

# Diagnosis and management of acute aortic dissection in the emergency department

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## Abstract

Acute aortic dissection is often misdiagnosed as a result of its atypical presentations. It affects 4000 patients a year in the UK of all ages, not just older patients, with increasing numbers of cases expected in the future because of the ageing population. Dissection of the aortic wall leads to sudden, severe pain, and commonly end-organ symptoms which must be recognised. Acute aortic dissection can be challenging to diagnose in the emergency department because of the multitude of possible presentations and the need for selective testing with Computed Tomography Angiography (CTA). Clinicians often miss acute aortic dissection because it is not considered in the differential diagnosis, and the challenge lies in identifying acute aortic dissection in a sea of complaints of chest, back and abdominal pain. There are several ways to improve diagnosis, including awareness campaigns, better education about patients in which to consider acute aortic dissection, and improved detection strategies including which patients should receive CTA. Clinical decision tools and biomarkers could help, but further research is required and is a research focus in emergency medicine. Once diagnosed, blood pressure control, analgesia and urgent surgery or transfer to enable this to occur with minimal delay is required.

**Key words:** Acute aortic syndrome; Aorta; Diagnosis; Dissection; Emergency

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## Introduction

Acute aortic dissection affects 4000 patients a year in the UK of all ages, not just older patients (Healthcare Safety Investigation Branch (HSIB), 2017). It causes sudden, severe pain and end-organ symptoms. Acute aortic dissection can be difficult to diagnose in the emergency department because it can have many different types of presentations, and selective testing with Computed Tomography Angiography (CTA) is needed to confirm the diagnosis. This article uses a case presentation to illustrate some of the difficulties in diagnosing acute aortic dissection in the emergency department and looks at how this could be improved in the future.

## Case presentation

Mr S was a middle-aged man whose visit to the emergency department 6 years ago is etched in my memory. At that time, I was stationed in our rapid access triage assessment area. This is essentially a triage area, common in many United Kingdom (UK) emergency departments, where a senior doctor is available to help identify potentially serious conditions early, and rapidly fast track anyone who is unwell or who could be redirected to be better managed by another specialty or service elsewhere. Mr S presented with a cold, pale left arm that had developed 2 hours earlier and no other accompanying symptoms, but my initial concern included the possibility of acute aortic dissection. However, after a thorough examination, it appeared to be a more straightforward case of an acute embolic event leading to acute limb ischaemia, a surgical emergency. Mr S went through to the emergency department, where I handed over his care to a colleague, an emergency department consultant, who concurred with my assessment and felt there was not anything else to suggest acute aortic dissection. Mr S was referred to the vascular surgery team, and later that afternoon underwent the removal of a brachial artery thrombosis in the operating theatre. At 3:00 am, Mr S had a cardiac arrest from which he was unable to be resuscitated. A postmortem

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examination showed that he had a type A acute aortic dissection. Two weeks before this, Mr S had presented to another department with chest pain. He had undergone a troponin test, which was normal, and was discharged with a diagnosis of non-specific chest pain.

## Misdiagnosing acute aortic dissection

Misdiagnosis of acute aortic dissection is relatively common. Data from the emergency departments of two Edinburgh hospitals showed that, between 2011 and 2020, diagnosis of acute aortic dissection was delayed or missed in 26 patients. More widely, 1 in 3 patients with acute aortic dissection are misdiagnosed (Harris et al, 2011; Lovatt et al, 2022), 1 in 4 patients are not diagnosed until over 24 hours after presentation to the emergency department (Lovy et al, 2013), and acute aortic dissection is a common cause of fatality-related negligence claims (NHS Resolution, 2022). The tragic thing is we know that prognosis is best when patients are treated early, with mortality increasing by 2% per hour of delay in diagnosis (Pape et al, 2015). Acute aortic dissection is a treatable condition with an 80% survival rate (Chukwu M et al, 2023) when diagnosed and treated on time.

Currently 4000 patients a year in the UK experience acute aortic dissection. About half of these patients die before they reach a hospital, but around 50% arrive at a hospital (Howard et al, 2013). Acute aortic dissection can affect people of any age, but becomes more common with age. However, it also affects young people, with 25% of patients aged under 50 years and half aged under 60 years (Trimarchi et al, 2010). Atherosclerosis is the predominant underlying cause in older patients with acute aortic dissection, whereas in younger patients connective tissue disease predominates. So, age should never be a reason to not consider acute aortic dissection.

While acute aortic dissection may not be as common as some other conditions such as acute myocardial infarction, it is not rare. About 15 patients a year present with acute aortic dissection to the two emergency departments in NHS Lothian, the commonest being a type A dissection. Most emergency department consultants will see at least one case of acute aortic dissection a year, equating to around 30–40 during a career. Another statistic to put the condition into perspective is that 2500 patients die within a month of being diagnosed with acute aortic dissection, more than the number of people who die in the UK from road traffic accidents (1800 per year), or from pulmonary embolism (2300 per year) (Howard et al, 2013). This large cause of mortality is put into perspective when considering the huge infrastructure available for managing trauma, with developed pre-hospital systems and established major trauma centres.

So, what does the future hold? The Oxford vascular study (Howard et al, 2013) tells us that UK cases of acute aortic dissection are set to rise over the next 10 years to over 5500 and if the trajectory remains the same, by 2050 almost 3500 people will die every year of acute aortic dissection, mainly as a result of the population ageing and acute aortic dissection being more predominant in the 40–70-year age group.

## Pathophysiology

The first thing to consider is the anatomy of the aorta. The aorta starts at the aortic root where the aortic valve is sited at the outflow track of the left ventricle and from where the coronary arteries originate. The aorta then continues as the ascending thoracic aorta up to the aortic arch where three main vessels originate, the combined right subclavian and right common carotid, followed by the left common carotid and finally the left subclavian artery. This vessel is especially important when considering the type of acute aortic dissection, whether it is type A or B, as this influences the patient's management once diagnosed. An acute aortic dissection that involves the aortic root or the ascending aorta up to the left subclavian artery is a type A acute aortic dissection and is managed surgically with cardiothoracic surgeons replacing the arch of the aorta. Any acute aortic dissection originating after this is a type B acute aortic dissection and is managed medically with blood pressure lowering to allow the acute aortic dissection to become chronic and to settle, normally under cardiology teams in a coronary care unit environment.

The aorta comprises three main layers: the inner intima, the media (a reasonably thick layer made up of more than 50 alternating layers of elastin and smooth muscle cells), and

the outer adventitia. In acute aortic dissection, a small tear occurs in the intimal inner lining of the aorta which allows blood into the middle media area. Because the media is weaker than the other two walls, the blood, coming straight out of the heart and therefore under pressure, tracks up and down through the media separating the layers of the intima and the adventitia. The blood in the aortic media then pushes the dissection flap into the middle of the aorta, separating the true from the false lumen.

Acute aortic dissection is now commonly referred to as acute aortic syndrome which is made up of four conditions: type A acute aortic dissection, type B acute aortic dissection, intramural aortic haematoma and penetrating aortic ulcer. In intramural haematoma, blood leaks into the aortic media at low pressure, forming a thrombus that pushes the outer wall of the aorta outward, leaving a relatively normal appearing aortic lumen. A penetrating atherosclerotic ulcer allows blood to enter the aortic media, but atherosclerotic scarring of the aorta typically confines the blood collection, often resulting in a localised dissection or pseudoaneurysm.

Acute aortic syndrome is a dynamic process, as the calibre of the true and false lumens is dependent on the pressure in both these lumens, and this will determine whether the dissection flap moves more towards the true lumen or the false lumen. Blood moving into the false lumen can cause a number of different effects. Pressure building up in the false lumen can lead to rupture, re-entry tear, branch vessel occlusion or true lumen collapse. If the false lumen blood ruptures out of the aorta through the adventitia layer, the result is a bloody pericardial effusion, mediastinal haematoma or haemothorax, all normally, fatal conditions. It is thought that 7% of out-of-hospital cardiac arrests are the result of type A aortic dissection (Gouveia e Melo et al, 2022).

In a re-entry tear, the blood tracks back into the true aorta through a further tear in the intima, creating a double lumen channel. This means that blood can go through either lumen and arrive back into the normal distal aorta. Branch vessel occlusion is essentially where the false lumen has blood within it which surrounds branches that come off the aorta. When there is a high enough pressure in the false lumen, it collapses the true lumen and restricts perfusion to the vessels coming off the aorta. This explains why acute aortic syndrome can present with bizarre symptoms like stroke as branch vessel occlusion can temporarily occlude the left carotid artery for example, or a ST elevation myocardial infarction presentation because the left coronary artery is occluded. The patient may also present with limb ischaemia, as in the case presented earlier, as a result of occlusion of the left subclavian artery. With acute aortic syndrome being a dynamic process, pressure changes between the true and the false lumens can lead to occlusion or reperfusion of different areas at different times. The pressure in the true and false lumen, and subsequent blood flow through these stabilises over a period of minutes, hours or at the most, a few days. Finally, in true lumen collapse, the pressure in the false lumen exceeds the pressure in the true lumen impeding distal perfusion in the true lumen resulting in distal organ ischaemia.

It is crucial to differentiate between acute aortic syndrome and abdominal aortic aneurysm. They are distinct pathologies, with differing presentations and treatments. In the emergency department, the two are often confused, leading to discussions about checking blood pressures in both arms when suspecting an abdominal aortic aneurysm. Acute aortic syndrome can result in varying blood pressures in each upper limb because of the obliteration of the true lumen in one arm. In contrast, an abdominal aortic aneurysm is a slow, gradual dilation of the aorta that occurs over several years. When it reaches a certain size, typically 5 or 6 cm, there is a significant risk of rupture, characterised by abrupt abdominal or back pain and haemodynamic collapse as a result of blood leakage. Patients with acute aortic syndrome, although their aorta may be abnormal, do not typically have an aneurysm.

## Acute aortic syndrome presenting symptoms and signs

The presenting symptoms (chest, back or abdominal pain, syncope or symptoms related to malperfusion) and signs of acute aortic syndrome ([Table 1](#)) are consistent with the three main pathophysiological processes at play and will vary depending on the extent of aortic involvement. First, dissection of the aortic wall is extremely painful. It is a sudden thing

**Table 1. Presenting symptoms and signs of acute aortic syndrome**

Presenting symptom or sign	Cause
Asymmetric blood pressure between limbs or pulse deficit	Malperfusion of one or more limb artery by dissection or compression
Bowel ischaemia	Malperfusion of the coeliac or superior mesenteric artery by dissection or compression
Dysphagia	Compression of the oesophagus
Gastrointestinal bleeding	Malperfusion of the coeliac or superior mesenteric artery by dissection or compression
Haemoptysis	Vascular rupture into the lung parenchyma
Hoarseness	Compression of the recurrent laryngeal nerve
Horner's syndrome	Compression of the sympathetic chain
Myocardial ischaemia or infarction	Coronary artery involvement by dissection or compression
New aortic regurgitation (early diastolic decrescendo murmur heard best at the left lower sternal border in expiration)	Incomplete aortic valve closure secondary to leaflet tethering by the dilated aorta or cusp prolapse because of dissection into the aortic root
Oliguria or gross haematuria	Malperfusion of one or both renal arteries by dissection or compression
Paraplegia	Spinal malperfusion secondary to intercostal artery involvement
Lower limb ischaemia	Malperfusion of iliac artery by dissection or compression
Shock	Cardiac tamponade Haemothorax Frank aortic rupture Acute severe aortic regurgitation Severe myocardial ischaemia
Shortness of breath	Pericardial effusion or cardiac tamponade Congestive heart failure from acute severe aortic regurgitation Haemothorax Compression of trachea or bronchus
Stroke symptoms	Carotid or vertebral artery involvement
Superior vena cava syndrome	Compression of the superior vena cava
Syncope	Carotid artery involvement Cardiac tamponade

that happens in seconds, with the dissection peeling away the aortic media. While the classic textbook portrayal of acute aortic syndrome typically describes an acute tearing or ripping pain, patients more frequently describe an abrupt onset of severe, sharp or stabbing pain in the chest, back, and occasionally the abdomen, which is most intense initially, may sometimes radiate and importantly might settle once the dissecting process has stopped. Second, a contained rupture leading to pericardial effusion, mediastinal haematoma or haemothorax will cause intense physiological instability and perhaps breathlessness and/or hypotension. Finally, there are the end organ symptoms associated with malperfusion,

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such as ST elevation myocardial infarction and stroke mimics, which may be transient, may recur and which may affect different organs ([Table 1](#)).

## Diagnosis

Diagnosis is by imaging. Direct echocardiography signs of an acute aortic syndrome such as an intimal flap, intramural haematoma or a penetrating aortic ulcer may be seen on occasions if timely point of care echo expertise is available. Otherwise, CTA is the preferred imaging modality in patients with acute aortic syndrome in their differential diagnosis, and should be performed in a timely fashion. The accuracy of CTA in the diagnosis of aortic dissection is high, with sensitivity and specificity ranging around 98–100% (Vardhanabhuti et al, 2016).

## Why is acute aortic syndrome so difficult to diagnose?

Chest pain is the most common presenting complaint for acute aortic syndrome (80%) (Erbel et al, 2014). Back (40%) and abdominal pain are not uncommon (Erbel et al, 2014), but there are 2 million chest, back or abdominal pain presentations to English emergency departments a year (NHS Digital, 2021), overwhelmingly for causes other than acute aortic syndrome. While 1 in 980 emergency department patients with atraumatic chest pain (Alter et al, 2015) will have acute aortic syndrome, 979 will have other causes.

NHS Lothian teamed up with Frimley Health NHS Foundation Trust to perform a retrospective review of missed acute aortic syndrome cases between 2011 and 2020 to better understand why we miss acute aortic syndrome is missed. A total of 43 cases were identified using Morbidity and Mortality (M&M) records (including postmortem reports and complaints), as well as reviewing results of CTA scans requested by downstream inpatient teams querying acute aortic syndrome following discharge from the emergency department with a different diagnosis. Electronic patient records were reviewed by two independent reviewers to establish the reason the diagnosis was missed (McLatchie et al, 2023). Chest pain was the presenting complaint in 27 patients (63%), with 28 describing symptoms being of sudden onset. The three most common alternative diagnoses made were acute coronary syndrome, pulmonary embolism and non-specific chest pain.

So why was acute aortic syndrome missed? In most cases, acute aortic syndrome was missed because it was never considered in the differential diagnosis. In some of these, the clinician was satisfied by an alternative clinical diagnosis or happy that acute coronary syndrome was excluded. In other cases, acute aortic syndrome was clearly considered but not pursued further with imaging because the clinician was inappropriately reassured by the absence of certain ‘textbook’ clinical symptom and signs, by resolved symptoms, or by a normal chest radiograph.

Lovatt et al (2022) reviewed 12 studies, including 1663 patients with acute aortic syndrome, with a misdiagnosis rate of 33.8%. Factors leading to the diagnosis being missed included the symptoms being attributed to other conditions, the reassurance of a normal chest radiograph, patients having walked into the emergency department and the absence of ‘typical’ acute aortic syndrome symptoms such as tearing or ripping pain, differential upper limb blood pressures, a pulse deficit or acute hypertension. These features do not reliably rule out acute aortic syndrome.

An NHS Resolution (2022) report examined 86 clinical negligence fatality claims in English emergency departments totalling £5.8 million. The most frequent causes of death included misdiagnoses of infection or sepsis, pulmonary embolism, suicide, acute coronary syndrome and acute aortic syndrome. The report revealed a lack of awareness and recognition of the significance of symptoms, and missed opportunities to use information from ambulance and triage notes.

## So, is there anything helpful?

Abrupt onset pain and/or worst ever pain are much more associated with acute aortic syndrome and are a useful start. If a patient does not have abrupt onset pain, this halves

the likelihood that they have acute aortic syndrome, but unfortunately still does not rule it out. If someone has abrupt onset pain, further investigations are needed. Worst ever pain is acute aortic syndrome until proven otherwise.

## How can we do better?

There have been campaigns and great educational resources such as those by the Aortic Dissection Charitable Trust. Think Aorta have also attempted to improve awareness of acute aortic syndrome in the emergency department. However, these have not led to improvements in mortality. Awareness is extremely important, and campaigns like these are vital, but they are not the only answer. A clinician survey of practice across the UK showed that only 12 of 56 emergency departments have a formal pathway for working up patients with potential acute aortic syndrome and no guideline predominated, probably because none are particularly simple to use in the emergency department, and there is no robust evidence-based method of ruling out acute aortic syndrome (McLatchie et al, 2022).

While CTA has high sensitivity and specificity to diagnose acute aortic syndrome, over-testing leads to diagnostic yields as low as 2% (Lovy et al, 2013; Ohle et al, 2018), significant costs and resource implications, ionising radiation risks, CT delays for non-acute aortic syndrome patients and the burden of ‘incidentalomas’. Clinicians need to use CTA selectively, yet there is no validated clinical decision tool for this scenario, despite several being proposed (Rogers et al, 2011; Ohle et al, 2020; Morello et al, 2021), and none that has been studied in undifferentiated emergency department populations. All clinical decision tools have low diagnostic yield for acute aortic syndrome, modest specificity and lead to higher rates of CTA.

D-dimer has been suggested as a rule-out biomarker in low pre-test probability patients (Bima et al, 2020; Yao et al, 2021) and is part of the Aortic Dissection Detection-Risk Score (ADD-RS) clinical decision tool, but it is currently unclear whether any acute aortic syndrome clinical decision tools have sufficient sensitivity to be acceptable to clinicians, which is the most accurate, and whether acute aortic syndrome clinical decision tools are likely to lead to over-investigation with CTA and D-dimer. The DASHED study (<https://clinicaltrials.gov/ct2/show/NCT05582967>) looked at 5548 adults attending 27 UK emergency departments with potential symptoms of acute aortic syndrome. This showed that 3 in 1000 patients presenting with possible symptoms had acute aortic syndrome, 74 in 1000 patients with possible acute aortic syndrome symptoms underwent CTA, with 2.9% being positive, 34 in 1000 patients where acute aortic syndrome was ‘most likely diagnosis’ had acute aortic syndrome and two patients where acute aortic syndrome was thought ‘not possible’ had acute aortic syndrome. This illustrates the diagnostic challenge of acute aortic syndrome, the limitations of methods for selecting patients for CTA and confirms that the best decision aid to facilitate decision to CTA and to outperform emergency department clinician gestalt is not yet clear.

## Management of acute aortic syndrome

Guidelines, both UK and international, offer advice on the management of acute aortic syndrome. In the UK, the Royal College of Emergency Medicine and Royal College of Radiologists (2024) best practice guideline primarily focuses on diagnosing acute aortic syndrome within the emergency department, while the NHS (2022) acute aortic dissection pathway toolkit lays out principles for timely image transfer and safe transfers, although it does not delve into the medical management of confirmed acute aortic syndrome in the emergency department. The Liverpool acute aortic syndrome pathway (Liverpool Heart and Chest Hospital) is another useful reference. Internationally, the European Society of Cardiology (ESC) guidelines on aortic diseases (Erbel et al, 2014) and the joint American College of Cardiology (ACC)/American Heart Association (AHA) guideline for the diagnosis and management of aortic disease (Isselbacher et al, 2022) provide comprehensive recommendations for acute aortic syndrome management.

Key principles of care include the imperative for rapid diagnosis and the recognition that acute aortic syndrome is a medical emergency. Definitive management of type A acute aortic syndrome involves surgery, where timing is crucial and interventions should not lead to significant delays. Blood pressure management is fundamental to care, with experienced clinicians ideally taking ownership, considering referral and potential urgent transfers for definitive care.

Blood pressure control is important because the main drivers of aortic dissection extension and rupture are hypertension and the force of left ventricular ejection. While no randomised studies have compared different medical treatments for acute aortic syndrome, extensive clinical experience has established the current standard of anti-impulse therapy. The goal is to reduce heart rate and blood pressure to minimise aortic wall stress, targeting systolic blood pressure below 100–120 mmHg. Expert opinion suggests the lowest blood pressure that does not compromise end-organ function should be sought, along with a target heart rate of 60–80 bpm. Achieving this often involves a combination of intravenous beta blockers (eg labetalol or esmolol) and intravenous vasodilators (eg glyceryl trinitrate GTN). The use of intravenous GTN alone is not recommended, as it can lead to compensatory increases in left ventricular contraction force. In cases where beta blockers are contraindicated, IV non-dihydropyridine calcium-channel blockers like verapamil or diltiazem should be considered.

Anti-impulse therapy and analgesia are essential aspects of the management of acute aortic syndrome. Pain associated with acute aortic syndrome can trigger elevated heart rate and blood pressure. To address this, morphine is typically administered IV and titrated to effect, with a subsequent transition to morphine patient controlled analgesia (PCA). In patients with renal impairment, fentanyl PCA may be preferred. Intravenous non-steroidal anti-inflammatory drugs (NSAIDs) like ketorolac should be used cautiously, as they can induce hypertension and lead to adverse renal effects.

In terms of patient transfer, it is advisable for the referring centre to arrange for a transfer with a medical escort who can interpret monitoring data and effectively manage the patient's blood pressure. At a minimum, the patient should have an arterial line, wide-bore intravenous access, and labetalol infusion available. Time sensitivity is paramount in such cases, prompting a blue light 999 ambulance call. In terms of family involvement, clear and effective communication is vital, ensuring that both the patient and their family grasp the life-threatening nature of acute aortic syndrome. Transfer to cardiac services allows for assessment and potential surgical intervention, although this carries high risks and is associated with a significant mortality rate.

For type B acute aortic syndrome, similar blood pressure and heart rate targets apply. In cases where patients develop leg weakness, interventions to prevent spinal cord ischaemia are considered. These may involve increasing the target blood pressure to avoid potential spinal cord infarction, implementing emergency cerebrospinal fluid (CSF) drainage, or conducting repeat Computed Tomography (CT) or Magnetic Resonance Imaging (MRI) as needed.

## Conclusions

Acute aortic syndrome is rare, devastating and often misdiagnosed, missed or delayed in diagnosis, because clinicians do not consider the condition or are not aware of it, atypical presentations, mimics of other disease, clinicians being falsely reassured by a normal chest radiograph, or lack of typical clinical signs. Its clinical features are highly unreliable but sudden and/or severe pain must always be taken seriously. Acute aortic syndrome is a dynamic process and symptoms may come and go. Better education and better detection strategies are needed to improve this care. The current focus will hopefully do this and mean that fewer acute aortic syndrome patients suffer misdiagnosis, missed or delayed diagnosis in the future.

## Key points

- The term acute aortic syndrome describes four conditions: type A acute aortic dissection, type B acute aortic dissection, intramural aortic haematoma and penetrating aortic ulcer.
- One in four patients with acute aortic syndrome is not diagnosed until over 24 hours after presentation to the emergency department.
- Age is not a reason to not consider acute aortic syndrome.
- Acute aortic syndrome can present with chest, back or abdominal pain, syncope or symptoms related to malperfusion.
- Abrupt onset and worst ever pain should be considered to be acute aortic syndrome until proven otherwise and investigated further.
- Aorta CT angiogram is the preferred imaging modality to diagnose acute aortic syndrome and should be performed in a timely fashion.
- It is unclear whether any clinical decision tools for acute aortic syndrome have sufficient sensitivity to be acceptable to clinicians.

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Not applicable.

### Author Contributions

MJR conceived and wrote the article. MJR was responsible for drafting and revision of content, approval of version to be published. MJR is accountable for all aspects of the work.

### Ethics Approval and Consent to Participate

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