Acute aortic syndrome

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Abstract

The concept of acute aortic syndrome (AAS) facilitates the early identification of patients with chest pain caused by an aortic condition, and expedites risk stratification and definitive treatment. Important differential diagnoses are acute coronary syndrome, pulmonary embolism and chest pain from trauma. Individuals with AAS should be evaluated using a combination of blood tests, electrocardiography and imaging, such as computed tomography CT or echocardiography initially, and magnetic resonance imaging at a later stage. Classic aortic dissection is the most common form of AAS. Risk factors include congenital factors such as hereditary connective tissue disorders, bicuspid aortic valve with aortopathy and, most importantly, acquired conditions such as untreated hypertension. Patients with evidence of rupture of one aortic layer, malperfusion syndrome or end-organ ischaemia require urgent intervention; in cases of proximal dissection including the arch this is typically open surgery, and in distal or type B dissection it is usually endovascular stent-graft placement with ancillary procedures. The optimal management of patients with AAS is challenging and requires a multidisciplinary team approach. Further studies are required to fully characterize conditions within the AAS spectrum and to offer an individualized, patient-centred treatment.

Keywords Aortic dissection; emergency; endovascular intervention; false lumen; intramural hematoma; stent-graft; stenting

Key points

- The incidence of acute aortic syndromes (AASs) is increasing
- Management of AAS should be based on low-threshold CT imaging
- Endovascular technology is encouraging in various types of aortic dissection
- Overall management of AAS should involve a multispecialty aortic team

Introduction

Acute aortic syndrome (AAS) encompasses acute aortic dissection, intramural haematoma and penetrating aortic ulcer and is a life-threatening condition of the aorta (Figure 1).[1] The main risk factors are systemic hypertension, early onset atherosclerosis, previous cardiac surgery, existing aortic aneurysm and a family history. AAS can also be associated with congenital cardiovascular defects such as bicuspid aortic valve with aortopathy, aortic coarctation and sole annulo-aortic ectasia, and hereditary genetic syndromes such as Marfan syndrome (*FBN1* mutations), Loeys–Dietz syndrome (*TGFBR1* or *TGFBR2* and *SMAD3* mutations), vascular Ehlers–Danlos syndrome (*COL3A1* mutations), and Turner syndrome (45, XO karyotype).[2,3]

Differential diagnoses

The most prevalent differential diagnoses are acute coronary syndrome (ACS) and pulmonary embolism, as well as pneumothorax, oesophageal perforation and trauma to the chest. If the diagnosis of an AAS is overlooked, a patient with aortic dissection may be given antithrombotic or anticoagulant therapy to treat suspected myocardial ischaemia or pulmonary embolism, and lose precious time to life-saving surgical repair; in the context of rupture, this can have devastating consequences. Patients with suspected AAS should be evaluated swiftly by a combination of blood tests, electrocardiography and early imaging.[1] The availability of non-invasive imaging (e.g. contrast-enhanced computed tomography (CT), echocardiography (transthoracic, transoesophageal)) in hospital emergency departments has enabled the confident and early diagnosis of even subtle forms of AAS.

Diagnostic tools

Electrocardiograph-gated CT is the most commonly used imaging modality owing to the fast acquisition of high spatial resolution images and should be used at a low threshold when clinically suspected, or clear-cut acute coronary syndrome has been ruled out. Unenhanced scans should be included to provide information regarding the presence of intramural haematoma, mediastinal haemorrhage, or haemorrhagic pericardial effusion and calcification.[4] A triple rule-out CT protocol including contrast agent to be given can ideally be used to differentiate between different causes of chest pain, namely AAS, ACS and pulmonary embolism.[5]

Echocardiography has the advantage of being portable and can be performed at the bedside for unstable patients in an emergency setting. Transthoracic echocardiography is useful for identifying aortic valve dysfunction, proximal aortic dissection

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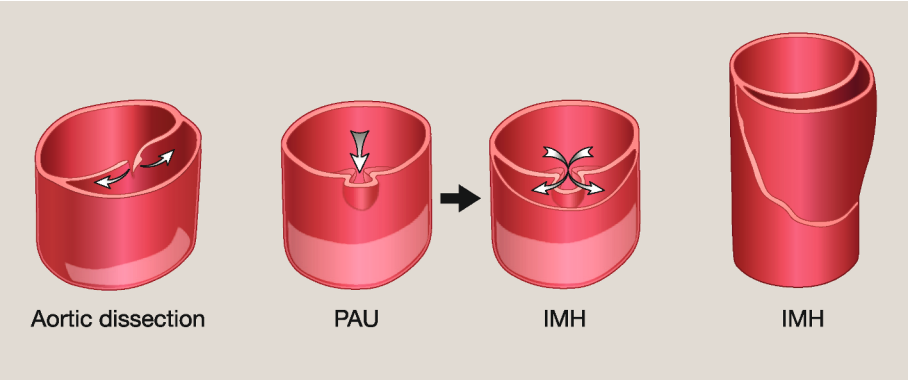

Figure 1 Schematic of aortic dissection (left), penetrating ulcer (PAU; middle), and intramural haematoma (IMH; right), all causing acute aortic syndrome. Reproduced from Nienaber CA, Powell JT. Management of acute aortic syndromes. *Eur Heart J* 2012; **33:** 26–35b with permission.

extending into the aortic root, pericardial tamponade and wall motion abnormalities at the left ventricle. Transoesophageal echocardiography is instrumental intraoperatively to confirm whether endovascular devices are located in the true or false aortic lumen, and to identify aortic side branches.

In aortic dissection, a dual-lumen aorta is usually formed, with the original aortic lumen defined as the true lumen and the new channel termed the false lumen. The estimated hospital-based incidence is 2.6–3.5 cases per 100,000 person–years; however, the real community-based incidence is higher – up to 30 per 100,000 at the age of 60–70 years. The hospital-based incidence underestimates the real incidence as hospital-based reports do not account for pre-admission deaths. The average age at presentation is 65 years and approximately 65% of patients are male.

Classification systems of aortic dissection

Aortic dissection is classified according to either the location of the entry tear or the part of the aorta affected by dissection, regardless of the position of tear using the DeBakey or Stanford system. This anatomical classification drives decisions regarding surgical and non-surgical management. (Figure 2). The Penn Abc classes and DISSECT are newer classification systems, focusing on acuity and complexity, but are less popular. Similar to Penn Abc, the newer TEM (type, entry, malperfusion) classification focuses on the location and evolution of complications in both type A and B aortic dissection.

The DISSECT system is a mnemonic-based classification system designed to divide cases into subsets according to anatomical involvement with relevance to endovascular management. Type B aortic dissection is traditionally divided into acute and chronic phases with reference to symptom onset: acute is within 2 weeks and chronic is in >2 weeks. This classification has more recently been revised to acute (<15 days), subacute (15–92 days) and chronic (≥93 days).

Complications of dissection lead to increased mortality owing to end-organ or limb malperfusion, impending aortic rupture, refractory hypertension, persistent pain, expansion >1 cm/year or an overall aortic diameter of >4.5 cm. Continued patency, rather than complete thrombosis of the false lumen, is associated with aortic dilatation during follow-up and poorer outcomes. More than 60% of deaths associated with acute type B aortic dissection result from the late rupture of an expanding false lumen.

In-hospital management

Patients with suspected/confirmed AAS should be admitted to an intensive care or high-dependency unit before definitive therapy. A systolic blood pressure of 100–120 mmHg and a heart rate of 60–80 beats per minute should be established while maintaining sufficient end-organ perfusion. Intravenous β-adrenoceptor blockers such as labetalol or metoprolol are recommended as first-line medical therapy, although multiple antihypertensive agents are often required.

The ideal imaging modality is CT angiography, which clearly delineates all the relevant information on the extent of dissection, location of the tear or rupture, pericardial or pleural effusion, and status of the true and false lumens. The location of all entry tears and the origins of all major branches should be identified to enable the dissection to be classified and the extent of surgical or endovascular repair planned. The condition of acute aortic dissection can be difficult to manage and suspicious images can be shared with a specialized aortic centre to ensure the best treatment. If it is appropriate, the patient should then be transferred at the earliest opportunity for best surgical or interventional management.

Dissection of the descending aorta is less frequently lethal early on than dissection of the proximal ascending aorta: medical management of uncomplicated type B dissection results in approximately 90% survival to hospital discharge at 7–10 days. These patients should be followed closely (regardless of the initial treatment) as they may require and benefit from deferred endovascular stenting within 3 months of onset. In complicated cases with malperfusion early endovascular repair is often indicated and can still result in an in-hospital mortality of 9% and a major complication rate of 7% (including stroke 3.1%, paraplegia 1.9%, conversion to type A dissection 2%, bowel infarction 0.9%, and major amputation 0.2%). Open repair is now rarely indicated for the management of type B aortic dissection because of the excellent results achieved with endovascular interventions with lower peri-interventional risks (Figure 3).

Nevertheless, optimal treatment of patients with AAS is challenging and requires a multidisciplinary team approach.

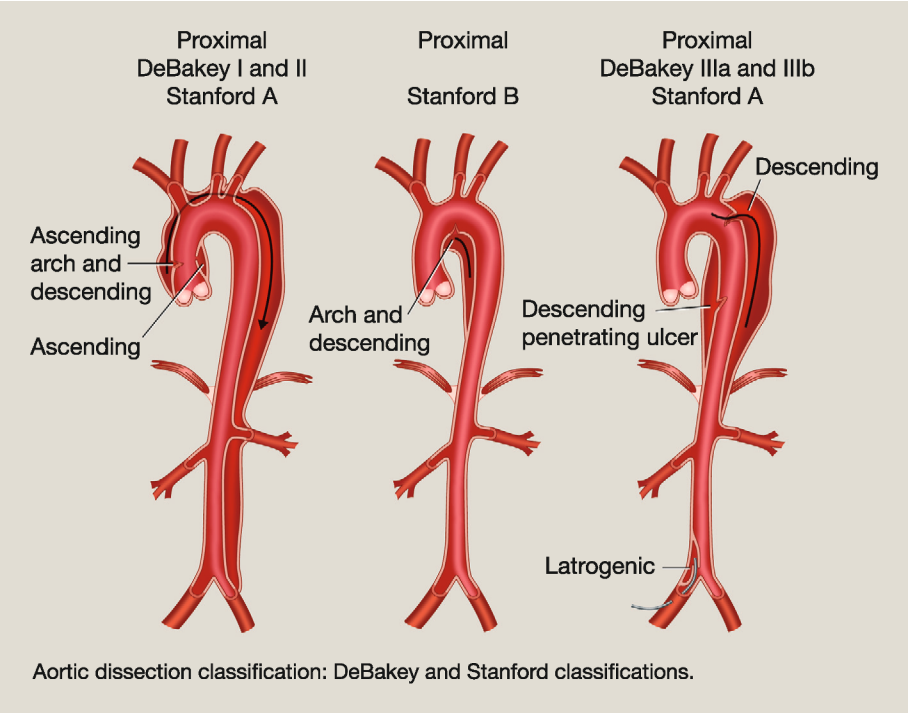

Figure 2 Reproduced from Nienaber CA, Kische S, Ince H, Fattori R. Thoracic endovascular aneurysm repair for complicated type B aortic dissection. *J Vasc Surg* 2011; **54**(5): 1529–1533 with permission.

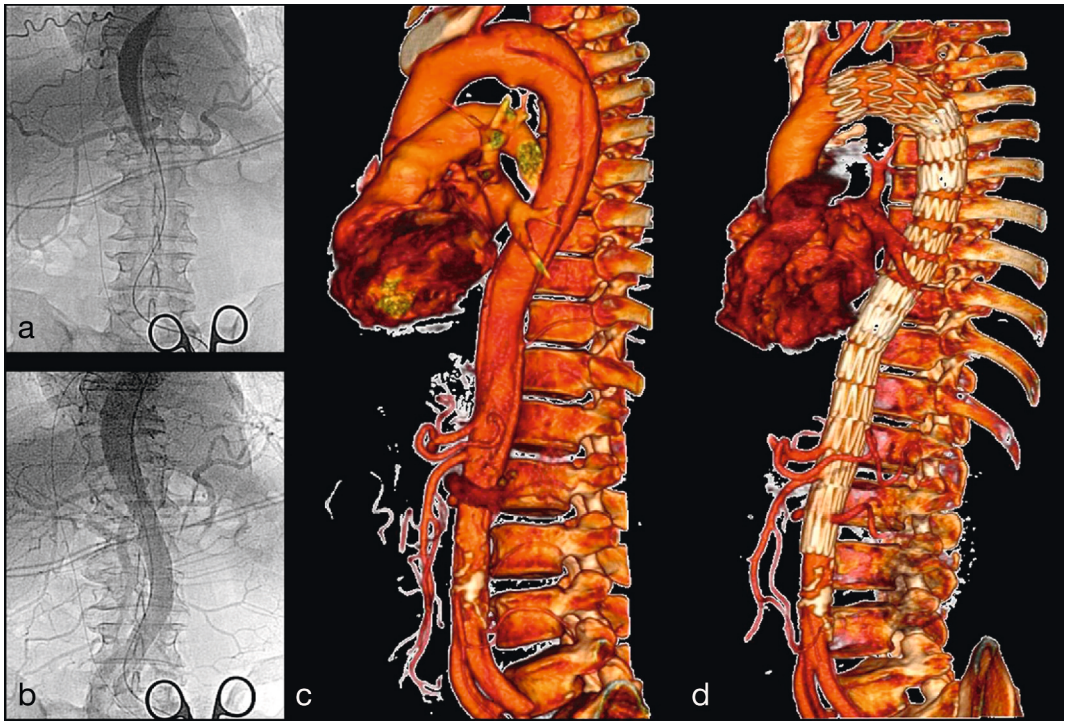

Figure 3 Malperfusion syndrome treated with an endovascular stent-graft and PETTICOAT. (**a**) Angiography of lower body malperfusion; (**b**) reperfusion after a proximal stent-graft insertion; (**c**) 3D CT reconstruction of an acute complicated dissection with malperfusion; and (**d**) a reconstructed aorta and abolished malperfusion after stent-graft and PETTICOAT treatment. Reproduced from Nienaber CA, Kische S, Ince H, Fattori R. Thoracic endovascular aneurysm repair for complicated type B aortic dissection. *J Vasc Surg* 2011; **54**(5): 1529–1533 with permission.

Further studies are required to fully characterize conditions within the AAS spectrum and to offer early affected patients an individualized, patient-centred treatment plan. ◆

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