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CASE REPORT

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An atypical type-A dissection of aorta: case report.

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ABSTRACT

Despite a dissecting aortic aneurysm being a surgical emergency, it is frequently misdiagnosed in the Emergency Department. It is no surprise that the thoracic aorta's walls can rupture catastrophically with over 200 million litres of blood travelling through it in an average lifetime. Chest pain +1 (symptom) should prompt a differential diagnosis of a thoracic aortic dissection. Unfortunately, up to 33% of aortic dissection patients are misdiagnosed in the ED. Although the diagnostic imaging of choice is the CT aortogram, the question is whether transthoracic Echocardiography (TTE) is a robust enough imaging modality to rule in or rule out the presence of a type A thoracic dissection on first presentation in the ED and if it is helpful in the ED without immediate access to a cardiothoracic surgeon? This case highlights the importance of Point-of-care ultrasonography (POCUS) in helping with risk stratification and patient streaming in the emergency department. The likelihood of missing an ascending thoracic aorta pathology is reduced if the image acquisition is of good quality.

KEY WORDS: Acute Aortic Syndrome (AAS), Emergency Medicine, Point-of-care Ultrasonography (POCUS), Emergency imaging, Case report.

INTRODUCTION

Over an average lifetime, nearly 200 million litres of blood travel through the thoracic aorta [1]. It comes to no surprise that if this vessel is exposed to further stresses, such as hypertension, ageing, diabetes or congenitally weak walls (Marfan's, Ehler Danlos), catastrophic rupture of the vessel walls can occur. Other major risk factors are the presence of a biscupid aortic valve, the use of cocaine, vascular inflammation and smoking. The rupture of the thin tunica intima layer of the vessel is called a dissection and can lead to rupture of the aorta or occlude branches originating from the aorta, leading to malperfusion of the target organs. The aortic dissection is one of the processes termed acute aortic syndrome (AAS). The other syndromes included in the umbrella term AAS are intramural haematoma (IMH) and penetrating aortic ulcer (PAU) and traumatic aortic injury (TAI). The incidence of a dissecting thoracic aorta has been reported to be approximately 2.7- 3.5 per 100000 per year [2] and is twice as common in men than women. Howard et al. (2014) [3] stipulated that the currently believed incidence is under-representative due to the high pre-hospital mortality in the elderly who do not get a post mortem. Furthermore, with an aging population, this incidence is predicted to rise over the next decade.

A commonly used classification for thoracic aneurysms/dissections is the Stanford classification: Type A affects the ascending aorta (with or without descending aorta involvement) and type B only affects the descending aorta. Type A is almost twice as common as type B. Unfortunately, the clinical presentation can be subtle and up to 33% of patients with an aortic dissection are misdiagnosed on initial presentation in the ED [4]. The textbook widened mediastinum on chest radiograph is only present in approximately 63% of patients [5].

Once suspected, the diagnostic imaging of choice is the CT aortogram (CTA) as it can clearly identify a dissection, thrombus, IMH, or PAU with excellent sensitivity and specificity of 100% and 98% respectively [1]. Access/time to CT scan can be variable but once started, the aortogram is a rapid investigation. MRI is another great imaging modality but due to the long duration required to acquire the images, it is not useful in the acute setting. Transoesophagal echocardiography (TOE) is a low radiation imaging tool that has been demonstrated to reach a sensitivity of 99% and a specificity of 89% but requires the patient to be sedated or intubated which is a limiting factor in the undifferentiated/unstable patient in the Emergency Department. The question therefore is whether Transthoracic Echocardiography (TTE) is a robust enough imaging modality to rule in or rule out the presence of a type A thoracic dissection on first presentation in the ED. Furthermore, if the presence of ASS is strongly suspected on TTE finding, can it help in the Emergency Department without direct access to a cardiothoracic surgeon on site?

CASE REPORT

PATIENT INFORMATION: At 12:32, London Ambulance Services (LAS) attended an 84 year old lady who had a pre-syncopal episode whilst shopping: she was standing at the tills at a superstore when she suddenly "felt hot and clammy" with mild blurring of her vision.

CLINICAL FINDINGS: With LAS,her blood pressure (BP) was 90/35, the heart rate (HR) of approximately 67 beats per minute (bpm). The rest of the initial assessment was normal. On the way to the hospital, she became bradycardic (HR 41-46) and the BP dropped to 77/55, which was responsive to a 500ml fluid bolus administered en route. On arrival to the ED at 13:44, her HR was 74, BP 115/65 and was assessed by the ED Senior House Officer(SHO) in the resuscitation room. Her primary survey and venous gas at that time were normal. She felt slightly nauseated but was pain free.

TIMELINE:

- 12:32 - Patient attended by LAS due to pre-syncopal episode and mild blurry vision.
- 13:44 - Arrival to ED, diagnosed new onset AF with Low cardiac output.
- 14:15 - ED Consultant review and TTE showing suspicion of Aortic Aneurysm.
- 14:30 - Patient transferred to the CT scanner.
- 14:40 - Patient lost cardiac output.

DIAGNOSTIC ASSESSMENT: Her first ECG showed a new diagnosis of atrial fibrillation (AF) (figure 1) and the working diagnosis of the treating doctor was new onset of AF with reduced cardiac output.

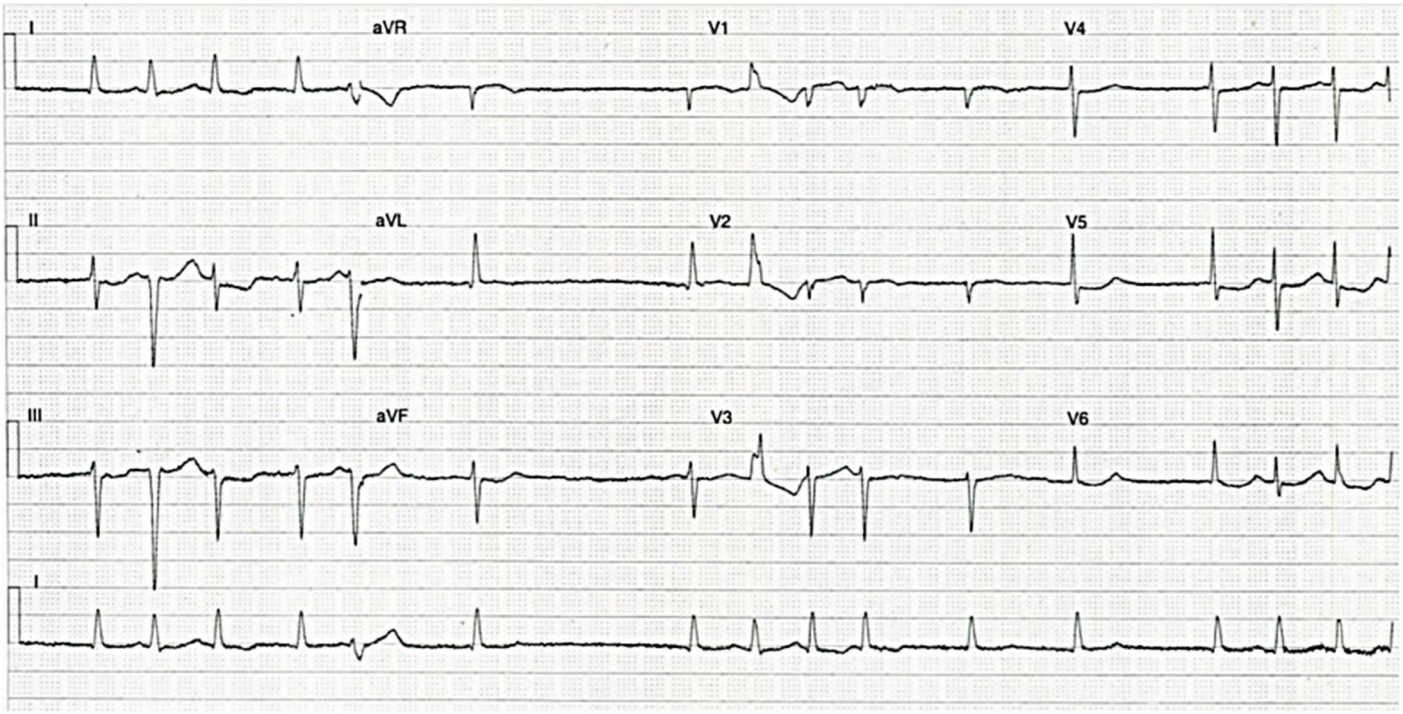

Figure 1. ECG on arrival to the ED. Demonstrating atrial fibrillation, left axis deviation, ventricular ectopics and some infero-lateral ischaemic changes.

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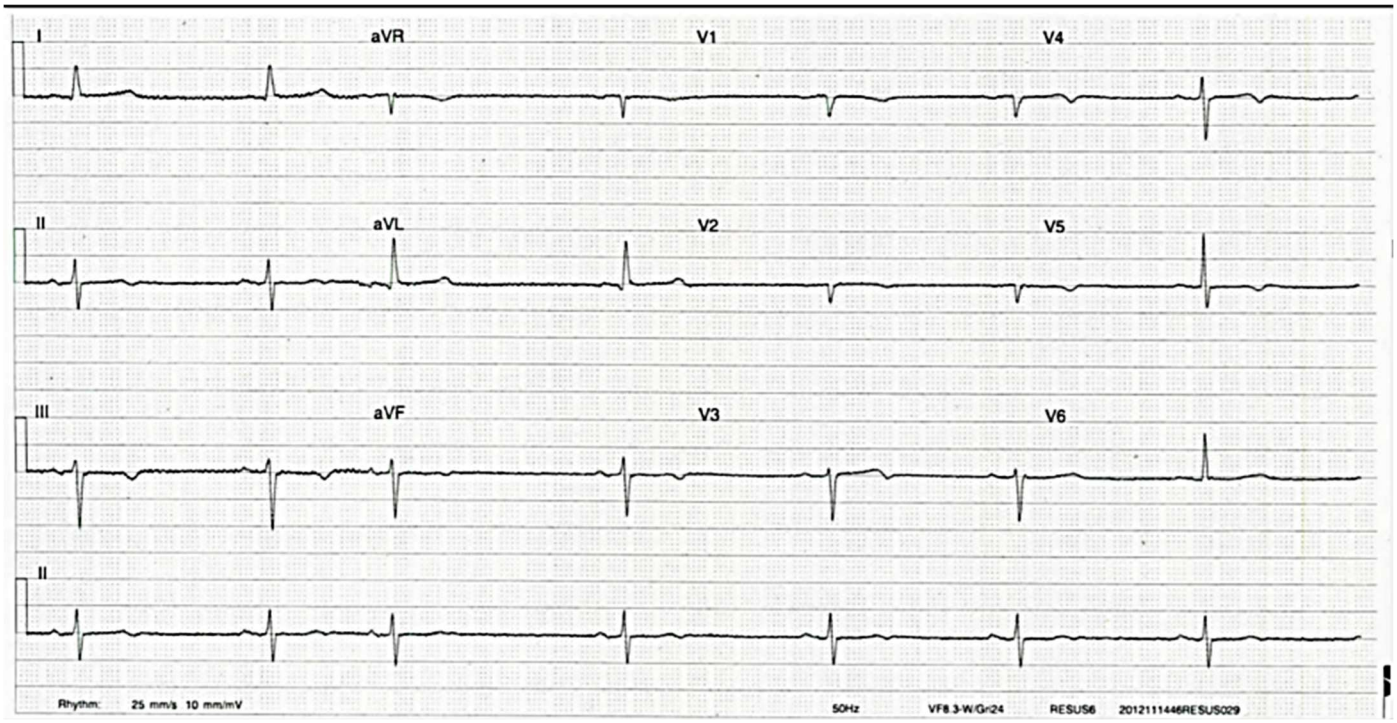
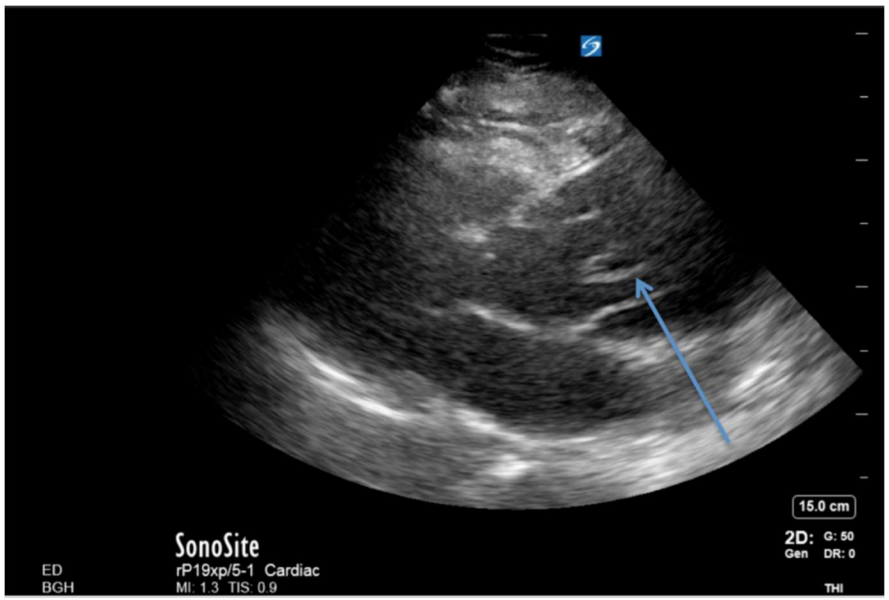


Figure 2. 2nd ECG showing sinus arrhythmia, biphasic T waves in v3-v5.

This case was presented to the A&E consultant at 14:15 who reviewed her second ECG (figure 2) which now showed a sinus rhythm with a sinus arrhythmia and small QRS complexes. The Consultant performed a routine bedside echocardiograph (echo) to look for a pericardial effusion. At this point she started to vomit and became more diaphoretic. Due to the acuity of the presentation, only the parasternal long axis view was obtained (picture 1).



Picture 1. Parasternal long axis (image capture from cineloop, video demonstrating mobility of flap available) demonstrating a dilated aortic root and intra-luminal flap (arrow).

The aortic root dilatation and visible intraluminal flap raised the concern of a type A thoracic aortic aneurysm dissection. Whilst the SHO organised the CT-Aortogram, consultant spoke to the local cardiothoracic team (residing in a different trust, 30 minutes away by blue light) to enquire whether they would accept the patient for transfer based on the echo finding. As she was asymptomatic and normotensive, the cardiothoraicic team advised to obtain a CT-Angiogram Aorta locally and proceed from there. The patient was transferred to the CT room at 14:30 and shortly after developed sternal and intrathoracic pain whilst the contrast infusion was connected. She described how the pain was radiating down her abdomen and then was unable to move her legs. She lost cardiac output at 14:40 just after the CT topogram (Picture 2).

Picture 2. CT topogram obtained just prior to loss of cardiac output. The mediastinum is widenend, further supporting the diagnosis of type A thoracic aneurysm.

OUTCOME: Due to the catastrophic nature of the underlying pathology (detected by the bedside echo), the team was in agreement prior to the cardiac arrest that cardiorespiratory resusciation were not to be started. The suboptimal view of the CT topogram does demonstrate a wide mediastinum and in combination of the echo, rapid clinical course, confirms the suspected diagnosis of type a dissecting thoracic aortic aneurysm. Thanks to POCUS, a futile resuscitation or post mortem were not necessary.

DISCUSSION

A dissecting thoracic aorta is a surgical emergency and every moment counts. It has a high pre-hospital mortality (17%) and once in the department, the mortality rises by one percent every hour [6] if left untreated. If the clinical picture is strongly suggestive of a dissection (characteristic pain, widened mediastinum on chest radiograph, new murmur), speed to organise imaging (CTA) and discussion with/ transfer to the cardiothoracic surgeon is advocated. Then why is such a significant pathology misdiagnosed in up to 33% of first presentations? The difficulty occurs in the subtle presentation: Hagan et al. (2000) [7] demonstrated how out of 289 patients with type A dissection, 12% presented with syncope and of these, 2.2% did not have any pain or neurological symptoms. 30% of all patients with a type A dissection were noted to have new ECG changes, with 15 % looking ischaemic. Hirata et al. (2010) [8] described ST elevation type changes in up to 8.2% in type A dissection presentations. It is therefore not unreasonable to consider the differential diagnosis of type A dissection in any patient presenting with syncope or acute ischaemic ECG changes. Performing a CT angiogram in all of these patients would be of low yield, expose the patient to unnecessary radiation and slow down the Emergency Department flow.

One of the accepted diagnostic tools, the TOE, has been demonstrated to rival the specificity of CTA and MRA but has a sensitivity ranging from 86% to 100% [5]. Kabirdas et al. (2010) [9] demonstrated in their prospective trial that the measurements of the visible aspect of the ascending aorta (proximal to mid ascending aorta) using a TTE were comparable to TOE. The body habitus, emphysematous lungs and even the difficult cooperation of the acutely unwell patient can limit acquisition of diagnostic images using TTE. Ceconi et al. (2012) [10] conducted an 8-year retrospective study looking at all patients presenting to the hospital who had a TTE as the initial diagnostic imaging. 270 patients had a TTE performed by a cardiologist with “high expertise in both TTE and TOE”. The image quality was defined as optimal or suboptimal. The optimal image was defined by full visualisation of valve structures and the ascending aorta within 5 cm of the valve without or with minimal clutter. A suboptimal image was when any of the above criteria were not met. Out of 270 patients, 67 patients had AAS diagnosed on TTE, later confirmed by surgery or autopsy. 244 patients had optimal views and suboptimal in 26. TTE was falsely positive in 19 cases (all had suboptimal views) and falsely negative in 9 (7 had suboptimal views). The findings are summarised in table 1 and 2.

Table 1. Sensitivity, Specificity, PPV and NPV of TTE vs final diagnosis when using both optimal and suboptimal images.

	Sensitivity (%) (95% CI)	Specificity (%) (95% CI)	PPV (%) (95% CI)	NPV (%) (95% CI)
All patients (n = 270)	87 (75-93)	91 (85-94)	75 (64-84)	95 (91-98)
Type A Aortic dissection (n = 62)	90 (79-96)	96 (92-98)	89 (78-95)	97 (92-100)
AAIH (n = 5)	40 (8-83)	94 (88-98)	14 (2-45)	98 (93-100)

NPV- Negative Predictive Value; PPV- Positive Predictive Value; AAIH- Type A Intramural Hematoma

Table 2. Sensitivity, specificity, PPV and NPV in patients with optimal views vs final diagnosis.

Sensitivity (%) (95% CI)	Specificity (%) (95% CI)	PPV (%) (95% CI)	NPV (%) (95% CI)
97 (87-99)	100 (97-100)	100 (92-100)	99 (96-99)

Sobzcyk and Nycz (2015) [11] conducted a 5-year retrospective study comparing the accuracy of TTE in diagnosing a type A aortic dissection to the CT and intraoperative findings. Out of 172 confirmed cases of type A dissection,159 (92.44%) were correctly identified with TTE. Statistically (chi square test), there was no difference between TTE and CT imaging. TTE was also able to detect high risk features such as aortic dilatation, cardiac tamponade, impaired left ventricular ejection fracture and regional wall motion abnormalities. Whilst the positive predictive value is high, a negative TTE still warrants further imaging if the pre-test probability of an type A dissection is high. The limitation of the study lies in the fact that it was done at a cardiovascular centre: all patients were referred in as suspected dissection (cognitive bias) and the TTEs were performed by "experienced echocardiographers' supported by an on call team of 5 cardiologists. This level of expertise is not comparable to a district general hospital that does not have a cardiothoracic or cardiologist resident.

The European society of cardiology [1] has incorporated TTE in the initial assessment of suspected AAS patients: it is rapidly available, has a strong positive predictive value and can facilitate process of the patient towards the final destination of being reviewed by a surgeon (Figure 3). Of note, the only time it is recommended to use TTE as a rule out test if both d-dimers and chest x-ray are normal in a low pre-test probability patient. Any other situation warrants an alternative imaging mode: CT or TOE.

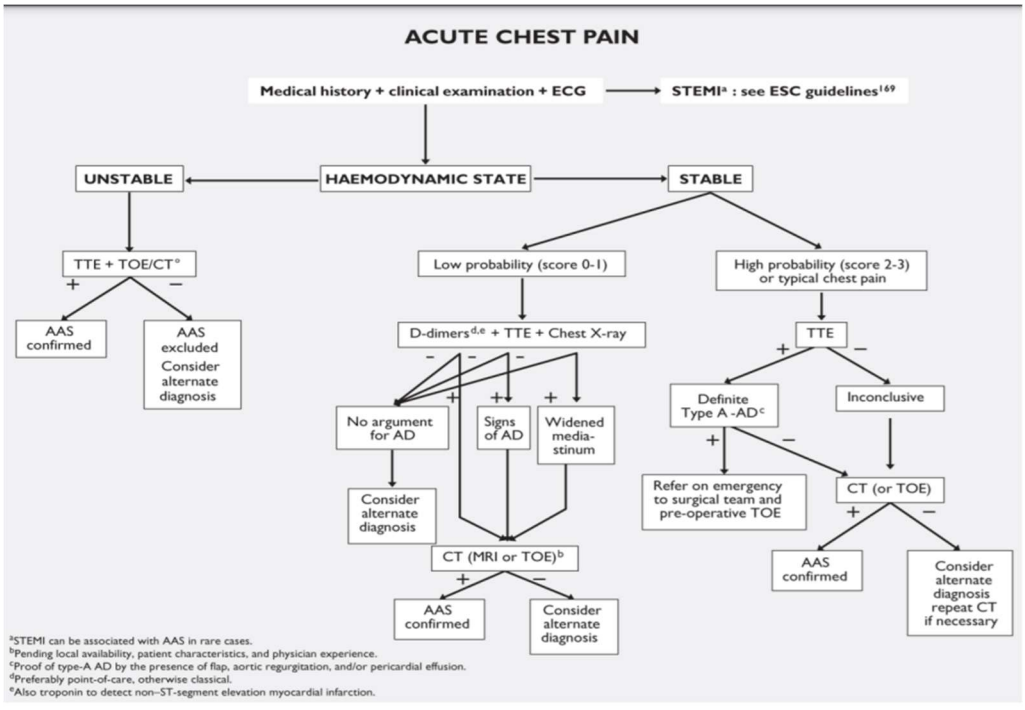


Figure 3. European Society of Cardiology [1] recommendation of approach to patient with suspected AAS.

Older papers show a lower specificity and sensitivity of TTE to rule out AAS but these studies had been done prior to the development of new ultrasound technology, in particular harmonic imaging. The quality of images attainable nowadays is not comparable to 20 years ago. To our knowledge, there have been no prospective diagnostic studies into the role of TTE at present, which makes further critical review difficult.

Based on the current evidence, it would be acceptable to “rule in” a type A aortic dissection when good images have been obtained. It does not have enough diagnostic strength to confidently rule out a type A aortic dissection. Performing a bedside TTE and immediately confirming a dissection would tremendously speed up the onward management in a trust where cardiothoracic surgeons are on site. Unfortunately, with the move to centralisation of specialist services in the UK, most of us work in trusts that do not have cardiothoracics on site. As we experienced first hand with the case described above, the surgeons required a diagnostic CTA prior to accepting the patient for transfer (a 30 minute blue light ambulance journey). Having the CTA images prior to arrival of the patient would allow time for surgical planning but ultimately delays arrival of the patient to the definite location. A direct transfer would not have altered my patient’s outcome as she succumbed to the pathology within 15 minutes of diagnosis.

The limitations of this paper are that although the patients were undifferentiated and TTE is the first imaging modality (no selection bias), the TTE is performed by a highly experienced cardiologist. Furthermore being a retrospectively collected data, detection bias might be present. The question that arises from this paper is whether a cardiothoracic surgeon would accept the critical transfer based on a TTE without definite gold standard imaging or not. If the patient is so unstable that a 15 minute delay via the CT scanner raises their mortality so drastically, it is unlikely they would make it in time to a different trust or survive the operation. They might therefore not be suitable for a transfer in the first instance. At present most trusts do not have an automatic acceptance agreement with the cardiothoracic surgeons (unlike PCI calls/major trauma patients), and a discussion with the lead at the local referral centre would be a place to start.

CONCLUSIONS

The use of point of care ultrasound (POCUS) can help risk stratifying/stream the patient in the Emergency Department. If the image acquisition is of good quality (good visualisation of aortic valve, 4-5cm of ascending aorta), the absence of a bicuspid aortic valve, lack of pericardial effusion, no aortic root dilatation (<4cm) reduces the probability of missing an ascending thoracic aorta pathology. As such if the pre-test probability of AAS was low and no further evidence is visible on adequate TTE views, it seems sensible to forego further investigations looking for AAS. Any patient with a high pre-test probability should undergo a TTE to possibly speed up investigations and referral to the cardiothoracic surgeon but a negative scan (even of good image quality) requires further definite imaging. In conclusion, this case report suggests that all patients with syncope or ischaemic ECGs would benefit from having a POCUS TTE to look for anomalies of the ascending aorta.

SUPPLEMENTARY INFORMATION

Funding: *No fund was received related to this study.*
Institutional Review Statement: *The study was conducted according to the guidelines of the Declaration of Helsinki.*
Informed Consent Statement: *Informed consent was obtained from all subjects involved in the study.*
Data Availability Statement: *The datasets generated and analyzed during the current study are available from the corresponding author on reasonable request.*
Conflicts of Interest: *The authors declare no conflicts of interest.*

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