Acute aortic syndromes in India: the need for a nationwide program to increase awareness

Shetty, V.; Shetty, D.; Grewal, N.

Indian Heart Journal 74 (2022) 76–78

Contents lists available at ScienceDirect

Indian Heart Journal

journal homepage: www.elsevier.com/locate/ihj

Opinion Paper

Acute aortic syndromes in India: The need for a nationwide program to increase awareness

Varun Shetty [a], Deviprasad Shetty [a], Nimrat Grewal [b, c, *]

[a] Department of Cardiac Surgery, Narayana Institute of Cardiac Sciences, Bangalore, India
[b] Department of Cardiothoracic Surgery, Leiden University Medical Center, Leiden, the Netherlands
[c] Department of Anatomy and Embryology, Leiden University Medical Center, Leiden, the Netherlands

ARTICLE INFO

Article history:
Received 20 July 2021
Received in revised form
28 November 2021
Accepted 30 November 2021
Available online 2 December 2021

Keywords:
Aorta
Aneurysm
Dissection
Aortopathy
Surgery

ABSTRACT

Acute aortic dissection is a rare, but potentially life-threatening and time-critical condition that is frequently misdiagnosed. Therefore, its prompt and proper diagnosis is vital to increase a patient's chance of survival and to prevent grievous complications. Raising awareness and educating the general population and healthcare professionals about an aortic dissection is mandatory, for early diagnosis and improving the chances of survival.

1. Introduction

Acute aortic syndrome (AAS) refers to a group of life-threatening aortic pathologies including aortic dissection, intramural hematoma and penetrating atherosclerotic aortic ulcer. This uncommon, but potentially lethal condition is challenging to diagnose. The low incidence of AAS, varied presenting symptoms, and lack of a standard diagnostic pathway has led to a high misdiagnosis rate.

An aortic dissection is a serious condition in which the inner layer of the aorta tears, blood surges through the tear, causing the inner and middle layers of the aorta to separate (dissect).[1] Based on their location and extent, an aortic dissection is classified as Stanford type-A aortic dissections (TAAD) (dissections involving the aortic root and/or ascending aorta, which can propagate to the aortic arch and further into the descending aorta), and type-B dissections (dissections that do not involve the ascending aorta).[2]

An acute type A aortic dissection is a medical catastrophe with a described estimated mortality of 26% in patients undergoing acute surgery and up to 58% in patients not receiving surgical treatment.[3–5] It is known that the associated mortality rate is 1–2% per hour immediately after symptom onset in historical untreated patients and approximately half of patients with a TAAD die before reaching a specializing centre.[3–5] Timely intervention, whether medical or surgical, is essential to yield the best short-term and long-term results for patients with an acute dissection. Patients initially presenting to non-tertiary hospitals experience delays in both recognition and surgery.[6] The relative infrequency of TAAD, coupled with clinical presentations that may mimic more common problems, such as acute coronary syndromes, can impede prompt establishment of the TAAD diagnosis.

Significant delays may exist between hospital arrival and definitive diagnosis and treatment. One of the reasons for the delay in diagnosis is the paucity of coronary care units and computerized tomography (CT) scan centres. Nowadays, the sensitivity and specificity of transthoracic echocardiography is close to CT.[7] With a good quality echo, the diagnosis of TAAD can be made certain and the patient can be transferred immediately to a centre with the requisite experience. However, the reality we face in clinical practice is quite different, often there is a delay in diagnosis and the patients who do reach the tertiary centres are in either cardiogenic shock, ischemic end organ injury or in tamponade. Consequently, surgical centres are reluctant to operate on such patients because of the high mortality.

* Corresponding author. Department of Cardiothoracic Surgery, Leiden University Medical Center (LUMC) Albinusdreef 2, 2333, ZA Leiden, the Netherlands.
E-mail address: N.Grewal@lumc.nl (N. Grewal).

https://doi.org/10.1016/j.ihj.2021.11.188

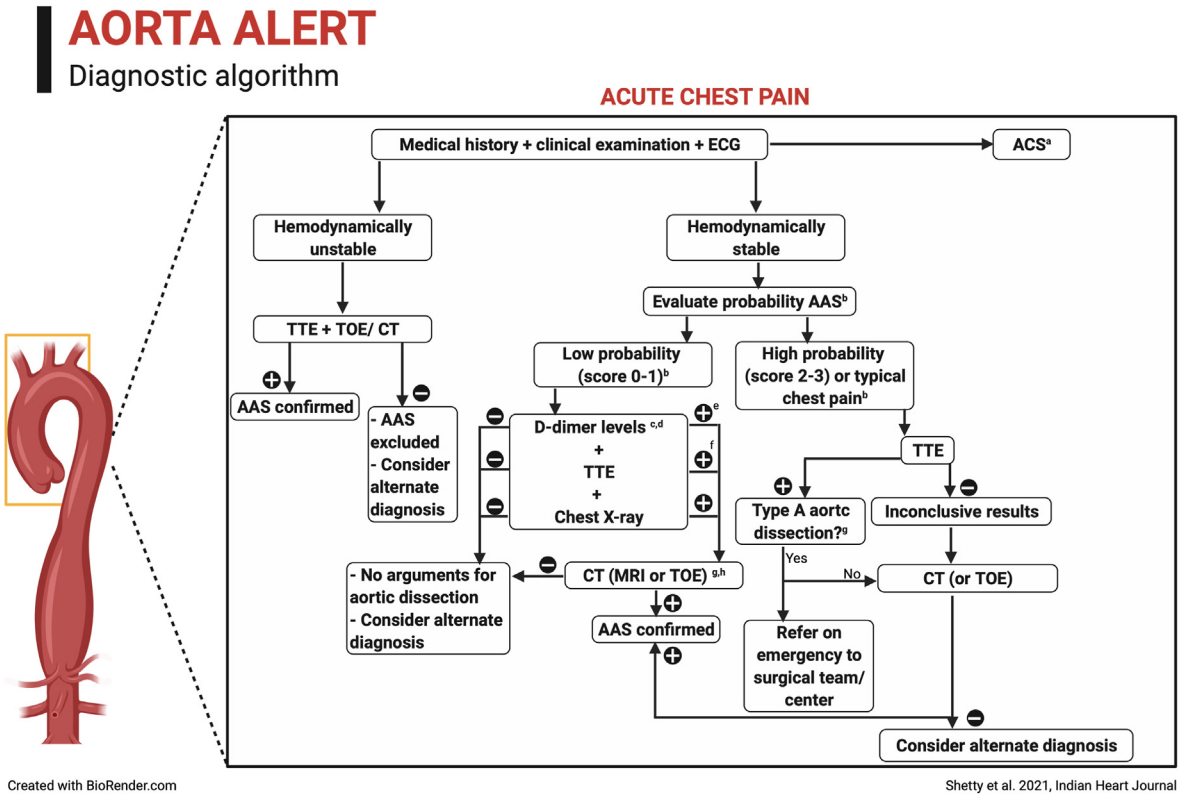

Fig. 1. a In rare cases ACS can be associated with AAS; b Factors increasing the probability of acute aortic syndrome. **Conditions:** Marfan syndrome (or other connective tissue disease); Family history of aortic disease; Known aortic valve disease; Known thoracic aortic aneurysm; Previous aortic manipulation (including cardiac surgery). **Pain features:** Chest, back, or abdominal pain characterized by any of: Abrupt onset, Severe intensity, Ripping or tearing. **Examination features:** Evidence of perfusion deficit: Pulse deficit, Systolic blood pressure difference in upper limbs, Focal neurologic deficit (in conjunction with pain); Aortic diastolic murmur (new and with pain); Hypotension or shock.The presence of features from one of the groups amounts to 1 point; from two groups, 2 points; and from three groups, 3 points. c Preferably point of care, otherwise classical d Also troponin levels to detect non-ST-segment elevation myocardial infarction e Features of aortic dissection f Enlargement of the mediastinum g Proof of type A dissection by the presence of a flap, aortic regurgitation, and/ or pericardial effusionh Depending on local availability, patients characteristics and physicians experience AAS: acute aortic syndrome; ACS: acute coronary syndrome; CT: computed tomography; MRI: magnetic resonance imaging; TOE: transoesophageal echocardiography; TTE: transthoracic echocardiography.

In order to address these challenges, national awareness programs are mandatory. The purpose is to support clinical decision making in cases of suspected aortic dissection, thereby minimizing diagnostic delays and misdiagnosis. As yet, there is a paucity of data describing the effect of time interval between first onset of symptoms, diagnosis and surgery for an acute Type A aortic dissection. Moreover, the precise clinical and diagnostic factors that contribute to these delays are unknown. An increased general awareness of acute aortic dissection is therefore necessary as early diagnosis, before the onset of irreversible ischaemic events, allows a high surgical success rate.

A nationwide quality improvement project to raise awareness and improve diagnosis of acute aortic dissection. Such a project aims to better reach out to the general population and healthcare professionals to equip them with the ability to recognize the signs of acute aortic dissection which is an emergency that demands immediate medical attention resulting in earlier diagnoses and appropriate intervention when indicated. Due to the variability of symptoms, patients may delay seeking help and treatment. Alternatively, patients who appear stable may first contact their general practitioner for advice and, due to the relative rarity and lack of awareness of the condition they may be misdiagnosed.

Furthermore, acute aortic Type A dissection is managed by specialist tertiary centers where there tends to be a greater awareness and knowledge of the typical presentation. Small district general hospitals may have early contact with such patients, but infrequently manage these patients beyond the initial stabilization period when compared to tertiary cardiac centers. This often causes a delay in the lack of specialist expertise. Through increased awareness, education and research one can however be able to save the lives of people at risk for an acute aortic dissection.

Based on the ESC Guidelines on the diagnosis and treatment of aortic diseases,[8] we highlight a protocol in this opinion paper to aid in decision making (Fig. 1) and also emphasize the requirements to implement this protocol to the Indian setting.

As the condition is often misdiagnosed as an acute coronary syndrome leading to fatal outcomes when high doses of anticoagulants are administered, we urge all medical professionals to use the diagnostic Flow Chart presented in this paper.

On basis of the medical history, clinical examination and ECG an acute coronary syndrome might be diagnosed (Fig. 1). In the emergency setting however it is of utmost importance to consider an AAS in all patients with typical chest, back or abdominal pain, syncope or symptoms consistent with a perfusion deficit.

If the patient is hemodynamically unstable a transthoracic echo is recommended as an initial imaging investigation. If there is a suspicion of an acute aortic syndrome, a transesophageal echo or CT scan is recommended according to local availability and expertise. A negative TOE/CT scan rules out the diagnosis. In hemodynamically stable patients the probability of an acute aortic syndrome should be evaluated. The diagnostic work-up to confirm or rule out a type a dissection is highly dependent on the *a priori* risk of this condition. The diagnostic tests can have different outputs according to the pre-test probability. A risk assessment tool based on three

groups of information is proposed: 1. Predisposing conditions, 2. Presenting pain features, and 3. Clinical examination findings. A scoring system is proposed that considers the number of groups involved ranging from 0 to 3. The presence of 0, 1, 2 or 3 groups of information is associated with increasing pre-test probability, which should be taken into account in the diagnostic approach to all AAS, as is shown in the diagnostic flow chart.

In high probability patients a transthoracic echo is performed. In case a type A dissection is diagnosed by the presence of flap, aortic regurgitation, and/or pericardial effusion, the patient is referred to a surgical team/center. If the transthoracic echo doesn't show signs of a TAD or the echo is inconclusive, a CT scan or transesophageal echo is performed. If that concludes the diagnosis the patient is referred, else an alternate diagnosis is considered. In case of initially negative imaging with persistence of suspicion of AAS, repetitive imaging (CT or MRI) is recommended.

In the low probability patients, with a score of 0–1, D-dimer levels can be checked. In case of suspicion of AAS, the interpretation of biomarkers should always be considered along with the pre-test clinical probability. In case of low clinical probability of AAS, negative D-dimer levels should be considered as ruling out the diagnosis. In case of intermediate clinical probability of AAS with a positive (point-of-care) D-dimer test, further imaging tests should be considered. In patients with high probability (risk score 2 or 3), testing of D-dimers is not recommended. If the TOE or chest X-ray raises suspicion of a TAD, additional imaging is recommended which can confirm the diagnosis or rule it out.

Summarizing the flow chart, considering an AAS in the differential diagnosis is the key. Followed by risk stratifying patients by the acute aortic syndrome risk score (low, intermediate or high) which is important, especially in community hospitals or clinics (usually not equipped with advanced imaging technology). Once the initial diagnostic decision is made in order to define the appropriate and available imaging test (transthoracic echo + CT scan or transesophageal echo), the aorta team should be alerted and the patient transferred to a tertiary centre with availability and expertise for imaging and management of aortic disease. Reconstructing the healthcare by sharing knowledge, educational material, collaborative data collection, educative webinars, and national guidance should lead to a structured assessment which would allow rapid confirmation of diagnosis and subsequently reduce the likelihood of mismanagement.

References

1. Grewal N, Velders BJJ, Gittenberger-de Groot AC, et al. A systematic histopathologic evaluation of type-A aortic dissections implies a uniform multiple-hit causation. *J. cardiovasc. dev. disease*. 2021;8.
2. Daily PO, Trueblood HW, Stinson EB, Wuerflein RD, Shumway NE. Management of acute aortic dissections. *Ann Thorac Surg*. 1970;10:237–247.
3. Hirst Jr AE, Johns Jr VJ, Kime Jr SW. Dissecting aneurysm of the aorta: a review of 505 cases. *Medicine*. 1958;37:217–279.
4. Mészáros I, Mórocz J, Szlávi J, et al. Epidemiology and clinicopathology of aortic dissection. *Chest*. 2000;117:1271–1278.
5. Tsai TT, Nienaber CA, Eagle KA. Acute aortic syndromes. *Circulation*. 2005;112:3802–3813.
6. Harris KM, Strauss CE, Eagle KA, et al. Correlates of delayed recognition and treatment of acute type A aortic dissection: the International Registry of Acute Aortic Dissection (IRAD). *Circulation*. 2011;124:1911–1918.
7. Cecconi M, Chirillo F, Costantini C, et al. The role of transthoracic echocardiography in the diagnosis and management of acute type A aortic syndrome. *Am Heart J*. 2012;163:112–118.
8. Erbel R, Aboyans V, Boileau C, et al. ESC Guidelines on the diagnosis and treatment of aortic diseases: document covering acute and chronic aortic diseases of the thoracic and abdominal aorta of the adult. The Task Force for the Diagnosis and Treatment of Aortic Diseases of the European Society of Cardiology (ESC). *Eur Heart J*. 2014;35:2873–2926, 2014.