

## CLINICAL REVIEW

# The diagnosis and management of aortic dissection

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Aortic dissection is caused by an intimal and medial tear in the aorta with propagation of a false lumen within the aortic media. It is part of the “acute aortic syndrome”—an umbrella term for aortic dissection, intramural haematoma, and symptomatic aortic ulcer (table 1).<sup>1</sup> Acute dissection is the most common aortic emergency, with an annual incidence of 3–4 per 100 000 in the United Kingdom and United States, which exceeds that of ruptured aneurysm.<sup>2 w1 w2</sup> The prognosis is grave, with 20% preadmission mortality and 30% in-hospital mortality.<sup>2</sup>

The best treatment depends on the anatomical and temporal classification of the disease. Aortic dissection is therefore categorised according to the site of the entry tear and the time between the onset of symptoms and diagnosis. A dissection is considered “acute” when the diagnosis is made within 14 days of onset, and thereafter it is termed “chronic.” The location of the entry tear plays a key role in treatment and outcome, and it is classified by being in the ascending aorta (Stanford type A dissection) or distal to the origin of the left subclavian artery (Stanford type B dissection) (fig 1).<sup>3</sup>

Type A dissection carries a far worse prognosis than type B dissection and urgent surgical intervention is often needed. By contrast, acute type B dissection is usually managed conservatively if uncomplicated and surgically if complicated.

We review the epidemiology, diagnosis, and management of aortic dissection drawing on evidence from population studies, randomised controlled trials, meta-analyses, and published guidelines.

### Who is at risk?

The causes of aortic dissection are multifactorial, and both inherited susceptibility and acquired degenerative disease have been implicated. Several modifiable and non-modifiable risk factors are recognised, the most important of which are discussed below.

### Hypertension

Systemic hypertension is one of the most important risk factors for aortic dissection and is present in 40–75% of patients

presenting with the condition.<sup>3</sup> Systolic hypertension exacerbates the differential haemodynamic forces acting on the relatively mobile aortic arch and the relatively fixed ascending and descending thoracic aorta. A cohort study of 175 patients identified physical exertion or emotional stress as the direct predecessor of acute pain in 66% of acute dissections, primarily as a result of acute changes in blood pressure during the event.<sup>w3</sup>

### Race and sex

A cross sectional study of 951 patients by the International Registry of Acute Aortic Dissection, comprising data from 12 international referral centres, showed that 68% of all patients presenting with the condition were male and 79% were white.<sup>5</sup>

### Connective tissue diseases

Various connective tissue diseases predispose to the inherent weakening of the aortic wall and subsequent dissection, and these diseases are especially important in patients under 40 years. These include Marfan’s syndrome with fibrillin defects, which is seen in 15–50% of patients under 40 years<sup>5 w4</sup>; Ehlers-Danlos type IV with abnormal synthesis of type III procollagen<sup>w5</sup>; and other connective tissue disorders associated with cystic medial necrosis.<sup>w6</sup>

### Congenital cardiovascular abnormalities

A cross sectional study described a fivefold to 18-fold increased risk of dissection in 516 patients with bicuspid aortic valves.<sup>5</sup> This increased risk was attributed to a coinherited developmental defect of the proximal aorta, which conferred a predilection towards apoptosis of the cellular components of the aortic media, and subsequent medial weakening and aortic dilation. The presence of a bicuspid aortic valve was also associated with dissection in a greater proportion of patients under 40 years (9% under 40 v 1% over 40; P<0.001). A prospective study of 631 patients from the adult congenital heart disease database showed that the coexistence of coarctation of the aorta with a bicuspid aortic valve significantly increases the risk of acute aortic complications such as dissection (odds ratio 4.7, 95% confidence

**Summary points**

- Aortic dissection is diagnosed and managed according to its anatomical extent and chronicity
- White men aged over 40 years with hypertension, or those under 40 with Marfan's syndrome or bicuspid aortic valves, are at highest risk
- Patients often present with acute onset sharp chest pain, sometimes with loss of consciousness or poor perfusion of end organs
- Computed tomography aortography is the first line diagnostic investigation, followed by transoesophageal echocardiography; magnetic resonance angiography is preferred for surveillance
- Manage proximal (type A) dissection surgically if possible
- Uncomplicated distal (type B) dissection is best managed with intensive drug treatment; complicated type B dissection requires surgical intervention
- All patients need lifelong antihypertensive therapy and surveillance imaging

**Sources and selection criteria**

We searched the Medline, Embase, Web of Science, and Cochrane databases for "aortic dissection" and used reference lists to identify key studies. Two authors independently performed the searches and mutual consensus was reached. Because of the lack of large well designed randomised controlled trials, we gave priority to systematic reviews, meta-analyses, and studies from the International Registry of Acute Aortic Dissection.

interval 1.5 to 15; P=0.01); this has been attributed largely to age, sex, aortic valve dysfunction, and the hypertension associated with coarctation.<sup>w7</sup> Several familial aneurysmal syndromes (such as congenital contractual arachnodactyly, familial thoracic aortic aneurysm or Erdheim's cystic medial necrosis, familial aortic dissection, familial ectopia lentis, and familial Marfan-like habitus) also predispose to aortic dissection.<sup>6 w8</sup>

**Miscellaneous risk factors**

Prevalence studies have shown that aortic vasculitic disease,<sup>w9</sup> cocaine misuse,<sup>w10</sup> and pregnancy<sup>w7</sup> are risk factors for aortic dissection. One report on 723 patients found a 5% rate of iatrogenic aortic dissection after cardiac interventions, including percutaneous revascularisation and coronary artery bypass grafting.<sup>w11</sup>

Although British national statistics show that dissection affects all ages (27% of patients aged 17-59 years, 40% aged 60-74 years,<sup>w12</sup> 33% aged >75 years), older patients (>40 years) are more likely to have concurrent hypertension or atherosclerosis, whereas younger patients are more likely to have Marfan's syndrome, a bicuspid aortic valve, or aortic intervention before presentation.<sup>5</sup>

**How do patients present?**

Patients typically present with the abrupt onset of sharp tearing or stabbing chest pain, which may improve slightly over time, although pain may be absent in 10% of patients.<sup>8</sup> Asymptomatic presentation is more common in patients with diabetes.<sup>w13-w15</sup>

The pain may radiate to the neck in type A dissection or to the interscapular area in type B aortic dissection.<sup>9</sup> Acute rupture or inadequate perfusion—depending on the site and extent of the dissection—may cause a patient to become unconscious.<sup>10</sup>

Interrupted perfusion may result in neurological deficits, symptomatic limb ischaemia, or visceral ischaemia. A cross sectional study of 617 patients with type A dissection found focal neurological deficits in 17% of patients.<sup>11</sup> One report from the International Registry of Acute Aortic Dissection showed that aortic regurgitation and pulse deficit were present in 32% and 15% of patients, respectively.<sup>w15</sup> Hypotension was seen in 25% of patients with type A dissection, whereas hypertension was typical in type B dissection.<sup>w15</sup>

Many differential diagnoses exist (box). Specific features, however, may alert clinicians to probable dissection. Consensus

guidelines from the American Heart Association describe three categories of high risk features to identify patients at greatest risk: predisposing conditions, pain features, and examination findings.<sup>12</sup> High risk predisposing conditions include Marfan's syndrome, recent aortic manipulation, or a known thoracic aneurysm. High risk pain features include an abrupt onset of ripping, tearing, or stabbing pain in the chest, back, or abdomen. High risk features of the examination include a pulse or blood pressure discrepancy, neurological deficit, a new murmur of aortic regurgitation, and shock. Urgent aortic imaging is needed in patients who have one or more high risk feature, but who present with no electrocardiographic changes of myocardial infarction and no history or examination findings that strongly suggest an alternative diagnosis. Although the specificity of this approach is unknown, a sensitivity of 95.7% has been reported.<sup>13</sup>

**How are patients initially managed?**

The emergency management of patients with suspected aortic dissection entails adequate resuscitation and optimisation for subsequent imaging and intervention. This includes ensuring adequate oxygenation and ventilation, with careful monitoring of respiratory function. Two large bore intravenous lines should be established for intravenous fluid resuscitation, with close monitoring of heart rate, heart rhythm, blood pressure, and urine output.  $\beta$  blockers may be given to reduce the rate of blood pressure changes and the shear forces on the aortic wall; aim for a heart rate of 60-80 beats/min and systolic blood pressure of 100-120 mm Hg.<sup>w16</sup> However, a careful balance must be maintained between suppressing tachycardia and hypertension and ensuring adequate end organ perfusion (by monitoring urine output; mental and neurological state; and peripheral vascular status, including the development or progression of carotid, brachial, and femoral bruits). Twelve lead electrocardiography is essential to exclude concurrent myocardial ischaemia, which would necessitate urgent discussion with cardiology colleagues about managing acute coronary syndrome in the context of a potential aortic dissection. Undertake definitive imaging and further intervention only once the patient is haemodynamically stable.

**How is aortic dissection diagnosed?**

Retrograde aortography was the gold standard for assessing patients in the 1970s and 1980s, but it has been superseded by

### A patient's perspective

I was relaxing at home one evening when I experienced sudden and extreme chest pain, which was followed by my left leg going numb. I called for an ambulance, which took me to the nearest hospital. After a computed tomography scan, I was diagnosed with a type A aortic dissection and transferred urgently to a regional vascular centre. The surgeons opened my chest and stitched a graft into the top of my aorta. Unfortunately, I had a mild stroke afterwards, which they had warned me about. I am still recovering from this but feel lucky to have survived, considering the high mortality rate.

I stayed well for a year but then developed chest pain and fever. This gradually worsened over four weeks and I was again admitted to my local hospital, where a computed tomography scan showed that I had pneumonia, pleural effusion, and now a type B dissection. They transferred me to the regional vascular centre, where the team decided that because this dissection was chronic and I was otherwise well, I did not need further intervention. I was investigated and treated for a bleeding stomach ulcer. I did well and was then discharged home but was readmitted a week later because I had recurrent chest pain and was coughing blood. A computed tomography scan found no clot in my lungs but showed that the aorta had increased in diameter from 5.9 cm to 7.9 cm. After three days in intensive care, where they controlled my blood pressure, I'm feeling better. I am still an inpatient at the regional vascular unit and am awaiting further surgery on my aorta. The surgeons have said that because of the complexity of my disease, I may be better suited to open rather than keyhole surgery.

### Differential diagnoses

#### *Patients with acute chest pain*

- Myocardial infarction
- Pulmonary embolism
- Spontaneous pneumothorax

#### *Patients with acute abdominal or back pain*

- Ureteric colic
- Perforated viscus
- Mesenteric ischaemia

#### *Patients with pulse deficit*

- Non-dissection related embolic disease

#### *Patients with focal neurological deficit*

- Stroke
- Cauda equina syndrome

cross sectional imaging, which performs better and has a better safety profile.<sup>14</sup>

Although chest radiography and electrocardiography are often ordered in the emergency care setting, these tests cannot establish or exclude the diagnosis of dissecting aortic aneurysm.<sup>w17</sup>

D-dimers are raised in aortic dissection and it has been suggested that a concentration below 500 ng/ml, which is already used to rule out pulmonary embolism, can exclude acute dissection (negative likelihood ratio of 0.07) in the first 24 hours.<sup>w18 w19</sup> However, these data were derived from a population of patients undergoing imaging for dissection in a tertiary centre. The high pre-test probability of dissection in this group limits the applicability of the study's findings, and the safety of using D-dimer testing to screen for dissection in all patients with non-coronary chest pain requires further study. Biomarkers such as smooth muscle myosin heavy chain protein have also proved to be less than useful in diagnosis.<sup>w17</sup>

Computed tomography can help the clinician rapidly confirm or exclude aortic dissection, classify its extent, and diagnose any complications. Correct categorisation of type A or type B dissection (fig 1) is imperative to plan treatment. Patients commonly need more than one non-invasive imaging test to acquire all necessary information. A cross sectional study of 464 patients reported computed tomography angiography as the initial investigation in 61% of cases, echocardiography in 33%, aortography in 4%, and magnetic resonance angiography in 2%.<sup>w15</sup>

## Computed tomography angiography

Multidetector computed tomography angiography is recommended by the European Society of Cardiology as the

first line of investigation for patients with suspected acute dissection.<sup>1</sup> This investigation can assess factors that are important in the planning of open or endovascular surgery, including the extent of dissection, the relative calibre of true and false lumens, and the involvement of aortic side branches. A meta-analysis of 1139 patients with aortic dissection found that multidetector computed tomography angiography had a sensitivity of 100%, specificity of 98%, and diagnostic odds ratio of 6.5.<sup>w20</sup> Outside the emergency setting, electrocardiogram gated computed tomography can provide dynamic information, although its spatial resolution is inferior to magnetic resonance imaging,<sup>w21</sup> which reduces its usefulness in planning complex aortic repairs. The disadvantages of computed tomography angiography include the need to use potentially nephrotoxic contrast media, exposure to ionising radiation, and inability to assess functional aortic insufficiency.

## Echocardiography

A small prospective cohort study showed that in patients presenting in shock, transthoracic echocardiography had a 78.3% sensitivity and 83.0% specificity for diagnosing proximal dissection.<sup>15</sup> However, the role of this modality is limited because it cannot accurately visualise the descending aorta in most patients, despite its ability to diagnose aortic incompetence. The combined use of transthoracic echocardiography and computed tomography is useful in the absence of multidetector computed tomography functional imaging.<sup>w22</sup> A meta-analysis of cohort studies (1139 patients) found that transoesophageal echocardiography accurately visualised the entire thoracic aorta (sensitivity 98.0%, specificity 95.0%, diagnostic odds ratio 6.1) and, despite the requirement for oesophageal intubation, can be performed at the bedside.<sup>w20</sup> Unlike static imaging, transoesophageal echocardiography detects aortic regurgitation

or pericardial effusion and can provide intraoperative assessment of operator position within the vessel lumen, although it cannot assess the abdominal aorta.<sup>9</sup> The operator dependency of transthoracic and transoesophageal echocardiography limits their accuracy and accessibility.

## Magnetic resonance angiography

A meta-analysis of diagnostic studies showed magnetic resonance angiography to have a sensitivity of 98% and specificity of 98%, with diagnostic odds ratio of 6.8, in the diagnosis of dissecting aortic aneurysm.<sup>w20</sup> Gadolinium contrast agents used in magnetic resonance angiography are less nephrotoxic than iodinated substances used for computed tomography angiography and there is no associated ionising radiation.<sup>w23</sup> Disadvantages include its limited use in patients with claustrophobia or metal devices, although it can be used in those with nitinol aortic stent grafts.<sup>1</sup> Long acquisition times and limited availability reduce its usefulness in the emergency setting, for which computed tomography angiography is ideal. Magnetic resonance angiography offers greater potential for long term surveillance of treated dissection and for the assessment of stable patients presenting with chronic dissection.<sup>1</sup>

## How is aortic dissection managed?

Owing to the paucity of evidence from randomised controlled trials, the management of aortic dissection is mainly guided by data from international registries, large series, and expert consensus.<sup>1 16 17</sup> The balance between medical and surgical management depends on the anatomical features of the lesion and its physiological sequelae.

## Type A dissection

Cross sectional studies from the International Registry of Acute Aortic Dissection have suggested that, if left untreated, proximal (Stanford type A or DeBakey type I or II) dissection carries a one week mortality of 50–91% owing to complications such as aortic rupture, stroke, visceral ischaemia, cardiac tamponade, and circulatory failure.<sup>w15</sup> Drug treatment alone results in a mortality of nearly 20% by 24 hours and 30% by 48 hours (fig 2).<sup>w15</sup> Urgent cardiac surgical consultation is therefore imperative. Surgery involves replacing the affected ascending aorta, with or without the aortic arch, with a prosthetic graft; this procedure has an in-hospital mortality of 15–35%.<sup>w24-w28</sup> A variety of techniques may be needed. For example, proximal extension of the dissection to the aortic valve or ostia of the coronary arteries may require replacement or resuspension of the aortic valve,<sup>1</sup> or coronary artery bypass.<sup>w29</sup> The International Registry of Acute Aortic Dissection reported these techniques in 24% and 15% of type A dissections, respectively.<sup>w30</sup> Together with adjunctive measures such as hypothermic circulatory arrest and perfusion of the head vessels,<sup>w31</sup> surgery for proximal aortic dissection has three year and five year survival rates of 75% (standard deviation 5%) and 73% (6%), respectively.<sup>14</sup>

## Acute type B dissection

The development of complicated dissection—defined by the presence of visceral or limb ischaemia, rupture, refractory pain, or uncontrollable hypertension—is the key factor that determines both intervention and outcome for patients with type B dissection.<sup>1 w13 w15 w17 w20 w32 w33</sup>

For uncomplicated acute type B dissection, series have shown that drug treatment alone can result in 78% three year survival after discharge from hospital.<sup>18</sup> Current guidelines deem this a

difficult benchmark to surpass,<sup>16</sup> and medical management remains the gold standard. Careful regulation of systolic blood pressure at 100–120 mm Hg is needed to minimise haemodynamic shear stress and discourage rupture.<sup>w17</sup>  $\beta$  blockers (such as propranolol and metoprolol) are first line agents. Calcium channel blockers (such as non-dihydropyridine agents) are useful in patients with chronic obstructive pulmonary disease and those who cannot tolerate  $\beta$  blockers.<sup>1 12</sup> Endovascular treatment is increasingly possible with low mortality,<sup>19</sup> and its role in uncomplicated acute type B dissection will be clarified by the results of the Acute Dissection Stent-grafting or Best Medical Treatment (ADSORB) trial (NCT00742274), which will randomise patients to best medical treatment, with and without stent grafting. Until these data are available, uncomplicated type B dissection should be medically managed. Intervention, usually endovascular repair using a stent graft, is necessary for complicated acute type B dissection.<sup>1</sup> A prospective cohort study of 159 patients reported that, if untreated, this pathology carries a mortality of 50%.<sup>w34</sup> Conventional open surgery for complicated dissection has a 30 day mortality of 30%,<sup>20</sup> whereas meta-analysis has shown that endovascular treatment has a 30 day mortality of 9.8%.<sup>21</sup> Long term postoperative surveillance is mandatory: a prospective cohort study of 125 patients suggested that complete thrombosis of the false lumen may be achieved in only 44% of cases, with 20% of dissections rupturing within five years from continual aortic expansion.<sup>w35</sup> Multiple cohort studies have shown that even after complete false lumen thrombosis, 16% of patients develop evidence of dissection in the unstenched distal aorta, which requires surgical reintervention.<sup>w35-w37</sup>

## Chronic type B dissection

Uncomplicated chronic type B dissection can be managed conservatively, but many of these patients develop complications, the foremost of which is formation of an aneurysm, which may require surgical intervention. Data on the natural course of the disease suggest that 15% of chronic type B dissections will be complicated by an aneurysm,<sup>w38</sup> and ongoing research is directed at predicting patients at high risk of this complication, so that they can be targeted for earlier intervention.<sup>22</sup>

Chronic dissection is difficult to treat. Conventional open surgery has an appreciable death rate and poses considerable physiological challenges, including the need for posterolateral thoracotomy, single lung ventilation, cardiopulmonary bypass, hypothermia, heparinisation, cerebrospinal fluid drainage, and circulatory arrest to prevent stroke and paraplegia.<sup>w39</sup> The endovascular approach is associated with less morbidity and mortality; a systematic review of 810 patients found that one year survival is higher after endovascular stenting than after open surgery (endovascular surgery 93%; open surgery 79%).<sup>23</sup> However, the long term efficacy of an endovascular approach to preventing long term aortic related death is still unclear.

## How should patients be followed up?

Ten year survival rates of patients who are discharged from hospital range from 30% to 60%.<sup>24 w25-w28 w40</sup> The underlying pathophysiology of aortic medial disease and defective wall structure confers an ongoing risk of further dissection, aneurysmal degeneration, and rupture.<sup>13</sup> A prospective cohort study of 721 patients found this risk to be higher in women and that annual mortality was 12% once the aortic diameter exceeded 6 cm.<sup>w41</sup> Consequently, the European Society of Cardiology recommends regular cross sectional imaging of the aorta,

preferably with magnetic resonance angiography, at one, three, and 12 months after discharge and every six to 12 months thereafter, depending on aortic size.<sup>1</sup> Various experts also advocate the combined use of echocardiography with axial imaging for routine surveillance.<sup>w42</sup> All patients should receive lifelong antihypertensive treatment, including β blockers, with a target blood pressure of 120/80 mm Hg.<sup>25-27</sup>

The sequelae of endovascular and open repair also merit surveillance. A small prospective cohort study reported that reintervention was needed in 27.5% of patients after open repair because of extension or recurrence of dissection, formation of a localised aneurysm remote from the original repair, graft dehiscence, aortic regurgitation, or infection.<sup>28</sup> A systematic review of the mid-term outcomes of endovascular treatment found high rates of reintervention for late morbidities, such as endoleak (8.1%), formation of a distal aneurysm (7.8%), and rupture (3.0%), thereby justifying mandatory postoperative surveillance.<sup>29</sup>

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**Competing interests:** All authors have completed the ICMJE uniform disclosure form at [www.icmje.org/coi\\_disclosure.pdf](http://www.icmje.org/coi_disclosure.pdf) (available on request from the corresponding author) and declare: no support from any organisation for the submitted work; no financial relationships with any organisations that might have an interest in the submitted work in the previous three years; no other relationships or activities that could appear to have influenced the submitted work.

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**Unanswered questions and ongoing research**

Which patients with uncomplicated type B dissection might benefit most from intervention? Research currently centres on defining a subgroup at greatest risk of future aneurysmal dilation despite best medical treatment (for example, aortic diameter >40 mm at presentation)<sup>22</sup>

The INvestigation of STEnt Grafts in Aortic Dissection (INSTEAD) trial will report the long term outcomes of endovascular stent grafting for uncomplicated chronic type B dissection (conducted across seven European centres)<sup>30</sup>

The Acute Dissection Stent-grafting or Best Medical Treatment (ADSORB) trial will report the success of endovascular stent grafting in patients randomised to best medical treatment with and without stent grafting for uncomplicated acute type B dissection

The mid-term success of stent grafting for dissection will be clarified by publication of the results of postmarket registries (CAPTIVIA (NCT01181947) and VIRTUE (NCT01213589))

**Additional educational resources***Resources for patients*

Patient UK information ([www.patient.co.uk/doctor/Aortic-Dissection.htm](http://www.patient.co.uk/doctor/Aortic-Dissection.htm))—A relatively in-depth summary for patients interested in the risk factors, diagnosis, and treatment of aortic dissection

Mayo Clinic ([www.mayoclinic.com/health/aortic-dissection/DS00605](http://www.mayoclinic.com/health/aortic-dissection/DS00605))—Detailed information about aortic dissection for patients explained in a stepwise fashion

*Resources for healthcare professionals*

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**Tips for non-specialists**

Refer patients with confirmed aortic dissection (or symptomatic high risk patients) to a regional cardiovascular unit for urgent diagnostic investigation and treatment

Young patients with a history of connective tissue disease (such as Marfan's disease) or congenital cardiovascular disease (such as bicuspid aortic valves) are at high risk

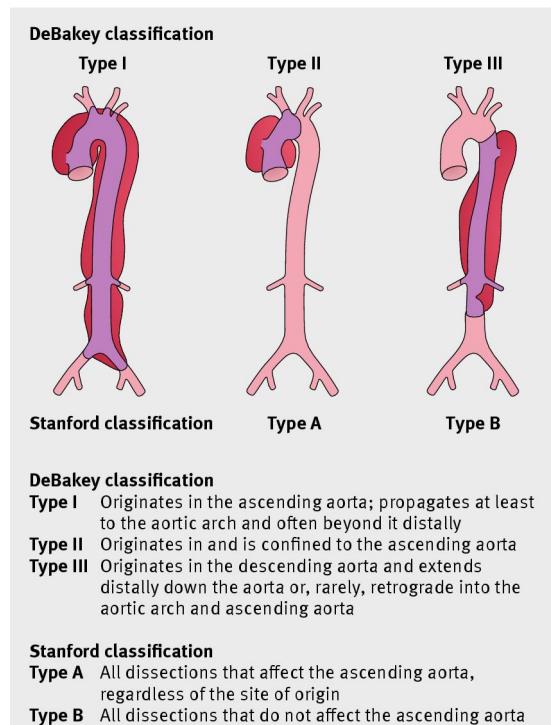
Maintain systolic blood pressure at 100-120 mm Hg in patients with a history of dissection; prescribe antihypertensive drugs (including  $\beta$  blockers) and deal with other modifiable cardiovascular risk factors

Ensure that patients with a history of dissection are enrolled in a surveillance programme at a regional cardiovascular unit

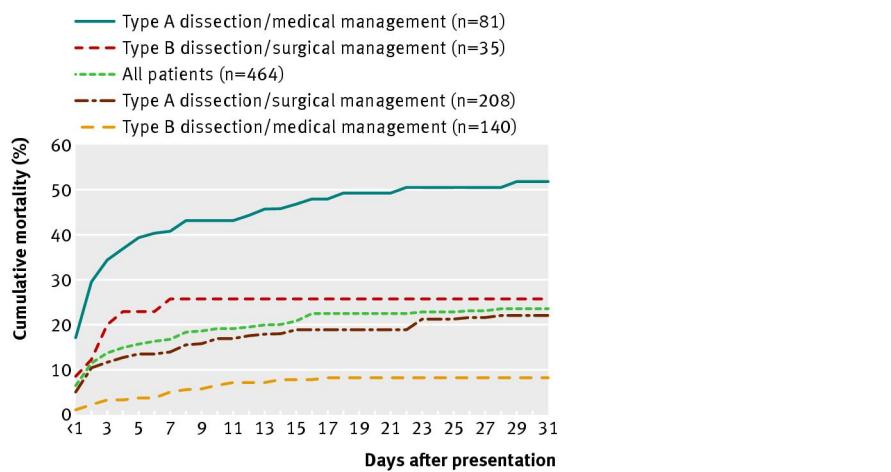
**Table****Table 1 | European Society of Cardiologists' classification of acute aortic syndrome**

Classification	Pathology
Type 1	Classic dissection with true and false lumens separated by the dissecting membrane
Type 2	Intramural haematoma
Type 3	Discrete dissection with a bulge at the tear site but no haematoma
Type 4	Penetrating aortic ulcer
Type 5	Traumatic or iatrogenic dissection

## Figures



**Fig 1** The Stanford and DeBakey classifications of aortic dissection. The dissection types are mainly differentiated by whether they affect the ascending aorta (the ascending aorta is affected in Stanford type A dissections, but not in Stanford type B dissections). Urgent surgical intervention is warranted when the ascending aorta is affected, and such cases are associated with higher mortality and morbidity than isolated descending aortic dissection<sup>4</sup>



**Fig 2** Thirty day mortality according to dissection type and management strategy<sup>w15</sup>