ABSITE Corner: Aortic dissection

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ABSTRACT

Key points: (a) Aortic dissection (AD) is the most common aortic emergency; (b) If untreated, 25-33\% of patients with AD die within the first 24 hours, 50-70\% die within 48 hours, and 75-80\% of patients with undiagnosed AD will die within two weeks; (c) The distinguishing feature of AD is the tearing of the aortic intima; (d) The most common site of intimal tear is within 2-3 centimeters of the aortic valve, with \textasciitilde 90\% occurring within 10 centimeters of the aortic valve; (e) There are two commonly used AD classifications – The Stanford classification and the DeBakey classification; (f) More than 30\% of acute dissections are not recognized on initial evaluation; (g) Only \textasciitilde 25\% of patients with AD have widened mediastinum on antero-posterior radiographs; (h) Contrast-enhanced spiral computed tomography is the initial investigation of choice for the evaluation of suspected AD; (i) Beta-adrenergic antagonists should be administered initially to reduce the rate of change of blood pressure (\Delta P/\Delta t) and the shear forces on the aortic wall, with target heart rate of 60-80 beats/minute and target systolic blood pressure around 100-120 mmHg; (j) Stanford Type A dissections are considered surgical emergencies and their prognosis is very poor when unrecognized or treated non-operatively; (k) Stanford Type B dissections may be treated non-surgically, which revolves around strict blood pressure control.

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INTRODUCTION

Aortic dissection (AD) is the most common aortic emergency. It is approximately two times more common than abdominal aortic aneurysm rupture. Best estimates of the incidence of AD are based on autopsy studies (AD is found on 1-3\% of autopsies). Population-based studies suggest that the incidence of AD is approximately 5-30 cases per million people per year. The diagnosis of AD is missed in \textasciitilde 40\% of cases on initial presentation, and \textasciitilde 30\% of AD are first diagnosed on post mortem exams.

Some authors report that \textasciitilde 20\% of patients with AD die before hospital admission. Untreated, 25-33\% of patients with AD die within the first 24 hours, 50-70\% die within 48 hours, and 75-80\% of patients die within two weeks. In fact, approximately 1-3\% of patients with proximal AD die every hour during the first 48 hours.

Aortic dissection is rare in individuals who are \textasciitilde 40 years old. Roughly 75\% of dissections occur between ages of 40-70 years (peak incidence, 50-65 years). Male to female ratio ranges from 1:1 to 3:1. Conditions associated with AD include hypertension, Marfan’s syndrome, Ehlers-Danlos syndrome, bicuspid aortic valve, annuloaortic ectasia, familial AD, adult polycystic kidney disease, Turner syndrome, Noonan syndrome, osteogenesis imperfecta, aortic coarctation, and other connective tissue disorders. Aortic dissection is also seen in heritable metabolic disorders such as homocystinuria and familial hypercholesterolemia, and its incidence may increase with pregnancy, syphilis, crack cocaine use, and cardiac catheterization procedures. It is important to differentiate AD from aortic aneurysms (abnormal dilation of the aorta) and aortic transections (usually due to high-energy trauma).

PATHOPHYSIOLOGY

The distinguishing feature of AD is the tearing of the aortic intima. Intimal tears lead to a false lumen formation that can progress in antegrade and/or retrograde direction via formation and propagation of a subintimal hematoma. The dissecting hematoma may occupy 50-100\% of the aortic circumference. Rupture can occur back into the aortic lumen or externally into the pericardium or the mediastinum. External rupture often results in fatal pericardial tamponade. Dissection can occlude aortic branches, most commonly involving renal, spinal, coronary, and iliac arteries.

Diseases that weaken the aortic wall predispose the patient to AD. Normal aorta contains collagen, elastin, and smooth muscle cells that form the aortic intima, media, and adventitia. With aging, degenerative changes lead to breakdown of the collagen, elastin, and smooth muscle, with concurrent increase in basophilic ground substance – a condition known as cystic medial necrosis (CMN). In early CMN, basophilic ground substance accumulates in the media, forming cyst-like pools. The media in these focal areas may show cellular loss/necrosis. Shearing forces then facilitate the separation of layers within the aortic media. Atherosclerosis that leads to occlusion of the vasa vasorum may also be contributory. Marfan syndrome, Ehlers-Danlos syndrome, and other connective tissue diseases affect the media of the aorta and make it especially prone to dissection. Intimal rupture subsequently occurs at points of fixation along the aorta where hydraulic stresses are maximal. Pulsatile flow and high blood pressure further contribute to propagation of the dissection. The most common site of intimal tear is within 2-3 centimeters of the aortic valve, with \textasciitilde 90\% occurring within 10 centimeters of the aortic valve. Approximately 5-10\% of aortic dissections do not have an obvious intimal tear and are usually attributed to rupture of the aortic vasa vasorum. Tears may also be seen distal to the left subclavian artery.

AORTIC DISECTIONS: CLASSIFICATION

There are two commonly used AD classifications. The first one, the Stanford classification, consists of two types: (a) Type A involves the ascending aorta; and (b) Type B involves the descending aorta (i.e., distal to the left subclavian artery). See Figure 1 for description of the Stanford classification of AD.
The second one, the DeBakey classification, involves three types: (a) Type I involves the ascending and the descending aorta; (b) Type II involves ascending aorta only; and (c) Type III involves the descending aorta only. DeBakey type III dissections are further subdivided into IIIa and IIIb subgroups. Type IIIa refers to dissections that originate distal to the left subclavian artery but extend both distally and proximally (mostly above the diaphragm). Type IIIb refers to dissections that originate distal to the left subclavian artery and extend only distally (may extend below the diaphragm). See Figure 1 for description of the DeBakey classification of AD.

Stanford type A dissection can be compared to DeBakey types I and II. Stanford type B dissection can be compared to DeBakey type(s) III. The Stanford system also helps determine the need for acute surgical intervention. Here, type A dissections usually require surgery and type B dissections may be managed non-surgically in many cases.

### DIAGNOSTIC CONSIDERATIONS

No single sign or symptom can positively identify acute AD. In fact, more than 30% of acute dissections are missed on initial evaluation. Aortic dissections usually present with “tearing” chest pain that radiates to the back. Patients may have reduced or absent peripheral pulses and/or soft early diastolic murmur. Clinical history may also include a syncopegal event. Diagnosis can be confirmed using advanced diagnostic imaging (see below). If aortic branches are involved, there may be clinical evidence of acute renal failure, paraplegia, limb ischemia, cerebral or myocardial infarction.

Chest pain is the most common presenting symptom in patients with AD. This highlights the need to include AD in the differential diagnosis of all clinically significant acute chest pain. The pain of AD typically is distinguished from the pain of acute myocardial infarction (MI) by its abrupt onset and maximal intensity from the time of onset. In fact, the sudden onset of chest pain has been shown to have a sensitivity of over 80% for AD. While the presence of “ripping” or “tearing” pain significantly increases the likelihood of AD, it is important to note that some patients present with no pain (~10%) or only mild pain (often mistaken for musculoskeletal conditions of the thorax, groin, or back).

Established clinical protocols for the assessment of chest pain should be followed. One such system is called the P-Q-R-S-T protocol (pain, quality, radiation, severity, and timing). In terms of specific clinical correlates, anterior chest pain and chest pain that mimics MI usually is associated with anterior arch or aortic root dissection. In this scenario, dissection interrupts blood flow to the coronary arteries, resulting in myocardial ischemia. Pain that radiates to the neck or jaw indicates that the dissection involves the aortic arch and extends into the area of great vessels. Pain that is “tearing” or “ripping” in nature and felt in the interscapular area may indicate that the dissection involves the descending aorta. The pain may change/evolve as the AD progresses. Painless dissection (~10%) is most often seen in patients with neurologic complications from the dissection and in Marfan syndrome.

Upper extremity systolic pressure differential of >20 mmHg should increase the suspicion of AD, but it does not rule it in. In fact, blood pressure differential between upper extremities may be found in ~20% of people without aortic dissection. Hypertension may result from catecholamine surge or may be a reflection of underlying essential hypertension. Hypotension is an ominous finding and may be the result of excessive vagal tone, cardiac tamponade, or hypovolemia/hemorrhage following aortic rupture. Dyspnea may be due to congestive heart failure or tracheal/bronchial compression. Dysphagia from esophageal compression may also be evident.

Neurologic deficits are seen in up to 20% of cases on presentation, and usually include syncope and non-specific mental status changes. Syncope may be associated with increased vagal tone, hypovolemia/hemorrhage, arrhythmia, cerebral infarction from compromised cerebral blood flow, or spinal ischemia from interruption of spinal blood supply. Horner syndrome (ptosis, miosis, anhidrosis) may be due to interruption of the cervical sympathetic ganglia. Peripheral nerve ischemia can manifest with extremity paresthesias. Hoarseness from recurrent laryngeal nerve compression also has been described. One must pay special attention to carotid, brachial, and femoral pulses on initial examination and look for development and/or progression of bruits on subsequent examinations.

Cardiac tamponade, often manifested by muffled heart sounds, hypotension, pulsus paradoxus, and jugular venous distension, must be recognized promptly. Hemothorax may be seen if the dissection ruptures into the pleura. Superior vena cava (SVC) syndrome (i.e., SVC compression from a large distorted aorta) has also been described.

### ELECTROCARDIOGRAPHY (ECG)

All patients with suspected thoracic AD should have an electrocardiogram (ECG). In acute thoracic dissection, ECG can mimic the changes seen in acute cardiac ischemia. In the presence of chest pain, these signs can make distinguishing dissection from MI very difficult, which is an important consideration when administering thrombolytic therapy to patients with chest pain. The incidence of abnormal ECG findings is greater in Stanford type A dissections than in other types of dissections. Stanford type A dissections can be associated with ST elevation because the dissection may interrupt coronary arterial blood flow. Moreover, the ECG abnormality is an ST depression. If the dissection involves the coronary ostia, the right coronary artery...
is the most commonly involved vessel, leading to inferior ST-segment elevation.

OVERVIEW OF DIAGNOSTIC IMAGING
With regards to radiographic appearance of AD on plain films, one must look for a mediastinal widening (width >8 cm on anteroposterior chest radiograph). However, only ~25% of patients with AD have widened mediastinum on anteroposterior radiographs. Tortuous aorta, common in hypertensive patients, may be difficult to distinguish from widened mediastinum. If doubts persist, a high-quality posterior-anterior radiograph may be helpful. Abnormal (i.e., blunted) aortic knob can be seen in >60% of patients. Ring sign (displacement of the aorta >5 mm past the calcified aortic intima) is considered a specific radiographic sign. Other abnormalities seen on chest radiography may include: (a) left apical cap; (b) tracheal deviation; (c) depression of left main bronchus; (d) esophageal deviation; and (e) loss of the paratracheal stripe. Of note, >10% of the chest radiographs of patients with aortic dissection are interpreted as normal. Thus, the presence of AD is not determined by any one sign, but rather by a combination of findings in conjunction with high degree of clinical suspicion.

The use of advanced medical imaging may help reduce the rate of missed AD. Contrast-enhanced spiral computed tomography (CT) is the initial investigation of choice for the evaluation of suspected AD. In terms of overall diagnostic accuracy, CT is comparable to magnetic resonance imaging (MRI/MRA) and trans-esophageal echocardiography (TEE). In fact, sensitivity and specificity of CT scanning in the setting of AD approaches 100%, and CT appears to be superior to both TEE and MRI in the assessment of aortic arch vessel involvement.

COMPUTED TOMOGRAPHY
Contrast-enhanced spiral CT is rapid, non-invasive, widely available, visualizes the entire aorta, demonstrates the extent of the dissection and organ involvement/ischemia, helps differentiate between Type A/Type B dissections, and allows for standardized, reliable, and reproducible follow-up. In terms of diagnostic accuracy, CT is comparable to MRI/MRA and TEE. Computed tomographic diagnosis of AD is based on the demonstration of an intimal flap that separates the true from the false aortic lumen. Other CT findings include internal displacement of intimal calcifications, delayed enhancement of the false lumen, and aortic widening. Computed tomography can also help identify atypical forms of AD, including those involving intramural hematoma, penetrating atherosclerotic ulcer, and various configurations of the intimal flap. Disadvantages and limitations of CT include: (a) the use if ionizing radiation; (b) the use of iodinated contrast agent; (c) limited visualization of the coronary vessels and the aortic valve (multislice CT overcomes this limitation); and (d) lack of information on aortic regurgitation. With the advent of multi-detector technology, CT scanning is quickly replacing angiography as the diagnostic test of choice. High-quality 2D and 3D reconstructions are possible with spiral CT scanning, which provides additional information regarding the type and location of the dissection, extent of the process (i.e., evaluation of the true and false lumen), and may help with operative/procedural planning.

MAGNETIC RESONANCE IMAGING
The overall accuracy of MRI for suspected AD is similar to that of CT and TEE. Magnetic resonance imaging has >90% sensitivity and >95% specificity for AD. Besides involving no ionizing radiation, MRI provides excellent anatomic visualization of the aortic tear, side branch involvement, aortic regurgitation, and dissection-related complications. In fact, MRI is the preferred modality for patients with renal failure and those with an allergy to iodine. Contrast-enhanced 3D magnetic resonance angiography (MRA) is the preferred technique for imaging chronic dissections and postoperative follow-up. It is very accurate and can evaluate the aortic valve much better than CT angiography. Major limitations of MRI include its high cost, long examination time, limited availability, and difficulty of monitoring hemodynamically unstable patients (i.e., limited physical access to the patient) during the examination. MRI is also contraindicated in patients with pacemakers, some heart valve prostheses, and other magnetically active implants/indwelling objects. However, most patients with prosthetic heart valves or coronary stents can safely undergo an MRI.

TRANSESOPHAGEAL ECHOCARDIOGRAPHY
Transesophageal echocardiography is comparable in diagnostic accuracy to CT. Advantages of TEE include: (a) portability and ability to perform the test at bedside; (b) evaluation for coronary artery involvement; and (c) ability to perform functional cardiac assessment. Limitations of TEE include its invasive nature, lack of visualization of the abdominal aorta (i.e., distal extent of the dissection), limited availability and/or expertise of TEE operators, and inadequate visualization of the proximal aortic arch because of signal interference due to tracheal air. TEE is contraindicated in patients with esophageal varices or stenosis. If TEE findings are negative and clinical suspicion remains high, additional confirmatory diagnostic testing is recommended.

CLINICAL MANAGEMENT OF AORTIC DISSECTION
Treatment of AD should begin as soon as the diagnosis is suspected. Management includes placement of two large-bore peripheral intravenous lines and/or a large-bore central venous introducer catheter, respiratory/hemodynamic monitoring, supplemental oxygen administration, and urine output assessment. Patients must be re-evaluated frequently for hemodynamic compromise, mental status changes, neurologic or peripheral vascular changes, and development or progression of carotid, brachial, and femoral bruits. Adequate pain control ensures patient comfort, promotes pulmonary toilet, and prevents exacerbations of tachycardia and hypertension. Intravenous opioid analgesia should be administered early. Pain control also reduces the force of cardiac contraction and the rate of rise of the aortic pressure (∆P/∆t). It also slows down the propagation of the dissection and may delay rupture.

Aggressive management of heart rate and blood pressure is crucial. Beta-adrenergic antagonists should be administered initially to reduce the rate of change of blood pressure (∆P/∆t) and the shear forces on the aortic wall. Target heart rate should be maintained around 60-80 beats/minute, with target systolic blood pressure around 100-120 mmHg or to the lowest level
commensurate with adequate end-organ perfusion. The best therapeutic approach includes administration of an intravenous beta-adrenergic blocker in incremental doses until a heart rate of 60-80 beats/minute is reached. Esmolol, an ultra-short-acting (elimination half-life of ~9 minutes) beta-2-adrenergic antagonist is particularly useful in patients with labile arterial pressure, especially if surgery is planned, because it can be discontinued abruptly if necessary. It may also be useful when testing beta-adrenergic antagonist safety and tolerance in patients with obstructive pulmonary disease who may be at risk of bronchospasm.

When beta-adrenergic antagonists are contraindicated (i.e., presence of second- or third-degree atrioventricular block), one should consider using calcium channel antagonists. Another group of anti-hypertensive agents used in AD are nitroglycerine derivatives. Nitroprusside is a peripheral vasodilator that acts directly on the venous and arteriolar smooth muscle, thus reducing peripheral vascular resistance. It has rapid onset and short duration of action, and is easily titratable to achieve desired therapeutic effect. Nitroprusside is sensitive to light and both the bottle and the tubing should be wrapped in aluminum foil. Prior to initiating nitroprusside, one should administer beta-blocker to counteract physiologic response of reflex tachycardia that occurs when nitroprusside is used alone. Thus, nitroprusside may increase shear forces acting on the aortic wall when administered alone or prior to initiating beta-adrenergic blockade.

At all times during antihypertensive management for AD, end-organ perfusion should be closely monitored. Balancing the risks of mechanical stress on the aortic wall versus the benefits of acceptable end organ perfusion may pose a difficult clinical problem.

**Stanford Type A dissections** are considered surgical emergencies and their prognosis is poor when treated non-operatively. In fact, up to 40% of patients die within 24 hours and the mortality rate of patients with aortic dissection is 1-3% per hour during the first 24-48 hours. Approximately 80% will die within two weeks if treated nonoperatively. There is high risk of acute aortic regurgitation, coronary artery occlusion, and pericardial rupture. Evolving stroke or established renal failure may constitute contraindications to surgery. Surgery is performed via median sternotomy, utilizing cardiopulmonary bypass. The area of dissection is excised and the aorta is replaced with a graft. Aortic valve is preserved whenever possible. However, when thoracic aortic dissections are associated with aortic valvular disease, the defective valve should be replaced. Operative mortality rate is usually <10% and serious complications are rare with ascending AD. Combined reconstruction and valve replacement carries operative mortality rate of ~5% and a late mortality rate of <10%. Using hypothermic circulatory arrest with retrograde cerebral perfusion, surgical morbidity and mortality rates have decreased substantially. Dissections involving the arch are more complicated that those involving only the ascending aorta because of the presence of innominate, carotid, and subclavian branch vessels. These more complex lesions carry an operative mortality rate of 10% despite the use of hypothermic circulatory arrest. With circulatory arrest rates times of <45 minutes, incidence of central nervous system complications is low (<10%).

Management of type B dissections is not defined as clearly as the management of type A dissections. Type B dissections may be treated non-surgically, which involves strict blood pressure control. Long-term medical therapy involves beta-adrenergic blockade combined with other antihypertensive medications. Antihypertensives that produce a hyperdynamic response (i.e., hydralazine, minoxidil) are associated with increased ΔP/Δt and should be avoided, especially without concurrent beta-adrenergic blockade. Distal dissections treated medically have mortality rates that are comparable to distal dissections treated with surgical therapy. Surgery should be considered if there is evidence of aortic expansion, leaking, rupture, or compromise in blood flow to vital organ(s). Inability to medically control hypertension is also an indication for surgery in patients with a distal thoracic AD. Surgery for Type B dissections is associated with risk of paraplegia (due to interruption of segmental blood supply to the spinal cord) and operative mortality of ~5%. Aggressive use of distal perfusion, cerebrospinal fluid drainage, and hypothermia with circulatory arrest improves early mortality and long-term survival rates. Following surgical therapy, patients should continue to receive beta-adrenergic antagonists.

Aortic stent grafting is a new, promising, but at the same time challenging technique. It has produced good results according to a limited number of clinical series and case reports. Aortic stent grafting may be a reasonable alternative in high-risk patients. In fact, some recommend that patients with complicated acute type B dissections undergo endovascular stenting as the primary surgical option, rather than open operative repair.

**CONCLUSION**

Aortic dissection is the most common aortic emergency. Early recognition and treatment are critical in successful approach to AD. Aortic dissection is missed on initial diagnosis in ~30% of cases. Multi-detector computed tomography is the initial imaging study of choice. Early management involves aggressive blood pressure and heart rate control. Beta-adrenergic blockade must be instituted early in order to decrease the stress on the aortic wall (ΔP/Δt). Stanford type A dissections are surgical emergencies and carry 1-3% mortality per hour during the first 24-48 hours. Stanford type B dissections can be managed non-surgically, with beta-adrenergic blockade for strict blood pressure and heart rate control.

**SELECTED REFERENCES**


