Acute Aortic Syndrome

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Acute aortic syndrome is a modern term that describes the acute presentation of patients with characteristic "aortic" pain caused by one of the life-threatening thoracic aortic conditions including aortic dissection, intramural hematoma, and penetrating atherosclerotic ulcer (Fig. 2.1). This entity involves acute lesions of the aorta involving the tunica media and can be distinguished by their etiopathogenesis and characteristic appearance using various diagnostic imaging modalities. Recent advances in imaging techniques and therapeutic interventions have increased awareness of these pathological conditions and emphasized the importance of early diagnosis and treatment.

Definition and Epidemiology

Acute aortic syndrome includes five classes of aortic disease (Table 2.1).[1] While being distinct pathological processes, there is the possibility of progression from one entity to another (Fig. 2.2). Each of these processes also shares the Stanford classification that defines the location and extent of aortic involvement. Stanford type A begins in the ascending aorta, while type B originates distal to the left subclavian artery to involve the descending thoracic aorta (Fig. 2.3). This classification system is popular since it directs different management strategies and correlates with patient prognosis. Aortic dissections can also be classified as acute or chronic, depending on whether the dissection is less or greater than 2 weeks old.

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Aortic Dissection

Classic aortic dissections (class I) are the most common cause of acute aortic syndromes (70%). Aortic dissection is the most common aortic catastrophe occurring two to three times more frequently than abdominal aortic rupture. The exact incidence is unknown but studies quote it to be 2.6–3.5 per 100,000 person-years.[2] Information gathered from the International Registry of Acute Aortic Dissection (IRAD) shows that two thirds of patients are male with a mean age of 63. Women are affected less often and present at a mean age of 67. Patients with dissections involving the ascending aorta tend to present at a younger age (50–55 years) than those with dissections of the descending aorta (60–70 years).

Table 2.2 outlines multiple risk factors for the development of aortic dissection with hypertension being the most common predisposing factor (72%). Following this is a history of atherosclerosis (31%), cardiac surgery (18%), and Marfan's syndrome (5%). Younger patients (<40 years of age) who present with aortic dissection most often have associated Marfan's syndrome, a bicuspid aortic valve or a history of aortic surgery.[3]

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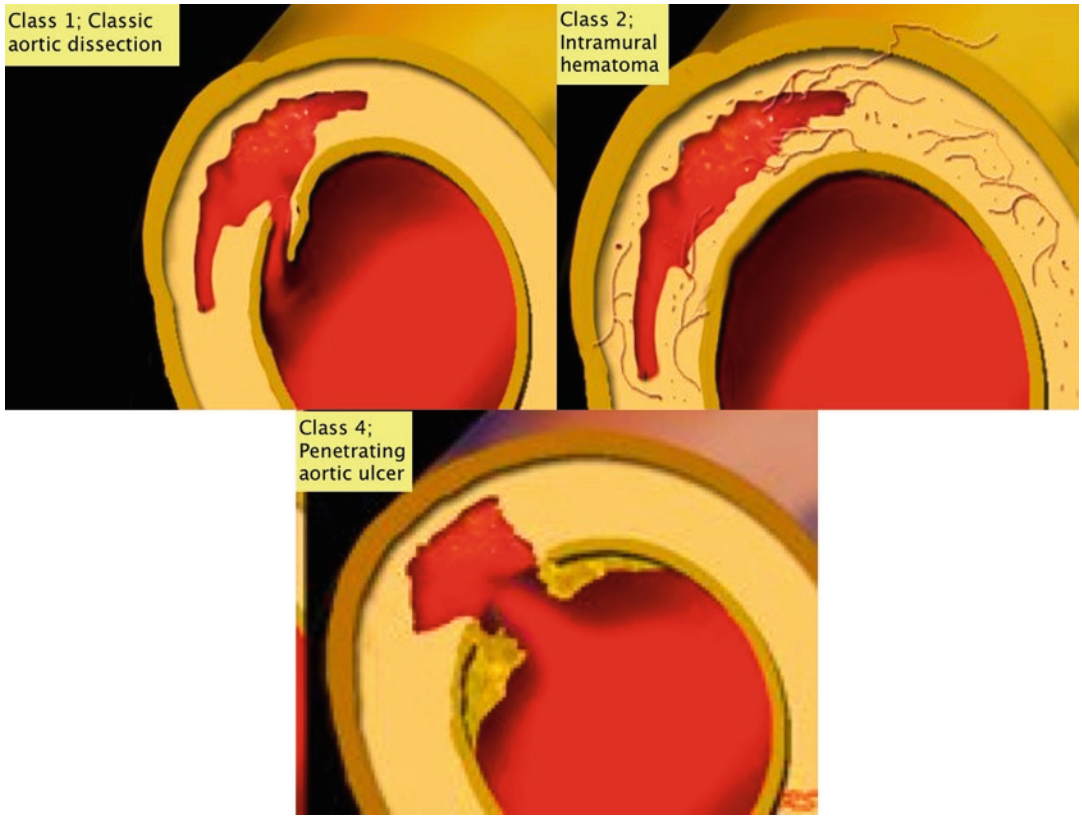

Fig. 2.1 Classification of acute aortic syndromes (Reprinted with permission from Berger F et al.[30])

Table 2.1 Classification of acute aortic syndrome

Class I – Classic aortic dissection
Class II – Intramural hematoma
Class III – Localized dissection, intimal tear without extensive intimal flap formation, localized bulge in the aortic wall
Class IV – Penetrating aortic ulcers
Class V – Iatrogenic or post-traumatic dissection

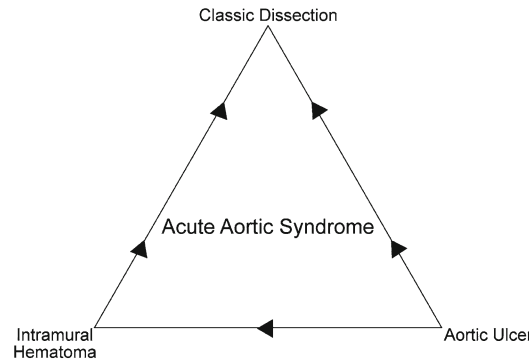

Fig. 2.2 Progression of one type of acute aortic syndrome to another type

Pathologically, classic aortic dissections (class I) are characterized by an intimal tear with separation of the aortic media into two layers, the inner two thirds and outer one third. This separation extends for a variable distance in both circumferential and longitudinal fashion. The vast majority of dissections originate from intimal tears in the ascending aorta within several centimeters of the sinuses of Valsalva where torsional movement of the aortic annulus provokes additional downward traction in the aortic root and increases longitudinal stress in that segment of aorta. The other common site for an intimal tear to originate is in the descending aorta just distal to the origin of the subclavian artery at the

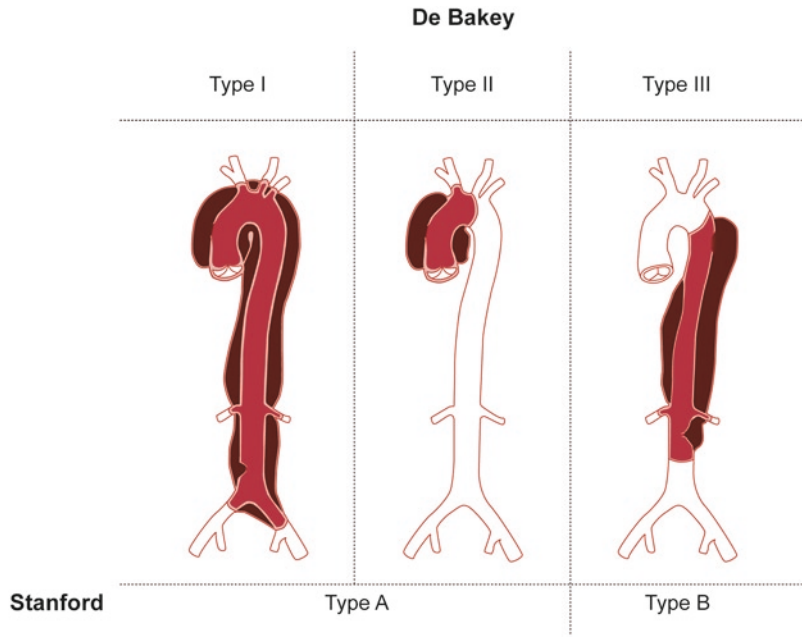

Fig. 2.3 Types of acute aortic syndromes based on the site of aortic involvement (Reprinted with permission from Springer[4])

Table 2.2 Risk factors for aortic dissection

Long-standing hypertension
Male
Advanced age
Prior cardiac surgery
Known aortic aneurysm or prior dissection
Cardiac catheterization/surgery
Connective tissue disorders Marfan's syndrome Ehlers–Danlos syndrome Bicuspid aortic valve Coarctation of the aorta Hereditary thoracic aortic disease
Vascular inflammation
Deceleration injury
Cocaine
Peripartum

site of the ligamentum arteriosum. Tears occur in the isthmus area because of increased tension at the union of the relatively mobile aortic arch with the fixed descending thoracic aorta. From this point, blood under pressure extends the dissection in an antegrade direction although retrograde extension is also possible, thus forming a false lumen and double channel aorta. A further reentrance tear allows blood to circulate in the false lumen and communicate with the true lumen. Reentry tears are often located in the abdominal aorta, iliac arteries, or other aortic branches. These small communications, normally less than 2 mm in diameter, represent intercostal and lumbar arteries severed by the dissection process.[4] The true lumen is most often the smaller of the two and is surrounded by calcifications if present. The false lumen is located along the outer curve of the aortic arch and from the aortic isthmus, a dissection adopts a spiral route involving the left border of the descending thoracic aorta and the left posterior region of the infradiaphragmatic and infrarenal aorta (Fig. 2.4). From there, a dissection may involve any branches of aorta including the arch vessels, the left intercostal, renal, and iliac vessels, most often, it is the left renal and iliac artery which are involved. The celiac, superior mesenteric, and right renal arteries usually communicate with the true lumen. Dissections rarely extend to the common femoral arteries.

Class III dissections are characterized by the presence of an intimal tear without a flap or hematoma formation. Patients often present with the classic symptoms of dissection and may have associated aneurysms, aortic regurgitation, or

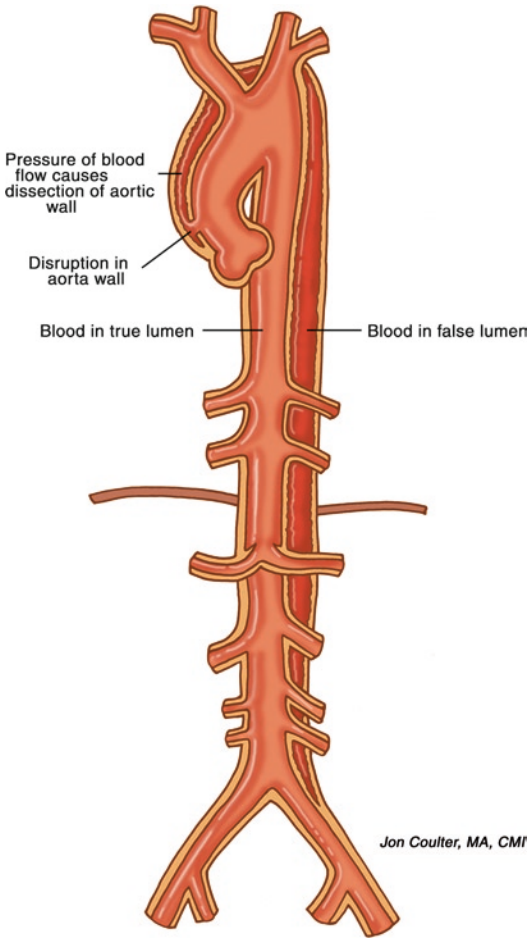

Fig. 2.4 CT reconstruction of a type A dissection following graft placement in the ascending aorta. The dissection that extends into the descending aorta demonstrates how the defects spiral around the aorta

pericardial effusion. Imaging techniques such as CT, MRI, or TEE may fail to detect this type of dissection since each of these modalities depends on the presence and identification of an intimal flap or true and false lumen for diagnosis. Aortography may show an eccentric bulge in the aorta, which should raise the suspicion of this class of dissection. Treatment is similar to that for classic aortic dissection.[1]

Class V aortic dissections are caused by traumatic or iatrogenic injury. Traumatic dissections are described in detail in another chapter. Iatrogenic aortic dissections may result from cardiac surgery, percutaneous coronary interventions, or endovascular procedures. Dissections resulting from cardiac surgical procedures may occur during the procedure, early in the postoperative period, or their presentation may be delayed for years.[5,6] The incidence varies between 0.12% and 0.35% of cardiac surgeries, but the mortality is high unless recognized early. Injury to the aorta can occur at the site of cross-clamp placement, cannulation and decannulation, aortotomy, cardioplegic cannulation, or aortocoronary anastomosis. Preexisting atherosclerotic or connective tissue disease, increased aortic diameter, previous heart surgery, a history of hypertension and elevated CPB pressures are all risk factors. There is a growing concern of an increased incidence of aortic dissection following off-pump coronary artery bypass grafting compared to on-pump CABG related to the application of a side-biting clamp under higher blood pressure and pulsatility.[7] Intraoperative TEE and epiaortic ultrasound may help in the prevention and early diagnosis of this potentially lethal complication. Type A dissections have been reported after diagnostic coronary angiography (0.01%) and other percutaneous interventions (0.03%).[8,9] Isolated coronary artery dissections and localized aortic dissections (less than 40 mm of ascending aortic involvement) can be treated with intracoronary or aortic stenting. However, a failed stenting procedure or progression of a dissection must be surgically treated. Retrograde type A dissections after endograft placement for type B dissections have been reported with an incidence of 10–27%.[10] The presentation may be acute or delayed for several months. Various etiological factors include wire and sheath manipulation in the arch, repeated balloon dilatations, oversizing of grafts, injury to a diseased aorta at the margin of inflexible grafts, and progression of disease unrelated to stent grafting. Surgical replacement with tube grafts is recommended in these patients and mortality is high (27%). Other procedures associated with an increased risk of aortic dissection include the insertion of intra-aortic balloon pumps, percutaneous angioplasty, and stenting for coarctation of the aorta.[11]

Intramural Hematoma

Aortic intramural hematomas (IMH) account for up to 20% of all cases of AAS and represent a variant of dissection characterized by the absence of an intimal flap, reentrant tear, or double channel with false lumen flow. IMH often occur in patients with severe atherosclerotic disease in which penetrating aortic ulcers or atherosclerotic plaques rupture causing intimal injury with blood entering the media. In patients with mild or no atherosclerosis, spontaneous rupture of the vasa vasorum may initiate aortic wall degeneration, which leads to hematoma formation in the aortic wall, splitting of the medial layer, and dissection formation without an intimal tear (Class II).[12] Patients in the first group tend to be older and have coexisting coronary and peripheral vascular disease. Absence of intimal tears is not mandatory in the diagnosis of IMH since small communications may be seen indicating rupture as a decompressing mechanism.[13] The presence of large intimal erosions or deep ulcer-like lesions in the intima is associated with a progressive disease course and poor outcome.[14]

IMH evolution is difficult to predict (Fig. 2.5). In some cases, the hematoma does not change in size. Resolution occurs in fewer than 10% of cases, but is more likely with less hematoma and aortic wall thickness.[2] When an IMH resolves, a localized aneurysm may develop because of a weakened media and remodeling, requiring close surveillance. Progression of IMH may lead to weakening and disruption of the intimal layer causing a classic dissection. Progression to dissection has been shown to occur in 16–47% of patients with IMH.[15] A few patients may demonstrate IMH and dissection in different segments of aorta, known as a hybrid case. Increasing aortic diameter (>50 mm) causing aneurysm formation is a poor prognostic factor associated with IMH.[16] In addition, increased permeability of the aortic wall may lead to pericardial or pleural effusions or a mediastinal hemorrhage. Most effusions will resolve. However, large and progressive fluid accumulations are an ominous signs. Weakening of the adventitial layer may lead to aortic rupture, while a contained rupture from disintegration of the outer medial layers with intact adventitia is not uncommon.[17] Similar to aortic dissections, the

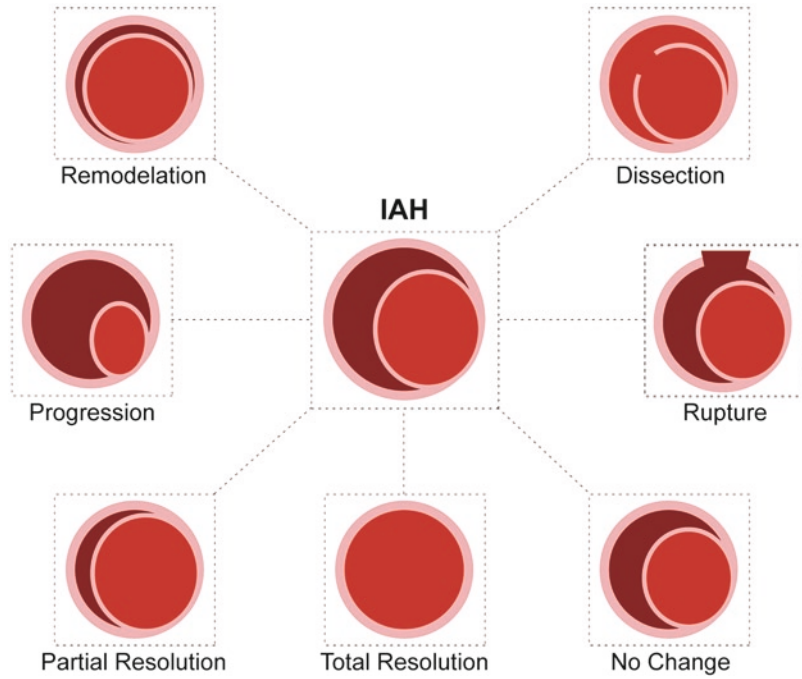

Fig. 2.5 Evolution of intramural hematoma (Reprinted with permission from Springer[4])

most common underlying condition associated with IMH is hypertension.

Penetrating Aortic Ulcer

The term penetrating atherosclerotic ulcer (PAU) describes a condition in which ulceration of an atherosclerotic lesion penetrates the intima and extends into the media, eroding the inner elastic layer of the aortic wall. The reason why most atherosclerotic ulcers do not penetrate the internal elastic lamina and few penetrate the media and adventitia is not clear. PAUs are focal lesions most often located in the descending thoracic aorta, which correlates with a greater disease burden in that region. These patients tend to be older with severe systemic atherosclerosis but without connective tissue diseases. Multiple PAUs are often found in a single patient. Imaging of PAU most often reveals extensive atherosclerosis with severe intimal calcification and plaque. A crater or extravasation of contrast is often visualized.[18]

Occasionally, PAU may progress (Fig. 2.6). This may precipitate a localized, intramural hemorrhage following progressive erosion and rupture of the vasa vasorum.[19] Further penetration to the adventitia may lead to the formation of a saccular aneurysm or pseudoaneurysm.[20] Symptomatic ulcers with signs of deep erosion are more prone to aortic rupture.[21] In rare cases, PAU may lead to aortic dissection, but those dissections arising from a PAU tend to be less extensive and demonstrate a thick, calcified static flap in a location atypical for entrance tears. Longitudinal spread of aortic dissections arising from PAU is limited by medial fibrosis and calcification.[20]

Acute Expanding Aneurysms

Also included in many discussions of acute aortic pathology are symptomatic aortic aneurysms which may represent impending rupture. The acute expansion of an aortic aneurysm accounts for a significant number of patients who present with the sudden onset of chest pain. As aneurysms increase in size, the dilatation results in greater wall tension and a more rapid rate of expansion. In addition, aneurysms may be associated with progressive dissections, IMH, or PAU, all of which cause weakening of the aortic wall.

Etiopathogenesis

The physiopathological mechanism that precipitates the appearance of each of these entities varies somewhat; however, both acquired and genetic conditions share an underlying pathology that involves breakdown of the intimal layer and weakening of the aortic media, both of which lead to higher wall stress. In addition, it is common for there to be progression from one

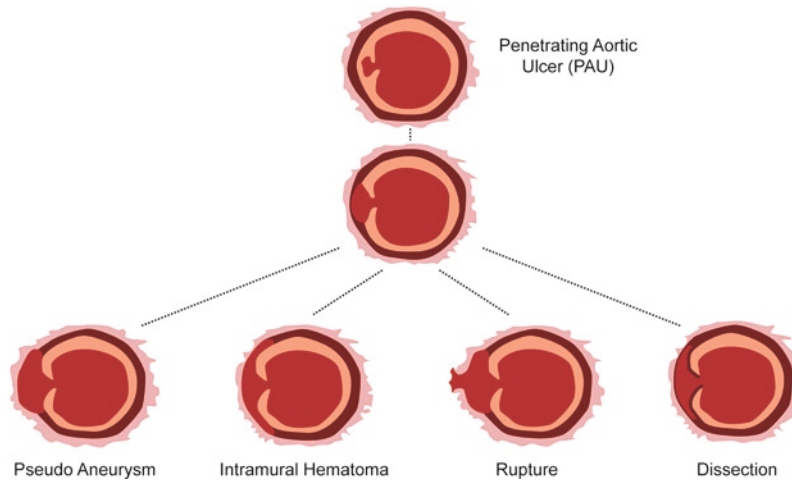

Fig. 2.6 Evolution of penetrating aortic ulcers (Reprinted with permission from Springer[4])

type of lesion to another and some patients may concurrently manifest multiple types of lesions.

Hypertension and Mechanical Forces

The most common risk factor for aortic dissection is hypertension. Chronic exposure of the aorta to high pressures leads to intimal thickening, fibrosis, calcification, and extracellular fatty acid deposition. The extracellular matrix also undergoes accelerated degradation, apoptosis, and elastolysis. Both mechanisms lead to intimal weakening and medial degeneration.[22] At any site where there is tissue thickening and fibrosis, there is compromise of nutrient and oxygen supply to the arterial wall. Eventually, there is necrosis of smooth muscle cells and fibrosis of elastic tissue in the vessel wall. The resulting stiffness increases the risk of aortic pathology when exposed to the chronic trauma of arterial hypertension.

Genetic Predisposition

Marfan's syndrome, Ehlers–Danlos syndrome, familial forms of aortic aneurysm and dissection, as well as bicuspid aortic valve are genetic conditions that predispose patients to develop an acute aortic syndrome and correlate with earlier presentations. Among these, Marfan's syndrome is the most prevalent connective tissue disorder with an incidence of one in 7,000. The underlying defect is a mutation on the fibrillin-1 gene, which results in defective fibrillin in the extracellular matrix.[22]

Ehlers–Danlos syndrome is another inherited connective tissue disorder characterized by tissue fragility. The familial form of aortic disease has been localized to mutations on the fibrillin-1 gene, similar to Marfan's syndrome. Characteristic to each of the different genetic disorders is a similar pathophysiology that includes a dedifferentiation of vascular smooth muscle cells and enhanced elastolysis of aortic wall components. In addition, increased expression of metalloproteinases promotes fragmentation of elastic tissue in the medial layer.[22]

Matrix Metalloproteins

The matrix metalloproteins are a group of proteins whose primary function is to degrade extracellular matrix. Patients with thoracic aortic disease have been shown to have increased levels of these enzymes, which favor proteolysis within the aortic wall. There are still many unanswered questions related to the role these enzymes play in AAS; however, they may serve as a marker for aortic pathology or their inhibition may slow the progression of disease.[23]

Clinical Presentation

AAS is characterized clinically by "aortic pain," most often in a patient with a coexisting history of hypertension. The early recognition of pain associated with progressive aortic lesions is of paramount importance. Table 2.3 lists various presenting symptoms and signs of AAS and Table 2.4 lists the most common associated conditions. The characteristic findings listed may

Table 2.3 Presenting features of acute aortic syndrome

Chest pain – abrupt, severe, tearing, radiating
Anterior pain or radiation to neck – associated with ascending aorta
Radiation to back or abdomen – associated with descending aorta
Syncope or cerebrovascular accident
Hypertension – SBP > 150 mmHg
Shock or tamponade
Diastolic murmur
Pressure differential in upper extremities
Pulse deficits
End-organ ischemia

Table 2.4 Comorbid conditions associated with acute aortic syndrome

History of hypertension
Smoking
Known coronary artery disease
Thoracic or abdominal aortic aneurysm
Peripheral arterial disease
Prior stroke
Chronic renal insufficiency

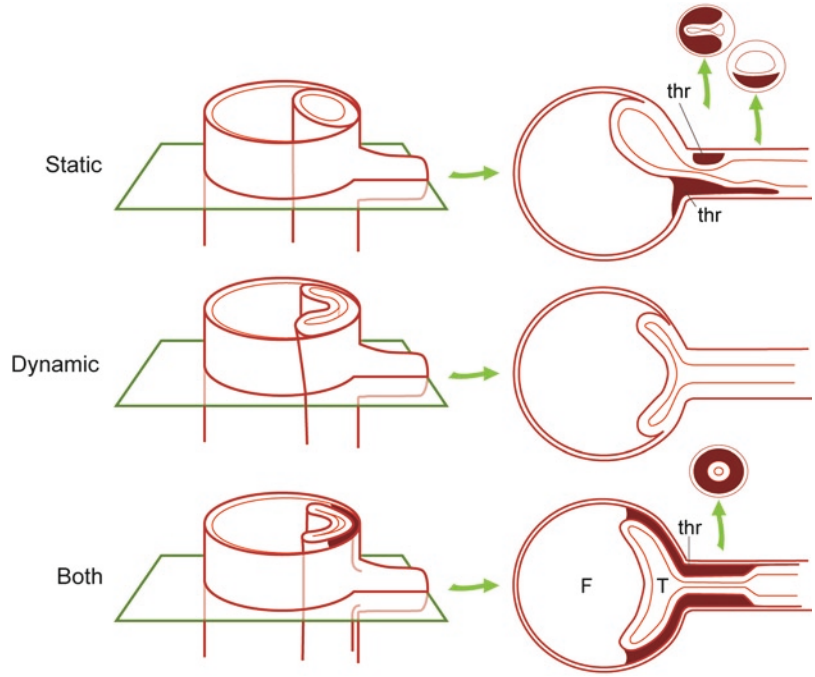

Fig. 2.7 Types of visceral organ malperfusion with aortic dissections (Reprinted with permission from Springer[4])

vary or be absent, emphasizing the necessity of a high level of suspicion in order to avoid diagnostic delays and poor outcomes.

The pain caused by PAU and IMH is similar to classic aortic dissection. The various types of AAS cannot be reliably differentiated on clinical grounds alone. Aortic pain is described as severely intense, acute, tearing, throbbing, and radiating.[24] In contrast to the increasing intensity of dull cardiac pain, AAS is described as being more abrupt and at maximal intensity from the onset. In addition, 4.5% of patients deny having any pain on presentation.[25] Pain located in the anterior chest and neck is related to involvement of the ascending aorta and may be easily confounded with that of ischemic syndromes. Back and abdominal pain may indicate that there is involvement of the descending aorta. Syncope may be the presenting symptom in up to 20% of cases and is associated with a proximal dissection. The onset of syncope or central neurological deficits indicates probable complications such as obstruction of cerebral vessels, cardiac tamponade, or activation of cerebral baroreceptors. Other presenting features of AAS may include end-organ ischemia or pulse deficits.[3] Two types of distal visceral organ malperfusion are described (Fig. 2.7). In static obstruction, the dissection flap enters the origin of the branch and encroaches on the lumen. If there is no reentry, then the true lumen will be narrowed to cause ischemia. In dynamic obstruction, the flap prolapses and obstructs the origin of the vessel without entering the vessel. Static and dynamic obstruction may be combined to contribute to branch obstruction.

Diagnostic Evaluation

As diagnostic modalities have improved over the past 2 decades, the awareness of AAS has increased significantly. PAU and IMH were virtually unknown in the prior era of aortic imaging by aortography. These disorders were not classified as being distinct entities until the mid-1980s. In the current era of three-dimensional, high-resolution imaging by computerized tomography (CT), magnetic resonance (MR) imaging, and

transesophageal echocardiography (TEE), these two disorders have become increasingly recognized. The sensitivity and specificity of different imaging techniques and their comparison are given in Table 2.5.[26] The goals of diagnostic imaging in patients with suspected AAS are confirmation of the diagnosis, classification and type of aortic pathology, tear localization, and identification of signs indicating the need for emergent intervention (pericardial, mediastinal, or pleural hemorrhage).[2] Additional information includes arch and branch vessel involvement. Each imaging modality has distinct advantages (Table 2.6) and most patients require multiple imaging studies to diagnose and characterize the underlying aortic pathology.[27]

Electrocardiogram

An ECG is an important first diagnostic test performed for all patients presenting with acute chest pain, whether typical or atypical for an acute aortic syndrome. The differentiation of AAS from acute coronary syndrome (ACS) is important; however, it must be remembered the ACS may occur as a result of AAS.

Chest Radiography

While rapid imaging plays a crucial role in the diagnosis of AAS, the role of chest radiography has become limited, especially for conditions

Table 2.5 Results of meta-analysis – diagnostic accuracy of different imaging modalities for suspected aortic dissection (Reprinted with permission from American Medical Association[16])

Imaging technique	Number of studies	Sensitivity	Specificity	Positive likelihood ratio	Negative likelihood ratio
TEE	10	98 (95–99)	95 (92–97)	14.1 (6.0–33.2)	0.04 (0.02–0.08)
Helical CT	3	100 (96–100)	98 (87–99)	13.9 (4.2–46.0)	0.02 (0.01–0.11)
MRI	7	98 (95–99)	98 (95–100)	25.3(11.1–57.1)	0.05 (0.03–0.10)

Data reported with 95% confidence intervals. Likelihood ratios greater than 10 and less than 0.1 are considered strong evidence to confirm or ruling out the diagnosis of aortic dissection

Table 2.6 Comparison of imaging modalities (Reproduced with permission from Oxford University press and the primary author Professor Raimund Erbel[56])

	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	++	++	+++	–	–
Intraoperative availability	+++	–	–	(+)	(+)

TTE/TEE – transthoracic/transoesophageal echocardiography; CT – computed tomography; MRI – magnetic resonance imaging; IVUS – intravascular ultrasound

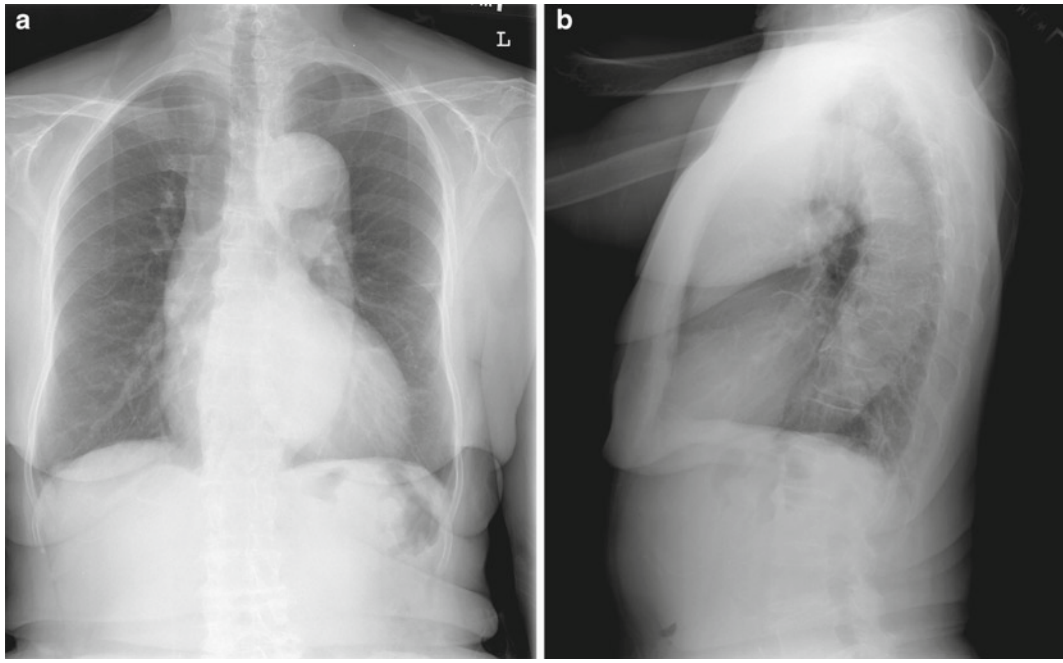

Fig. 2.8 PA (**a**) and lateral Chest X-Rays (**b**) demonstrating a dilated thoracic aorta and widened mediastinum

confined to the ascending aorta. In a study of 216 patients over a 6-year period, the sensitivity for aortic disease was 64%, with a specificity of 86%. The sensitivity for lesions of the ascending aorta was 47%, while that for disease involving distal aortic segments was 77%.[28] A chest X-ray may show widening of the aortic contour, displaced calcification, aortic kinking, or opacification of the aortopulmonary window (Fig. 2.8a and b).

Computerized Tomography (CT)

When AAS is suspected based on clinical presentation and an acute coronary syndrome has been excluded, cardiac-gated, contrast-enhanced multidetector CT angiography is nearly 100% sensitive and specific in the diagnosis, differentiation and staging of AAS.[29] Modern CT machines have up to 64 rows of detectors, and this enables them to generate multiple simultaneous images with a slice thickness of less than 1 mm. Multidetector CT is also extremely fast with spiral imaging of the entire thorax can be done in a single breath-hold, which eliminates respiratory motion artifact. Cardiac gating is done to avoid artifacts produced because of the imaging being obtained during different phases of cardiac cycle. CT allows for a complete diagnostic evaluation of the thoracic aorta, including the lumen, aortic wall, and periaortic region. Both unenhanced and enhanced images are valuable. Unenhanced scans depict intramural hematoma (crescent shaped or circumferential wall thickening) and thrombosis of the false lumen. Contrast-enhanced imaging allows demonstration of an intimal flap, which is the most reliable finding in the diagnosis of dissection (Fig. 2.9a and b). Contrast differences between arterial and venous phase can help differentiate true and false lumens. Interestingly, it can be difficult to differentiate an aneurysm with thrombus from a dissection with a thrombosed false lumen. Intimal calcifications are displaced by a false lumen in the latter case.[30]

The diagnosis of IMH and PAU are also highly accurate with CT.[31,32] The diagnosis of PAU is made by the demonstration of an outpouching of the aortic wall with rough edges (Fig. 2.10).

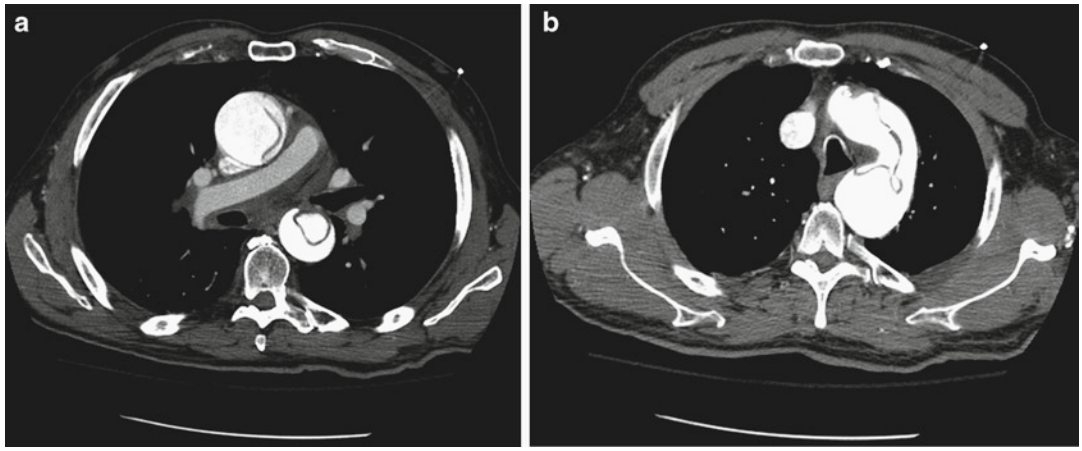

Fig. 2.9 Contrast-enhanced chest CT clearly depicting a type A dissection. The intimal flap is seen in the ascending and descending aorta (**a**) as well as the aortic arch (**b**)

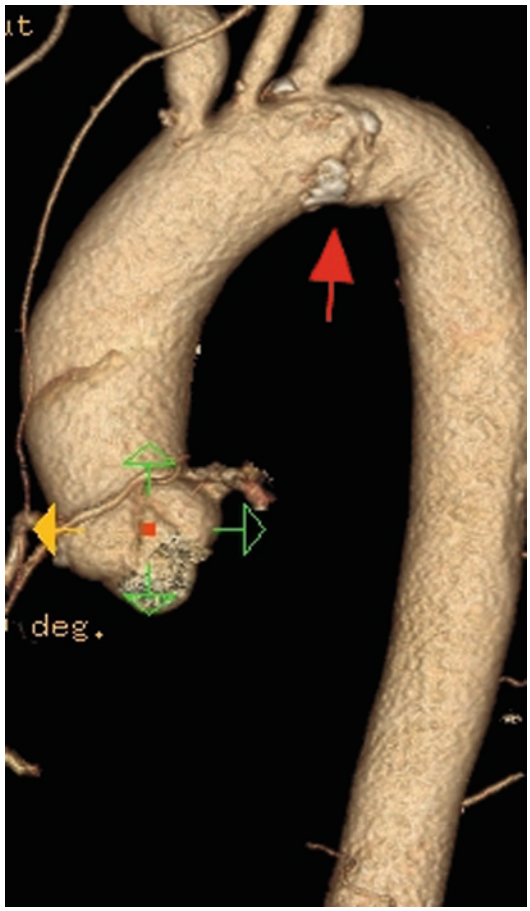

Fig. 2.10 CT reconstructions demonstrating PAU of the arch and descending aorta as indicated by arrows

The ulcer is usually surrounded by extensive atherosclerotic plaques. CT imaging of the aorta should include the iliac arteries for possible endovascular intervention and aortic arch branches to evaluate the extent of dissection and possible neurological complications. Other benefits of CT are its availability and noninvasiveness. The major drawbacks of CT are the exposure to large doses of ionizing radiation and contrast agents.[29] In addition, CT does not offer the capability to assess for aortic insufficiency or involvement of the coronary arteries.

Magnetic Resonance Imaging (MRI)

MRI is a valuable diagnostic tool for the diagnosis of acute aortic syndrome. MRI can be highly accurate even without the use of contrast. Although MRI has the highest sensitivity and specificity for the detection of all forms of aortic pathology and provides superior anatomic detail when compared to other imaging modalities, it is limited by availability, expense, and patient restrictions such as pacemakers, aneurysm clips, or other metal devices. Prolonged scanning time and limited access to unstable patients are other limitations. For these reasons, MRI is not a widely used tool (<5% of patients in IRAD)[3] (Fig. 2.11).

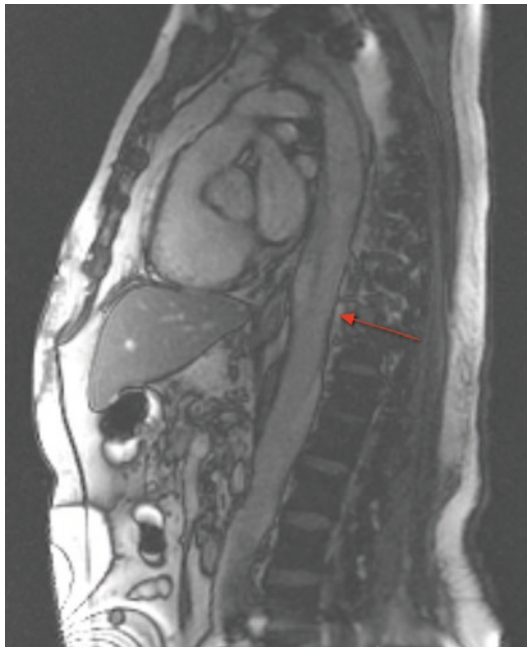

Fig. 2.11 MRI of the chest showing a descending aortic dissection

Echocardiography

Transthoracic echocardiography (TTE) can be used as screening tool for the diagnosis of AAS of the proximal aorta. TTE is useful in the rapid evaluation of aortic insufficiency, pericardial tamponade, arch vessel involvement, and left ventricular systolic function. Hemodynamically unstable patients in whom TTE has shown pericardial effusion to be present should be taken to the operating room for airway stabilization and further TEE evaluation. Thus, cardiac surgeons rely on echocardiography more than other imaging modalities including CTA in emergency situations.[33] A TEE can be done while the surgeon simultaneously prepares to open the chest.

Transesophageal echocardiography (TEE) is highly sensitive and specific in diagnosing thoracic aortic pathologies. The only limitation is in the distal ascending aorta and proximal arch which are not clearly visualized by TEE in most patients. Evaluation of the aortic valve and suitability for repair can be done with TEE. The diagnosis of an IMH is characterized by crescentric aortic wall thickening, the absence of an intimal flap and a lack of false lumen color flow typical of dissection. The primary limiting factor in the use of TEE is the requirement for a skilled echocardiographer to be immediately available to perform and interpret results in emergency situations.[29] A further detailed description of TEE examinations for aortic pathology is described elsewhere in this textbook.

Aortography

Since the development of newer, noninvasive imaging modalities, there has been a shift away from invasive techniques for imaging the aorta. Traditionally, aortography had been the gold standard for the diagnosis of aortic dissection. Its primary limitations include its invasive nature and further risk of intimal damage, the use of contrast agents, as well as limited visualization of thrombosed dissections, IMH, and occluded branch vessels. The specificity for diagnosing aortic dissection is >95%, but the sensitivity only averages 90%.[2] Intravascular ultrasound (IVUS) with high-frequency transducers (8–10 MHz) has been used to complement conventional angiography in the diagnosis of acute dissection. IVUS probes are advanced through guidewire and guiding catheters under fluoroscopy to evaluate the aorta from inside and provide useful information about the vessel wall and pathology.

Management

Once there is a suspicion of AAS, patients should be rapidly stabilized and transported to a tertiary care center with adequate aortic surgical and endovascular surgical expertise. It is there that further imaging and management should take place. Figure 2.12 provides an algorithm for the rapid evaluation and treatment of patients presenting with a suspected AAS.[29,34]

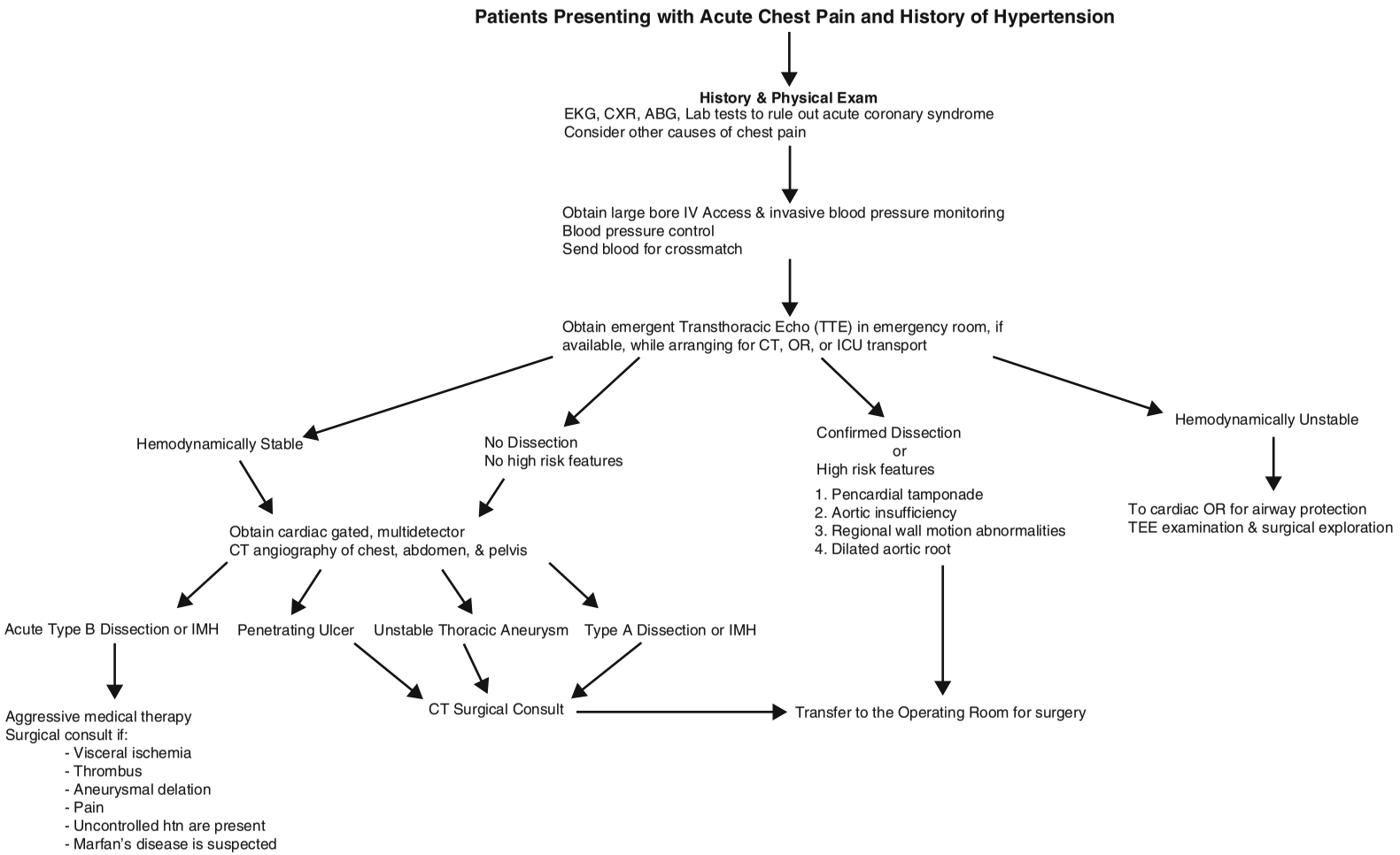

Fig. 2.12 Acute aortic syndrome management algorithm (Modified and reprinted with permission from the following resources[29,34])

Although the treatment of AAS remains a therapeutic challenge, diverse surgical and percutaneous strategies continue to evolve. There are many factors that affect management decision as listed in Table 2.7. One third of the mortality associated with AAS is the result of end-organ failure, which emphasizes the importance of early intervention. The goal of treatment is to prevent the progression of the disease and its lethal complications.

The initial management of all patients with AAS involves pain relief and aggressive blood pressure control. In normalizing the blood pressure, the goal is to reduce the force of left ventricular ejection (dP/dt), which is the primary cause of dissection extension and aortic rupture. Beta-blockers are the preferred agents because they not only reduce systemic pressure but also lower heart rate. For most patients, the goal is a systolic pressure between 100 and 120 mmHg and heart rate <60 bpm or the lowest tolerable levels that provide adequate cerebral, coronary, and renal perfusion. Less is known about the role of calcium channel blockers for patients who are β-blocker intolerant, but they should reduce blood pressure without causing reflex tachycardia. If neither of the above agents is adequate to control the blood pressure, vasodilators may be added. However, they should never be used as an initial form of therapy before starting β-blockers because of the reflex tachycardia and increase in the force of left ventricular ejection leading to increased aortic wall stress.

Table 2.7 Patient factors affecting management decisions

Disease location – type A or B
Retrograde extension into the arch and/or ascending aorta
Rupture or impending rupture
Site of entry and reentry
Involvement of side branches
Branch origin from true or false lumen
Risk of end-organ damage
Complications – rupture, coronary occlusion, aortic insufficiency, neurological
Diameters of the true and false lumens
Areas of normal vessel for stent-graft placement

Type A Aortic Dissection

Acute ascending aortic pathology (acute dissection, IMH, and PAU) should be treated as a surgical emergency because of the possible risk of life-threatening complications including aortic rupture, pericardial tamponade, and high early mortality (Fig. 2.13). Surgery is also aimed to relieve aortic regurgitation and reestablish coronary and branch vessel perfusion. Acute type A dissection has a mortality rate of 1–2% per hour during the first hours of symptom onset and without surgical treatment, the mortality rate is 20% by 24 h, 30% by 48 h, 40% at 1 week, and 50% at 1 month.[35] Even with surgical treatment,

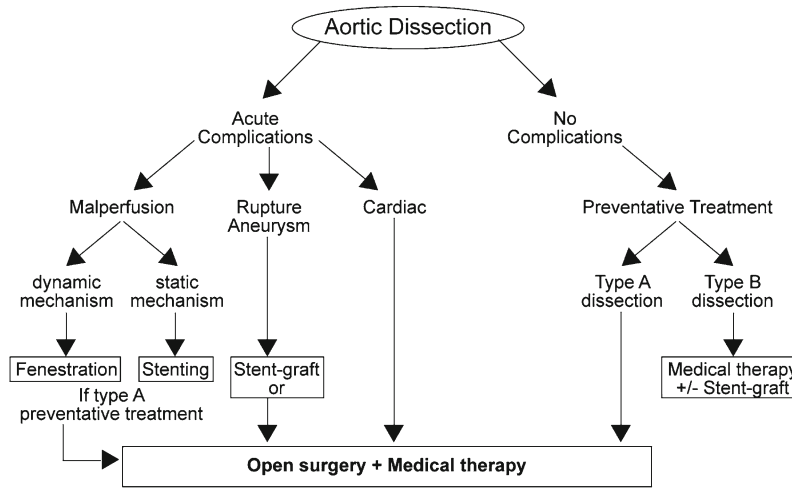

Fig. 2.13 Management of acute aortic dissection (Reprinted with permission from Springer[4])

the mortality rate is as high as 10% by 24 h and 20% at 1 month.[36,37]

Aortic arch and descending thoracic intimal tears are seen in 20–30% of patients with type A dissection and if left untreated predisposes to later distal reoperation.[38,39] Patients who require partial or total arch replacement with reconnection of supraaortic vessels to the graft must often undergo deep hypothermic circularoty arrest with antegrade or retrograde cerebral perfusion. If a dissection extends into the descending thoracic aorta, an elephant trunk extension of the arch graft is an option.[40] In a later procedure, the elephant trunk portion of the graft may be connected to the distal descending aorta, a tubular graft or endovascular graft. There are reports of endovascular repair of the ascending aorta in highly selected patients, but this has most often been for the purpose of temporizing symptoms and preventing disease progression before a more definitive open repair could be performed. It is most often not possible due to the anatomic restrictions of securing the graft in the ascending aorta. The placement of stent grafts has been applied to treat aortic branch occlusions in both type A and B acute aortic dissections. In type A dissections with visceral malperfusion, treatment of the leaking aorta and aortic valve take precedence over visceral malperfusion. However, patients with prolonged limb or bowel ischemia may be unsuitable for proximal aortic surgery, and endovascular treatment to restore perfusion to critical organs is recommended.

Type B Aortic Dissection

Uncomplicated Type B Dissection

Medical management (analgesics and antihypertensive therapy) still remains the main stay of therapy for patients with uncomplicated type B disease. It is safe to treat these patients medically with close follow-up for ischemic complications, disease progression, or aneurysmal enlargement. In a group of 384 patients with type B dissections, 73% were treated medically with a 10% in-hospital mortality. Long-term survival is 60–80% at 5 years.[3] Surgical repair has not been shown to improve outcomes in this group of patients. Recently, a completed randomized study (Investigation of stent grafts in patients with type B aortic dissection-INSTEAD) examined the use of endografts versus medical therapy in uncomplicated dissections. The results did not show any survival benefit at 2 years.[41]

Complicated Type B Dissections

Complicated type B aortic disease is differentiated by the presence of a distal malperfusion syndrome or rapid disease progression. Indications for intervention are similar to those for type A diseases; the prevention of life-threatening complications such as organ or limb ischemia, aneurysm expansion and risk of rupture, periaortic blood collection, intractable pain, aneurysm expansion or uncontrolled hypertension. The mortality rate for open surgical repair (graft replacement, fenestration, or bypass procedures) is 30–35% and even higher with the presence of visceral malperfusion.[42,43] Aortic endovascular grafting may be particularly beneficial in this group and has shown improved mortality rates (16%). The goals of endograft treatment include reconstruction of the aortic segment containing the entry tear, induction of thrombosis in the false lumen, and the reestablishment of flow in the true lumen and branches.[44] In one series, false lumen thrombosis was achieved in 85–100% of patients.

An additional benefit of stent grafts is the ability to relieve dynamic and combined static and dynamic obstructions successfully in complicated acute type B dissections.[45,46] Static obstructions are relieved by placing the graft in the branch vessel and dynamic obstructions may benefit from stents in the true lumen. Endografts have also been used in patients with retrograde type A dissection with intimal tear in the descending aorta to induce false lumen thrombosis.[47] Surgical treatment may be the only option in patients with failed endografts or patients unsuitable for this less-invasive technique.

With either surgical, endograft, or medical management, the risk of further dissection is never eliminated. Therefore, close, regular monitoring, most likely with CT imaging, is necessary to assess for progression or complications.

Intramural Hematoma

Type A Disease

The recommended treatment for patients with Type A IMH is prompt surgical intervention. Proximal IMH are independently associated with potential progression to dissection, aneurysm, and rupture as well as poor clinical outcomes.[17] The risk of a nonsurgical approach to type A IMH demonstrates an early mortality of 55% compared with 8% following surgical repair.[17] However, for patients with significant comorbidities and uncomplicated type A IMH (no dissection or intimal tear, thickness less than 11 mm, aortic diameter less than 50 mm), medical treatment with follow-up imaging and timed surgical intervention has been recommended.

Type B Disease

The management of IMH involving the descending aorta is similar to that recommended for type B dissections. The current literature supports medical management. However, if complications arise (ulceration, expansion, and dilatation), endograft placement may be considered although limited data exists. Endografts may cause erosion of the intima during the acute phase. IMH of the descending aorta have been associated with an in-hospital mortality rate of 10%, similar to that of type B aortic dissection, further emphasizing the importance of correct diagnosis and proper treatment.

Penetrating Aortic Ulcers

There are multiple important factors to identify when diagnosing PAU. One must determine the number (single or multiple), location (type A or B), and associated complications (IMH, dissection, pseudoaneurysm, and rupture). Type A PAU should be treated surgically. Medical therapy is indicated in stable patients with type B PAU.[48] Very few centers advocate any surgical intervention in uncomplicated type B patients because there is a high risk of organ failure and poor prognosis due to the high likelihood of extensive atherosclerotic disease. For patients with symptomatic or progressive disease, focal PAU in the descending aorta are ideal targets for endograft placement. In a meta-analysis of 58 patients from 13 studies, complete sealing of the ulcer was possible in 94% of patients. Neurologic complications were present in 6% and the in-hospital mortality rate was 5%.[49] Long-term results are not known at this time.

Natural History and Prognosis

The outcomes of patients treated for AAS have improved significantly, although there still remains a high mortality rate in the acute phase. Table 2.8 lists predictors of in-hospital death.[50] Early clinical suspicion and greater surgical expertise appear to be the most important factors in reducing mortality.

Type A aortic dissections are highly lethal. Overall, mortality at 1 month is 20% with and 50% without surgical treatment for type A dissections. The risk of death is higher if there are complications of pericardial tamponade, involvement of the coronary arteries causing acute myocardial ischemia, or a malperfusion syndrome. Age greater than 70 has been identified as an independent risk factor for hospital death for acute type A dissection. Shock, hypotension, and tamponade are other risk factors for increased mortality.[37]

For type B dissections, the overall mortality rate is 10% with medical treatment. Circulatory shock and visceral ischemia predispose to a higher mortality in type B dissection.[51]

Table 2.8 Predictors of in-hospital death

Age >70
Abrupt onset of pain
Hypotension/cardiac tamponade/shock
Abnormal EKG
Kidney failure
Pulse deficits
Iatrogenic cause

Following the acute-phase, mid- to long-term survival depends not only on the underlying aortic disease but also other comorbidities. In patients with surgically corrected type A dissections, survival differences were based on the presence or absence of distal false lumen flow. In one study, the absence of a false lumen was achieved in 53% of patients compared to 10–20% in most series. In patients with absent false lumen flow, the survival was 85% at 6 years compared to 62% in patients with persistent false lumen flow.[37] At 10 years, the survival rate was 44% for corrected type A dissections compared to 32% in medically treated type B dissections.[51] The primary reasons for higher, long-term mortality in type B dissections were aneurysmal expansion and rupture. Dilatation in the descending aorta was greater in medically treated type B dissections compared to operated type A dissections. Dilatation occurs at a faster rate in patients with false lumen flow when compared to absent false lumen flow in type B dissections.[52] Junoven et al. described rupture in 18% and rapid expansion requiring surgery in 20% of patients with type B dissection at 3 years.[53] The impact of early endovascular treatment on the long-term survival of type B dissections is yet to be determined.

In patients with type A IMH, Kaji et al.[54] reported that surgery was required in 43% of patients during the acute phase and, among those discharged without surgery (57%), complete resolution occurred in 40% of patients. Type B IMH has a better long-term prognosis than type B dissections with 5-year survival reported between 43% and 90%.[17,55] Close follow-up with imaging techniques is recommended in patients undergoing medical treatment to look for complications (dilatation, pseudoaneurysm formation, and dissection).

Prevention and Follow-Up

At a time when there is an increasing elderly population, the awareness of such conditions as AAS is likely to continue to rise. This is due to improvements in diagnostic modalities, longer life expectancy, and longer exposure to elevated blood pressure. In order to best treat these patients, continued improvements in diagnostic imaging and therapeutic strategies are necessary, and a focus on surveillance and prevention will further reduce the morbidity and mortality associated with aortic pathology.

One possibility for the surveillance of patients at risk for the development or progression of AAS is the development of biomarkers, which would enable serum diagnosis. In addition, this would provide a fast and economic means of differentiating patients who present to the emergency room with chest pain. Possible markers currently include an assay for circulating smooth muscle myosin heavy chain protein, soluble elastin fragments and acute-phase reactants such as C-reactive protein, fibrinogen, and D-dimer.

All patients with a known aortic disease require close surveillance following discharge. Lifelong treatment of hypertension is required and regular assessments of the aorta should be performed at 1, 3, 6, 9, and 12 months as well as every 6–12 months thereafter, depending on the aortic size. The most important findings on imaging are aortic diameter, signs of aneurysm formation, and hemorrhage at surgical anastamosis or stent-graft sites. The close follow-up emphasizes the fact that aortic disease progression is not easy to predict. Repeated surgery is required in 12–30% of patients due to extension or recurrence of dissection, aneurysm formation, graft dehiscence, aortic insufficiency, or infection.[26]

Conclusion

Significant advances have been made in the diagnosis and management of acute aortic dissections over the past 2 decades. These advancements have led to a better understanding of aortic pathology and led to the discovery of variants that are collectively termed acute aortic syndrome. Despite a persistent level of uncertainty in the diagnosis and management of this lethal disorder, advances are being made and patient outcomes are improving.

Key Notes

1. The clinical progress of patients with AAS is unpredictable. A high level of suspicion is required for early diagnosis and crucial for patient survival.
2. The most common risk factor for AAS is hypertension, with men being affected more often.
3. Any mechanism that causes intimal damage and leads to weakening of the medial layers of the aortic wall can result in dissection, IMH, or PAU.
4. Acute aortic dissection, the most common etiology of AAS, is characterized by an intimal tear which is often preceded by medial wall degeneration or cystic medial necrosis.
5. IMH, a variant of aortic dissection, originates from a disruption of the vasa vasorum within the media. They are treated similar to dissections and have a similar prognosis.
6. PAU is associated with atherosclerotic disease and can lead to dissection or perforation. Both IMH and PAU are found most often in the descending aorta.
7. AAS may present in many ways. Most often there is the sudden onset of severe, sharp chest pain or back pain.
8. Type A aortic dissections are associated with high mortality rates and without surgery, 30-day mortality exceeds 50%.
9. Uncomplicated type B dissections have a 30-day mortality of 10% and may be managed medically. Complications require surgical intervention or endovascular stenting.
10. CT, MRI, and TEE are all accurate in the diagnosis of AAS.
11. The initial treatment of all patients with AAS is blood pressure control in order to decrease the force of left ventricular contraction and lower the risk of dissection extension or rupture. Beta-blockers are the preferred first-line agent to achieve a systolic pressure <120 mmHg and heart rate <60 bpm.
12. Surgery is the definitive treatment of type A acute aortic pathology. The goal is to prevent life-threatening complications such as aortic rupture or pericardial tamponade.
13. Medical therapy with β-blockers and antihypertensive agents is the recommended therapy for patients with uncomplicated type B aortic dissection, IMH, or PAU.
14. The advent and incorporation of less-invasive endovascular treatments has opened up new perspectives in the treatment of acute aortic disease and continued advances will result in improved patient outcomes.

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