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). 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. The prognosis is grave, with 20% preadmission mortality and 30% in-hospital mortality.

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).

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. 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.

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.

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; Ehlers-Danlos type IV with abnormal synthesis of type III procollagen; and other connective tissue disorders associated with cystic medial necrosis.

Congenital cardiovascular abnormalities
A cross sectional study described a fivefold to 18-fold increased risk of dissection in 516 patients with bicuspid aortic valves. This increased risk was attributed to a co-inherited 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 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.

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.

Miscellaneous risk factors
Prevalence studies have shown that aortic vasculitic disease, cocaine misuse, pregnancy and hypertension 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.

Although British national statistics show that dissection affects all ages (27% of patients aged 17–59 years, 40% aged 60–74 years; 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.

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. Asymptomatic presentation is more common in patients with diabetes. The pain may radiate to the neck in type A dissection or to the interscapular area in type B aortic dissection. Acute rupture or inadequate perfusion—depending on the site and extent of the dissection—may cause a patient to become unconscious. 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. 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. Hypotension was seen in 25% of patients with type A dissection, whereas hypertension was typical in type B dissection.

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...
intravenous fluid resuscitation, with close monitoring of
tiltation, with careful monitoring of respiratory function.
Two large bore intravenous lines should be established for
aortic dissection entails adequate resuscitation and
sensitivity of 95.7% has been reported.
Although the specificity of this approach is unknown, a
findings that strongly suggest an alternative diagnosis.
but who present with no electrocardiographic changes
needed in patients who have one or more high risk feature,
aortic regurgitation, and shock. Urgent aortic imaging is
sure discrepancy, neurological deficit, a new murmur of
features of the examination include a pulse or blood pres
pain features include an abrupt onset of ripping, tearing,
manipulation, or a known thoracic aneurysm. High risk
Aortic Dissection (CONTRIBUTION) trial will report the success of endovascular stent
grafting in patients randomised to best medical treatment
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))

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)\(^2\)
The INvestigation of STEnt Grafts in Aortic Dissection
(INSTEAD) trial will report the long term outcomes of
devascular stent grafting for uncomplicated chronic type B dissection (conducted across seven European centres)\(^3\)
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 clarification of these questions will be
clarified by publication of the results of postmarket registries
(CAPTIVIA (NCT01181947) and VIRTUE (NCT01213589))

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 ven-
tilation, 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 out-
put. \(\beta\) 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.\(^4\) 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 devel-
opment 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 haemo-
dynamically stable.

How is aortic dissection diagnosed?
Retrograde aortography was the gold standard for assessing
patients in the 1970s and 1980s, but it has been super-
seeded by cross sectional imaging, which performs better
and has a better safety profile.\(^5\)

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.\(^6\)

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.\(^7\) However, these data were derived from
a population of patients undergoing imaging for dissection
in a tertiary centre. The high pre-test probability of dissec-
tion in this group limits the applicability of the study’s
findings, and the safety of using D-dimer testing to screen
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
useful in diagnosis.\(^8\)

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-
vasive 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%.\(^9\)

Computed tomography angiography
Multidetector computed tomography angiography is rec-
commended by the European Society of Cardiology as the
first line of investigation for patients with suspected acute
dissection.\(^1\) 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. Outside the emergency setting, electrocardiogram gated computed tomography can provide dynamic information, although its spatial resolution is inferior to magnetic resonance imaging, 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. 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. 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. 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. 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. 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. Disadvantages include its limited use in patients with claustrophobia or metal devices, although it can be used in those with nitinol aortic stent grafts. 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.

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. 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. Drug treatment alone results in a mortality of nearly 20% by 24 hours and 30% by 48 hours (fig 2). 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%. 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, or coronary artery bypass. The International Registry of Acute Aortic Dissection reported these techniques in 24% and 15% of type A dissections, respectively. Together with adjunctive measures such as hypothermic circulatory arrest and perfusion of the head vessels, surgery for proximal aortic dissection has three year and five year survival rates of 75% (standard deviation 5%) and 73% (6%), respectively.

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. For uncomplicated acute type B dissection, series have shown that drug treatment alone can result in 78% three year survival after discharge from hospital. Current guidelines deem this a difficult benchmark to surpass, and medical management remains the gold standard. Careful regulation of systemic blood pressure at 100-120 mm Hg is needed to minimise haemodynamic shear stress and discourage rupture. β 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 β blockers. Endovascular treatment is increasingly possible with low mortality, and its role in...
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.

Chronic type B dissection

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. A prospective cohort study of 159 patients reported that, if untreated, this pathology carries a mortality of 50%. Conventional open surgery for complicated dissection has a 30 day mortality of 30%, whereas meta-analysis has shown that endovascular treatment has a 30 day mortality of 9.8%. 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.

Multiple cohort studies have shown that even after complete false lumen thrombosis, 16% of patients develop evidence of dissection in the unstented distal aorta, which requires surgical reintervention.

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. 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%). 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%. The underlying pathophysiology of aortic medial disease and defective wall structure confers an ongoing risk of further dissection, aeurysmal degeneration, and rupture. 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. 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. Various experts also advocate the combined use of echocardiography with axial imaging for routine surveillance. All patients should receive lifelong antihypertensive treatment, including β blockers, with a target blood pressure of 120/80 mm Hg.

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 aeurysm remote from the original repair, graft dehiscence, aortic regurgitation, or infection. 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 aeurysm (7.8%), and rupture (3.0%), thereby justifying mandatory postoperative surveillance.

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Patient consent obtained.

CLINICAL REVIEW

Operative treatment won’t Neurovascular compromise, Extension type (caused by fall under anaesthesia with fracture but will be for Gartland and brachial artery. especially to the median nerve to a flexed hand).

PICTURE QUIZ
An injury in a child’s elbow

1 Displaced supracondylar fracture; a visible fat pad (posterior) (fig) indicates an undisplaced fracture.
2 Secondary ossification centres.
3 Extension type (caused by fall on an outstretched hand) and flexion type (caused by fall on to a flexed hand).
4 Neurovascular compromise, especially to the median nerve (anterior interosseous nerve) and brachial artery.
5 Operative treatment won’t be needed if it’s a Garland I fracture but will be for Garland II or III fractures (manipulation under anaesthesia with or without K wires).
Complications include cubitus varus, Volkman’s contracture, and ulnar nerve injuries.

A visible posterior fat pad

ANSWERS TO ENDGAMES, p 48
For long answers go to the Education channel on bmj.com

CASE REPORT
Recurrent fever after a holiday in Turkey

1 Brucellosis.
2 Identification of the bacterium with biochemical and molecular methods; serological assays.
3 Transmission is via consumption of unpasteurised dairy products.
4 Fever, arthralgia, fatigue, sweating, chills, hepatomegaly, splenomegaly, anaemia, and raised C reactive protein.
5 Treatment is with a combination of antibiotics that are effective on bacteria that live inside phagocytic cells.

STATISTICAL QUESTION
Allocation concealment
Allocation and selection bias (answers a and d) were minimised by allocation concealment, whereas ascertainment and detection bias (answers b and c) were not.