danlos syndrome,24 and Takayasu arteritis.27 In pregnant women with acute undifferentiated chest pain or dyspnea presentations, AAD should be high on the differential diagnosis for evaluation and management in the ED.

AAD: SUMMARY

For emergency physicians assessing acute undifferentiated chest pain, the diagnosis of AAD remains one of the most sinister challenges. The key clinical features and initial diagnostic tests do not always lead to this diagnosis, and the potential mimics involving cardiovascular, abdominal, neurologic, and other systems are numerous and easily misleading. Astute physicians should always have an awareness of potential risk
factors for AAD, classic versus atypical presentations, careful physical examination findings, and a high index of suspicion/low threshold for moving on to advanced imaging techniques to reliably include or exclude this diagnosis. An ongoing awareness of new diagnostic modalities to facilitate the AAD diagnosis should be maintained. Once properly diagnosed, initial management steps targeting pain, heart rate, and blood pressure control can be undertaken to optimize hemodynamic stability, and subsequent definitive surgical consultation can then be undertaken. Failure to consider AAD in these situations (and document risk assessments accordingly) can lead to clinically adverse outcomes for patients and medicolegal liability for physicians.

REFERENCES