Warning at emergency room: The role of diagnostic test in acute aortic dissection

Alfonso Lagi*

Chief, Department of Emergency, Hospital Villa Donatello, Florence, Italy

Abstract

Background: The aortic diseases are life-threatening diseases which ask to be diagnosed and treated as soon as possible. The main thing is to lose no time. In the Emergency Room, the most appropriate investigations should be performed, so that the accurate decisions can be taken.

Methods: We composed a narrative review analysing the most important prospective population studies (1,641 patients) either retrospective studies (over 14,000 patients) and evaluated guidelines in the filed. Both sensitivity and specificity and the chances for doing of the diagnostic test were evaluated.

Results: The specificity and sensitivity of the diagnostic tools was evaluated to decide the priority of test to utilize in emergency room. The diagnostic work up is founded on chest pain associated with age, sex, previous diseases, after which some significant signs and symptoms are measured such as aortic valvular regurgitation, pleural or pericardial effusion, syncope, neurological or vascular symptoms. At the end, one set of diagnostic tests confirming the diagnosis are taken into account. It is imaging tests Computed tomography (CT), Magnetic Resonance Imaging (MRI), and Transoesophageal Echocardiography (TOE) are equally reliable for confirming or excluding the diagnosis of AAD and they are considered as diagnostic gold standard.

Conclusion: The right balance between availability of the test and its diagnostic sensitivity and specificity is the goal of a proper work up. The whole of the prospective and retrospective population-based ascertainment showed that hospital-based registries indicate as diagnostic gold standard test CT scan or Transoesophageal Echocardiography to make as soon as possible in selected population of high risk patients.

Key-message

The aortic diseases are life-threatening and time consuming. To take the appropriate diagnostic test, it may be very expensive. Therefore, in the emergency room health workers should rank the high risk clinical conditions and features which select the patients very likely to suffer from Acute Aortic Disease.

Introduction

The Acute Aortic Syndromes (AAS) are a severe, life threatening and time related disease, which ask fast diagnosis and therapy. The common denominator of AAS is disruption of the media layer of the aorta with bleeding within, along the wall of the aorta resulting in separation of the layers of the aorta (dissection), or transmurally through the wall in the case of ruptured of penetrating atherosclerotic ulcer or trauma. They are defined as emergency conditions with similar clinical characteristics involving the aorta: acute aortic dissection (AAD), intramural haematoma, penetrating aortic ulcer, complete rupture of the aorta, traumatic aortic injury, iatrogenic aortic dissection. The clinical progress of patients with AAS is unpredictable. A high level of suspicion is required for early diagnosis and crucial for patient survival. The aim of this manuscript is to rise a red flag for the health workers of emergency room to improve their reaction and conduct in front of AAD which is the common pathology of all AAS [1,2].

Methods

The intention was to do a narrative review. Progressive and retrospective population-based studies and guidelines on Acute Aortic Diseases were evaluated for a whole of 1,641 and over 14,000 patients respectively. All retrieved articles were screened for eligibility based on title, abstract, and full content. The specificity and sensitivity of the diagnostic tools was evaluated to decide the priority of test to utilize in emergency room. Guidelines on Acute Aortic disease were evaluated in comparison.

Results

Epidemiology

Up-to-date data on the epidemiology of AAD is scarce. In the Oxford Vascular study, its incidence is estimated at six per hundred thousand persons per year. This incidence is higher in men than in women and increases with age [3-5].

A review of 464 patients from IRAD reported a mean age at presentation of 63 years, with significant male predominance (65%) [5]. The incidence of dissection appears to be increasing, independent of the ageing population, either It may be that two to three times as many patients die from aortic dissection than from ruptured abdominal aortic aneurysm; with an unknown number of patients dying before diagnosis, the true prevalence is not precise.

The most common risk factor but not the most important, associated with AAD is the acute hypertension, observed in 65 – 75% of individuals [6-9].

Correspondence to: Alfonso Lagi, Chief, Department of Emergency, Hospital Villa Donatello, Florence, Italy, E-mail: alfonso.lagi1@tin.it

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*Correspondence author.
Other risk factors, less frequent but more important, include aortic valve disease, history of cardiac surgery, cigarette smoking, use of intravenous drugs (e.g. cocaine and amphetamines), Marfan syndrome or other connective diseases.

The most common cause of traumatic aortic dissection or rupture is direct blunt chest trauma, by road traffic accidents or deceleration. An autopsy study of road accident fatalities found that ≥80% of the patients had a ruptured aorta, emphasizing the importance of traumatic rupture of the aorta [10].

Clinic in acute aortic dissection

Many signs and symptoms can be associated with chest or back pain, that should solicit the attention of physicians.

Chest pain: Chest pain is the most frequent symptom of AAD. Abrupt onset of severe chest and/or back pain is the most typical feature. The pain may be sharp, ripping, tearing, knife-like, and not different from Acute Myocardial Infarction (AMI); the abruptness of its onset is the most specific characteristic [11,12].

The most common site of pain is the chest (80%), while back and abdominal pain are experienced in 40% and 25% of patients, respectively. Anterior chest pain is more commonly associated with Type A of AAD, whereas patients with Type B dissection present more frequently with pain in the back or abdomen [13]. The pain may migrate from its point of origin to other sites, following the dissection path as it extends through the aorta.

Pain radiating to the neck, throat, and/or jaw may indicate the involvement of the ascending aorta, particularly when associated with murmur of aortic regurgitation, pulse differentials, or signs of tamponade; conversely, pain in the back or abdomen may herald dissection of the descending aorta.

Classification

Acute thoracic aortic dissection can be classified according to either the origin of the intimal tear, it drives decisions regarding surgical vs. non-surgical management. The two most commonly used classification schemes are the DeBakey and the Stanford systems. For purposes of classification, the ascending aorta refers to the aorta proximal to the brachiocephalic artery, and the descending aorta refers to the aorta distal to the left subclavian artery.

DeBakey classification

- **Type I**: Dissection originates in the ascending aorta and propagates distally to include at least the aortic arch and typically the descending aorta (surgery usually recommended).
- **Type II**: Dissection originates in and is confined to the ascending aorta (surgery usually recommended).
- **Type III**: Dissection originates in the descending aorta and propagates most often distally (non-surgical treatment usually recommended).
  - Type IIIa: Limited to the descending thoracic aorta.
  - Type IIIb: Extending below the diaphragm.

Stanford classification

- **Type A**: All dissections involving the ascending aorta regardless of the site of origin (surgery usually recommended).
- **Type B**: All dissections that do not involve the ascending aorta (non-surgical treatment usually recommended). Note that involvement of the aortic arch without involvement of the ascending aorta in the Stanford classification is labelled as Type B.

The dissection can spread from the intimal tear in a anterograde or retrograde fashion, often involving side branches and causing malperfusion syndromes, tamponade, or aortic insufficiency.

Aortic regurgitation

Aortic regurgitation may accompany 40 – 75% of cases with Type A and may be so severe to influence the prognosis [14]. Pericardial tamponade is observed in 20% of patients with acute Type A [9].

Heart failure

Registry IRAD data show that this complication occurs within 10% of cases of AAD and is commonly related to aortic regurgitation [9]. Although more common in Type A, heart failure may also be encountered in patients with Type B, suggesting additional aetiologies of heart failure, such as myocardial ischemia, pre-existing diastolic dysfunction or uncontrolled chronic hypertension.

Hypotension and shock may result from aortic rupture, acute severe aortic regurgitation, extensive myocardial ischemia, cardiac tamponade, pre-existing left ventricular dysfunction, or major blood loss.

Plural effusions

Smaller plural effusions may be detected in 15–20% of patients, with almost equal distribution between Type A and Type B patterns, and are believed to be mainly the result of an inflammatory process. Large plural effusions resulting from aortic bleeding into the mediastinum and plural space are rare because these patients usually do not survive up to their arrival at the hospital [15].

Syncope

Syncope is an important initial symptom, occurring in approximately 15% of patients with Type A and in 5% of those presenting with Type B. This feature is associated with an increased risk of in-hospital mortality because it is often related to life-threatening complications, such as cardiac tamponade or supra-aortic vessel dissection.

Neurological symptoms and peripheral vascular diseases

They may result from cerebral malperfusion, hypotension, distal thromboembolism, or peripheral nerve compression. The frequency of neurological symptoms range from 15–40%, and in half of the case may be transient. Acute paraplegia, due to spinal ischaemia caused by occlusion of spinal arteries may be a serious disease. Radial or femoral pulses may be absent. Cerebral ischemia and acute myocardial infarction may associate with AAD.

Mesenteric ischemia may also result from the involvement of a major arterial orifice in the dissection process. The perfusion disturbance can be intermittent if caused by a dissection flap prolapse, or persistent in cases of obliteration of the organ arterial supply by false lumen expansion. Renal failure may be encountered at presentation or during hospital course in up to 20% of patients with acute Type A and in approximately 10% of patients with Type B AAD [7]. This may be the result of renal hypoperfusion or infarction, secondary to the involvement of the renal arteries, or may be due to prolonged hypotension.
In conclusion, in order to rank the patients referring to the risk to be affected from AAS it considered low and high - risk referred conditions, pain and examinations features.

Clinical data, useful to assess the a priori probability of AAD, are resumed in table 1.

**Laboratory testing**

If D-dimers are elevated, the suspicion of AAD is increased [15-17]. Typically, the level of D-dimers is immediately very high, compared with other disorders in which the D-dimer level increases gradually. D-dimers yielded the highest diagnostic value during the first hour If the D-dimers are negative, Intramural Hematoma (IH) and Penetrating Aortic Ulcer (PAU) may still be present; however, the advantage of the test is the increased alert for the differential diagnosis [15].

**Diagnostic imaging**

The target of imaging in AAD is to identify the evidences reported in table 2 [18].

So, it needs to avoid ineffective exams, cause of time consuming, but to use the most sensitive and specific ones. A flowchart for the emergency room has been developed in order to enhance the standardization of decision making in AAS, because survival is strongly related to time. In the diagnostic work-up clinical data are useful to assess the a priori probability of AAS.

Computed tomography (CT), Magnetic Resonance Imaging (MRI), and Transoesophageal Echocardiography (TOE) are equally reliable for confirming or excluding the diagnosis of AAD so that they are considered as diagnostic gold standard.

**Table 1. Clinical data useful to assess the probability of acute aortic dissection.**

<table>
<thead>
<tr>
<th>High-risk conditions</th>
<th>High-risk pain features</th>
<th>High-risk examination features</th>
</tr>
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<tbody>
<tr>
<td>Marfan syndrome or other connective tissue diseases</td>
<td>Chest, back, or abdominal pain described as any of the following: - abrupt onset - severe intensity - ripping or tearing</td>
<td>Evidence of perfusion deficit: • pulse deficit • systolic blood pressure difference • focal neurological deficit (in conjunction with pain)</td>
</tr>
<tr>
<td>Family history of aortic disease</td>
<td>Aortic diastolic murmur (new and with pain)</td>
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<tr>
<td>Known aortic valve disease</td>
<td>Hypotension or shock</td>
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<tr>
<td>Known thoracic aortic aneurysm</td>
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**Table 2. Imaging evidences in acute aortic dissection.**

<table>
<thead>
<tr>
<th>Aortic Dissection</th>
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<tbody>
<tr>
<td>Visualisation of intimal flap</td>
<td></td>
<td></td>
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<tr>
<td>Extension of diseases</td>
<td></td>
<td></td>
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<tr>
<td>Identification of true or false lumen</td>
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<tr>
<td>Localisation of entry an re-entry tears</td>
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<tr>
<td>Involvement of the side branches</td>
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<tr>
<td>Detection of organ ischemia, pericardial, pleural effusions</td>
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<td></td>
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<tr>
<td>Detection of peri-aortic bleeding</td>
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<td></td>
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<tr>
<td>Intramural haematoma</td>
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<tr>
<td>Localisation and extent of aortic wall thickening</td>
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<td></td>
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<tr>
<td>Presence of intimal tear</td>
<td></td>
<td></td>
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<tr>
<td>Penetrating aortic ulcer</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Localisation of the lesion</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Co-existence of intramural hematoma</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Involvement of peri-aortic tissue and bleeding</td>
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</tbody>
</table>

**CT** an MRI have to be considered superior to TOE for the assessment of AAD extension and branch involvement, as well as for the diagnosis of IH, PAU, and traumatic aortic lesions [17].

The sensitivity of TOE reaches 99%, with a specificity of 89% [19] The positive and negative predictive values are 89% and 99%, respectively, based on surgical and/or autopsy data that were independently confirmed [18].

The diagnosis of AAD by standard transthoracic M-mode and two-dimensional echocardiography (TTE) is based on detecting intimal flaps in the aorta. The sensitivity and specificity of TTE range from 77–80% and 93–96%, respectively, for the involvement of the ascending aorta. The tear is defined as a disruption of flap continuity, with fluttering of the ruptured intimal borders. Smaller intimal tears can be detected by colour Doppler, visualizing jets across the flap, which also identifies the spiral flow pattern within the descending aorta.

TTE is very useful in identifying aortic valve dysfunction, pericardial tamponade, or wall motion abnormalities, and may screen for proximal 4 to 8 mm of the ascending aorta to just above the sino - tubular junction in patients with good echocardiographic windows and a short segment of the descending aorta in patients with shock. It may show the intimal flap and a thickened aortic wall. The proximal ascending aorta can usually be seen in long- and short-axis parasternal views. TTE is limited, however, in visualizing the distal ascending aorta and the arch. Proximal aortic dilation is usually seen in ascending aortic dissection. The presence of normal aortic dimensions and geometry and absence of aortic regurgitation on TTE are evidence against the presence of an ascending aortic dissection, but this does not fully exclude the diagnosis.

TTE is restricted in patients with abnormal chest wall configuration, narrow intercostal spaces, obesity, pulmonary emphysema, and in patients on mechanical ventilation.

A negative TTE, therefore, does not exclude aortic dissection [18]. To avoid a delay in diagnosis, TTE is not the modality of choice in suspected acute aortic syndrome. Its utility, therefore, in the emergency setting is in the rapid assessment of complications of dissection, such as aortic valve dysfunction, pericardial tamponade, or wall motion abnormalities.

**Prognosis**

The risk of death is increased in patients who present with or develop complications of pericardial tamponade, involvement of coronary arteries causing acute myocardial ischaemia/infarction, or malperfusion of the brain [20].

Other predictors of increased in-hospital death include age ≥70 years old, hypotension or cardiac tamponade, kidney failure, and pulse deficits. Less appreciated predisposing factors for Type A dissection include prior cardiac and valvular surgery (15%) and iatrogenic dissection occurring during cardiac surgery or catheterization (5%); iatrogenic aortic dissection carries a mortality that is slightly higher than non-iatrogenic (35 vs. 24%) [21,22].

**Conclusion**

Scientific societies advice to utilize a decisional flow chart which rank the patients to be affected from a AAD. Whether the patient is included in high-risk for referred conditions or pain or examinations features (Table 1) CT scan or TOE must be applied as soon as possible to avoid the delay in the diagnosis.
References

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