The role of imaging in acute aortic syndromes

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The classic entity of life-threatening aortic dissection represents one pathology of a spectrum of acute conditions coined the acute aortic syndrome comprising dissection, intramural haematoma, penetrating atherosclerotic ulcer, and contained aortic rupture of any cause. The common denominator is disruption of the aortic media layers associated with severe pain and a variety of other symptoms. Any clinical suspicion of acute aortic syndrome should prompt immediate action and confirmatory non-invasive imaging; with respect to sensitivity and specificity for acute aortic pathology modern contrast-enhanced CT technology, MR imaging and ultrasound techniques have similar diagnostic accuracy near 100%. Since the prognosis of most patients with acute aortic dissection is related to undelayed diagnosis and (often surgical) treatment swift diagnostic imaging should be the primary goal in the work-up of any patient with suspected acute aortic syndrome; transfer and in-hospital logistics and local expertise for the differential use of various imaging modalities should be constantly improved.

Key words: Aortic dissection • CT angiography • MRI • Diagnostic algorithm

Introduction

Acute aortic syndrome is a modern term and consists of interrelated emergency conditions with similar clinical characteristics and challenges. These conditions include aortic dissection, intramural haematoma (IMH), penetrating atherosclerotic ulcer (PAU and aortic rupture); trauma to the aorta with intimal laceration may also be considered. The common denominator of AAS is disruption of the media layer of the aorta with bleeding within the layers as IMH, 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 PAU or trauma. In the majority of patients (90%), an intimal disruption is present that results in tracking of the blood in a dissection plane within the media potentially rupturing through the adventitia or back through the intima into the aortic lumen (Figure 1). Relatively simple classification systems are still being used to share both descriptive and prognostic information in scenarios of aortic dissection (Figure 2). The Stanford classification divides aortic dissection in type A and B. Type A involves the ascending aorta, and eventually progresses distally. Type B dissection starts at the level of the descending aorta. The DeBakey classification distinguishes types I, II, and III, with type I involving both the ascending and descending aorta, type II only the ascending aorta and the arch, and type III sparing the ascending aorta and the arch. Further subdivision in Class 1 for classic dissection and in Class 2 for intramural haematoma is less popular.

Confirmatory imaging for acute aortic syndrome

Diagnostic imaging studies in the setting of the clinical suspicion of dissection have important primary goals such as confirmation of clinical suspicion, classification of dissection, localization of tears, and the assessment of both extent of dissection and indicators of urgency (e.g. pericardial, mediastinal, or pleural hemorrhage); in addition biomarkers (such as myocardial markers, D-dimer elevation >500 μg/L, and smooth muscle myosin heavy chain) may be used strategically in concert with swift aortic imaging, although an ideal algorithm has yet to be determined. A concise selection of imaging modalities is summarized in Table 1. The suspicion of acute aortic syndrome is high with abrupt or severe retrosternal or interscapular chest pain often migrating down the back; associated findings can produce signs of acute aortic insufficiency, pericardial effusion, or occluded aortic sidebranches causing ischaemia or pulse differential. With predisposing factors, such as hypertension, connective tissue disorders, bicuspid aortic valve, coarctation, and previous cardiac surgery or recent percutaneous instrumentation, undelayed diagnostic imaging is required for any of the above

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symptoms. While transthoracic ultrasound provides vital information (new-onset aortic insufficiency, pericardial effusion, or even visualization of proximal dissection), additional transoesophageal (TOE) interrogation of the thoracic aorta is the logical next step, or MD-CT scanning of the entire aorta if considered safe.\textsuperscript{11–15} Both imaging modalities provide further detail beyond classification as type A and B (or distal) dissection and allow for strategic planning: ultrasound technology is portable, avoids transport of a critically ill patient and may even be hold in the operating theatre.\textsuperscript{15} MRI has no place in urgent diagnostic work-up of acutely symptomatic patients. Additional information not essential for immediate management decisions such as coronary, arch vessel, and side-branch involvement is usually depicted on CT angiograms without the need for invasive angiography even in the presence of ST changes.

**Choice of imaging modality**

Considering the excellent accuracy of all modalities, imaging protocols for both chronic and suspected acute aortic diseases should adapt to local expertise and to specific questions about the target of interest.\textsuperscript{11,12,16} For unstable patients with suspected aortic syndromes in the emergency room often unfit for transportation, bedside echocardiographic techniques combined with transoesophageal echogram and colour-Doppler interrogation have first priority, even though abdominal segments of the aorta
may not be seen from standard subcostal views. Conversely, rapid acquisition CT technology using 16-, 64-, and even 256-slice CT scanners has essentially replaced invasive diagnostic angiography for large- and medium-sized vessels of both the chest and the abdomen (Figure 3). The technology is robust and rapidly performed with high spatial resolution to differentiate intramural haematoma from ulcers and dissection of the aorta, but requires transportation and stable haemodynamic conditions.13,14

MR angiography (MRA) is also capable of high-resolution aortic imaging with three-dimensional post-processing; delayed imaging allows the evaluation of venous structures without additional contrast. The ability to image thin intimal flaps, intramural processes, and the morphology of aortic wall inflammation may offer new insight into vascular disease detection and classification. Indeed, intramural haematoma and asymptomatic aortic flaps, aortic ulcers, and aneurysms are reported at increasing frequency with access to tomographic imaging.8,17

In contrast to both CT and MR technology, modern ultrasound equipment is mobile and especially attractive at the bedside for unstable cases. TEE interrogation added to transthoracic suprasternal screening ultrasound is superb for acute aortic dissection (type A) even intraoperatively with near perfect sensitivity and specificity,12,15 even considering a blind spot at the proximal arch from bronchial air. Colour Doppler is instrumental to assess entry sites and false lumen flow in real time in order to confirm proximal dissection (Figure 5). In addition, important prognostic information, such as pericardial effusion, acute aortic regurgitation, and proximal coronary obstruction, can be visualized. For patients in

Table 1 Diagnostic Algorithm (Aortic Dissection)

<table>
<thead>
<tr>
<th>Clinical suspicion of AAS</th>
<th>Unstable/critical conditions</th>
<th>Follow-up evaluation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Compulsory:</td>
<td>TEE with the colour-Doppler flow</td>
<td>MRI with Gd enhancement</td>
</tr>
<tr>
<td>ECG, chest radiography, biomarkers (TNT, TNI, D-dimer &gt; 500 μg/L)</td>
<td>MD-CT with CT angiography or MRI with Angiography rarely required</td>
<td>MD-CT with CT angiography</td>
</tr>
<tr>
<td>Stable clinical condition</td>
<td>Optional:</td>
<td></td>
</tr>
<tr>
<td>ECG, chest radiography, biomarkers (TNT, TNI, and D-dimer)</td>
<td>TEE with the colour-Doppler flow</td>
<td>MRI with Gd enhancement</td>
</tr>
<tr>
<td>ECG, chest radiography, biomarkers (TNT, TNI, and D-dimer)</td>
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</tr>
<tr>
<td>Angiography rarely required</td>
<td>Angiography rarely required</td>
<td>Angiography rarely required</td>
</tr>
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AAS, acute aortic syndrome; TNT, troponin t; TNI, troponin I; MD-CT, multidetector computer tomography; Gd, gadolinium; TEE, transoesophageal echocardiography.
shock and very high clinical suspicion of ascending aortic dissection, transthoracic echocardiography (TTE) alone seems reasonable prior to surgery, with TEE performed prior to sternotomy even in the operating room. Although both ultrasound techniques are important bedside tools for acute dissection, both fail to provide sufficient anatomic detail to plan endovascular interventions.13

Catheter-based and digital subtraction angiography as well as ultrasound-based techniques have recently been replaced by contrast-enhanced CT and MRA; MRA requires no iodinated contrast or ionizing radiation, allows three-dimensional multiplanar acquisition, and is particularly useful for patients unable to tolerate contrast due to allergy or renal failure. With acute aortic dissection linked to young patients with fibrillinopathies, diagnostic evaluation during pregnancy and lactation becomes an issue. Whereas foetal radiation exposure needs to be avoided (in the initial 20 weeks of pregnancy) for teratogenic and carcinogenic reason, any iodine component of contrast media given during pregnancy has potential to initiate neonatal hypothyroidism. Even with MR contrast agents a minimal teratogenic risk cannot be excluded since paramagnetic agents like gadolinium cross the placenta with unknown long-term effects. Although exposure during the first trimester has not been associated with an adverse impact on the foetus, controversy is ongoing. While European guidelines consider gadolinium ‘probably safe during pregnancy’ since distributed extracellularly and eliminated into urine, most centres in the USA discourage gadolinium during pregnancy in fear of retention in amniotic fluid and toxicity.19 For suspected aortic disease in pregnancy, non-contrast MRI using steady-state free precession (SSFP) techniques ensures both maximum safety and high diagnostic precision.20

Specific considerations in the choice of a modality

MRA is not as severely affected by calcification as is CT. Heavily calcified arteries may still induce an artefact on MR angiograms, but luminal narrowings and intramural haematoma are depicted even in the presence of atherosclerotic calcification. Thus, MRA is better suited for evaluating occlusive disease of medium-sized arteries and, therefore, modality of choice for patients with lower or upper extremity vascular disease (Figure 4). Despite easier post-processing MRA has lower spatial resolution than CT and images are sensitive to metal (implanted clips or stents) causing distortion and artefact. Patients with pacemakers, defibrillators, or older mechanical valves are confined to CT angiography (contraindications for MRI). CT is less sensitive to small implants, but large metallic objects, such as hip prostheses degrade image quality. With recent scanners complex vessel morphology as seen in dissections, irregular aneurysms, and vascular tortuosity is better delineated ECG gated by CT, whereas MR imaging may produce an artefact in areas of the turbulent blood flow.21,22 In cases of iodinated contrast allergy, CT angiography can also be performed after i.v. injection of gadolinium (60–80 mL); although gadolinium provides less intense enhancement, diagnostic images are feasible. The status of branch arteries and quantitative morphology of both aneurysm or dissection on CT or MRI is essential for strategic planning and thus either modality is recommended prior to endovascular repair of aneurysm and dissection.13,22

Both MR and CT angiography are useful for extra-aortic pathologies, often vascular in nature such as mycotic aneurysms or traumatic pseudoaneurysm, and even for inflammatory vascular disease. Moreover, the loss of elastic properties, aortic shear

Figure 4 (A) Coronal MIP 3D MR-angiogram shows significant stenosis of the right common carotid artery (small arrow). The left subclavian artery reveals two stenotic segments (large arrows) with some post-stenotic irregularities of the left common carotid artery. Axial unenhanced (C) and gadolinium-enhanced (B) T1-weighted MR-images show wall thickening of the ascending aorta (arrows).
stress, and increased wall tension can be quantified with flow-sensitive MR sequences. For serial follow-up after surgical or endovascular repair, three-dimensional MRI sequences are preferred particularly when stent-graft components consist of nitinol; in the presence of stainless steel MD-CT is better to identify endoleak or confirm complete isolation of an aneurysm since steel causes MRI artefact from magnetic disturbances.

CT angiography of the aorta

Whereas invasive angiography visualizes two-dimensional luminograms, non-invasive multi-detector CT is the result of rapid image acquisition and three-dimensional reconstruction in high resolution during brief contrast opacification. The technical evolution with MD-CT was the cone-shaped X-ray beam generating image data over a volume, rather than from an axial section per rotation, being displayed as three-dimensional image along the centreline of the flow (useful for evaluating vascular disease morphology and planning endovascular procedures). The ability to view vessels in multiple projections and orientations helps evaluating three-dimensional anatomy of the aorta accentuated by tortuosity, dilation, or spiralling dissection (Figure 3). The advantages of modern CT include rapid image acquisition, post-processing flexibility, and less image noise resulting in high-definition CTA from the neck to the abdomen in <20 s. A significant drawback of MD-CT is a radiation dose of 10–25 mSv, especially of concern in young patients often subject to serial cardiovascular imaging.12,14,23

In suspected acute aortic syndrome, a non-contrast scan through the chest is useful to screen for acute haemorrhage or intimal vascular calcifications, particularly in the aortic wall (intramural haematoma or separation of wall layers). Subsequently, to facilitate three-dimensional reformatting a thin (1 mm) scan is acquired during rapid bolus administration of contrast at 4–5 mL/s. Since vascular imaging is dependent on the iodine flux (iodine concentration multiplied by the flow rate) excellent vascular images require injections via venous catheters or implanted ports.24 Arterial phase imaging has to coincide with arterial contrast opacification; delayed venous phase imaging is also useful to evaluate solid organs for mass lesions or for endoleaks in patients with stent-grafts (by the use of three-dimensional multi-planar reconstruction). In aortic sidebranches such as coronaries or renal arteries, CT angiography tends to overestimate calcified stenoses and underestimate luminal narrowing by a non-calcific plaque,
potentially relevant when correlating clinical signs with imaging findings. Occasionally, CT artefacts occur in obese or uncooperative patients with noise and motion causing image distortion. ECG-gated CT acquisition can improve image quality especially in the aortic root, where pulsation artefacts are common and likely to mimic dissection. Meanwhile, first reports emphasize the potential of virtual vascular endoscopy based on gated acquisition for both detailed anatomic evaluation and therapeutic guidance.25,26

Ultrasound/transoesophageal echocardiography
TTE has limited value for the evaluation of the entire aorta, but is highly useful in identifying aortic valve dysfunction, pericardial tamponade or wall motion abnormalities, and may screen for proximal and descending aortic dissection in patients with shock in the emergency room. It is limited, however, in visualizing the distal ascending and transverse aorta. Advantages of TEE for the detection of acute aortic syndromes result from close proximity of the oesophagus to the thoracic aorta and its ability to visualize both the ascending and the descending aorta and parts of the arch with high spatial resolution in real time. Although TEE requires oesophageal intubation, it is portable and images are acquired at bedside and immediately interpreted (Figure 5). Aortic dissection is confirmed when two lumens are separated by an intimal flap visualized within the aorta. Tears can be identified and differentiation between true and false lumen is often easy and diagnostic with optional colour-Doppler flow mapping or contrast ultrasound27; intimal tear(s) can be localized in the majority of patients.7,8 Furthermore, variants of acute aortic syndromes such as IMH, atherosclerotic penetrating ulcers, and side-branch obstruction can also be identified.17 Overall, the European Cooperative Study Group and others showed that TEE can reach a sensitivity of 99% with a specificity of 89%, positive-predictive accuracy of 89%, and negative predictive accuracy of 99% findings later confirmed in IRAD.1,2,11 Although TEE is swiftly performed in unstable patients at bedside, an experienced operator is needed for image acquisition and interpretation. Yet, the adjunctive use of colour-Doppler interrogation is instrumental to confirm the blood flow in both true and false lumen, to identify communication sites, to visualize dynamic side-branch obstruction and other aortic emergencies15; in selected cases contrast-enhanced echocardiography (including TEE) may facilitate the differentiation of true and false lumen27,28 and three-dimensional echograms may soon further improve image interpretation. TEE is limited in assessing abdominal side branches and unpleasant for patients who cannot tolerate topical anaesthesia or conscious sedation. Given these issues, and considering availability, excellent quality, and scanning speed of multi-detector CT angiography, TEE may be advantageous for urgent cases of suspected type A dissection. Never-theless, TEE is an important imaging adjunct to perform endovascular stent-grafting in complicated type B dissection and to document immediate procedural success.27

Magnetic resonance imaging of the thoracic aorta
MRA is a complementary, rather than competing imaging modality for the thoracic aorta. With neither ionizing radiation, nor iodinated contrast required, MRA is ideal for patients with multiple follow-up scans and/or contrast allergies.
Spin echo T1-weighted imaging provides best patho-anatomical detail of intramural haematoma, intimal flaps, or atheromas, whereas T2-weighted images allow tissue characterization of aortic wall or blood compounds (Figure 6). ECG triggering is essential to minimize pulsatility artefact. With additional preparatory radiofrequency pulses superior black blood fast T1- and T2-weighted sequences are generated and improve image quality in any plane.26 Dynamic and functional information is derived from gradient-echo MRI based on flow-related signal enhancement. Although MR angiographic methods without contrast enhancement have been available for a long time, gadolinium-enhanced MRA has dramatically shortened examination time and emerged as preferred MR modality for aortic disease; adequate images result from only 15 mL of gadolinium.29 Although MRA was
considered ideal in renal failure. Discovery of nephrogenic systemic fibrosis in patients with renal dysfunction receiving gadolinium has tempered enthusiasm and contributed to a renaissance of non-enhanced MRA. Among established non-enhanced sequences beyond ECG-gated partial Fourier fast spin echo, balanced SSFP has emerged as a central technique to provide vivid imaging of flowing blood. Non-contrast SSFP imaging enables rapid exclusion of dissection in the ‘single shot’ mode and a more detailed evaluation (for entry location and flow pattern) in the cine mode both visualizing dissection in any plane. The high signal-to-noise and contrast-to-noise ratio (due to cardiac and respiratory gating) renders SSFP particularly useful for patients incapable of breath holding, or in suspected aortic syndrome with better detection of aortic wall pathologies such as intramural haematoma than by MRA alone. True MRA without gadolinium is also feasible, but inferior to gadolinium-enhanced MRA and three-dimensional volume-rendering or maximum intensity reconstruction (Figure 7).

The ability of MRI acquisition in any plane and three dimensions enables swift and high-resolution imaging for both chronic pathologies or even acute aortic dissection. Because of the closed bore design of the magnet and the need for monitoring and resuscitative equipment close to magnetic field, MRI is less suitable for unstable patients than CT. Both for CT and MRI, real-time video sequences and serial examinations allow assessing instantaneous haemodynamics and longitudinal evolution in transition from acute to chronic aortic dissection; four-dimensional imaging including time domain is eventually becoming standard. Post-processing involves multi-planar, volume-rendered, and MIP reconstructions, as well as virtual endoscopy for complex evaluation of aortic dimensions, coarctation, parietal thrombus and ulcers, dissection, intramural haematoma, and perivascular fat.

**Catheter angiography and intravascular ultrasound**

Although no longer used for an initial diagnosis of aortic dissection, catheter angiography and IVUS may be useful during endovascular procedures or when non-invasive modalities were inconclusive. The sensitivity and specificity of angiography for diagnosing aortic dissection are 88 and 95%, respectively, with a relatively high rate of false-negative studies because of occasionally inadequate opacification of the false lumen or in the stage of IMH. Incomplete visualization of the false lumen can also occur when the catheter tip is placed distal to the primary tear. Other important features of aortic dissection that cannot be visualized by a lumino-graphical modality include aortic wall thickness in patients with aortitis or the aortic diameter in the presence of mural thrombus and of course extraluminal findings. Moreover, cost and time required for angiography is considerably greater than for cross-sectional imaging. With almost no role in the diagnostic work-up in the setting of suspected dissection angiography is being used during endovascular management of these conditions, primarily to seal communications between the true and false lumen, and for the placement of endovascular stent-grafts.

**Integration of imaging and intervention**

In contrast to both CT and MR technology, modern ultrasound equipment is mobile and especially attractive at the bedside for unstable emergency cases. TEE interrogation added to transthoracic suprasternal screening ultrasound is superb for acute aortic dissection (type A) even intraoperatively with near perfect sensitivity and
such as elevated myosin heavy chain concentration, D–dimer and MR imaging protocols, will not only further improve diagnostic specificity, but has a blind spot confined to the proximal arch due to bronchial air. Colour Doppler is instrumental to assess entry sites and the false lumen flow in real time in order to confirm proximal dissection (Figure 5). In addition, prognostic information such as pericardial effusion, acute aortic regurgitation, and proximal coronary obstruction can be visualized. For patients in shock and with a very high clinical suspicion of ascending aortic dissection, TTE alone is reasonable prior to immediate transfer to surgery, with TEE performed prior to sternotomy. Although TTE and TEE are important bedside tools in dissection, both fail to provide sufficient anatomic detail to plan endovascular interventions. Intravascular ultrasound may emerge as a useful adjunct to endovascular procedures, but is of little value for primary diagnosis.

Future trends in aortic imaging

Although molecular imaging is largely confined to neoplasia and degenerative diseases, vascular applications of molecular imaging are emerging with the goal of identifying early stages of atherosclerosis by an endothelial expression of vascular adhesion molecules. Additionally, biochemical markers of acute dissection, such as elevated myosin heavy chain concentration, D–dimer levels, and soluble elastin fragments, may assist in preselecting patients for imaging. The rapid evolution of CT and MR technology supports the investigational, but emerging ‘triple rule-out’ concept by simultaneously evaluating coronary arteries (acute coronary syndrome), thoracic aorta (dissection, ulcer), and pulmonary arteries (pulmonary embolism). Further technological advances are on the horizon with multiple X-ray sources around the patient to minimize CT scan time to seconds; 3 T MR units are already clinical reality and provide an improved signal/noise ratio with better MRA image contrast than 1.5 T magnets. ECG-gated CTA can accurately determine aortic distensibility in both phantom models and real patients, and 3 T MRA with three-dimensional velocity mapping offers haemodynamic evaluation of normal and diseased thoracic aorta. Both techniques attempt to visualize stress and strain impacting early on aortic wall. Intravascular ultrasound imaging or optical coherence tomography may be particularly useful in conjunction with endovascular procedures and visualize wall architecture, associated plaques, haematoma, and microdissections. Moreover, real-time MR-guided vascular interventions have been demonstrated in animals and in patients.

Conclusion

Acute aortic syndrome is not an uncommon condition. Currently, TEE and non-invasive tomographic imaging play a leading role in both primary diagnosis and treatment planning. In the near future, new approaches based on refined multi-detector MD-CT and MR imaging protocols, will not only further improve diagnostic precision, but allow risk stratification as part of diagnostic imaging.

Conflict of interest: none declared.

References


Anomalous origin of right coronary artery from left coronary sinus

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A 58-year-old lady presented with atypical chest pain, increasing breathlessness, and reduced exercise tolerance. There was no other significant past medical history or risk factors. The systemic examination was normal and so were the biochemical and haematological parameters. An electrocardiogram showed normal sinus rhythm. As she was unable to perform exercise test, coronary CT angiogram was organized, which revealed the anomalous origin of the right coronary artery (RCA) from the left aortic sinus running a potentially malignant course with narrowing between the aorta and right ventricular outflow tract before regaining normal calibre (Panels A and B). The calcium score was raised at 616. Subsequent angiogram revealed minor disease in the left coronary artery (LCA) and the circumflex artery but the RCA could not be cannulated. Myocardial perfusion scan for functional testing demonstrated no evidence of ischaemia. After a multi-disciplinary meeting, decision was made to treat patient medically with a regular clinic follow-up.

The incidence of the anomalous origin of coronary arteries is reported to be ~1%, anomalous RCA being more common than the left. This anomaly has been associated with dyspnoea, palpitations, syncope, angina, myocardial infarction and even sudden death. The proposed mechanisms include compression by great vessels especially when the aberrant pathway is between the aorta and the pulmonary artery, origin at an oblique angle resulting in a slit-like orifice leading to collapse on exertion and coronary spasm. Anomalous RCA generally follows a benign course and is therefore treated conservatively, whereas surgical intervention which includes translocation, ostioplasty, or bypass graft is recommended for anomalous LCA.

(A) The grey-scale image is an oblique axial maximal intensity projection (MIP) at the level of the left coronary sinus showing the anomalous RCA (arrow) between the aorta and main pulmonary artery. The proximal RCA is narrowed before resuming a normal calibre. (B) The colour image is a volume rendering technique (VRT) image of the coronary tree showing the anomalous course of the RCA (arrow).

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