Aortic dissection is one of the most common causes of aortic emergency and may represent a life-threatening condition requiring immediate diagnosis and treatment, including thoraco-abdominal aortic aneurysm rupture, intramural haematomas and penetrating atherosclerotic ulcers.

Clinical presentation of acute aortic dissection is typically characterised by chest pain, which is the most common symptom. Typically, in acute dissection the pain is tearing and intractable, and is primarily anterior in type A dissection and posterior in type B disease. The pain may extend from the chest to the interscapular region, down to lower back or up into the neck and head (migratory pain). Other manifestations include congestive heart failure, syncope, cerebrovascular accident, shock, paraplegia, shortness of breath, peri-cardial effusion, pleural effusion and peripheral pulse deficit. However, also described in the literature are cases of acute aortic dissection presenting with Otten’s syndrome with dysphonia or hoarseness, and others with mesenteric compromise, including abdominal pain, haematochezia, melena and obstructive jaundice.

Recently, one case of type A aortic dissection presenting with testicular pain has been reported in the literature. In intramural haematomas and penetrating atherosclerotic ulcers, clinical manifestations are similar to those in aortic dissection. Nevertheless, in 10–33% of patients with acute dissection chest pain is absent, and the correct diagnosis is not made before post mortem examination in between 28 and 38%.

In 2006, Hayter et al. demonstrated that in 67 patients who tested positive for acute aortic disorders, the textbook symptom of tearing chest pain was not present in all of the patients with acute aortic disorder. Furthermore, in 1998 Armstrong et al. concluded that symptoms in patients with acute aortic dissection are more variable than commonly admitted. Therefore, we can deduce that the clinical presentation of acute aortic dissection – as well as the other acute aortic disorders – is variable and sometimes misleading.

Although these diseases are aetiologically different, the explanation of this variability is due to a common character: all of these conditions affect the wall of the aorta and its layers. In fact, aortic dissection is a separation of the aortic intima from the adventitia; aortic aneurysm rupture is the most important complication of the aortic aneurysms and consists of seepage of blood in clefts in the aortic wall. Intramural haematoma is due to the subintimal haemorrhage into the aortic wall from rupture of the vasa vasorum without a demonstrable intimal tear. A penetrating atherosclerotic ulcer is an ulcerated plaque penetrating the intimal lamina resulting in haematoma formation within the media.

The intrinsic anatomical structure of the aortic wall generates a confusing patient symptomatology. In fact, the tunica adventitia is primarily composed of connective tissue that encloses the vasa vasorum and the nervus vasorum. Chest and/or aortic pain is elicited by the stimulation of nerves ending in the adventitia. However, nerve impulses from other strutures (i.e. skin, oesophagus and heart) also converge in a pool of neurons in the posterior horn of the spinal cord. This may explain why many different clinical conditions may mimic an aortic dissection or opposite condition.

Another cause of an unexpected and diverting presentation in the emergency setting is that patients may present directly with systemic signs and symptoms related to malperfusion of the brain, limbs or solid or visceral abdominal organs. More than one-third of patients with aortic dissection show signs and symptoms indicative of systemic involvement.

In 1972, Anagnostopoulos et al. reported mortality within 48 hours of about 50% of patients suffering acute type A aortic dissection. Actual mortality rate in these patients remains about 40%. The mortality rate for patients with untreated proximal aortic dissections has been reported to increase by 1% to 2–3% per hour after presentation and is approximately 25% during the first 24 hours after the initial presentation, 70% during the first week and 80% at two weeks.

Despite medical and surgical progress and technological evolution of diagnostic procedures, one-third of cases are still not diagnosed and misdiagnosis occurs in 30–39% of patients ultimately diagnosed with, longer time to correct diagnosis.

If 12.7% of patients with acute aortic type A dissection appear with syncope, it is legitimate to wonder how many patients come to our hospitals.
Cardiothoracic Imaging

Figure 1: Unsuspected B-type Aortic Dissection

The case was a 71-year-old man presenting at our emergency department for paraquat intoxication and mild dyspnoea. A: At the level of the left main pulmonary artery, multidetector computed tomography (MDCT) scan shows descending aorta dissection with intimal flap (arrow) and signs of competition between false (f) and true lumen. B: At the level of the renal arteries the MDCT scan shows abdominal aortic dissection with left renal artery (white arrow) arising from the false lumen and ischaemia of the homolateral kidney (arrowheads). Note also the gross haemorrhage at the left paranephric space (open arrow) with ventral dislocation of the kidney. A small amount of fluid is also visible at the perirenal space (black arrows). C: An MDCT scan depicts a distal abdominal aortic dissection with intimal calcifications (black arrow) and retro-peritoneal haemorrhage (arrowheads) at the left iliac space for vessel rupture at the level of the aortic bifurcation. A small quantity of free fluid is also visible at the right paracolic gutter (open arrow).

Attention for syncope with a clinically unacknowledged acute aortic dissection, having undergone a brain computed tomography (CT) and after being discharged or treated for ischaemic stroke. Therefore, it is clear that aortic dissection is a complicated condition: if severe chest pain is the most common symptom, atypical clinical manifestations are not so sporadic and this can make the diagnosis of aortic dissection particularly challenging in the emergency setting.

Due to these considerations, we can classify patients suffering from aortic dissection into three categories: in the first category, death occurs immediately and therefore they never come to our attention; the second category includes the smallest group of patients, and both signs and symptoms are so immediately recognisable that the patient comes to our attention with specific indication of aortic dissection; the third category includes patients with subtle symptoms, where only the suspicion of dissection can ‘save’ them.

For patients belonging to the first group, the pathologist will make a correct diagnosis only with a post mortem examination. For patients belonging to the second and third groups, the anatomical–pathological reality lies in the radiologist’s hands: in the first case he or she should not attempt more than to simply confirm the correct clinical indication, while in the second case he or she will have to interact with individuals who could walk into the emergency room in good clinical condition with a vague, apparently inconsistent symptomatology that can be properly framed, without delaying the correct diagnosis, only with the use of appropriate imaging techniques. In fact, despite the clinic presentation, some circumstances suggest a likely diagnosis; however, we could be faced by a discrepancy between the symptoms and the first-level imaging techniques. Described in the literature, as previously mentioned, are atypical presentations of aortic dissection. For example, in acute abdominal pain aortic dissection is one of the many conditions we have to consider in differential diagnosis; however, the symptoms are often not so clear and can render the diagnosis of aortic dissection particularly challenging for the emergency physician, thus delaying the correct diagnosis and increasing mortality. Consequently, the question is as follows: how can we change the natural history of a disease that causes thousands of deaths every year (two to four per 100,000 deaths per year) despite an improvement in the field of technology and medical sciences? The answer is to develop a high index of suspicion. This notion is crucial and becomes of vital importance in catastrophic emergencies, especially in challenging cases.

We can suspect something only if we are faced by something we know. Therefore, we must know the natural history of the disease. Some risk factors predisposing to aortic dissection are extremely important: long-standing arterial hypertension, connective tissue disorders (such as Marfan’s syndrome and Ehlers-Danlos syndrome, but also bicuspid aortic valve or giant cell arteritis, Takayasu arteritis), deceleration trauma or iatrogenic factors (valvular or aortic surgery) should be considered for framing patients with an unclear symptomatology. However, in many cases the typical clinical picture does not correspond at all to the anatomo-pathological condition and vascular emergencies are often an ‘unsuspected killer’. For instance, consider the following case. A 71-year-old male with no history of hypertension or cardiac condition came to our emergency department because of paraquat intoxication, with mild dyspnoea. Preliminary laboratory exams, X-rays and abdominal plain film were negative. In order to determine the reason for the unexplained symptomatology, a total-body multidetector computed tomography (MDCT) scan was performed (see Figure 1). B-type dissection was unsuspected based on the clinical picture and presentation. As with this case, there are many situations in which clinical presentation is not in agreement with evidenced first-level imaging. Thus, in challenging cases we should always think about a second-level imaging approach in order to explain unclear clinical manifestations of a case. However, achieving a challenging diagnosis is possible only if we are familiar with the disease.

The core of the question lies in the true knowledge of disease. The suspicion arises only from knowledge of this entity, with its common
clinical aspects and its atypical presentations and if we consider that in 1999 dissection occurred nearly three times as frequently as rupture of an abdominal aortic aneurysm in the US,22 maybe aortic dissection is a more common disease than we thought. Subsequently, the suspicion should be confirmed or denied by diagnostic imaging that plays an essential role not only to identify but also to classify aortic dissections, regardless of the clinical context. In his review in 2002, Klompas maintained that the clinical examination is insufficiently sensitive to rule out aortic dissection given the high morbidity of missed diagnoses.23 Classification of aortic dissection is necessary to decide on treatment either with a surgical approach or with conservative therapy.

Echocardiography, MDCT and magnetic resonance imaging are all useful techniques in the evaluation of the presence of intimal flap, the site of intimal disruption, re-entry points, true and false lumen, the status of the aortic valve, the presence of myocardial ischaemia and aortic branch vessel involvement with high sensitivity and specificity and diagnostic accuracy in the assessment of aortic dissection.24 However, depending on its clinical variability, the choice of the best imaging technique for the case depends on the clinical picture.

Therefore, the only way to identify this life-threatening and sometimes insidious condition is to research as many techniques as possible in order to find one that is quick in its execution and that provides us with as much information as possible even on the other districts. Currently, the only technique presenting these features are MDCT. The wide availability in the field, the possibility of acquiring large volumes of data in a short time with high spatial resolution and the possibility of reconstructing the images in post-processing makes MDCT the primary investigation, even for an atypical presentation of an aortic dissection. A further advantage of MDCT is the possibility of making rapid differential diagnosis with a wide spectrum of clinical entities that can mimic this condition. Acute chest and abdominal pain are clinical expressions of a huge variety of diseases. In the series by Thoongsuwan et al.,25 in 28 out of 130 patients (21.5%) CT provided an alternative diagnosis in patients initially suspected of aortic dissection.

Published data on CT show it to have 100% sensitivity and specificity for detection of aortic dissection.12 The introduction and diffusion of MDCT make it the first-line imaging technique representing a valuable diagnostic tool in the emergency setting if aortic dissection26,27 is suspected and, more generally, in cases of suspected vascular emergency.

Nevertheless, we maintain that a clinical examination can sometimes be insufficiently sensitive, especially in cases of misleading signs and symptoms. It is also true that it is unrealistic to submit all patients with unclear presentation who come to our attention to CT. This means that the passage from the suspicion of the illness to the final diagnosis can happen only with a dedicated emergency team working in the emergency departments. Emergency medicine is a separate chapter in the whole book of medicine, which requires knowledge of a series of illnesses that an inexperienced physician cannot know in depth.

Finally, we can conclude that emergency medicine is a sensitive subject, but aortic dissection is a complex and insidious condition that continues to claim lives despite diagnostic and surgical progress.