Acute Aortic Syndromes: Pathophysiology and Management

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The acute aortic syndromes carry significant morbidity and mortality, especially when detected late. Symptoms may mimic myocardial ischemia, and physical findings may be absent or, if present, can be suggestive of a diverse range of other conditions. Maintaining a high clinical index of suspicion is crucial in establishing the diagnosis. All patients with suspected aortic disease and evidence of acute ischemia on electrocardiogram should undergo diagnostic imaging studies before thrombolytics are administered. The demonstration of an intimal flap separating 2 lumina is the basis for diagnosis. Tear detection and localization are very important because any therapeutic intervention aims to occlude the entry tear. The goals of medical therapy are to reduce the force of left ventricular contractions, decrease the steepness of the rise of the aortic pulse wave, and reduce the systemic arterial pressure to as low a level as possible without compromising perfusion of vital organs. Surgical therapy still remains the gold standard of care for type A aortic dissection, whereas in type B dissection, percutaneous aortic stenting and fenestration techniques have been developed and are sometimes used in conjunction with medical therapy in certain situations.
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The acute aortic syndromes formerly known as *aortic dissection* encompass several entities that include classic aortic dissection, intramural hematoma/hemorrhage (IMH), subtle-discrete aortic dissection, plaque rupture/ulceration, and iatrogenic/traumatic dissection. Acute aortic syndromes are associated with acute chest pain with a high rate of complications and mortality. Many patients with acute aortic syndrome die before presentation to a hospital and/or diagnosis. The symptoms of acute aortic syndrome may mimic

myocardial ischemia, and physical findings in acute aortic syndrome may be absent or, if present, can be suggestive of a diverse range of other conditions. Therefore, keeping a high clinical index of suspicion is crucial in establishing the diagnosis of aortic dissection.

The term *aneurysme dissequant* (ie, dissecting aneurysm) was first coined by Laennec in 1819 and had been used to refer to the condition before it was replaced by the term *acute aortic syndromes*. Although the aorta is frequently enlarged, it is rarely aneurysmal, and it is in fact a hematoma that may dissect or rupture.

Etiology

Aortic dissection is usually caused by weakening of the intima layer of the aorta that leads to aortic dilatation and aneurysm formation, eventually resulting in intramural hemorrhage, aortic dissection, or rupture. The etiology can be broadly classified into 2 groups, congenital and acquired conditions.

Congenital Causes

Marfan syndrome is the most common connective tissue disorder associated with the diagnosis of aortic dissection. It is the most common cause of aortic dissection in patients younger than 40. It has an estimated incidence of 1 in 5000, with autosomal dominant inheritance and variable penetrance.[1,2] Ehlers-Danlos syndrome is a group of connective tissue disorders characterized by hypermobility, skin hyperextensibility, and tissue fragility. Aortic dissection is usually seen in autosomal dominant type IV Ehlers-Danlos syndrome. Congenitally bicuspid and unicommissural aortic valves, aortic coarctation, annuloaortic ectasia, other chromosomal abnormalities (Turner syndrome and Noonan syndrome),[3,4] and aortic arch hypoplasia are also associated with aortic dissection.

Acquired Causes

Hypertension is the most important predisposing factor for aortic dissection. It coexists in 70% to 90% of patients in most series[5-7] and is more common in distal dissections than in proximal ones. It affects the composition of the arterial wall, causing intimal thickening, fibrosis, calcification, and extracellular fatty acid deposition.

Direct iatrogenic trauma that occurs during arterial cannulation for cardiac surgery or during catheter-based diagnostic and therapeutic interventions accounts for about 5% of cases of aortic dissection. The majority of iatrogenic dissections have been reported in the descending thoracic and abdominal aorta.[8-11] There is a relationship between the severity of atherosclerosis and the risk of developing an iatrogenic dissection. Indirect trauma, such as sudden deceleration, may also result in transection of the aorta, typically at the ligamentous arteriosus.

Cocaine use has also been recognized as a cause of aortic dissection. The mechanism of dissection involves a profound elevation of the blood pressure induced by acute catecholamine release, causing a rapid rise in pressure in the aortic wall resulting in an intimal tear. Rapid discontinuation of β-blockers has also been reported to cause aortic dissection via similar mechanisms.

Inflammatory diseases weaken and destroy the medial layers of the aorta, which results in expansion and dissection of the aortic wall. Dissection has been associated with giant-cell aortitis, systemic lupus, and relapsing polychondritis.[12,13]

Dissection occurs about 3 times more often in men than in women.[14] In women, dissection is associated with pregnancy.[15] Half of the dissections in women younger than 40 occur during pregnancy, usually in the third trimester[16,17] or the first stage of labor. The most common site is the proximal aorta, and intimal tearing occurs within 2 cm of the aortic valve in 75% of cases. Recently, in men and women, dissection has been linked to extreme physical exertion, as seen in weightlifters, and profound emotional distress.

Classification

New studies have suggested that intramural hemorrhage, intramural hematoma, and aortic ulcers may be signs of evolving dissections or dissection subtypes. Thus, a new classification has been proposed:

- Class 1: Classic aortic dissection with an intimal flap between the true and false lumens.
- Class 2: Medial disruption with formation of IMH.
- Class 3: Discrete/subtle dissection without hematoma, eccentric bulge at tear site.
- Class 4: Plaque rupture leading to aortic ulceration, penetrating atherosclerotic ulcer with surrounding hematoma, usually subadventitial.
- Class 5: Iatrogenic and traumatic dissection.

Classic Aortic Dissection

Acute aortic dissection is defined by the accelerated formation of an intimal flap separating the true and false lumens. Due to a pressure difference, the true lumen is usually smaller than the false lumen. Intimal tears characterize communicating dissections. The dissection can spread either antegradely or retrogradely and involve side branches and cause other complications.

In classic aortic dissection, 3 major classification schemes are used to

define the location and extent of aortic involvement. All 3 schemes share the same basic principle of distinguishing aortic dissections with and without ascending aortic involvement for prognostic and therapeutic reasons.

In the Stanford classification, type A dissection involves the ascending aorta, regardless of the site of origin, and type B dissection includes all dissections not involving the ascending aorta.[18] The DeBakey classification consists of 3 types. Type I dissection originates in the ascending aorta and often involves the entire aorta. Type II originates in the ascending aorta but is confined to the arch. Type III originates in the descending aorta and extends distally down the aorta or, rarely, retrogradely into the aortic arch and ascending aorta (Figure 1).[19]

Anatomically, aortic dissection can also be classified into 2 types: proximal and distal. Proximal dissections include DeBakey types I and II and Stanford type A. Distal dissections include DeBakey type III and Stanford type B.

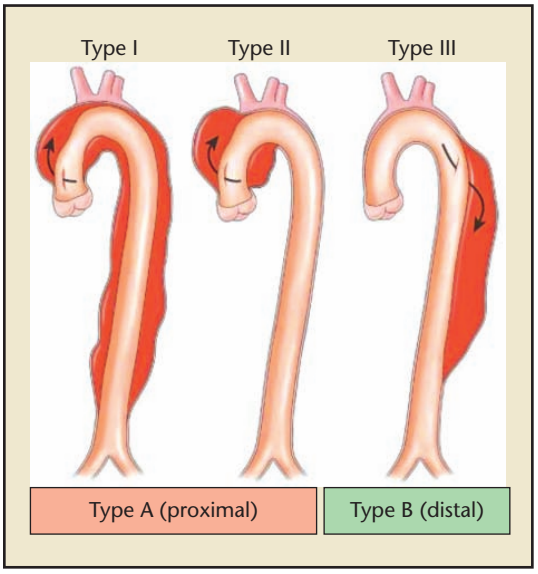

Figure 1. *Classification of aortic dissection according to the DeBakey and Stanford criteria. Reprinted with permission from Braunwald E et al.*[101]
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Intramural Hematoma/Hemorrhage

IMH is a localized hematoma within the aortic wall and is considered the precursor lesion in overt aortic dissection. It is probably the initial lesion in the majority of cases of cystic medial degeneration. This condition leads to aortic dissection in which the intimal tear seems to be secondary to preceding intramural dissection.[20-29] It seems to originate from rupture of vasa vasorum that appears either normal or is diseased. It may extend along the aorta and progress, regress, or reabsorb.

The prevalence of IMH in acute aortic syndromes is between 10% and 30%.[27,29-31] It can be classified into 2 types. Type I is characterized by a smooth inner aortic lumen, diameter of less than 3.5 cm, and wall thickness exceeding 0.5 cm. Type II is seen in aortic atherosclerosis and is characterized by a rough inner surface with severe aortic sclerosis, aortic dilatation greater than 3.5 cm (Figure 2), frequent calcium deposits, and a mean wall thickness of 1.3 cm. Predictors of disease progression include involvement of the ascending aorta, maximum aortic diameter of 56 mm or more, persistent pain, progressive maximal aortic wall thickness, increase in associated pleural effusion, and a large penetrating atherosclerotic ulcer on top of the IMH. Other factors for disease progression include age younger than 55 years, no β-blocker treatment, progressive aortic diameter, symptomatic penetrating atherosclerotic ulcer, and progressive aortic wall thickness.

IMHs are found more often in the descending aorta than the ascending aorta. Acute aortic dissection as a consequence of intramural hematoma (Figure 3) develops in 28% to 47% of patients and is associated with aortic rupture in 21% to 47% of patients; regression is seen in 10% of patients.[27,29-31]

Subtle-Discrete Aortic Dissection

This type has been described as a partial stellate or linear tear of the vessel wall that is covered by a thrombus. When the partial tear forms a scar, this constellation is called an *abortive discrete dissection*. The partial tear may rupture the inner layer of the aorta, allowing blood to enter the already damaged media, leading to dissection along the aortic wall. A second lumen is thus formed within the wall; it may rupture or heal during follow-up.

Plaque Rupture/Ulceration

Ulceration of atherosclerotic plaques can lead to aortic dissection or perforation.[32-37] The emergence of imaging modalities such as intravascular ultrasound, transesophageal echocardiography (TEE), magnetic resonance imaging (MRI), and spiral computed tomography (CT) has provided new insights into the pathophysiology of the disease. Ulcers commonly affect the descending thoracic as well as the abdominal aorta, with extensive longitudinal

propagation. The ulcer may penetrate beyond the intimal border (Figure 4), often with a nipple-like projection with subadjacent type II intramural hematoma formation. Valvular, perivalvular, and other vascular complications are rare. The internal elastic membrane may eventually be breached by the progressive erosion of the atherosclerotic plaque.[33] False aneurysms, aortic rupture, or dissection may occur.[36,37]

Traumatic/Iatrogenic Aortic Dissection
Blunt chest trauma usually causes transection of the ascending aorta and/or of the region of the ligamentum arteriosus at the aortic isthmus. Iatrogenic dissection of the aorta is rarely seen during cardiac catheterization but regularly seen following angioplasty of aortic coarctation in adults. It can also be seen after cross-clamping of the aorta and after intra-aortic balloon pumping.[38-41] Most catheter-induced dissections are retrograde and usually self-limited.

Epidemiology, Natural History, and Prognosis

Aortic dissection has an annual prevalence of 0.5 to 2.95 per 100,000.[42-45] Annual mortality is between 3.25 to 3.6 per 100,000.[46] Due to the high mortality of aortic dissection in the acute stage, the survival rate in both type A and B dissection is very low. Forty years ago, the 24-hour mortality was 21%. After 30 days, 8% of patients were alive, and only 2% were alive after 1 year.[22] Ten years later, the 48-hour mortality was still reported to be 50%, or approximately 1% per hour. Up to 20% of patients died before reaching the hospital.

Even recently, a mortality of 68% within 48 hours and 1.4% per hour was reported in a survey spanning 27 years.[47] In this study, the annual incidence of dissection was 2.95 per 100,000, and the important observation was that the diagnosis was established in only 15% before autopsy. The most common cause of death was aortic rupture, which occurred in 80% of patients.[47] The natural history of the disease is best characterized by differentiating between patients with proximal or distal aortic dissections.

Type A (Proximal) Dissection
Acute aortic dissection of the ascending aorta is a highly lethal condition with a mortality rate of 1% to 2% per hour early after symptom onset.[22,48] It is characterized by sudden onset of severe chest (85%) and/or back pain (46%); other common symptoms include abdominal pain (22%), syncope (13%), and stroke (6%).[22,49] Rupture into the pericardium leading to a pericardial effusion, involvement of the coronary arteries causing acute myocardial ischemia/infarction, and dissection compromising brain perfusion carry a particularly high risk. Additionally, aortic valve disruption leading to acute congestive heart failure, extensive aortic involvement as manifested by multiple pulse deficits and/or renal failure, and advanced age also correlate with increased risk.

Acute type A dissection is a surgical emergency. Medical management alone is associated with a mortality rate of nearly 20% by 24 hours after presentation, 30% by 48 hours, 40% by day 7, and 50% by 1 month. Even with surgical repair, mortality rates are 10% by 24 hours, 13% by 7 days, and nearly 20% by 30 days.[48] Aortic rupture, stroke, visceral ischemia, cardiac tamponade, and circulatory failure are the most common causes of death.[50-53]

Type B (Distal) Dissection
Type B aortic dissection affecting the descending aorta is less lethal than type A dissection, but it has a similar presentation. Sudden onset of severe pain in the back (64%) and/or chest (63%) are frequently reported symptoms. Stroke is less common (21%), and patients sometimes present with an ischemic leg or peripheral ischemic neuropathy.[48,53,54]

The 30-day mortality from uncomplicated type B aortic dissection is 10%. Conversely, in the setting of the development of ischemic leg, renal failure, or visceral ischemia of contained rupture often requiring urgent repair, the mortality rates are 20% by day 2 and 25% by day 30.[49,51,53] Independent predictors of early mortality are advanced age, rupture, shock, and malperfusion.

Similar to myocardial infarction, acute aortic dissection exhibits significant circadian and seasonal variations, with onset of dissection preferentially in the morning between 6 and 10 o'clock and in the early afternoon.[55] Moreover, the risk of dissection is higher in the cold weather than in the summer.[55]

A dramatic improvement due to medical and surgical therapy has been seen over the last 3 decades. The European Cooperative study group reported a 1-year survival rate of 52%, with 69% in type A (type I and II) and 70% in type B (type III) dissection. This survival rate decreased to 48%, 50%, and 60%, respectively, after 2 years. Similar results have also been reported that show a better prognosis with type B dissection.

Among 464 patients in the International Registry of Acute Aortic Dissection (IRAD), mortality after surgical therapy was 27% for type A dissection and 29% for type B dissection; mortality after medical therapy was 53% for type A dissection and 9% for type B dissection.[48] Spontaneous healing of aortic dissection can occur during medical therapy, but it is rare. The false lumen

disappears and circumscript wall thickening develops. Another form of healing, complete thrombosis of the false lumen, is sometimes seen and appears to be a prerequisite for complete healing.

Clinical Features and Diagnosis

Approximately 90% of patients with an acute aortic syndrome presenting to the emergency department will complain of pain of abrupt onset that was most intense at onset. With further extension of the dissection process, the pain may change its location accordingly. The pain is described as sharp more often than tearing, ripping, or stabbing.[14,48,56] In proximal dissections, the pain is usually located retrosternally, whereas distal dissections are characterized by interscapular and back pain. The absence of chest pain is rare and usually indicative of chronic aortic dissection. Up to 20% of patients with acute aortic dissection may present with syncope and without a history of typical pain or neurological findings.[14,28,56] Following a period of pain, cardiac failure may become the main symptom and is usually related to severe aortic regurgitation. Cardiac tamponade may result in hypotension and syncope. Syncope may also result from severe pain, obstruction of cerebral vessels, or activation of aortic baroreceptors. Paraplegia may suddenly develop as intercostal arteries are separated from the aortic lumen by the dissection.

Further propagation of the dissection at a later point in time will usually result in another attack of the same acute pain as occurred with the initial event. It is often associated with a deteriorating clinical picture.[48] High fever, although not common, can occur due to release of pyrogenic substances. Persistent abdominal pain, elevation of acute phase reactants, and increased lactate dehydrogenase usually signify involvement of the celiac artery. This involvement is seen in about 8% of patients, and mesenteric artery involvement is seen in 8% to 13%. Anuria or oliguria usually signifies involvement of the renal arteries.[57,58] Clinical features and symptoms of patients presenting with acute aortic dissection are summarized in Table 1.

Pulse deficits are seen in only 20% of cases, as documented in the IRAD registry of patients.[48] They often signify progression to complications and bad outcome. They may be transient due to the intimal flap's changing position. Neurological deficits (loss of consciousness, ischemic paresis) occur in 40% of patients with proximal dissection.[56,58]

Table 1
Clinical Features of Acute Aortic Dissection

Anterior chest pain—ascending aortic dissection
Neck or jaw pain—aortic arch dissection
Interscapular tearing or ripping pain—descending aortic dissection
Myocardial infarction
Syncope
Stroke symptoms
Altered mental status
Limb paresthesias, pain, or weakness
Hemiparesis or hemiplegia
Horner syndrome
Dyspnea
Dysphagia
Orthopnea
Flank pain
Anxiety and premonitions of death
Dyspnea and hemoptysis if dissection ruptures into the pleura
Anuria or oliguria
Persistent abdominal pain, elevated lactate dehydrogenase

Rare comorbidities include vocal cord paralysis, hematemesis, hemoptysis, superior vena cava syndrome, Horner's syndrome, and Leriche's syndrome. A diastolic murmur of aortic regurgitation is heard in about 50% of patients with proximal aortic dissection.[14,48,59] Signs of pericardial involvement, such as pericardial friction rub, jugular venous distention, and pulsus paradoxus, are indications for rapid surgical intervention.

Pleural effusions occur secondarily to rupture of the aorta into the pleural space and are usually left-sided effusions. Sometimes the effusions are secondary to the expression of an exudative inflammatory reaction. Pleural effusions should be drained, and the hematocrit, hemoglobin, and red cell count of the fluid measured to determine if it is frank blood.

Up to 30% of patients later found to have acute aortic syndrome are initially suspected of having other conditions such as acute coronary syndromes, nondissecting aneurysms, pulmonary embolism, and aortic stenosis.[56,59] Table 2 outlines the prominent symptoms and physical findings in 464 patients as described by Eagle.[60]

Initial Diagnostic Steps in the Emergency Room

An electrocardiogram must be obtained in all patients suspected of having an acute aortic syndrome because it can help distinguish acute myocardial infarction, for which thrombolytic therapy or percutaneous coronary intervention with antiplatelet agents may be life-saving, from acute aortic syndromes, in which thrombolytic therapy may be detrimental. However, one-third of patients with coronary involvement in acute aortic syndromes will have a normal electrocardiogram (EKG), and most of these patients have nonspecific ST-T changes.[61] About

Table 2
Prominent Symptoms and Physical Findings in 464 Patients in the IRAD

Category	Total Number Reported (%)	Type A (%)	Type B (%)
Symptoms			
Any pain reported	443/464 (95.5)	271 (93.8)	172 (98.3)
Abrupt onset of pain	379/447 (84.8)	234 (85.4)	145 (83.8)
Anterior chest pain	262/430 (60.9)	191 (71.0)	71 (44.1)
Posterior chest pain	149/415 (35.9)	85 (32.8)	64 (41)
Back pain	240/451 (53.2)	129 (46.6)	111 (63.8)
Abdominal pain	133/449 (29.6)	60 (21.6)	73 (42.7)
Severe/worst ever pain	346/382 (90.6)	211 (90.1)	135 (90)
Sharp pain	174/270 (64.4)	103 (62)	71 (68.3)
Tearing/ripping pain	135/267 (50.6)	78 (49.4)	57 (52.3)
Radiating	127/449 (28.3)	75 (27.2)	52 (30.1)
Migrating	74/446 (16.6)	41 (14.9)	33 (19.3)
Syncope	42/447 (9.4)	35 (12.7)	7 (4.1)
Physical Examination			
Hypertensive SBP > 150 mm Hg	221 (49.0)	99 (35.7)	122 (70.1)
Normotensive SBP 100-149 mm Hg	156 (34.6)	110 (39.7)	46 (26.4)
Hypotensive SBP < 100 mm Hg	36 (8.0)	32 (11.6)	4 (2.3)
Shock or tamponade SBP < 80 mm Hg	38 (8.4)	36 (13.0)	2 (1.5)
Auscultated murmur of aortic insufficiency	137/434 (31.6)	117 (44)	20 (12)
Pulse deficit	69/457 (15.1)	53 (18.7)	16 (9.2)
Cerebrovascular accident	21/447 (4.7)	17 (6.1)	4 (2.3)
Congestive heart failure	29/440 (6.6)	24 (8.8)	5 (3.0)

IRAD, International Registry of Acute Aortic Dissection; SBP, systolic blood pressure. Data from Eagle KA.[60]

20% of patients with type A dissection have EKG evidence of acute ischemia or acute myocardial infarction.[61] All patients with suspected aortic disease and evidence of acute ischemia on EKG should undergo diagnostic imaging studies before thrombolytics are administered.[61] The role of the chest x-ray is currently unclear; a routine chest x-ray will be abnormal in 60% to 90% of patients with suspected aortic dissection.[62] In the unstable patient, invasive blood pressure monitoring should be provided by placing an arterial line to the right radial artery. Large bore IV access must be obtained, with simultaneous type and cross of blood and an emergency cardiothoracic surgery consultation.

Pain control and reduction of systolic blood pressure to values between 100 and 120 mm Hg are essential. The most appropriate drug for pain relief is morphine sulphate, and β-blockers are the drugs of choice for reduction of the force of left ventricular ejection and blood pressure control.[54]

Diagnosis

In the IRAD patients, the initial diagnostic tests were transthoracic echocardiography (TTE) and TEE in 33%, CT in 61%, angiography in 4%, and MRI in 2%.[60] As secondary techniques, TTE/TEE was used in 56%, CT in 18%, angiography in 17%, and MRI in 9%. An average of 1.8 methods were used to diagnose aortic dissection. In situations in which 3 methods were utilized, CT was used in 40%, MRI in 30%, and angiography in 21%.[60] The choice of which technique to use depends on the availability in emergency situations and the experience of the emergency department and imaging staff. With all of these imaging techniques, the demonstration of an intimal flap separating 2 lumina is the basis for diagnosing aortic dissection. If the false lumen is completely thrombosed, then central displacement of the intimal flap, calcification, or separation of intimal layers can be regarded as definitive signs of aortic dissection/intramural hematoma. Erbel and colleagues[63] summarized the recommendations of the task force on aortic dissection regarding diagnostic imaging (Table 4).

Tear Localization and Disease Extent

Tear detection and localization are very important because any therapeutic intervention aims to occlude the entry tear. Frequently, multiple tears are seen, and the pressure difference between the true and false lumen forces the blood from the true lumen to the false lumen and back. Multidirectional flow jets can also be seen.

The full extent of aortic dissection can be analyzed with ultrasonographic techniques alone, if echocardiography is combined with duplex sonography, abdominal sonography, or, even better, intravascular ultrasound. Of all the ultrasonic

Table 3
Chest Radiography and Electrocardiography Results in Patients With Acute Aortic Syndrome Seen in the IRAD

Category	Total Number (%)	Type A (%)	Type B (%)
Radiography findings (N = 427)	427 (100)	256 (88.6)	171 (97.7)
No abnormalities noted	53 (12.4)	26 (11.3)	27 (15.8)
Absence of widened mediastinum or abnormal contour	91 (21.3)	44 (17.2)	47 (27.5)
Widened mediastinum	263 (61.6)	169 (62.6)	94 (56)
Abnormal aortic contour	212 (49.6)	124 (46.6)	88 (53)
Abnormal cardiac contour	110 (25.8)	69 (26.9)	41 (24.0)
Displacement/calcification of aorta	60 (14.1)	29 (11.3)	31 (18.1)
Pleural effusion	82 (19.2)	46 (17.3)	36 (21.8)
Electrocardiogram findings (N = 444)			
No abnormalities noted	139 (31.3)	85 (30.8)	54 (32.1)
Nonspecific ST-segment or T-wave changes	184 (41.4)	116 (42.6)	68 (42.8)
Left ventricular hypertrophy	116 (26.1)	67 (25)	49 (32)
Ischemia	67 (15.1)	47 (17.3)	20 (13.2)
Myocardial infarction, old Q waves	34 (7.7)	19 (7.1)	15 (9.9)
Myocardial infarction, new Q waves or ST-segment elevation	14 (3.2)	13 (4.8)	1 (0.7)

IRAD, International Registry of Acute Aortic Dissection. Data from Eagle KA.[60]

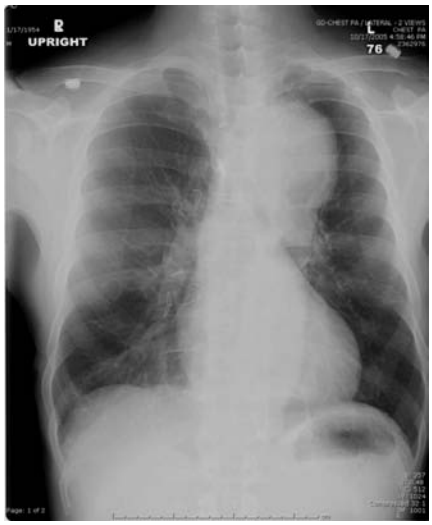

Figure 2. *Chest radiograph revealing marked dilatation of the arch of the aorta in a patient with aortic dissection.* www.medreviews.com

techniques, intravascular ultrasound (IVUS) has gained the highest accuracy[25,26,64]; it eliminates the blind spots in the ascending aorta and in the abdomen that previously limited examinations.

It is important to differentiate between the true lumen and the false lumen. The true lumen is usually compressed by the false lumen in diastole, and it demonstrates systolic expansion and systolic forward flow. The true lumen is usually close to the inner curvature of the aortic arch. Spontaneous echocardiographic contrast is a sign of slow flow, and thrombus formation is rare in the true lumen. The false lumen exhibits systolic compression. It is located next to the outer portion of the aorta involving the take-off of the great vessels, and signs of slow flow are regularly described. Thrombus may be present.[65-67] When the flap extends distally, it commonly involves the left kidney and the left femoral artery.

Imaging Modalities

Direct and indirect methods, as well as invasive and noninvasive techniques, can be employed to diagnose aortic dissection. Options include transthoracic/transesophageal echocardiography, computed tomography, MRI, aortography, and IVUS.

Transthoracic/Transesophageal Echocardiography

The aorta and the major vessels/side branches can be studied using different scanning fields. The diagnosis of aortic dissection is confirmed when 2 lumina separated by an intimal flap can be visualized within the aorta (Figure 5). Positive criteria are complete obstruction of a false lumen, central displacement of intimal calcification, separation of intimal layers from the thrombus, and shearing of different wall layers during aortic pulsation.[65] Smaller tears can be detected by color Doppler registering jets transversing the dissection membrane.[66] The true lumen is identified on echocardiography by systolic expansion and diastolic collapse, the absence or low intensity of spontaneous echocontrast, systolic jets directed away from the lumen, and systolic forward flow.[65,68]

A thrombus is diagnosed when a mass separate from the intimal flap and the aortic wall is imaged in the free space of the false lumen or, rarely, the true lumen.[65] Pericardial effusion is considered to exist when an echo-free space between the pericardium and the epicardium is present. Transthoracic echocardiography can detect involvement of the ascending aorta with a sensitivity of 77% to 80% and a specificity of 93% to 96%.[69-71] The European Cooperative Study Group showed that the sensitivity of single-plane TEE (occasionally

Table 4
Diagnostic Imaging in Aortic Dissection: Diagnosis and Management of Acute Aortic Syndromes*

Recommendation	Class I	Class IIa	Class IIb	Class III	Level of evidence
TTE followed by TEE	X				C
Computed tomography	X				C
If detection of tears is crucial			X		C
Contrast angiography					
To define anatomy in visceral malperfusions and to guide percutaneous interventions	X				C
In stable patients		X			C
In routine preoperative coronary angiography				X	C
In hemodynamically unstable patients			X		C
Magnetic resonance imaging		X			C
In hemodynamically unstable patients				X	C
Intravascular ultrasound		X			C
To guide percutaneous interventions			X		C

*Recommendations of the European Task Force on Aortic Dissection.
TTE, transthoracic echocardiography; TEE transesophageal echocardiography.
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biplane, but not multiplane) reaches 99% and specificity reaches 89%, with a positive predictive accuracy of 89% and a negative predictive accuracy of 99%. Multiplane TEE can provide accurate diagnosis of aortic dissection with sensitivity of 99% and specificity of 98%.[67]

Computed Tomography

The use of helical CT has dramatically improved CT imaging because it minimizes motion artifacts and eliminates respiratory misregistration. The diagnosis is based on the demonstration of an intimal flap that separates the true lumen from the false lumen (Figure 6). The flap is identified as a low attenuation linear structure in the aortic lumen.[72,73] Secondary findings include internal displacement of the intima, linear calcifications, delayed enhancement of the false lumen, and aortic widening. Reported limitations of CT are attributable to artifacts, usually venous streaks and aortic motion artifacts. The sensitivity and specificity of CT are 83% to 94% and 87% to 100%, respectively.[67,72] With the use of helical CT, sensitivity is 93% and specificity is 98%, with an overall accuracy of 96%.[72-74] CT is the most commonly used imaging modality in cases of suspected aortic dissection.[60]

Magnetic Resonance Imaging

Although MRI is both highly sensitive and specific in diagnosing aortic dissection,[72,74,75] it is not often available on an emergency basis. MRI clearly demonstrates the extent of the disease and depicts the distal ascending aorta and the aortic arch in more detail than even TEE.[76] The localization of entry and reentry is nearly as accurate as with TEE, and the sensitivity approaches 90%.[76] Flow in the false lumen and the true

Table 5
Comparing the Diagnostic Value of Imaging Modalities in Aortic Dissection

	TTE/TEE	CT	MRI	Angiography	IVUS
Sensitivity	++	++	+++	++	+++
Specificity	+++	++	+++	++	+++
Classification	+++	++	++	+	+++
Tear localization	+++	–	++	+	++
Aortic regurgitation	+++	–	++	++	–
Pericardial effusion	+++	++	++	–	–
Mediastinal hematoma	++	+++	+++	–	+
Side branch involvement	+	++	++	+++	+++
Coronary artery involvement	++	–	+	+++	++
X-ray exposure	–	++	–	+++	–
Patient comfort	+	++	+	+	+
Follow-up studies	++	++	+++	–	–

TTE, transthoracic echocardiography; TEE, transesophageal echocardiography; CT, computed tomographic scan; MRI, magnetic resonance imaging; IVUS, intravascular ultrasound.
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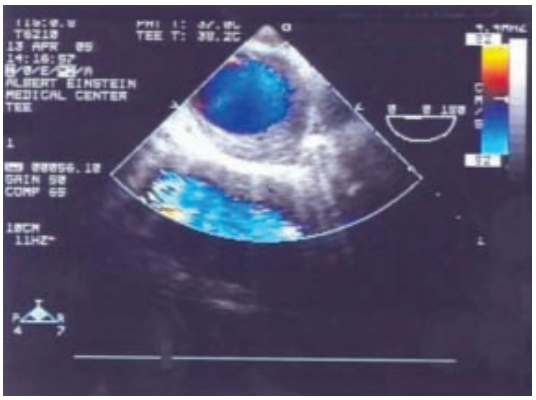

Figure 3. *Transesophageal echocardiogram showing an intramural hematoma in a patient with aortic dissection.* www.medreviews.com

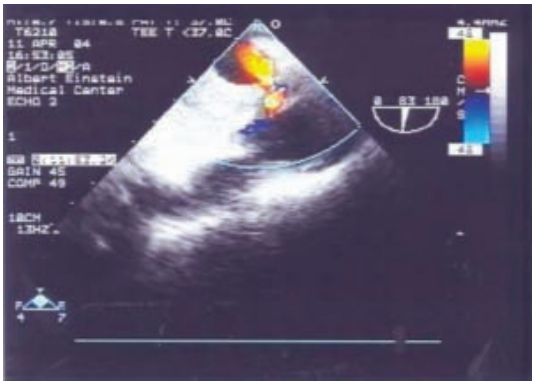

Figure 4. *Transesophageal echocardiogram showing a penetrating atherosclerotic ulcer in a patient with aortic dissection.* www.medreviews.com

lumen can be quantified using phase contrast cine MRI or by tagging techniques.[77,78] MRI permits detection of acute from subacute aortic intramural hemorrhages.

Aortography

Retrograde aortography was the first accurate diagnostic test for aortic dissection, since reports of the condition began to appear in 1939.[79,80] It has remained the diagnostic gold standard.[81] The angiographic diagnosis of aortic dissection is based upon direct angiographic signs, such as the visualization of the intimal flap or the recognition of 2 separate lumens, or indirect signs, including aortic lumen irregularities, rigidity or compression, branch vessel abnormalities, thickening of the aortic walls, and aortic regurgitation.[71] Aortography is able to localize the site of origin of the dissection.[82] The true lumen is typically compressed and tends to adopt a spiral shape. Contrast angiography accurately identifies branch vessel involvement; in particular, angiography is an excellent technique to define renal or mesenteric compromise.

The specificity of aortography for diagnosing aortic dissection is better than 95%, but its sensitivity is lower than that of other techniques.[83] In the European Cooperative Study, the sensitivity and specificity of aortography for the diagnosis of acute aortic syndrome was 88%.[67] Aortic intramural hematoma (class 2 dissection) has been considered the principal reason for false-negative findings on aortography.[84] In patients with suspected aortic dissection based on the history, aortography should be performed when noninvasive techniques are negative. Limitations of the technique include the invasiveness of the procedure, the toxicity of the radiopaque dye, and the time interval required to set up the procedure.

Intravascular Ultrasound

The use of IVUS has been advocated to complement angiography; it can overcome the potential limitations and pitfalls of angiography.[25,26] It directly visualizes the vessel wall architecture from inside the aortic lumen. It therefore allows accurate recognition of aortic wall characteristics and pathology. Sensitivities and specificities of close to 100% have been reported.[64] The shape of the true and false lumen is readily displayed, whereas false lumen thrombosis is detected with a higher sensitivity and specificity than with TEE.[64] Table 5 illustrates the diagnostic

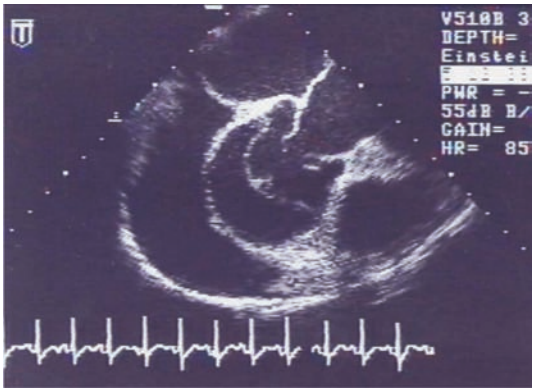

Figure 5. *Transesophageal echocardiogram revealing a proximal aortic dissection with an intimal flap separating the true and false lumen.* www.medreviews.com

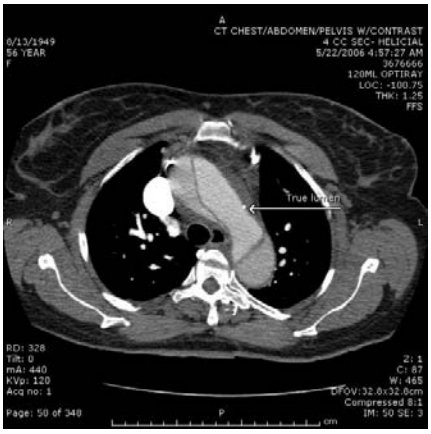

Figure 6. *Contrast-enhanced computed tomographic scan of the chest revealing a proximal aortic dissection involving the aortic arch and showing the true and false lumen.* www.medreviews.com

value of different imaging modalities in acute aortic dissection.

Therapeutic Management

Medical Therapy

Intravenous antihypertensive therapy should be started emergently in all patients, except those who are hypotensive, as soon as the diagnosis is suspected. The aims of medical therapy are to reduce the force of left ventricular contractions, decrease the steepness of the rise of the aortic pulse wave, and reduce the systemic arterial pressure to as low a level as possible without compromising perfusion of vital organs.

Experimental models of dissection revealed that laminar nonpulsatile flow is associated with the cessation of the advancement of dissection, whereas pulsatile flow of increasing acceleration results in the continuation of dissection in both directions from the initial intimal tear. Thus, reducing the rate of rise of the aortic pulse by decreasing the force of the left ventricular contractions should retard propagation of the dissection.

Standard medical therapy involves the use of a β-blocker and a vasodilator such as sodium nitroprusside. The β-blocker should be started before the vasodilator, so as to prevent the reflex catecholamine release secondary to the direct vasodilation that is caused by potent vasodilators, which may increase the force of the left ventricular contraction. Labetalol, an α- and β-adrenergic blocker, is an alternative to the combination of a β-blocker and nitroprusside.

Patients with uncomplicated distal aortic dissection can be initially managed with medical treatment. A survival rate of 75% is seen in the initial phase whether patients are treated medically or surgically. The goals of medical therapy in aortic dissection are to stabilize the dissection, prevent rupture, accelerate healing, and reduce the risk of complications. The main causes of death in patients treated medically are aortic rupture and organ malperfusion.

Surgical and Interventional Therapy

Surgical intervention is indicated in all patients with proximal dissections, with the exception of patients with serious comorbid conditions that preclude surgery. Stroke is often a contraindication to surgery because there is real concern that anticoagulation therapy and reperfusion can result in further neurologic deterioration by converting the ischemic stroke to a hemorrhagic stroke.

The aim of any surgical intervention in type A dissection is the prevention of aortic rupture or the development of pericardial effusion, which may lead to cardiac tamponade and aortic regurgitation. In type B dissection, the main goal is to prevent aortic rupture.

Approach to the Proximal Aorta in Acute Type A Dissection

The size of the aortic root and the condition of the aortic valve determine the most appropriate technique for repairing an acute type A dissection. If the ascending aortic and aortic root diameters are normal—without downstream displacement of the coronary ostia, commissural detachment of the aortic valve leaflets, or acute or chronic pathological changes of the leaflets—a tubular graft is usually anastomosed to the sinotubular ridge. Whenever 1 or more commissures are detached, the valve must be resuspended prior to graft insertion. If valve reconstruction appears unsafe, or if obvious congenital or acquired anomalies are present, it is generally better to replace the valve.

The approach in acute type A dissection in a previously ectatic aorta, as seen in Marfan syndrome, involves implantation of a composite graft (aortic valve plus ascending aortic tube graft) with coronary reimplantation.[85,86] Implantation of allografts and xenografts should be restricted to elderly patients or patients with certain other indications because late postoperative degeneration may require reoperation on the aortic root.

Approach to the Aortic Arch in Acute Type A Dissection

This approach remains a much debated subject. The debate is centered around when and to what extent the arch should be replaced. At present, there is a broad consensus that any dissected arch should be explored during a short period of hypothermic circulatory arrest. In the absence of an arch tear, an open distal anastomosis of the graft and the conjoined aortic wall layers at the junction of the ascending and arch portions is made.

Arch tears occur in 30% of patients with acute type A dissection. If an entry tear traverses the aortic arch, the distal graft to aortic anastomosis is usually made in such a manner as to replace the arch beyond the

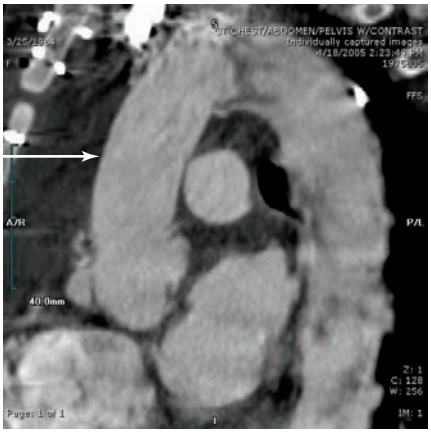

Figure 7. *Magnetic resonance imaging of the thorax showing a repaired proximal aortic dissection with a graft.* www.medreviews.com

entry-bearing portion. In patients with extensive tears that continue beyond the junction of the transverse and descending aortic segments or who had acute dissection of a previously aneurysmatic arch, subtotal or total arch replacement may be required, with reconnection of some or all of the supraaortic vessels to the graft during hypothermic circulatory arrest and antegrade head perfusion. Patients are usually cooled to about 15°C (59°F) for up to 40 minutes; the risk of transient cerebral dysfunction after this period is 15% at 40 minutes, 30% at 50 minutes, and 60% at 60 minutes. Stroke rates of 7% to 11% have been reported following deep hypothermic circulatory arrest. In dissecting and nondissecting aneurysms extending to the downstream aorta, an elephant trunk extension of the graft is an option, as first described by Borst and colleagues.[87,88]

Surgery in Type B Dissection

Indications for surgery in type B dissection include relief of intractable pain; treatment of a rapidly expanding aortic diameter; periaortic, mediastinal, or pleural hematoma; and limb or gut ischemia. The standard approach to the dissected descending aorta is the replacement of affected portions with a tubular graft of appropriate length and size. Presently, uncomplicated type B dissections are managed medically because surgical intervention has no proven benefit.

Interventional Therapy by Percutaneous Stenting and/or Percutaneous Fenestration

Aortic stent grafts were first used to treat abdominal and thoracic aneurysms.[89-92] Their use in the treatment of aortic dissection evolved slowly because of initial concerns that paraplegia would be a complication. However, with the advent of improved techniques, a large series of patients with aortic dissection have now been successfully treated by stent grafts (Figure 7) to cover the entry tear in the descending aorta and even the aortic arch.[93]

The role of percutaneous fenestration and stent placement in the treatment of aortic dissection has yet to be determined. However, a clear-cut indication for fenestration and stent placement is for the treatment of static or dynamic obstruction of aortic branches. Vessels compromised by static obstruction of a branch artery are treated by placing endovascular stents across the vessel of origin.[94] Vessels compromised by dynamic obstruction are treated by placing percutaneous balloon fenestration, with or without stents, in the aortic true lumen. A second indication for fenestration is to provide a reentry tear for the dead-end false lumen back into the true lumen. The aim of this procedure is to prevent thrombosis of the false lumen, which might compromise branches that derive their supply exclusively from the false lumen or jointly from the false and true lumen.

The technical goal in percutaneous balloon fenestration is to create a tear in the dissection flap that separates the true lumen from the false lumen. It is preferable to perform fenestration from the smaller (usually the true lumen) into the larger false lumen.

In a study of 57 patients who underwent interventional therapy, flow was restored in more than 90%, the average 30-day mortality was 10%, and no patient required additional surgical revascularization.[45,94-96] Most patients remained asymptomatic over a mean follow-up time of about a year. Potential problems may arise from unpredictable hemodynamic alterations in the true and false lumen after fenestration and stenting. These alterations may result in the loss of both previously well perfused arteries and desired salvage of compromised arteries.

It is evident that percutaneous interventions are safer and produce better results than surgical graft stent application.[92] Although paraplegia may occur, it is related to extensive graft stenting and is extremely rare when short (< 15 cm) stents are used. Results of short-term follow-up are good, with the 1-year survival rate exceeding 90%. The tears become well occluded and aortic diameters decrease after complete thrombosis of the false lumen. Patients can develop an inflammatory reaction after implantation, marked by elevated C-reactive protein and fever. Both signs may disappear spontaneously as the healing progresses.[97,98]

Interventional therapy in aortic dissection may provide new approaches to management of complications. Aortic fenestration with or without stent placement allows immediate relief of organ malperfusion for visceral, renal, and limb ischemia before and after surgical treatment. Graft stent implantation is an evolving technique that opens new

avenues to treat type B dissection. Occlusion of entry tears induces thrombosis and vessel wall healing.

Follow-Up

Patients with a history of aortic dissection or aortic dissection repair need close follow-up. The main task is to control blood pressure to less than 135/80 mm Hg in order to reduce wall stress. β-Blockers are the medical treatment of choice, and combination therapy with other drugs is usually needed to control the blood pressure. The most common complication after aortic dissection repair is pseudoaneurysm at the proximal or distal anastomosis of the tube graft. MRI appears to be the radiologic method of choice to follow-up patients with dissection because it avoids exposure to the ionizing radiation or nephrotoxic agents used for CT and is less invasive than TEE. Also, when serial studies are done, images can be compared with ease. After hospital discharge, regular outpatient visits are recommended at 1, 3, 6, and 12 months, and thereafter every year.

Reoperation

The majority of late deaths following primary surgery of acute dissection are caused by rupture of the aorta.[99] Therefore, timely reoperation is required whenever necessary. Indications for reoperation include aneurysm of the dissected aorta (5 to 6 cm in diameter) remote from the site of initial repair and a pseudoaneurysm at the proximal or distal anastomosis site. The rate of reoperation for proximal dissection is 10% at 5 years and up to 40% at 10 years after the primary surgery.[65,100] The proximal aorta, particularly the aortic root and ascending portion, is the most common site requiring reoperation.

Conclusion

The acute aortic syndromes still remain a diagnostic dilemma, but the advent of new imaging techniques has made establishing the diagnosis easier for clinicians. They carry significant morbidity and mortality, especially when detected late. As we look ahead into the next 100 years of clinical medicine, in order to beat this devastating disease process, we must achieve near perfection in prevention, rapid noninvasive diagnosis, and safe and effective medical management and surgical techniques. ■

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Main Points

- Aortic dissection is usually caused by weakening of the intima layer of the aorta that leads to aortic dilatation and aneurysm formation, eventually resulting in intramural hemorrhage, aortic dissection, or rupture.
- Hypertension is the most important predisposing factor for aortic dissection.
- The natural history of the disease is best characterized by differentiating between patients with proximal or distal aortic dissections.
- Up to 30% of patients later found to have acute aortic syndrome are initially suspected of having other conditions such as acute coronary syndromes, nondissecting aneurysms, pulmonary embolism, and aortic stenosis.
- The demonstration of an intimal flap separating 2 lumina is the basis for diagnosing aortic dissection.
- Intravenous antihypertensive therapy should be started emergently in all patients, except those who are hypotensive, as soon as the diagnosis is suspected.
- Newer surgical methods and techniques are now being incorporated into management of type B aortic dissections.

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