

# Acute Aortic Syndromes

The spectrum of penetrating aortic ulcers, intramural hematomas, and dissections.

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**A**ortic dissection is the disruption of the intima and media of the aortic wall. Blood flow within the resulting false lumen can have devastating consequences for end-organ perfusion, leading to significant morbidity and mortality. Aortic dissection is estimated to occur annually in 3.5 per 100,000 patients, with a 5:1 predominance in men.<sup>1</sup> Approximately one-third of all dissections affect only the descending aorta and are therefore classified as Stanford type B. As imaging technology and medical knowledge advance, intramural hematomas and penetrating aortic ulcers distinguish themselves as separate entities. Although dissections, intramural hematomas, and penetrating aortic ulcers may be considered acute aortic syndromes, the natural history of each may require significantly different management.

## AORTIC DISSECTION

Generally, aortic dissections are classified according to the location of the entry tear, the extent of the dissection, and the presence or absence of complications (Figure 1).<sup>2</sup> The dissection is considered Stanford type A if the ascending aorta and/or the aortic arch are involved in the dissection. Stanford type B dissections are limited to the descending aorta (Figure 2). Type A dissections require prompt surgical intervention to avoid cardiac complications. Surgery for type B dissections is reserved for patients who do not respond to medical management of hypertension.

Classically, in type B dissections, the transverse entry tear occurs immediately distal to the left subclavian artery on the posterolateral aspect of the aorta. Hypertension, connective tissue diseases (such as Marfan syndrome or Ehlers-Danlos syndrome type IV), trauma, and surgery

can weaken the aortic wall, creating the origin of dissection. This intimal flap, known as the *lamella*, divides the aorta into a true and false lumen. The lamella can have points of entry and re-entry along the length of the dissection. Pain between the shoulder blades in a hypertensive patient is the most common presenting symptom. The broad spectrum of symptoms on presentation, including weakness of one limb or mesenteric ischemia causing abdominal pain, can cause delay in diagnosis.

Whether acute (< 2 weeks) or chronic (> 2 weeks), the determination of whether a dissection is complicated or uncomplicated has important prognostic implications.<sup>3</sup> Complicated dissections involve impending rupture, malperfusion, uncontrollable pain, uncontrollable hypertension, or rapid growth. The patient's response to anti-

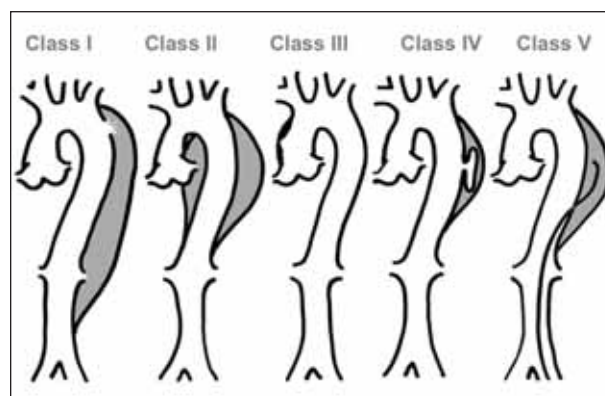
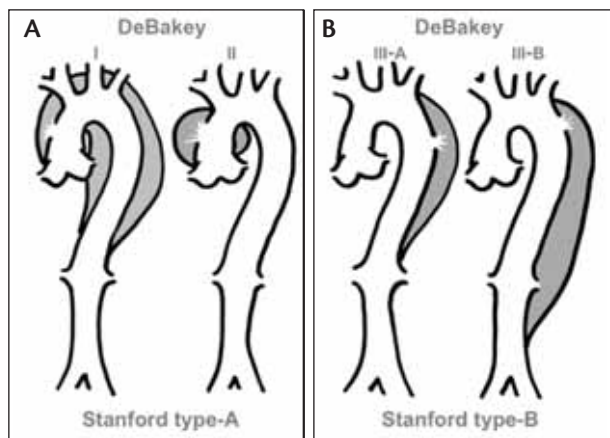


Figure 1. Classification proposed by Svensson et al<sup>2</sup> and later adopted by the European Society of Cardiology. Class I, classic double-barrel dissection; class II, intramural hematoma without intimal disruption; class III, intimal tear; class IV, penetrating aortic ulcer; class V, iatrogenic injury.



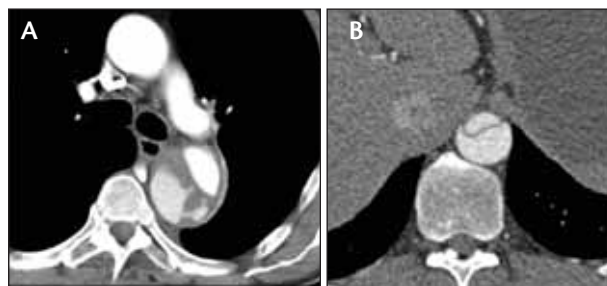
**Figure 2.** Anatomical classification of aortic dissection according to Stanford and DeBakey: Stanford type A/DeBakey I and II (A) and Stanford type B/DeBakey IIIa and IIIb (B).

hypertensive therapy will determine the need for surgical intervention.

Computed tomographic (CT) angiography and transesophageal echocardiography are most commonly used to diagnose dissections (Figure 3).<sup>2,4,5</sup> Depending on availability, magnetic resonance angiography may add valuable hemodynamic characterization of the lesion.

The International Registry of Acute Aortic Dissection (IRAD) reports an in-hospital mortality rate of approximately 30% for open surgical repair of dissection.<sup>4,6</sup> Medical treatment alone carries an in-hospital mortality rate of 10%.<sup>3,4</sup> Medical management to limit propagation should be directed toward a heart rate of 60 bpm and a maximal systolic blood pressure of 120 mm Hg using intravenous beta blockade in an intensive care unit.<sup>7</sup> If clinically stable, the patient should transition to long-term oral antihypertensive medications<sup>8</sup> and serial outpatient evaluation.

Approximately one in five uncomplicated acute type B dissections become complicated.<sup>9</sup> Four general strategies exist for repair of complicated dissections: open surgical repair,<sup>3</sup> surgical fenestration,<sup>10</sup> stent graft coverage of the entry tear with surgical revascularization of ischemic organs as necessary, and endovascular fenestration and branch vessel stenting.<sup>11</sup> The goal of intervention is to obliterate the entry tear and achieve end-organ perfusion. The extent of coverage of the abnormal aorta must be balanced with the risk of paraplegia. Proper placement of a stent graft in a normal aorta often requires coverage of the left subclavian artery. Selective revascularization of the subclavian artery should be considered when coverage of long aortic segments is required and in those with previous infrarenal aortic surgery, renal insufficiency, and hypoplastic right vertebral



**Figure 3.** CT angiogram of aortic dissection. A chronic dissection showing complex false and true lumen communication (A) and a typical image of an acute dissection (thin lamella) (B).

artery.<sup>12</sup> Before stent placement, each branch vessel perfusion should be characterized with a clear plan and material preparation for revascularization should ischemia occur after stent graft implantation. Results from IRAD comparing stent grafting to open surgery for treating complicated acute type B dissections showed a significant decrease in in-hospital mortality (from 33.9%–10.6%).<sup>7</sup> Nonrandomized studies are necessary to determine the superiority of endografting, open surgery, and medical management.<sup>9,13</sup>

Furthermore, the question of whether to apply stent graft technology to uncomplicated acute dissections remains unanswered. Tsai et al showed that approximately one in four medically treated patients will die within 3 years from onset of disease. Additionally, approximately one in four survivors treated medically will develop dilatation of the false lumen.<sup>14</sup> Obliteration of the entry tear may improve aortic wall remodeling and healing.<sup>15,16</sup> Initial results with early endograft implantation show a technical success of 70% to 100% in false lumen thrombosis (Figure 4), but long-term outcomes have yet to be published.<sup>17,18</sup>

Two-year results of the INSTEAD trial show no significant differences in mortality between stent grafting of uncomplicated acute dissections and medical treatment alone. Intervention actually resulted in an increased adverse event rate.<sup>19</sup> Sixteen percent of patients under medical management required a stent graft. Favorable aortic remodeling with stent grafting did not affect mortality at 2 years. Endovascular intervention for aneurysmal dilatation must be weighed against the risk of paraplegia and the risk of branch vessel revascularization. Stent grafting is believed to be more effective in the acute setting before septal fibrosis of the dissecting membrane occurs, limiting aortic remodeling. Endovascular treatment of chronic dissections is questionable due to the likely persistent pressurization of the false lumen from the noncompressible fibrotic septum.

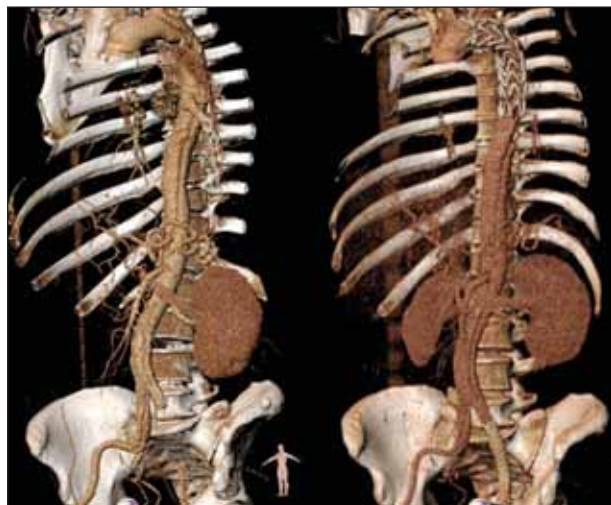


Figure 4. Preintervention and postintervention CT angiogram after processing reconstruction of aortic dissection. Notice the reperfusion of the right kidney after sealing the entry tear. Iliac patency was achieved by subsequent placement of a covered stent.

A large aortic diameter, large false lumen diameter, patent entry site, presence of blood flow in the false lumen, Marfan syndrome, chronic obstructive pulmonary disease, age older than 60 years, and female gender predict aortic enlargement after conservative treatment. Similarly, predictors of aortic rupture after conservative treatment are old age, chronic obstructive pulmonary disease, hypertension, patent false lumen (especially partial thrombosis), and aortic diameter > 55 mm.<sup>14,20-23</sup> Patients with uncomplicated type B dissection but with large initial aortic diameters, a patent false lumen, or persisting hypertension will likely benefit most from early endovascular treatment.

### INTRAMURAL HEMATOMA AND PENETRATING AORTIC ULCER

O'Gara first described acute aortic syndrome, including intramural hematoma, penetrating ulcer, and dissection, as well as aortic aneurysm rupture.<sup>24</sup> These entities share acute pain and disruption of aortic wall integrity. Intramural hematoma and penetrating aortic ulcers account for 5% to 30% of acute aortic syndromes. IRAD classified 58 of 982 patients (6%) as having intramural hematoma.<sup>25</sup> *Intramural hematoma* refers to the accumulation of blood within the media layers of the aorta without disruption of the intima. Atherosclerotic penetrating ulcers disrupt the internal elastic lamina. Malperfusion is rare with both of these aortic pathologies.

Despite similarities in presentation and management, the disease mechanisms are thought to be different.<sup>25-27</sup>

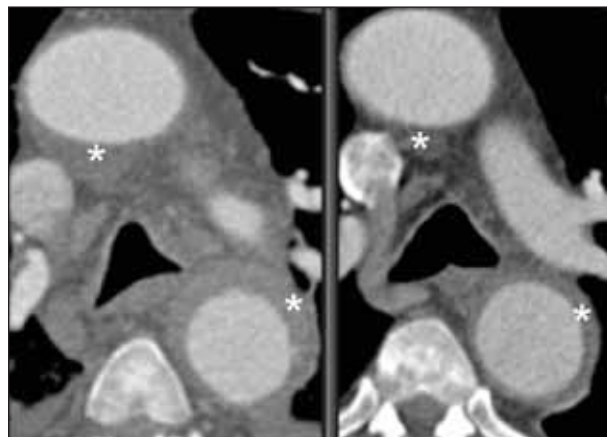


Figure 5. Regression of intramural hematoma (\*) documented by computed tomography 3 weeks after diagnosis (treated conservatively). Notice the re-expansion of the true lumen as the hematoma regresses.

Although the initiating mechanism for penetrating aortic ulcers appears to be principally atherosclerotic, intramural hematoma may be caused by rupture of the vasavasorum, causing hematoma within the media. This vasavasorum theory has been challenged by observations that minor intimal disruption or penetrating ulcers feed the hematoma. This would explain why coverage of a localized intimal defect results in reabsorption of hematoma in the entire aorta.<sup>28-30</sup> The IRAD investigators also showed that intramural hematoma and penetrating ulcers occur more often in the descending aorta (approximately two-thirds of cases) as opposed to classic dissections.<sup>25</sup>

The compressed aortic lumen surrounded by crescent intramural hematoma within the media can often be evaluated by noncontrast CT scanning. Calcifications usually remain internal to the accumulated blood in the media wall. Associated small ulcers disrupting the intima can be frequently detected in high-resolution imaging studies, making the distinction between intramural hematoma and penetrating aortic ulcers difficult. In typical penetrating ulcers, a contained intramural hematoma is limited by heavy atherosclerotic burden of the intima.

IRAD demonstrated that one in five patients will die during their first admission for intramural hematoma, especially if the ascending aorta is involved.<sup>25,31</sup> For hematoma limited to the aortic arch or the descending aorta, the mortality rate is < 10%. Conservative treatment will result in regression of the hematoma in more than half of patients (Figure 5). However, 28% to 47% of patients will progress to a double-lumen dissection, and 20% to 25% will progress to aortic rupture.<sup>25,32-35</sup>

For penetrating aortic ulcers, the dreaded complication of disruption of the aortic layers is aortic rupture or

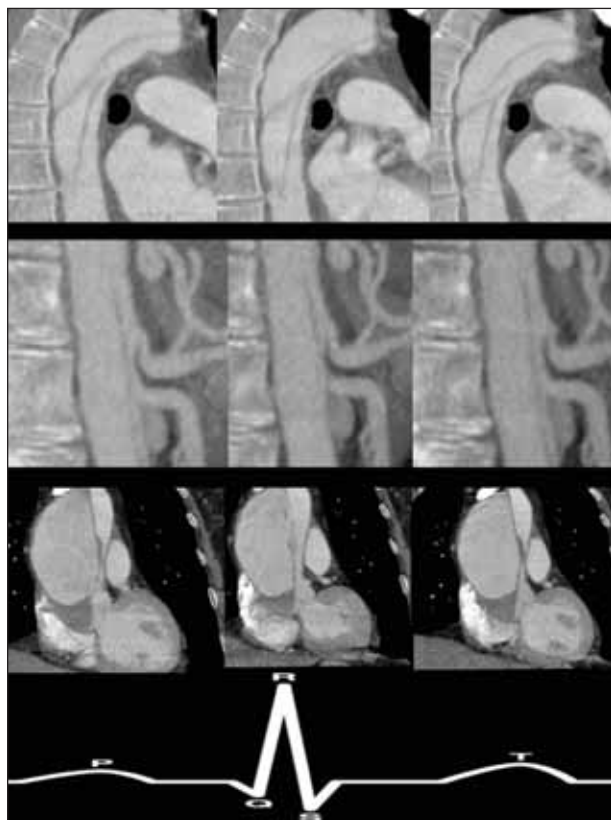


Figure 6. Dynamic electrocardiography-gated CT angiogram of acute aortic dissection showing the mobility of the intimal flap.

pseudoaneurysm formation. One in four symptomatic aortic ulcers will progress to pseudoaneurysm, and more than one-third will eventually rupture.<sup>36</sup> Risk factors for progression of these lesions are the presence of symptoms, old age and advanced atherosclerosis, evidence of inflammatory plaque, and interval increase in size.<sup>28,36,37</sup>

Similar to type B dissections, the mainstay of treatment for intramural hematoma and penetrating aortic ulcers is blood pressure and heart rate control with beta-blocker therapy. Penetrating ulcers are frequently short, localized lesions that permit minimal aortic coverage. Intramural hematoma, on the other hand, may involve a long segment of aorta with mismatch of the lumen compressed by hematoma and an enlarged outer aortic diameter. This mismatch may complicate endograft sizing and branch vessel management. Most experts agree that coverage of identified ulcers will result in regression of the hematoma. Catheter and sheath manipulations should be minimized to avoid additional fenestrations in the diseased intima. Patients with localized lesions at high risk for progression with adequate seal zones may be particularly amenable to stent graft implantation to prevent future complications.

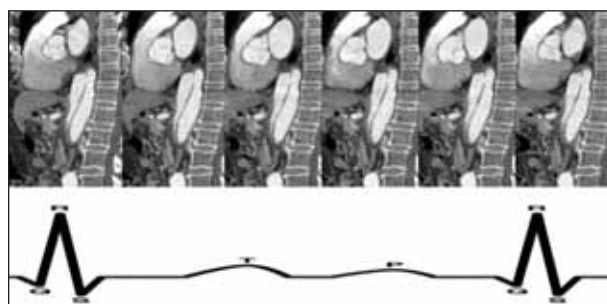


Figure 7. Dynamic electrocardiography-gated CT angiogram of chronic aortic dissection showing no mobility of the intimal flap.

## THE FUTURE

Dynamic CT angiography shows that the intimal flap is very mobile in the acute phase of dissection, whereas it is thick and rigid in latter stages. Electrocardiographic gating also significantly diminishes artifacts in pulsatile structures, such as the aorta and the dissection flap, and shows the distensibility of the aorta, enabling more accurate sizing (Figures 6 and 7).<sup>23</sup> Additionally, functional vascular magnetic resonance imaging techniques are able to indirectly measure intra-arterial pressure and tissue perfusion. Intravascular ultrasound may also prove useful in the dynamic evaluation of acute aortic syndromes.

Aortic dissections are currently treated with endovascular devices designed for treating aneurysms. The ideal stent graft for acute type B dissections should be maneuverable, flexible, and conformable to even the most fragile aorta. Using bioabsorbable materials may also promote successful aortic healing and remodeling, normal aortic physiology, and the absence of late device-related complications.

## CONCLUSION

Rather than distinct entities, aortic dissection and variants—intramural hematoma and penetrating aortic ulcer—likely represent a spectrum of aortic pathology. As the understanding of this spectrum progresses and medical and endovascular technology advance, acute aortic syndrome treatment will improve. On the basis of current knowledge, all patients require immediate and continued antihypertensive therapy that includes beat blockade. Intervention should be reserved for those with complications and those considered to be at high risk for complications. When indicated, endograft implantation offers better overall results than open surgery or endovascular fenestration. Exclusion of the intimal entry point in acute lesions may promote aortic remodeling. Close and lifelong follow-up is mandatory for all patients with dissections, intramural hematoma, and penetrating aortic ulcers. ■



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