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Mini Review

Acute Aortic Syndrome: Current Perspectives and Future Directions

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ABSTRACT

Acute Aortic Syndrome (AAS) is a life-threatening condition. This review covers important issues about clinical features, classification, epidemiology, diagnostic and therapeutic strategy for these patients. The role of the aorta code, aorta team and aorta centers has been highlighted in the management of these patients to improve clinical outcomes. Treatment approach is discussed according to the type of aortic lesion and patient's clinical profile.

Keywords: Acute aortic syndrome; Multidisciplinary teams; Aortic surgery; Endovascular treatment

INTRODUCTION

Initially described in 1998, and revisited in 2009 the term 'Acute Aortic Syndrome' (AAS) includes four entities with a common clinical presentation: Classic aortic dissection, intramural aortic hematoma, penetrating atherosclerotic ulcer and incomplete dissection. Although the pathological substrate is different in each one, the aortic wall is acutely injured in all of them [1,2].

They typically present separately in a particular patient, but can also appear together in the same patient. The main purpose of the present document is review the current status of AAS and future directions on the diagnosis and management of this serious condition.

CLASSIFICATION

Many anatomic, prognostic and etiological classifications for AAS have been proposed [3-5]. Among them, Stanford and DeBakey classifications are the most widespread as a result of their simplicity and prognostic value [4,5]. However, the aortic arch involvement is disregarded in these classifications. The term 'non-A non-B' has been used to describe those cases where the lesion is confined to the aortic arch alone or the arch is involved by retrograde propagation of a lesion originating distal to the left subclavian artery without reaching the ascending aorta (Figure 1) [6-8]. Aortic arch involvement is a challenging condition in the therapeutic approach to the AAS, both for open surgery and for endovascular treatment.

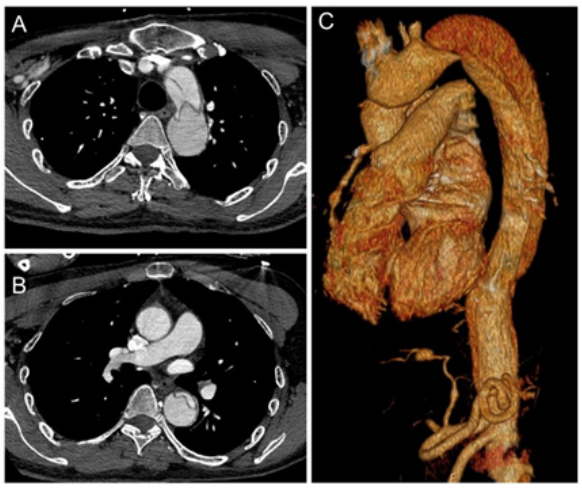

Figure 1: 'Non-A Non-B' aortic dissection. Series of CT scan images showing a patient with a previous Bentall-De Bono surgery. A) New-on set'non-A non-B' aortic dissection with a large portal of entry located at the aortic arch. B) Prior graft tube at the ascending aorta and aortic dissection extending to the descending aorta. C) Three-dimensional reconstruction of the 'non-A non-B' aortic dissection.

EPIDEMIOLOGY

Data on the epidemiology of AAS are scarce and imprecise [3,9]. Hospital-based registries underestimate the true incidence of AAS, as none of them include out of hospital deaths [10,11]. Population

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based studies report an incidence of 6-7.7/100000 person-year [12,13]. Although most registries focused on classic aortic dissection, the Olmstead County population-based study included all AAS [14]. Their results showed that the incidence of penetrating aortic ulcer (2.1/100000 person-years) and intramural hematoma (1.2 per 100 000 person-years) were much lower than that of aortic dissection (4.4). This study revealed two interesting issues: The incidence of AAS remained stable over 20 years and patients with AAS had twice the mortality rate at 5, 10 and 20 years compared to controls. Thus, a strong effort should be made in order to improve diagnosis and prognosis of patients with AAS.

CLINICAL PRESENTATION AND DIAGNOSIS

Diagnosis of AAS requires a high level of clinical suspicion. Three main features make AAS diagnosis challenging: a low prevalence, the absence of specific biomarkers and a highly variable clinical presentation [15,16]. Therefore, maintaining a high level of clinical suspicion is the cornerstone of AAS diagnosis. In that sense, several initiatives such as AAS training sessions, education programs, electronic diagnostic tools, and specifically designed algorithms for patients with chest pain may increase the level of alertness.

We propose a three-step algorithm to facilitate diagnosis in the emergency room [17]. In a first step we evaluate the probability of AAS based on three categories (Figure 2): Risk factors, pain features and physical examination. Risk factors for AAS are well known [3,17,18]. Among them, severe chronic hypertension is the most frequent, and strongly related to development of AAS [19]. In fact, hypertensive patients have more than two-fold increased risk of AAS and hypertension confers a population-attributable risk of 54% for AAS [19]. Some other risk conditions such as prior aortic aneurysm, aortic valve disease, and bicuspid aortic valve, past family history of aortic disease and connective tissue diseases (i.e., Marfan, Loeys-Dietz syndrome) should be considered. Added to risk factors, chest pain is the most common clinical symptom among patients with AAS [3,10,8,20]. The so-called "aortic pain", is described as abrupt, sharp, intense, tearing, frequently irradiated in the direction of the lesion's progression and according to the aortic branches involved. The chest is the most frequent location of pain in patients with type A AAS, whereas aortic pain is usually located in the back in patients with type B AAS. It is less frequent having an AAS without pain. Finally, comprehensive physical exam may reveal typical signs of AAS, such as pulse deficit/asymmetric pulses or an aortic regurgitation murmur. However, AAS must also be suspected in patients admitted with severe hypotension, syncope or shock, especially if they have chest pain. This pretest probability assessment is a class I recommendation [3,18]. Detection of ≥ 1 risk factors should raise suspicion of a potential AAS and lead us to the next step.

The second step includes the usual complementary tests that must be performed in any patient with chest pain, (i.e., ECG, chest X-ray and laboratory test/biomarkers) [17,21]. This is a key part of the diagnostic algorithm, where we make the differential diagnosis with the two most frequent life-threatening diseases causing chest pain: Acute coronary syndrome and pulmonary embolism. Combination of ECG and troponin levels has a high sensitivity in the diagnosis of Acute Coronary Syndrome (ACS) [21]. Differential diagnosis between AAS and ACS is essential, considering that antiplatelet drugs needed for ACS may complicate the treatment and deeply worsen prognosis of patients with AAS. Nevertheless, a high troponin level does not absolutely rule out AAS. Increased troponin values in AAS may be secondary to involvement of a coronary artery by the dissection flap or due to myocardial ischemia exacerbated by acute aortic regurgitation or hypotension.

The usefulness of D-dimer in diagnosis of AD has been widely proven in numerous studies [22-24]. When included in an algorithm based on the a priori probability of AAS, D-dimer levels increase the value of the diagnostic strategy [23,25]. Thus, when a particular patient has none or one risk factor for AAS and the D-dimer is negative, the negative predictive value of having an AAS is almost 100%, and the sensitivity and specificity are 98.8% and 57.3%, respectively (Figure 3) [23].

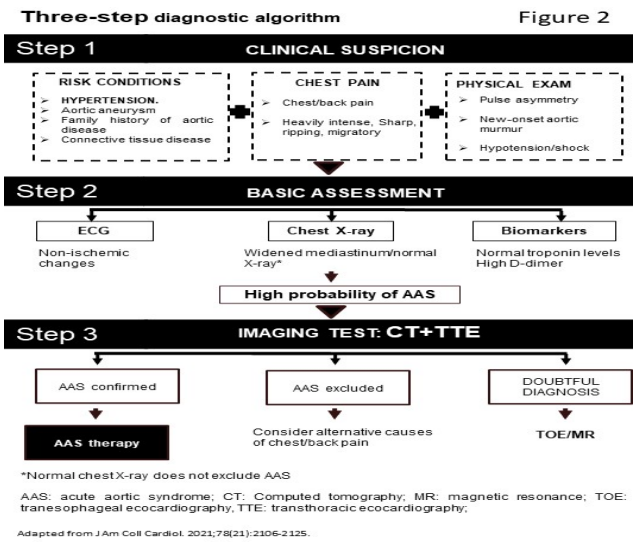

Figure 2: Three-step diagnostic algorithm for patients with suspected acute aortic syndrome.

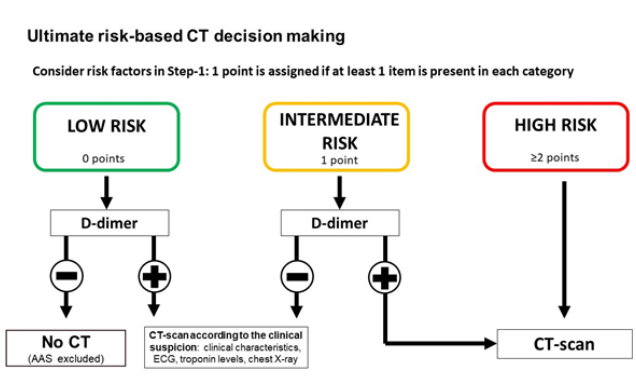

Figure 3: Ultimate risk-based CT decision making.

The second step is key step, since in patients with "aortic pain", the combination of a normal ECG, normal troponins, and increased levels of D-dimers, AAS/pulmonary embolism must be suspected and should move us directly to the third step, imaging tests, which will give us the confirmation diagnosis [18,26].

To confirm the presence of an AAS, Computed Tomography (CT) is the imaging test of choice in the emergency department. This technique has several advantages that makes it ideal: rapid acquisition times, extensive availability in emergency departments of most hospitals and full anatomic assessment of the whole aorta and aortic branch vessels [26,27]. A CT scan, with and without contrast, of the entire aorta (from the cervical branches to

the iliofemoral arteries) should be performed to all patients with intermediate/high suspicion of AAS (Figure 3) [28].

Additionally, focus TTE may contribute to the assessment of other relevant issues (pericardial effusion, aortic valve regurgitation, and ventricular function) [3,29]. The combination of bedside TTE and CT scan is an excellent diagnostic approach, as it allows us to examine the full aorta length and to evaluate potential AAS-related complications.

Transesophageal echocardiography should be considered intra operatively to help in the procedure guidance and for immediate evaluation of the result for both open surgery and endovascular therapy [30].

MULTIDISCIPLINARY TEAMS IN THE MANAGEMENT OF AAS

Healthcare networks have proven their utility and prognostic impact in acute cardiovascular diseases [31,32]. Acute aortic syndrome is not an exception and there is strong evidence on the volume-outcome relationship in AAS surgery [33-35]. Thus, centralization of AAS patient management must be a primary goal in cardiovascular healthcare organizations [36].

Success in AAS management begins with early recognition and quick transfer to the aorta referral center. In that sense, initiatives such as an 'Aorta Code' pursue a triple target (Figure 4) (A) to increase awareness of AAS among emergency care providers to improve diagnosis of AAS, (B) to decrease the time from diagnosis to treatment, and (C) to provide the optimal treatment by activation of a 24/7 available highly specialized aorta team. The final purpose of an 'Aorta Code' protocol is improving clinical outcomes and survival of AAS patients. An Aorta Team is a necessary step in the standard of care of patients with AAS [17,18,36]. Early evaluation of patients in the acute phase, proper medical management in the intensive cardiovascular care unit both pre and post-surgery, surgical/endovascular treatment of aortic patients, all of them require a high level of expertise. Establishing specialized aorta teams, including clinical cardiologists, experts in cardiac imaging, cardiac surgeons, vascular surgeons, radiologists, vascular interventional radiologists, and anesthesiologists, decrease variability in diagnosis and treatment decision making (Figure 5) [17,18,36]. Finally, low incidence diseases demanding highly complex treatment techniques justify centralization of care in high-volume centers with high-volume surgeons, as it offers the best opportunity to improve outcomes [34,35].

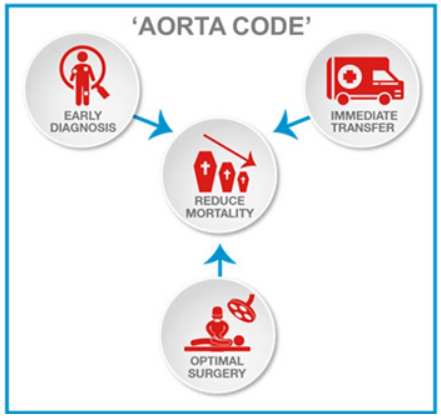

Figure 4: 'Aorta code'.

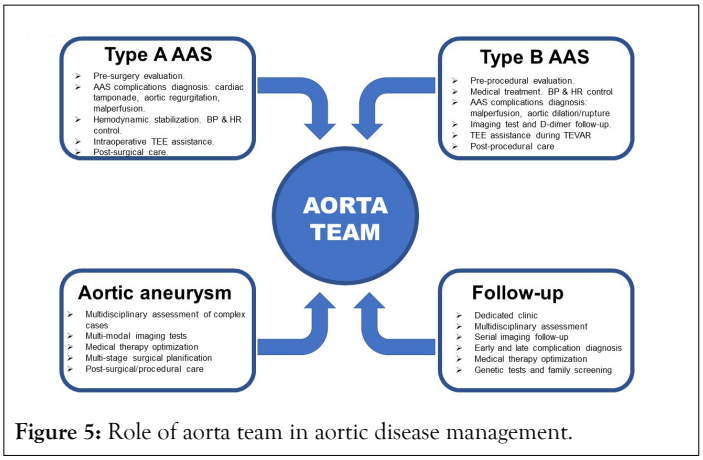

Figure 5: Role of aorta team in aortic disease management.

MANAGEMENT OF PATIENTS WITH AAS

Emergent open surgical repair is the standard approach for patients with type A-AAS. The main purpose is to prevent aortic rupture, repair aortic regurgitation and re-direct the blood flow to the true lumen to avoid organ malperfusion [17]. When the aortic root is not affected, a supra-coronary tube-graft with or without valve replacement/repair may be sufficient to achieve this goal (Figure 6). In cases of aortic root involvement, a Bentall-De Bono procedure is the most frequently used approach, either with mechanical or biological valve prosthesis [37,38]. However, several alternative techniques should be considered in high-volume centers/experienced surgeons. Valve-sparing aortic root operations, such as David or Yacoub techniques (including modifications of both), should be considered in selected patients (normal aortic valves, young patients, hemodynamic stability, absence of peripheral severe complications) (Figure 7) [38,39]. Patients with aortic annulus enlargement, entrance tear at the aortic root and most patients with connective tissue diseases will require an aortic root replacement with a composite valve tube or aortic valve resuspension.

Regarding the aortic arch, 'hemiarch' replacement without involvement of the supra-aortic trunks continues to be the standard surgical approach in patients with type A AAS. There is growing evidence on the safety and long-term benefits of total arch replacement [40-42]. Thus, in either of the following circumstances: a dilated aortic arch, an entrance tear at the aortic arch, arch or proximal descending re-entrance tears, a dilated proximal descending aorta, or branch vessel dissection, a total arch replacement surgery should be considered in experienced centers [17,41,43]. This approach will allow distal aortic remodeling and prevent further reoperations. The "frozen elephant trunk" technique using a hybrid prosthesis has emerged as a feasible approach in the AAS scenario with good results (Figures6 and 7). It is also possible to perform an endovascular repair of the distal descending and abdominal aorta by splicing additional endoprosthesis to the distal end of the hybrid prosthesis (Figure 8).

Regarding type B AAS, Thoracic Endovascular Aortic Repair (TEVAR) is advisable when complications appear, i.e. hemodynamic instability, malperfusion, quick aortic dilation and aortic rupture. In other risky circumstances such as an aortic diameter>44 mm, a false lumen diameter>22 mm, a large proximal entrance tears, and refractory pain or hypertension (Figure 9) should also be considered [3,7]. When possible, TEVAR should be deferred as it has shown good results and fewer periprocedural complications [44,45]. All other patients with type B AAS should be treated.

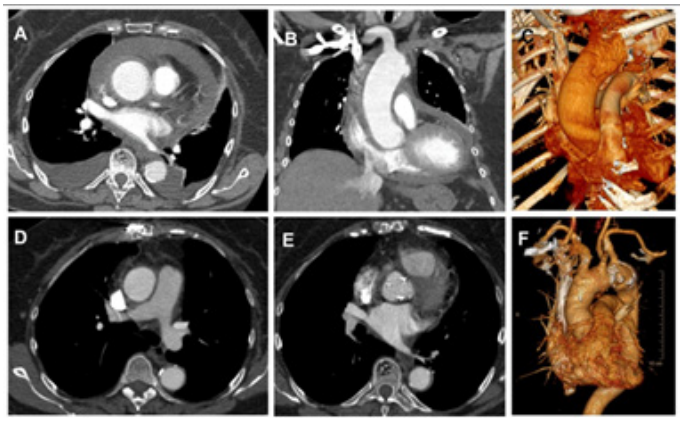

Figure 6: (A-C)Acute aortic dissection with thrombosed false lumen; portal of entry at the distal portion of the ascending aorta. (D-F) Post-surgery CT scan showing supra-coronary tube graft and aortic bioprosthesis.

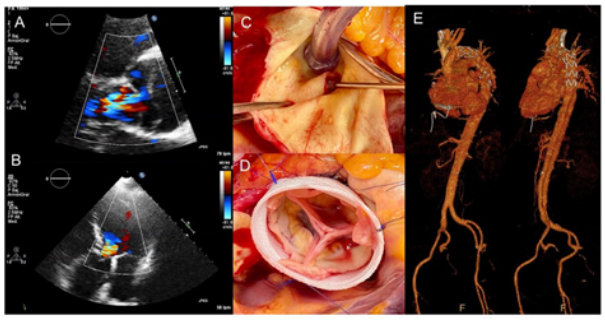

Figure 7: (A and B)Marfan patient with a type A acute aortic dissection and severe aortic regurgitation. C) Portal of entry in the ascending aorta. D) David operation. E) E-vita open prosthesis with flow redirection to the true lumen.

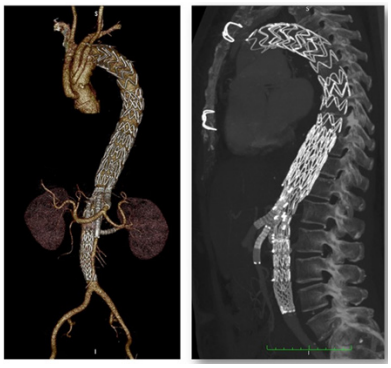

Figure 8: T-branch endoprosthesis attached to a previously implanted frozen elephant trunk. Complete thoraco-abdominal aortic repair.

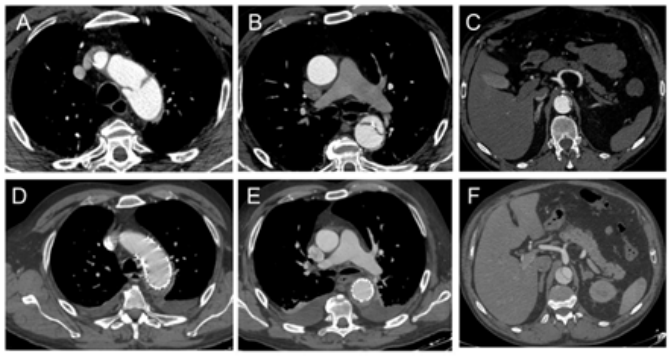

Figure 9: (A-C)Acute thoracic endovascular aortic repair in a patient with type B acute aortic dissection with collapsed true lumen and visceral malperfusion. (D-F)Control CT scan after TEVAR showing true lumen re-expansion.

Aortic Intramural Hematoma (AIH) deserves several special considerations. First, initial medical management with a watchful waiting strategy might be a safe option for stable patients with type A AIH. In this situation, serial clinical and radiological control must be obtained and surgery performed in case of hemodynamic deterioration or when high-risk imaging signs appear [17,46,47]. Second, absence of a clear entrance tear to be sealed or the absence of a healthy aortic proximal landing zone are usual unknowns in TEVAR in patients with type B AIH. Medical management is the standard approach of patients with type B AIH and the presence of ulcer-like projections deserve a close imaging follow-up, as they are associated with a higher risk of complication, especially when they are present on the initial CT [48-50].

CONCLUSION

AAS continues being the most dreaded cardiovascular syndrome. Over the last years, important improvements in the imaging techniques and mostly in the "surgical/endovascular" treatment have been achieved. Initiatives such as the aorta teams, aorta codes and referral aorta centers should be pursued. We look forward to hearing from emergent technologies and more specific biomarkers that will help us to perform an early diagnose of patients with AAS.

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