

## Acute Aortic Syndrome

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### Case Presentation

An 83-year-old woman presented with mild, nonradiating, central chest pain. The results of her physical examination were unremarkable, and her pulses were symmetrical and bilaterally palpable. Her ECG revealed normal sinus rhythm with no acute ST/T-wave changes.

Her transthoracic echocardiogram showed good left ventricular systolic function, with mild dilatation of the ascending aorta (4.5 cm) (Figure 1A), without evidence of pericardial effusion.

Two hours later, she developed severe chest pain and collapsed. She was cold, clammy, and her blood pressure dropped to 60/30 mm Hg. Her jugular venous pressure was elevated. An urgent bedside transthoracic echocardiogram revealed cardiac tamponade (Figure 1B and 1C). Urgent computed tomography (CT) demonstrated a penetrating atheromatous ulcer (Figure 1D) with a large intramural hematoma.

### Background

Acute aortic syndrome (AAS) refers to the acute presentation of closely related life-threatening entities having similar clinical features and challenges, including the aortic dissection, intramural hematoma, and penetrating atheromatous ulcer<sup>1</sup> (Figure 2).

It has been suggested that aortic dissection, intramural hematoma, and penetrating atheromatous ulcer are variations on a continuum of aortic disease (Figure 3). Coady et al<sup>2</sup> suggested that in patients suspected of aortic dissection, 5% to 17% actually have either intramural hematoma or penetrating atheromatous ulcer. The International Registry for Acute Aortic Dissection, which reviewed 464 patients, revealed that two-thirds of patients were male, with a mean age of 63 years.<sup>3</sup> Women were found to be affected less often, and their mean age was 67 years.<sup>4</sup>

The classic aortic dissection is initiated by an intimal tear, with resultant propagation within the middle third of the medial layer of the aorta. Intramural hematoma originates owing to a rupture of vasa vasorum<sup>5</sup> or hemorrhage within an atherosclerotic plaque,<sup>6</sup> followed by aortic wall infarction, which, in turn, weakens and may rupture, sometimes resulting in aortic dissection.<sup>7</sup> Penetrating atheromatous ulcer is thought to be caused by the rupture of atheromatous plaque through the internal elastic lamina, with subsequent localized medial disruption and potential dissection, pseudoaneurysm formation, or free rupture.

In the International Registry for Acute Aortic Dissection, hypertension

was the most common predisposing factor (72%) for aortic dissection, followed by atherosclerosis (31%), history of cardiac surgery (18%), Marfan syndrome (5%), and iatrogenic causes (4%).<sup>3</sup>

### Clinical Manifestations

A high index of suspicion is required to promptly establish the diagnosis of this life-threatening entity, because the symptoms can be variable and may mimic those of more common conditions. The pain caused by intramural hematoma and penetrating atheromatous ulcer is similar to aortic dissection.

Other clinical features at initial evaluation, occurring with or without associated chest pain, may include congestive cardiac failure (7%), syncope (9%), acute stroke (6%), acute myocardial infarction, ischemic peripheral neuropathy, paraplegia, and cardiac arrest or sudden death.<sup>8</sup>

### Classification Systems

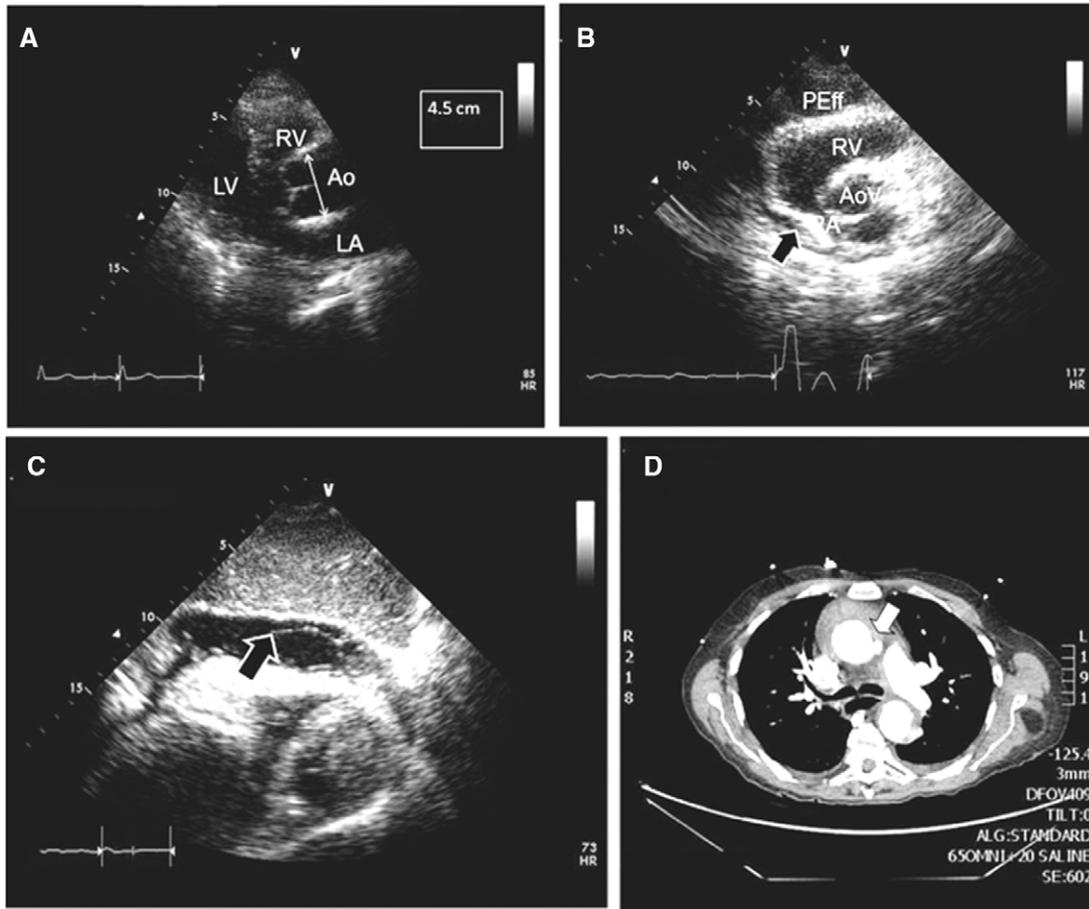
The most commonly used classification schemes for AAS are the De Bakey and the Stanford systems (Figure 4). For classification purposes, the ascending aorta refers to the part of the aorta proximal to the brachiocephalic artery, and the descending aorta refers to the aorta distal to the left subclavian artery.

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**Figure 1.** **A**, Transthoracic echocardiogram (parasternal long-axis view) showing mildly dilated proximal aorta at 4.5 cm. **B**, Transthoracic echocardiogram (parasternal short-axis view at aortic valve level) showing large pericardial effusion with the collapse of the right atrium in diastole (thick arrow) suggesting cardiac tamponade. **C**, Transthoracic echocardiogram (subcostal view) showing a large pericardial effusion (thick arrow). **D**, Axial contrast-enhanced CT scan shows penetrating atheromatous ulcer of the aorta (thick arrow) filling with contrast material. Ao indicates the proximal aorta; AoV, aortic valve; CT, computed tomography; LA, left atrium; LV, left ventricle; PEff, pericardial effusion; RA, right atrium; and RV, right ventricle.

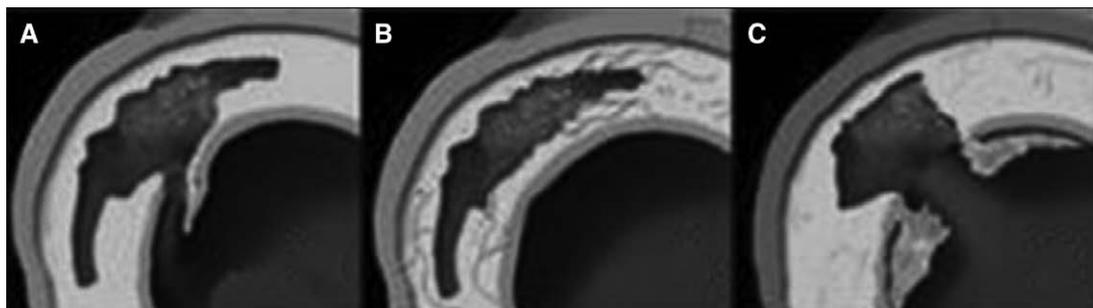
The acute dissection is defined as occurring within 2 weeks from the onset of initial symptoms to the time of first presentation; subacute between 2 and 8 weeks from the onset of symptoms, and chronic >8 weeks from the onset of symptoms.

**Diagnostic Workup**

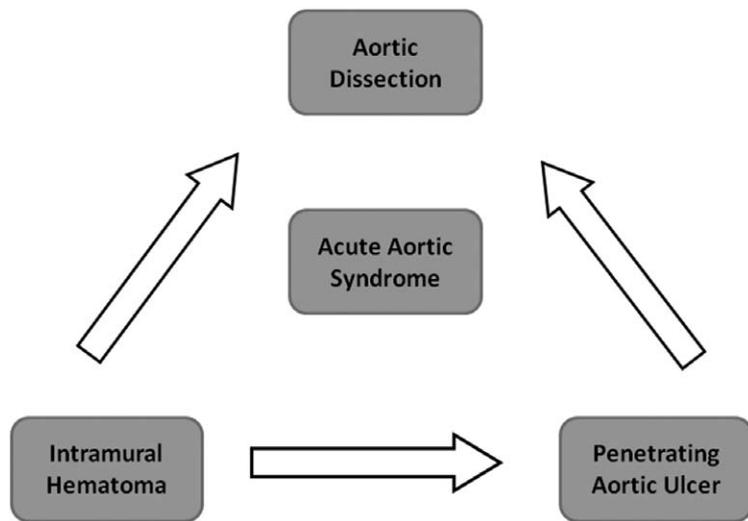
In the setting of suspected aortic disease, the diagnostic workup may include ECG, chest radiography, myocardial markers, transthoracic echocardiogram, transesophageal echocardiogram, CT, and MRI.

**Electrocardiography**

The electrocardiographic findings in patients with aortic dissection are nonspecific. Three-quarters of ECGs with dissection are normal or demonstrate nonspecific ST-segment or T-wave changes, and 25% have left ventricular hypertrophy.<sup>3</sup>



**Figure 2.** Classification of acute aortic syndrome. **A**, Aortic dissection. **B**, Intramural hematoma. **C**, Penetrating atheromatous ulcer. Reprinted from Berger et al<sup>1</sup> with permission from the publisher. Copyright © 2006, Radiological Society of the Netherlands.



**Figure 3.** Acute aortic syndrome. Progression of one type to another is shown by arrows.

### Chest Radiography

A chest radiograph may show widening of the aortic contour; other features may include displaced calcification, aortic kinking, or opacification of the aortopulmonary window.<sup>9</sup>

### Biomarkers

Currently, there are no reliable biomarkers diagnostic of aortic dissection, although a number of markers are under investigation. Release of smooth muscle proteins, soluble elastin fragments, and myosin heavy chain and

creatine kinase BB isoforms have been reported after aortic dissection.<sup>10</sup>

D-Dimers, when elevated >500 µg/L, appear to correlate with the extent and severity of acute aortic dissection, but fail to distinguish AAS from pulmonary embolism. Elevated D-dimers in the clinical context of suspected aortic dissection should prompt an urgent CT or transesophageal echocardiography for confirmation of either life-threatening entity.<sup>10</sup>

### Diagnostic Imaging

In the acute setting, the primary goals of diagnostic imaging in patients with suspected AAS are confirmation of clinical suspicion, classification of dissection, localization of the tear, assessment of the extent of dissection, and identification of signs indicating the need for an emergency intervention (pericardial, mediastinal or pleural hemorrhage).

### Computed Tomography

Contrast-enhanced CT scanning has become the most commonly used modality in evaluating aortic dissection. With the advent of spiral CT, studies may be performed in less time than before, with less patient discomfort, greater accuracy, and lower iodine load. Spiral CT has been proposed as the diagnostic test of choice, because it has

a sensitivity of 94% and specificity of 87% in diagnosing aortic dissection.<sup>11</sup>

### Echocardiography

For stable patients, any of the imaging modalities may be useful depending on the local expertise. Transthoracic echocardiography is less sensitive (59%–83%) and less specific (63%–93%) for the diagnosis of aortic dissection than other modalities.

In hemodynamically unstable patients, transesophageal echocardiography may be the first diagnostic modality, but it may not be able to provide any details of the abdominal segments. Willens and Kessler<sup>12</sup> reported a higher accuracy of transesophageal echocardiography for diagnosing aortic disease with a sensitivity of 97% to 100% and a specificity of 77% to 100%.

### Magnetic Resonance Imaging

MRI is a high-precision diagnostic modality for the diagnosis of aortic dissection. The sensitivity and specificity of MRI for the diagnosis of aortic dissection has been reported to be between 95% and 100%.<sup>13</sup>

### Aortography

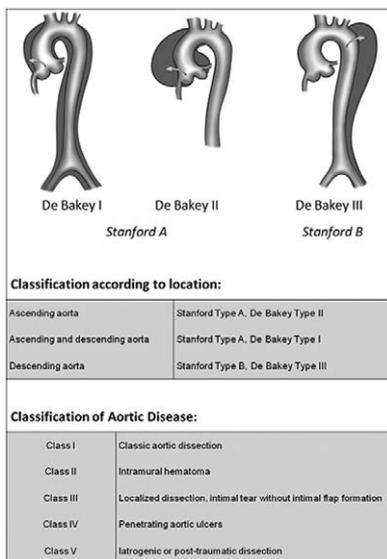
Aortography is now rarely used for the diagnosis of acute aortic dissection. Comparative studies found the sensitivity of aortography to be 88% and the specificity 94%.<sup>14</sup>

### Intravascular Ultrasound

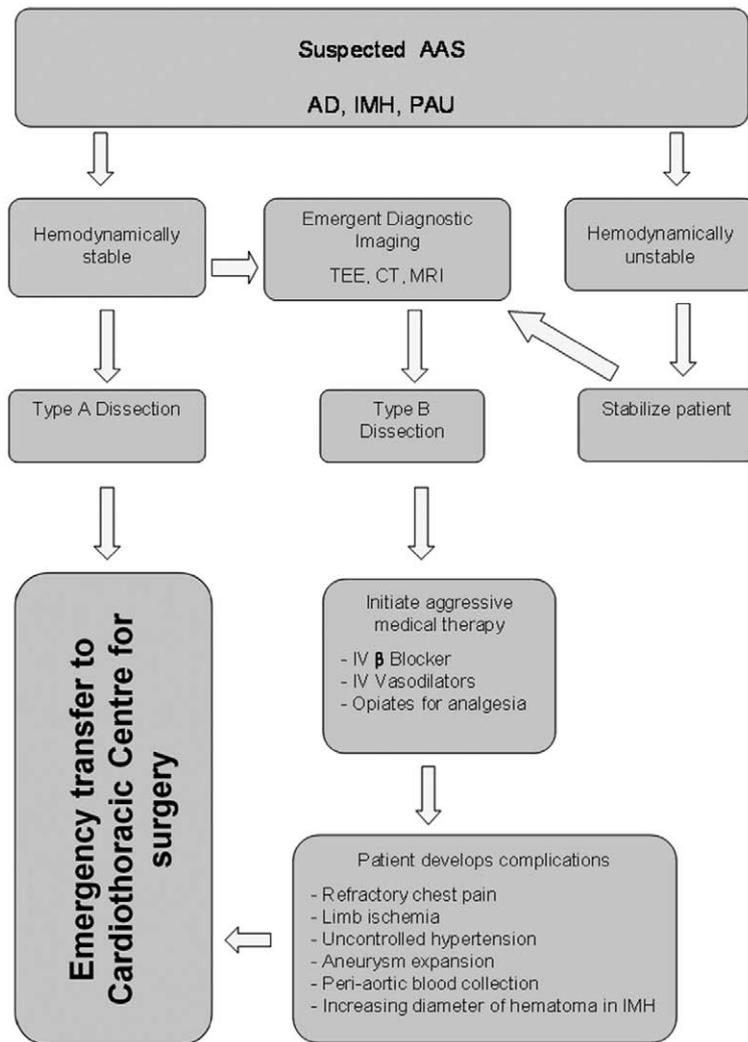
Intravascular ultrasonography is a promising tool for the accurate determination of the location and extent of dissection and the assessment of branch vessels. It may be particularly helpful to differentiate aortic dissection from penetrating atheromatous ulcer, but it has not been widely used.

### Coronary Angiography

Routine coronary angiography is not recommended before surgery for type A aortic dissection because of the concern about the delay in emergency surgery.<sup>15</sup>



**Figure 4.** Classification of aortic disease.



**Figure 5.** The algorithm for the management of AAS. AD indicates aortic dissection; CT, computed tomography; IMH, intramural hematoma; IV, intravenous; PAU, penetrating atheromatous ulcer; and TEE, transesophageal echocardiogram.

**Management Issues**

The therapy for aortic dissection aims to halt progression of the dissecting hematoma. Lethal complications arise not from the intimal tear itself, but rather from the subsequent course taken by the dissecting aorta, such as vascular compromise or aortic rupture.

**Initial Medical Management**

The initial management of patients with AAS involves adequate pain relief and an aggressive control of blood pressure. This helps to limit the propagation of dissected wall components and to reduce dP/dt. Reduction in pulse pressure with a target systolic

pressure of 100 to 120 mmHg and a heart rate of 60 to 80 bpm, to maintain adequate cerebral, coronary, and renal perfusion, is a priority. Use intravenous  $\beta$ -blockade as first line of therapy.

Vasodilators may be used to control blood pressure, but they should never be used as an initial form of therapy before commencing  $\beta$ -blockers because of the reflex tachycardia and increase in the force of left ventricular ejection leading to increased aortic wall stress. Opiate analgesia should be used, because it helps attenuate the release of catecholamines in response to pain, with resultant tachycardia and hypertension.

**Type A Aortic Dissection**

The main aim behind any surgical intervention in type A aortic dissection is the prevention of life-threatening complications including aortic rupture and pericardial effusion, which may lead to cardiac tamponade. Acute type A dissection has a mortality rate of 1% to 2% per hour during the first 24 to 48 hours of the onset of symptoms. Without surgical treatment, the mortality rate is 20% by 24 hours, 30% by 48 hours, 40% at 1 week, and 50% at 1 month.<sup>3</sup> The operative mortality for ascending aortic dissections at experienced centers varies widely, between 10% and 35%, but is less than the 50% mortality with medical therapy.<sup>16</sup>

**Uncomplicated Type B Aortic Dissection**

Medical management remains the mainstay of treatment for patients with uncomplicated type B disease. In 384 patients with type B dissection, 73% were treated medically with 10% in-hospital mortality. Long-term survival was 80% at 5 years.<sup>3</sup>

**Complicated Type B Aortic Dissection**

Complicated type B aortic dissection is differentiated from uncomplicated type B aortic disease by the presence of a distal malperfusion syndrome or rapid disease progression. Indications for emergency intervention include the prevention organ or limb ischemia, aneurysm expansion and risk of rupture, periaortic blood collection, intractable pain, or uncontrolled hypertension.

**Intramural Hematoma**

Surgery is advocated in patients with type A intramural hematoma, and a trial of medical therapy is advocated in patients with type B intramural hematoma, similar to type A and B aortic dissection. The indications for surgical intervention in type B intramural hematoma include recurring, refractory chest pain and increasing extent or diameter of the hematoma.

The complete resolution of type B intramural hematoma has been documented in 50% to 80% of patients.<sup>17</sup> However, these patients can also progress to frank dissection or late aneurysm.<sup>18</sup> Resolution is more likely in young patients,<sup>17</sup> aortic diameter <4.0 to 4.5 cm,<sup>7</sup> thickness of the hematoma <1 cm,<sup>7</sup> and  $\beta$ -blocker usage.<sup>18</sup>

### Penetrating Atheromatous Ulcer

Penetrating atheromatous ulcer involving the ascending aorta is uncommon; however, the ulcer usually ruptures and is commonly lethal. Patients with penetrating atheromatous ulcer involving the descending aorta can initially be treated conservatively with aggressive medical therapy and with close observation, similar to descending aortic dissection.

Like intramural hematoma, penetrating atheromatous ulcer is seen more often in the descending thoracic aorta. Stanson et al<sup>6</sup> advocate an aggressive surgical approach. These ulcers often arise in the mid-descending thoracic aorta, and, therefore, any surgical resection should include this portion of the aorta. Hussain and associates<sup>19</sup> challenge this approach and present evidence that nonoperative management was successful in many cases. Our algorithm for the management of AAS is shown in Figure 5.

### Prognosis

Prompt diagnosis and appropriate and timely management reduce the frequency of fatal outcomes.

Coady et al<sup>2</sup> demonstrated that the rate of aortic rupture is much higher for intramural hematoma (35%) and penetrating atheromatous ulcer (42%) in comparison with aortic dissection (type A 7.5%, type B 4.1%).

### Long-Term Follow-Up

All patients with known aortic disease require close surveillance after discharge and annually thereafter. The 10-year actuarial survival rate in this group of patients has ranged between 30% and 60%.<sup>16,20</sup>

Lifelong treatment of hypertension is required, and regular assessments of the aorta should be performed at 1, 3, 6, 9, and 12 months, and every 6 to 12 months thereafter, as well, depending on the aortic size.

After medically or surgically treated acute aortic dissection, MRI appears to be the imaging modality of choice. The most important findings on imaging are aortic diameter, signs of aneurysm formation, and hemorrhage at surgical anastomosis or stent-graft sites.

### Conclusions

AAS should be considered in all patients presenting with chest pain. The etiologies include aortic dissection with a false lumen and intimal flap, intramural hematoma, and penetrating atheromatous ulcer. The surgical and percutaneous strategies to treat AAS continue to improve and evolve. These disorders can be assessed by MRI, CT, or transesophageal echocardiography with high diagnostic accuracy. There is a higher risk of rupture with intramural hematoma and penetrating atheromatous ulcer, especially when located in the ascending aorta. Currently, the recommendations are to treat type A lesions surgically, whereas type B lesions are initially managed medically. All patients with AAS require close surveillance after discharge.

### Case Resolution

The patient was immediately transferred to a Cardiothoracic Surgical Center where she underwent emergency surgery. She had cardiac tamponade with significant bloody pericardial effusion and an extensive intramural hematoma of the ascending aorta. The ascending aorta was replaced with a Vacutek Vascular graft, reinforced with bovine pericardium.

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### Disclosures

None.

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KEY WORDS: aortic dissection ■ pericardium  
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 ■ surgery