16.14 Diseases of the arteries 3674 16.14.1 Acute

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16.14 Diseases of the arteries CONTENTS 16.14.1 Acute aortic syndromes 3674 James D. Newton, Andrew R.J. Mitchell, and Adrian P. Banning 16.14.2 Peripheral arterial disease 3680 Janet Powell and Alun Davies 16.14.3 Cholesterol embolism 3688 Christopher Dudley 16.14.1 Acute aortic syndromes James D. Newton, Andrew R.J. Mitchell,
and Adrian P. Banning ESSENTIALS The acute aortic syndromes are acute dissection, intramural haema- toma, and penetrating ulcer, and all involve disruption of the wall of the aorta with potentially devastating consequences. Although relatively uncommon, left unrecognized and untreated they can carry a mortality rate of up to 2% per hour and 50% within the first few weeks. Clinical presentation—the pain of an acute aortic syndrome is typ- ically of instantaneous onset, cataclysmic in severity, pulsatile and tearing in quality, located either in the anterior thorax or back, and migrating if a dissection extends through the thorax. Patients usu- ally appear shocked, but blood pressure may be normal or raised and heart rate relatively slow. Physical signs typically reflect the re- gion of the aorta involved and effects of pressure on adjacent struc- tures: evidence of new aortic regurgitation or development of pulse deficits should be actively sought.

Diagnosis—abnormalities on the chest radiograph and ECG are common, but neither investigation is diagnostic and further imaging is always necessary by MRI, contrast-enhanced CT, or transoesophageal echocardiography, depending on local availability and the clinical condition of the patient. Management—every patient with a clinical suspicion of an acute aortic syndrome should receive effective pain relief and antihypertensive medication (intravenous labetalol or esmolol), aiming to maintain systolic blood pressure less than 120 mm Hg. For confirmed intramural haematoma or dissection of the as- cending aorta (type A), emergency surgery is indicated. Penetrating ulcers can be treated with endovascular stenting. When the as- cending aorta is spared (type B), aggressive control of blood pres- sure is the usual initial management, with surgery being considered if there is evidence of further progression of dissection or ischaemic complications. In the long term, strenuous efforts to control blood pressure are indicated for all patients who have survived aortic dis- section, with repeat imaging at least once a year.
Introduction An acute aortic syndrome should be considered, even if only briefly, in the differential diagnosis of any patient complaining of acute chest pain and other symptoms (Box 16.14.1.1). A careful history and physical examination will often secure the diagnosis, which is then confirmed by appropriate noninvasive investigations. The consequences of missing an acute aortic syndrome can be disas- trous: when managing a patient with acute chest pain it is always prudent for clinicians to ask themselves ‘could this be an acute aortic syndrome?’ to ensure it features in the differential diagnosis before any antiplatelet or anticoagulant agents are administered. The three mechanisms of acute aortic syndromes are acute aortic dissection, acute intramural haematoma, and penetrating ulcer of the aortic wall (Fig. 16.14.1.1). Box 16.14.1.1 Symptoms that warrant consideration of an acute aortic syndrome • Chest pain • Syncope • Thoracic back pain • Neurological, mesenteric, or limb ischaemia • Abdominal pain • Symptoms of pericardial tamponade

16.14.1 Acute aortic syndromes 3675 Pathogenesis The aortic wall is composed of three layers: a thin intimal lining, a thicker medial layer (largely composed of elastin fibres that provide strength), and a thinner adventitial outer layer from which small blood vessels (the vasa vasorum) arise to nourish the outer layers of the media. Acute aortic dissection occurs when a breach in the integrity of the intima allows blood at high pressure to penetrate through and into the media. Through this tear, pulsatile blood flow can then propagate distally, parallel to the lumen, often spiralling and split- ting the arterial wall into an inner (intima–medial) and outer layer (media–adventitial). This process of tearing within the wall results in the formation of a false lumen, parallel to the original true lumen, and commonly of a similar or larger size (Fig. 16.14.1.2). Further communication between the lumens (or re-entry tears) can occur and may reduce the pressure within the false lumen, thus limiting propagation of the dissection. However, the process often extends along the entire length of the aorta to the common iliac ar- teries, threatening the origins of branch vessels that may be avulsed or narrowed by the mass effect of the false lumen, and leading to ischaemia in the dependent vascular territories. When dissection extends retrogradely towards the heart it can cause occlusion of a coronary artery and distortion of the aortic valve, resulting in acute aortic regurgitation. Dissection may also rupture into the pericar- dial space, causing cardiac tamponade. The weakened aortic wall can rupture at any point along its length; this is usually fatal. Acute intramural haematoma was described by pathologists in 1920. It usually occurs when the small arterioles that run in the outer media of the aorta (the vasa vasorum) rupture and bleed, rarely it can occur following trauma. It is a medial/adventitial event, with the intima remaining intact, and there is no false lumen (Fig. 16.14.1.3). The clinical presentation is very similar to that of acute

aortic dissec- tion, with thoracic pain being the commonest presenting symptom. The diagnosis can only be made by exclusion of an intimal tear or a penetrating atherosclerotic ulcer. The intramural haematoma is not readily identifiable by aortography; but using noninvasive imaging, a circular or crescentic thickening of the aortic wall of more than 7 mm in depth associated with central displacement of any intimal calcification supports the diagnosis. There is increasing evidence that spontaneous intramural haematoma may be a precursor of aortic dissection. Clinical studies have supported this assertion: des- pite aggressive blood pressure control, up to 50% of patients with an intramural haematoma develop dissection or aortic rupture. Surgery is generally indicated when the ascending aorta is involved. Penetrating atherosclerotic ulcer presents with similar symptoms to aortic dissection, usually in elderly patients with disseminated atheroma. Intimal disruption caused by atheroma results in perfor- ation and secondary haemorrhage into the media. Imaging demon- strates an out-pouching from the lumen into the aortic wall with localized haemorrhage and evidence of diffuse atheroma. Rarely, this can cause a localized dissection, but the main threat is the (b) (a) Penetrating aortic ulcer Spontaneous intramural haematoma Thrombus L Mural haemorrhage Intact intima Disrupted intima Medial haemorrhage Fig. 16.14.1.1 Mechanism of acute thoracic aortic syndromes: (a) spontaneous intramural haematoma; (b) penetrating atherosclerotic ulcer. Dissection flap true false Fig. 16.14.1.2 Transoesophageal echocardiography of the descending aorta showing a dissection flap separating the true and false lumens. Haematoma Aorta Fig. 16.14.1.3 Transthoracic echocardiography of the ascending aorta. The aorta is dilated and there is a posterior intramural haematoma.

section 16 Cardiovascular disorders 3676 high incidence of rupture. Pseudoaneurysm formation can occur (Fig. 16.14.1.4). Treatment is usually with endovascular stenting to cover the ulcer, or with high-risk surgery. Classification The commonest sites for thoracic aortic dissection to originate are in the ascending aorta, just above the sinuses of the aortic valve, and in the upper descending aorta just beyond the origin of the left sub- clavian artery. The Stanford group proposed a classification that is directly linked to patient management (Fig. 16.14.1.5). Aortic dis- section that involves the ascending thoracic aorta is classified as type A and demands consideration of immediate surgery, whereas dissection that spares the ascending aorta is classified as type B and initial management is usually medical. Aetiology The most common predisposing risk factor (70% of patients) for aortic dissection is hypertension. Although the processes involved in the initiation of dissection remain incompletely understood, medial haemorrhage from rupture of vasa vasorum appears to be important. When this process is self-limiting and there is no ex- pansion of the resultant haematoma by recurrent bleeding, healing may occur with reabsorption of the haemorrhage. Alternatively, and particularly when the bleeding is extensive or recurrent, a large intramural haematoma may form around the circumference of the aorta. This alters the distribution of tensile stresses within the aorta, with much of the redistributed stress affecting the intima/endothe- lium overlying the mass. An intimal tear may then result in split- ting and separation of the media, propagation of a false lumen, and dissection. Specific risk factors Patients with Marfan syndrome (see Chapter 16.11) may present with aortic dissection or aortic root dilatation and aortic regurgi- tation (Fig. 16.14.1.6). Abnormal fibrillin within the aortic media results in intimal instability, particularly when aortic dilation leads to increased wall stress. Although the absolute risk of dissection rises with increasing size of the ascending aorta, it is important to remember that all patients with Marfan syndrome are at risk, particularly when there is a family history of aortic dissection (Box 16.14.1.2). Loeys–Dietz syndrome is an autosomal dom- inant connective tissue disorder caused by genetic mutations Pseudoaneurysm Fig. 16.14.1.4

Emergency aortography during endovascular closure of a pseudoaneurysm occurring due to a penetrating atherosclerotic ulcer. aortic regurgitation dilated aorta Fig. 16.14.1.6 Transthoracic echocardiography in a patient with Marfan syndrome. The aortic root is significantly dilated with a central jet of aortic regurgitation. Type A Type B Fig. 16.14.1.5 The Stanford classification of aortic dissection. Type A dissection involves the ascending aorta, irrespective of the distal extent of dissection.

16.14.1 Acute aortic syndromes 3677 in transforming growth factor-β and leads to aortic dilation at a younger age than the Marfan syndrome. Patients with Ehlers– Danlos syndrome are also at risk of spontaneous dissection, not only of the aorta but of its principal branches, including the cor- onary arteries. Patients with coarctation of the aorta and those with bicuspid aortic valves also appear to be at increased risk of dissection, pos- sibly related to defects in aortic wall composition. Dissection may also occur in patients with Turner's syndrome, Noonan's syn- drome, and in the later stages of pregnancy, particularly in pa- tients with Marfan syndrome. In high-risk patients with Marfan syndrome and a dilated aorta (or a family history of dissection) deferring pregnancy until after elective aortic root replacement may be advisable. Clinical features Most patients present with characteristic symptoms and clinical findings, in which case the diagnosis of dissection can be made with reasonable assurance. However, a few present atypically and it is worth considering the possibility of aortic dissection in any patient who is haemodynamically unstable without satisfactory explanation. The pain of acute dissection of the aorta can be described in terms of its (1) instantaneous onset, (2) cataclysmic severity, (3) pulsatile and tearing quality, (4) location either in the anterior thorax or back, and (5) migration as it follows the course of the dissection through the thorax. Careful interrogation about the presence of these five features will usually allow differentiation from other causes of chest pain. The instant onset, tearing/pulsatile quality, and migratory pattern contrast with the pain of cardiac ischaemia, which is usu- ally gradual in onset (over minutes), tight or crushing, and more unchanging in its distribution in the anterior chest. Syncope shortly after the onset of typical pain is not common, but is another char- acteristic presentation of dissection, often caused by rupture of the false lumen into the pericardial cavity. Other uncommon modes of presentation include stroke and limb ischaemia, with or without pain, and very occasionally congestive heart failure resulting from severe aortic regurgitation. Although patients with dissection usually appear shocked, their blood pressure may be normal or raised and their heart rate rela- tively slow. The distribution of the abnormalities detected by phys- ical examination usually reflect the region of the aorta involved in the dissection and pressure on adjacent structures. Signs of aortic regurgitation or tamponade are likely to be found in a patient with dissection involving the ascending aorta, whereas absent upper limb pulses and cerebral abnormalities suggest involvement of the aortic arch. Expansion of the arch may compress venous return and cause engorgement of one or both jugular veins. Similarly, hoarseness and Horner's syndrome can follow pressure on the left recurrent laryn- geal nerve and superior cervical ganglion, respectively. Tenderness over a carotid artery may be due to dissection extending up the ar- tery from the arch. Involvement of the descending aorta can result in visceral and lower limb ischaemia. Although traditional teaching emphasizes the relevance of blood pressure discrepancy between the arms, this is not a particularly sen- sitive sign, particularly when dissection spares the ascending aorta and arch. However, evidence of new aortic regurgitation or develop- ment of pulse deficits are specific signs of dissection and should be actively sought by the examining physician. Clinical investigation Abnormalities of the chest radiograph and electrocardiogram (ECG) are common in patients with dissection, but neither investi- gation is diagnostic and further imaging is always

necessary. Chest radiograph Potential abnormalities on the chest radiograph include a widened aortic contour, aortic kinking, tracheal deviation, left pleural effu- sion, and a widened mediastinum (Fig. 16.14.1.7). The ‘calcium sign’ is medial displacement of the calcium in the aortic knuckle by more than 6 mm and occurs in 20% of cases. The chest radiograph is normal in 10% of patients with acute aortic dissection. Urgent portable anterior–posterior chest radiographs are often of insuffi- cient quality to comment on the mediastinal contours and cannot be relied upon. Box 16.14.1.2 High-risk conditions for acute aortic syndromes • Marfan syndrome • Loeys–Dietz syndrome • Bicuspid aortic valve • Recent aortic manipulation • Prior aortic dissection • Known thoracic aortic aneurysm • Family history of aortic dissection or thoracic aneurysm • Hypertension Fig. 16.14.1.7 Chest radiograph in aortic dissection showing mediastinal enlargement.

section 16 Cardiovascular disorders 3678 ECG Nonspecific ST-segment and T-wave changes on the ECG are often found, as are changes of left ventricular hypertrophy related to pre- vious hypertension. The ECG is normal in one-third of patients. Actual involvement of a coronary artery is relatively uncommon, presentation is usually with features of right coronary occlusion since involvement of the left main stem is usually rapidly fatal. An atypical distribution of ST-elevation changes (i.e. generalized acute changes, affecting the anterior and inferior leads) is well recog- nized and should always alert the physician to the possibility of a diagnosis other than acute myocardial infarction and thereby re- duce the possibility of inadvertent administration of thrombolytic treatment. Blood tests The diagnosis of aortic dissection should not be delayed while the results of blood tests are awaited. Immunoassay of serum smooth muscle myosin heavy chains has a high sensitivity and specificity for the diagnosis of aortic dissection, but is not used in routine clin- ical practice. Cardiac enzymes are usually normal, but an elevated cardiac troponin on admission is a marker for a worse in-hospital outcome. If there is haemolysis of blood in the false lumen, lactate dehydrogenase may be elevated. Haemoglobin may be reduced if there has been significant leakage of blood from the aorta. A mildly raised leucocyte count, and raised C-reactive protein are common. D-dimer is often elevated in dissection but is a nonspecific finding. A normal D-dimer test has been used in low risk patients to identify those unlikely to benefit from further aortic imaging. Key imaging studies The priorities when imaging a patient with suspected dissection are to confirm the diagnosis and to decide if the ascending aorta is involved (Stanford type A), as this will determine whether emer- gency surgery is required. The surgeon wants to know the entry site of the dissection, if the aortic valve is competent, if there is a pericardial effusion or tamponade, and if there is involvement of the coronary arteries. Several diagnostic techniques are available (Table 16.14.1.1). Historically, aortography was the investigation of choice, but it has several disadvantages. These include delay during the as- sembly of the catheter laboratory team, the risk of aortic rup- ture during catheter manipulation, and the nephrotoxicity of radiological contrast media when renal function may already be compromised by hypotension or renal artery involvement. CT, MRI, and echocardiography all have proven advantages over aortography. However, with the advent of primary percutaneous coronary intervention some patients do present to the cardiac catheter laboratory with a diagnosis of acute coronary syndrome. Aortic dissection should always be considered in those in whom coronary angiography is normal and further imaging performed if appropriate. Contrast-enhanced CT is noninvasive, but requires the use of radiological contrast medium. In sensitivity and specificity, it is at least equivalent to aortography, but its accuracy is inferior to MRI, although this has been improved by the use of newer multislice CT scanners. MRI is noninvasive and provides excellent images of the whole aorta. Its sensitivity and specificity for dissection are up to 100% in some series, and the

addition of cardiac gated and cine techniques can give information on luminal blood flow and valvular regurgitation (Fig. 16.14.1.8). MRI is therefore the investigation of choice for most diseases affecting the aorta, but it has several limita- tions in patients with suspected acute dissection of the aorta. These include the requirement for patient transfer to the scanner, with at- tendant delays, restricted access to the patient during scanning, and the high degree of patient cooperation required to obtain artefact- free images. The limited sensitivity and specificity of transthoracic echocardi- ography mean that it cannot be used to exclude aortic dissection. However, in some cases dissection of the ascending aorta can be confidently diagnosed using parasternal and suprasternal imaging, mandating urgent transfer to a surgical centre where additional in- formation can be obtained by transoesophageal echocardiography in the anaesthetic room. Transoesophageal echocardiography pro- vides detailed anatomical information about the morphology of a dissection and can also demonstrate the consequences of proximal extension, including the presence of aortic regurgitation, pericardial effusion, and involvement of the coronary artery ostia, thus making complementary investigations such as angiography unnecessary (Fig. 16.14.1.9). Table 16.14.1.1 Sensitivity and specificity of investigations for the diagnosis of aortic dissection Investigation Sensitivity (%) Specificity (%) MRI 99–100 99–100 CT 96–100 96–100 Transoesophageal echocardiography 98 95 Transthoracic echocardiography 59–85 63–96 Aortography 77–88 94 Fig. 16.14.1.8 MRI of the chest. A dissection flap in the descending aorta and a left-sided pleural effusion (large arrow) are visible.

16.14.1 Acute aortic syndromes 3679 Management Emergency management Lowering systolic blood pressure and limiting shear stress re- duces the likelihood of progression of dissection. Every patient with a clinical suspicion of dissection should therefore receive ef- fective pain relief (intravenous morphine is usually required) and antihypertensive medication pending a definitive diagnosis by imaging. Patients should be cared for in a high-dependency area with continuous monitoring of the ECG and regular blood pres- sure and urine output measurement. Ideally, systolic blood pres- sure should be maintained below 110 mm Hg and heart rate to less than 60 bpm, using intravenous labetalol (initial dose 50 mg bolus followed by 1–2 mg/min) or intravenous esmolol. Both these agents produce a rapid and titratable reduction in blood pressure, with β-blockade particularly appropriate in this context because it reduces the force of cardiac contraction and the rate of rise of the arterial pressure (dP/dt). If blood pressure control remains suboptimal, an additional infusion of sodium nitroprusside may be used (0.5–8 micrograms/kg per min). Intravenous nitrates and oral calcium antagonist are alternatives in patients who are in- tolerant of β-blockers. Patients presenting with or developing cardiogenic shock should undergo immediate echocardiography for investigation of pericardial tamponade. Emergency surgery is the treatment of choice, as peri- cardiocentesis can accelerate bleeding and is usually ineffective. The optimal management of patients with aortic dissection re- quires close liaison between those who admit patients as medical emergencies and cardiac surgical centres, using local guidelines for investigation that should reflect the available expertise and surgical opinion. Patients with a low clinical index of suspicion of dissection who are in a stable cardiovascular state should undergo prompt in- vestigation in their local hospital, using a nominated noninvasive technique—usually CT scanning. Unless noninvasive imaging is available immediately, unstable patients with a high clinical index of suspicion should receive medical treatment and be transferred immediately to a surgical centre for both diagnostic imaging and management. This approach minimizes delay, a critical aspect of the management of acute aortic dissection. Surgery When the dissection involves the ascending aorta (type A), imme- diate surgery is required as there is a high risk of proximal exten- sion causing dissection of the coronary arteries, incompetence of the aortic valve, and

rupture into the pericardium. Surgery usually involves excision of the intimal tear in the ascending aorta and interposition of a Dacron graft. This procedure protects the lower ascending aorta and valve from progressive dissection and prevents distal extension by reducing pressure within the false lumen. The false lumen may subsequently thrombose, or—in cases with mul- tiple intimal tears—may remain patent but decompressed. Replacement of the aortic valve is usually performed only when resuspension of the valve is not possible. However, in patients with Marfan syndrome the ascending aorta and valve are usually replaced with a composite graft to prevent subsequent annular dilatation. In cases where dissection extends into the aortic arch, some surgeons advocate that the arch and great vessels should be included in the initial repair as arch involvement is a strong predictor of a require- ment for repeat surgery. However, extended surgery can increase the duration of the operation and the risk of damage to the central ner- vous system, hence inclusion of the arch in dissection repair is gen- erally restricted to expert centres. Spinal cord damage and paraplegia is a common complication of aortic dissection repair, resulting from cross-clamping of the aorta. Techniques to improve distal aortic perfusion can reduce the inci- dence of this complication to less than 5%. The overall operative mortality for surgical repair is between 10 and 20%. Further management of descending aortic dissection Proximal extension towards the heart is less likely when the dissec- tion begins distal to the left subclavian artery (type B). These patients (a) (b) Dissection flap Dissection flap aorta RCA Fig. 16.14.1.9 Transoesophageal echocardiography at the level of the aortic valve: (a) view along the aorta; (b) cross-sectional view. There is a large dissection flap in the ascending aorta (type A) that nearly involves the ostium of the right coronary artery (RCA).

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