Thoracic and Cardiovascular Surgery

Acute Aortic Syndromes: New Insights from Electrocardiographically Gated Computed Tomography

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The development of retrospective electrocardiographic (ECG)-gating has proved to be a diagnostic and therapeutic boon for computed tomography (CT) imaging of patients with acute thoracic aortic diseases, such as aortic dissection/intramural hematoma (AD/IMH), penetrating atherosclerotic ulcer (APU), and ruptured/leaking aneurysm. The notorious pulsation motion artifacts in the ascending aorta confounding regular CT scanning can be eliminated, and involvement of the sinuses of Valsalva, the valve cusps, the aortic annulus, and the coronary arteries in aortic dissection can be clearly depicted or excluded. Motion-free images also allow reliable identification of the site of the primary intimal tear, the location, and extent of the intimomedial flap, and branch artery involvement. ECG-gated CTA also allows the detection of more subtle lesions and variants of aortic dissection, which may ultimately expand our understanding of these complex, life-threatening disorders.

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Computed tomography (CT) has a long tradition of providing valuable clinical imaging of the thoracic aorta; CT angiography (CTA) has replaced diagnostic angiography for the evaluation of acute thoracic aortic diseases. Modern multiple detector-row CT technology is widely available, often 24 hours a day, 7 days a week, in close proximity to the emergency room, with sensitivity and specificity approaching 100% for the detection of acute thoracic aortic disorders, and with the ability to detect many other findings.[1] The recent development of CTA with retrospective electrocardiographic (ECG) gating provides additional benefits because it eliminates cardiac pulsation motion artifacts and has extended the clinical applicability of CTA to also include the aortic root. ECG gating allows cardiac phase-resolved ('time-resolved') cine imaging and visualization, adding yet another "fourth dimension" (4D) to this technique.

The first major clinical benefit of motion-free 3D imaging of the thoracic aorta is the elimination of notorious ascending aortic pulsation motion artifacts, which may mimic dissection (Fig. 1). Motion-free 3D imaging improves visualization and, thus, characterization of acute aortic dissection. In addition to the ability to visualize the dissection flap extending into the aortic root and the coronary arteries, it is possible to precisely localize, characterize, and visualize the site of the primary intimal tear, which is increasingly important in the stent-graft era.[2,3] Motion-free images allow the detection of less-common subtle but clinically equally important intimal lesions, which have been notoriously difficult to detect in vivo with any current imaging modality.[4] Finally, the improved ability to see subtle abnormalities and variants of aortic dissections, and their evolution over time, may ultimately expand our understanding of these disorders.

CT Imaging Strategy and Postprocessing

We routinely use ECG gating at our institution in all patients with acute aortic syndromes undergoing CTA. Until recently, we have used 16-channel CT systems; we now use 64-channel scanners. A complete CT evaluation of patients with suspected acute aortic abnormalities also includes an initial nonenhanced scan to detect intramural hematomas or blood within the chest, abdomen, or pelvis. The contrast-medium enhanced CTA acquisition uses ECG gating through the chest, and immediately continues through the abdomen and

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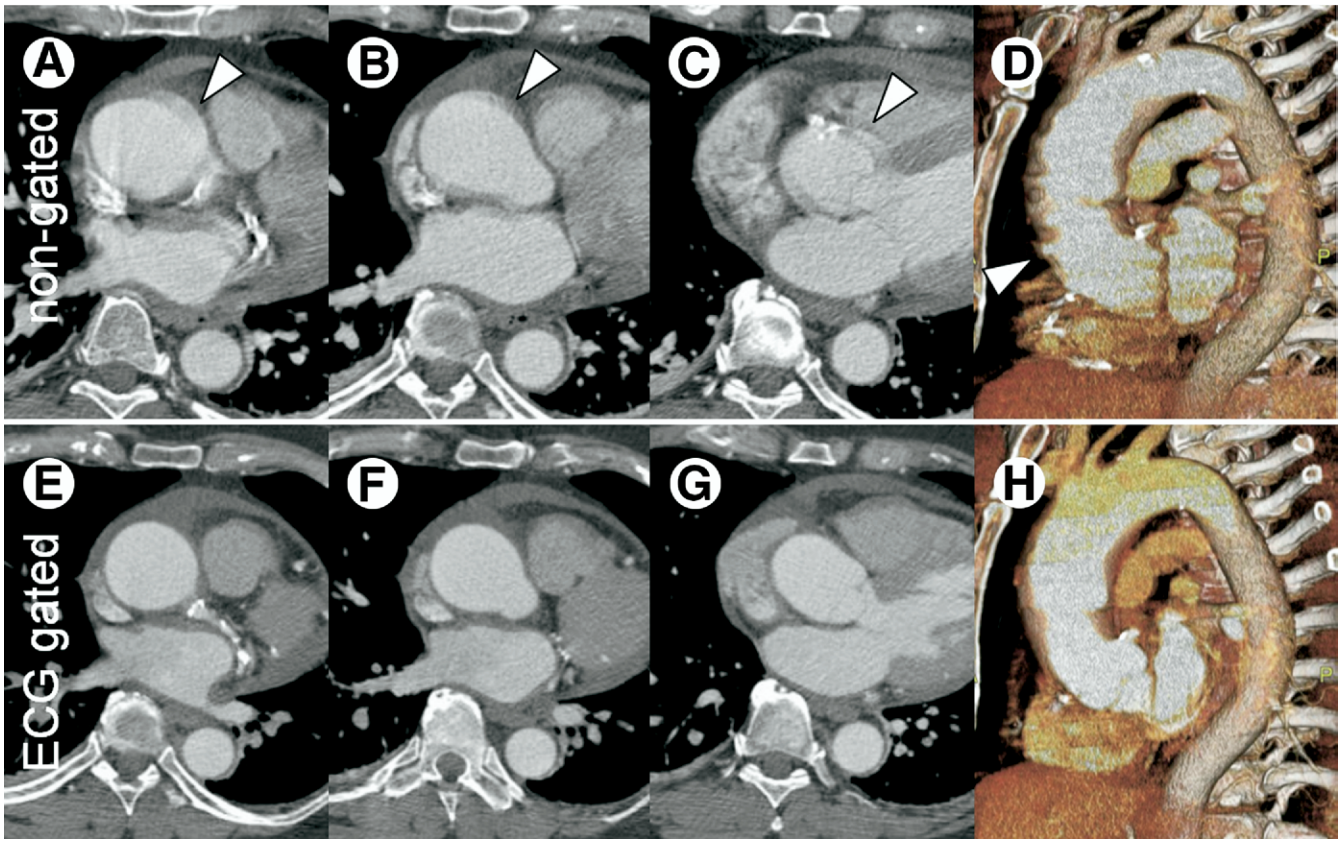

Figure 1 Aortic motion-artifact suppression with ECG-gated CTA. Three transverse CT images obtained with a non-gated 16-channel CT show typical motion artifacts with double contours (arrowheads) at the aortic root (A), the proximal (B), and mid (C) ascending aorta. Corresponding volume-rendered image (D) shows artifactual contour abnormalities (arrowhead) in the ascending aorta due to transmitted cardiac pulsation. This study was repeated 12 hours later with ECG gating, which eliminates motion artifacts in the corresponding transverse (E-G) and volume rendered (H) images.

pelvis down to the level of the common femoral arteries without ECG gating. Complete visualization of the aorta and iliofemoral arteries is of critical importance to completely evaluate complications of dissection and also for access route planning if endovascular treatment is an option.

The technical aspects of ECG gated CT of the thorax have been described elsewhere in this issue. The ability to generate motion-free images of the aorta, and the ability to "cine"-loop cardiac motion, aortic pulsation, and motion of an intimal flap are an asset in the evaluation of aortic emergencies. This allows visualization of more subtle lesions; it allows substantially improved assessment of the aortic root, including the valve apparatus and coronary artery involvement and, in times of expanding endovascular treatment options, this technique easily depicts not only the intimal flap, but also the site of the primary intimal tear, which may be an accessible endovascular stent-graft target, e.g., patients with retrograde type A dissection ("retro-A") when the tear is in the descending thoracic aorta.

Transverse CT source images remain the most important images in the acute setting to make the diagnosis, characterize a lesion, and assess potential complications. Postprocessing is often required, specifically for stent-graft treatment planning, and both 3D and 4D visualization provide detailed and unprecedented preoperative visualization of complex and unusual ascending aortic lesions and dissections such as completely circumferential intimal flaps (Fig. 2) and intima-intimal intussusception.

Imaging Manifestations in Acute Aortic Syndrome

The acute aortic syndrome is a contemporary term referring to acute life-threatening abnormalities of the aorta associated with intense chest or back pain, and traditionally includes classic aortic dissection (AD), intramural hematoma (IMH), and penetrating atherosclerotic ulcer (APU).[5] While the presumptive diagnosis of acute myocardial infarction is usually considered first (due to its much greater prevalence), excluding acute thoracic aortic disease is imperative to expedite appropriate treatment and to avoid deleterious thrombolysis and the delay associated with emergency coronary angiography. While this "classification" of acute aortic lesions is almost universally used in the literature, it does not truly reflect all forms of aortic pathology, which often leads to confusion when it comes to lesion classification, which is today mainly an imaging domain.

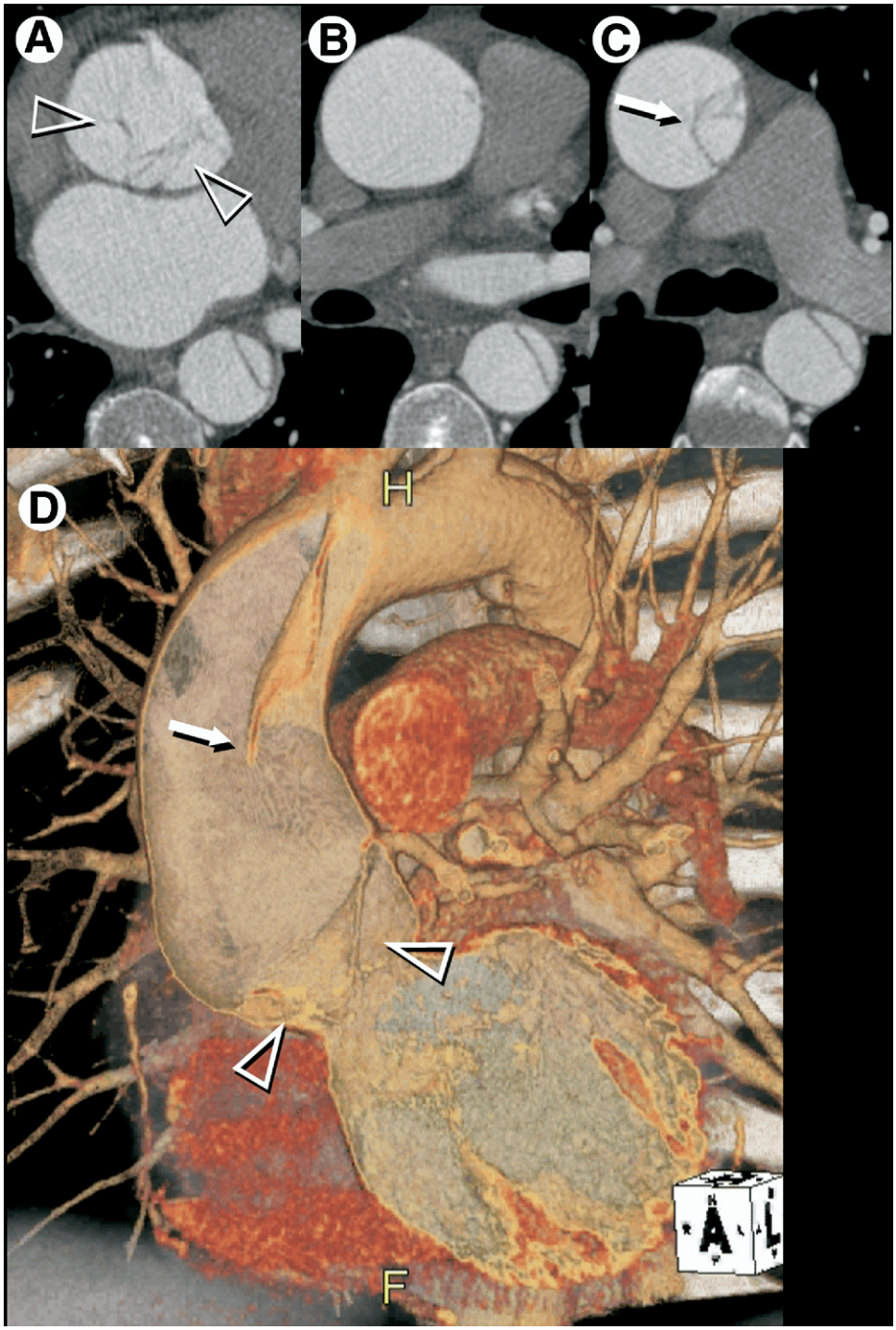

Figure 2 Acute type A dissection in a 45-year-old man. Transverse CT images (A)-(C) show irregular linear opacities (arrowheads) at the level of the aortic root (A), which are difficult to interpret on this transverse image, but no clear dissection flap in the mid ascending aorta (B). In the distal ascending aorta (C), a dissection flap (arrow) between a true and false lumen is identified. Volume rendered image (D) clearly depicts an extensive dissection with the proximal portion of the dissection flap torn off and prolapsing onto and through the aortic valve (arrowheads). Arrow points to dissection flap in the distal ascending aorta.

The main conceptual shortcoming of the classification into "AD–IMH–APU" is the fact that it lumps diseases and imaging findings together, which has led to several unconvincing attempts to explain the dynamic evolution of one into the other in the past.

In the opinion of the authors, it is more intuitive to regard the acute aortic syndromes as the manifestation of two major entities, which may occasionally coexist: (1) Diseases of the aortic media, and (2) a diseased intima, in the form of atherosclerosis. The first entity, a diseased media, historically

Table 1 Acute Aortic Syndromes

1. Dissection and variants
a. Classic AD
b. IMH
c. Limited intimal tear
2. APU
a. APU with IMH
3. Rupturing aortic aneurysm

AD, aortic dissection; IMH, intramural hematoma; APU, atherosclerotic penetrating ulcer.

but erroneously described as "Erdheim's cystic medial necrosis" results in separation of aortic media layers (within the media), which is present in both classic aortic dissection, and in the dissection-variant of intramural hematoma. The etiology of "cystic media necrosis" is diverse and a typical manifestation of the Marfan syndrome and related heritable connective tissue disorders in young individuals, but most commonly is a consequence of hypertension. While classic aortic dissection has an entry tear, a false lumen within the aortic media, and an exit tear, the "classic" IMH has—per original definition—no obvious communication with the true lumen. The second cause of acute aortic syndromes, a diseased intima, is the result of advanced atherosclerosis, where an atherosclerotic ulcer penetrates through the internal elastic lamina into the aortic media, which, in the acute setting, is typically associated with an intramural hematoma. Intramural blood can thus be a manifestation of two very different entities; it can be a variant of aortic dissection, and it can be a manifestation of an atherosclerotic penetrating ulcer (APU). In short, an intramural hematoma is not a disease or pathologic entity, but simply an imaging finding, and the patient demographics, prognosis, and potential progression are most likely quite different in these entities and should not be lumped together. The main justification of using IMH as one of the acute aortic syndromes comes from the fact that the treatment of an acute thoracic aortic lesion is more dependent on its location (Stanford type A versus type B), and the presence or absence of complications than on the underlying etiology.

The improved spatial resolution of modern CT also challenges several other "classical" concepts. The spectrum of aortic dissection appears to be wider than just classic dissection and IMH, and more subtle lesions can now be detected in vivo, such as limited superficial tears.[4] If this latter lesion is truly a dissection variant or perhaps a manifestation of acute dilation/impending rupture of an aneurysmal aorta is speculative at this point. Not infrequently we encounter lesions that have features of both, classic dissection and "classic" IMH, which is traditionally defined as blood in the aortic wall with no communication to the true lumen (at least macroscopically), and the evolution of such lesions is hard to predict on any given imaging study. With modern CT it is not uncommon to see an IMH associated with a small local primary intimal tear but no double-barrel aorta and exit tear. We also frequently observe cases of IMH with one or several small distal communications ("natural fenestrations") with the aortic true lumen typically at the site of torn side-branch ostia (e.g., intercostals arteries), which should not be confused with "ulcers."

Conceptually, we classify acute aortic syndromes as shown in Table 1. We use the term intramural hematoma not as a disease but as a manifestation of either a dissection variant or a APU, and define it as the presence of hyperdense blood within the aortic wall irrespective of the presence or absence of small communications with the true lumen. It seems also justified to add rupturing aneurysms into the abnormalities manifesting as acute aortic syndrome.

Morphology of Classic Aortic Dissection and Intramural Hematoma

Aortic dissection is a relatively uncommon yet potentially catastrophic clinical event. The incidence has been estimated at from 5 to 30 million/y (versus 4.400/million/y for acute

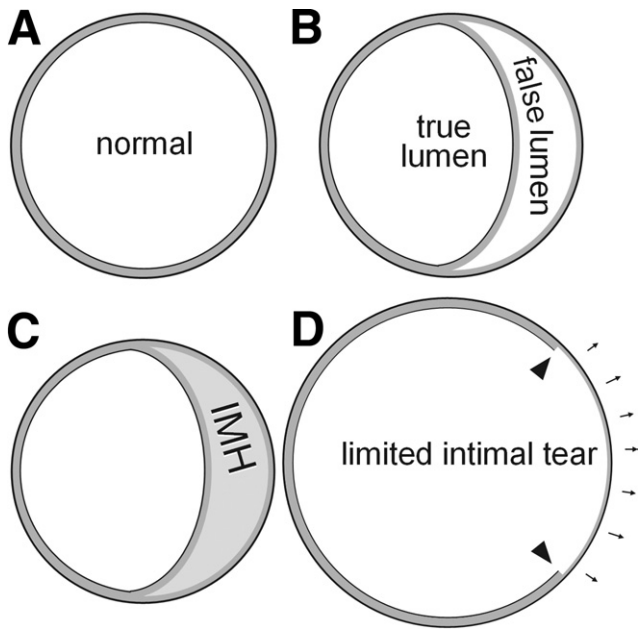

Figure 3 Schematic of aortic dissection variants. (A) the layers of the normal extra-pericardial thoracic aortic wall consist of the intima, the media, and the adventitia. Most of the substance of the aortic wall is media (gray). Both, the intima and the adventitia (drawn schematically as black inner and outer contours of the aortic wall) are not visible on CT. All dissection variants have abnormal media in common, historically and erroneously referred to as "cystic medial necrosis" (see text for details). Classic aortic dissection (B) occurs within the outer third of the medial layer, resulting in two channels of blood flow. Note, that the tissue separating the true and false lumen is mostly made of media tissue, and correctly should be termed the intimomedial flap (in lieu of intima flap). (C) When the separation plane within the media is filled with stationary blood, instead of flowing blood, this is an intramural hematoma (IMH). A limited intimal tear (D) is a partial thickness tear (arrowheads) through the intima and inner portion of the media, exposing the residual media/adventitia, which tends to 'bulge out' (small arrows) relative to the remainder of the aortic circumference.

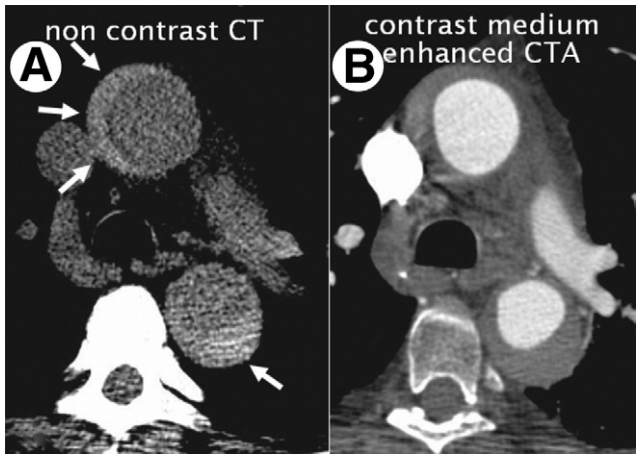

Figure 4 Acute intramural hematoma (type A). Nonenhanced CT image (A) in a 62-year-old hypertensive man with squeezing chest and back pain shows hyperdense crescent shaped abnormality (arrows) in the ascending and descending thoracic aorta, consistent with intramural hematoma. (B) Corresponding postcontrast medium CTA image at the same anatomic level.

myocardial infarction in the U.S.) Acute aortic dissection is the most common acute aortic condition requiring emergency operative repair. While the initiating event leading to aortic dissection is unknown, most patients have a structural abnormality of the aortic media. Elastic fiber degeneration of the aortic media is seen in patients with the Marfan syndrome and other heritable disorders leading to aortic aneurysms and dissections, whereas it is a medial smooth muscle cell substrate problem (a normal feature of the aging process) in older individuals and those with long-standing hypertension. Intimal disease (atherosclerosis) is not a prerequisite for aortic dissection.

Classic aortic dissection is characterized by the presence of an entry tear in the intima, and a clear separation between layers of the aortic media resulting in two separate flow channels, the true lumen and the false lumen (a channel entirely within the media) (Fig. 3). The relative flow of blood and the shape of the true and false lumens are extremely variable, and probably depend on the size of the entry tear, the degree of media degeneration, the presence, location, number, and size of re-entry tears (where false lumen blood flow re-enters the true lumen). The size of the two channels, orientation, and degree of arterial enhancement therefore do not necessarily allow reliable identification of the true and false lumen, respectively. Modern CT technology allows reliable identification of flow channels, however, by analyzing the entire aorta and tracing the respective lumens. This is particularly important when ischemic complications occur (cerebral, renal, mesenteric, or lower limb).

Involvement of the ascending aorta in acute dissection is a surgical emergency owing to its high mortality rate if untreated (1% to 3% per hour). Blood seeping through the diseased wall can lead to pericardial tamponade. Ascending dissection can extend into the sinuses of Valsalva and compromise the coronary artery ostia causing aortic regurgitation or myocardial ischemia, and also lead to frank intrapericardial rupture.

There is considerable overlap between intramural hematoma (IMH) and aortic dissection, in terms of underlying

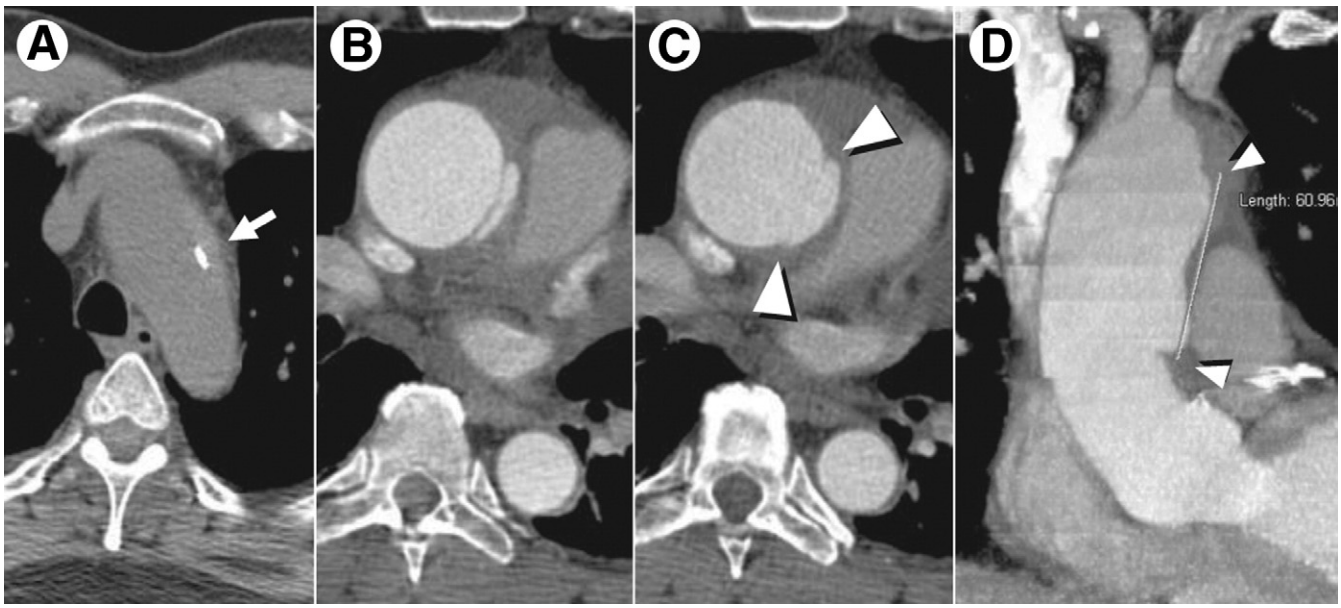

Figure 5 Limited intimal tear of the ascending aorta. Nonenhanced CT image (A) and ECG-gated axial CTA images (B) and (C) show a small intramural hematoma at the level of the proximal aortic arch [arrow in (A)]. Note the displaced small calcification, indicating the location of the intima. In the proximal ascending aorta, a single image (B) demonstrates a small flap, consistent with an undermined edge of a limited intimal tear. Image (C), immediately superior to (B) shows the edges of the limited intimal tear (arrowheads in (C) and (D)) and bulge of the exposed residual aortic wall (small arrows). The full longitudinal extent of the lesion is shown in a thin-slab MIP image (D): the tear extends from the proximal ascending aorta to the proximal arch, with a length of more than 6 cm.

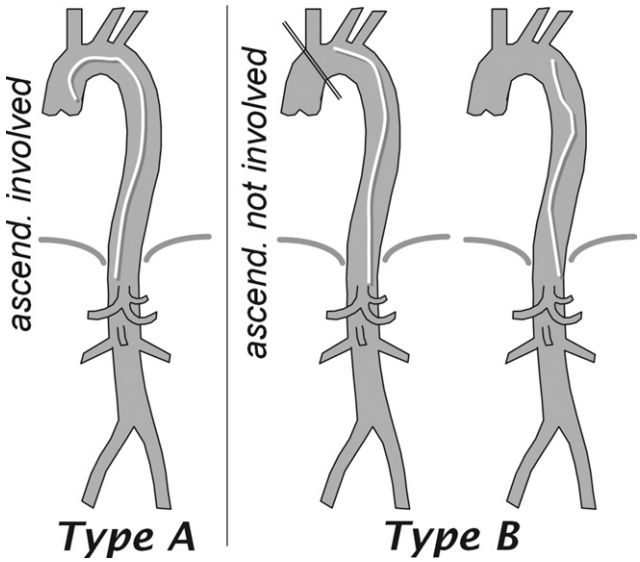

Figure 6 Stanford classification of aortic dissection. Type A dissection (left panel) is defined by the presence of an intimal flap in the ascending aorta (meaning proximal to the origin of the brachiocephalic artery). Type B aortic dissection is defined by the absence of the dissection flap in the ascending aorta and therefore also includes dissections that begin in the transverse aortic arch. Importantly, it should be realized that most type B dissections have some degree of retrograde false lumen extension back into the transverse arch; this does **NOT** make it a type A dissection, which is a common clinical misconception. ascend. = ascending aorta.

media degeneration, patient demographics, and risk of rupture. IMH is considered a variant of classic dissection, where the above-mentioned layer within the aortic media is not filled with flowing blood, but with stationary blood (Fig. 3). Presenting features are similar, progression to dissection may occur, and treatment considerations in our minds are identical to patients with a classic dissection.[6]

Intramural Hematoma

Intramural hematoma, as discussed above, is an imaging finding, and not specific for a disease. It occurs both as part of the dissection complex and with atherosclerotic penetrating ulcers. The imaging criterion for intramural hematoma is a hyperdense, often crescent shaped lesion within the aortic wall seen on nonenhanced CT images (Fig. 4). Symptomatic intramural hematoma of any cause is an aortic emergency.

Limited Intimal Tear (Limited Dissection)

So-called isolated or limited intimal tears are probably the least common and less well known intimomedial lesions of the aorta. The true prevalence and spectrum of these lesions is unknown, but may be up to 5% of patients undergoing acute ascending aortic repair, if specifically searched for.[4] These may resemble "mushroom cap" lesions or very localized dissections. The clinical implication is identical to other acute aortic syndromes, and surgical repair is indicated in these cases, which affect the ascending aorta (type A), which can also be aneurysmal. Chronic limited tears without medial propagation have also been described, notably in patients with Marfan syndrome.

Owing to their morphology, these lesions are particularly difficult to detect on imaging studies. In fact, the nine patients described in Svensson's original description were all diagnosed intraoperatively despite multiple preoperative noninvasive and invasive studies.[4]

Pathologically, these lesions represent partial-thickness linear or stellate tears through the intima and underlying superficial media, exposing the deeper media and adventitia.[4] The edges of the tear can show some limited undermining, but oddly enough, this does not result in a more extensive separation between the torn and intact layers of the aortic wall as one might expect (Fig. 3). Intramural blood has not been described as a typical feature of this lesion, but we have observed both a tiny intimal flap and a small intramural hematoma at both ends of a longitudinal tear in the same patient imaged with ECG-gated CTA, respectively (Fig. 5). The edges of the tear are separated from each other, probably due to stretching of the residual aortic wall (consisting of remaining intact media and adventitia) or some elastic recoil of the torn layer, resulting in an eccentric bulge of the aorta. The eccentric bulge can be the only imaging sign of this important lesion, which has been underdiagnosed with all imaging modalities (US, CT, MR). It is not clear what determines whether a classic dissection with a septum or flap, intramural hematoma, or limited intimal tear develops. It can also be speculated that such longitudinal intimal tears reflect a consequence rather than the cause of rapid dilation/impending rupture of a diseased aorta.

While it is reasonable to assume that the sensitivity of ECG gated CTA will improve the detection of these subtle ascending aortic intimal lesions, this has not been established conclusively. The main implication for the diagnostic imager is therefore to be aware of the fact that such subtle lesions exist, and be prepared to find them with appropriate technique and in the right clinical setting.

Anatomic Classification of Aortic Dissection

We use the 1970 Stanford functional classification to describe the type of dissection and the extent of the intimal (intimo-medial) flap.[7] In addition, we specifically describe the site of the primary intimal tear (PIT) as advocated by Griepp's Mt. Sinai cardiovascular surgical group,[8] which was not included in the original Stanford classification scheme. The definition of a Stanford type A dissection is based on the presence of a dissection flap in the ascending aorta (anything proximal to the origin of the brachiocephalic artery), whereas a Stanford type B dissection, irrespective of the location of the PIT (arch, descending, or abdominal aorta), is defined by the absence of a dissection flap in the ascending aorta. Note that the definition of type A versus type B is thus exclusively based

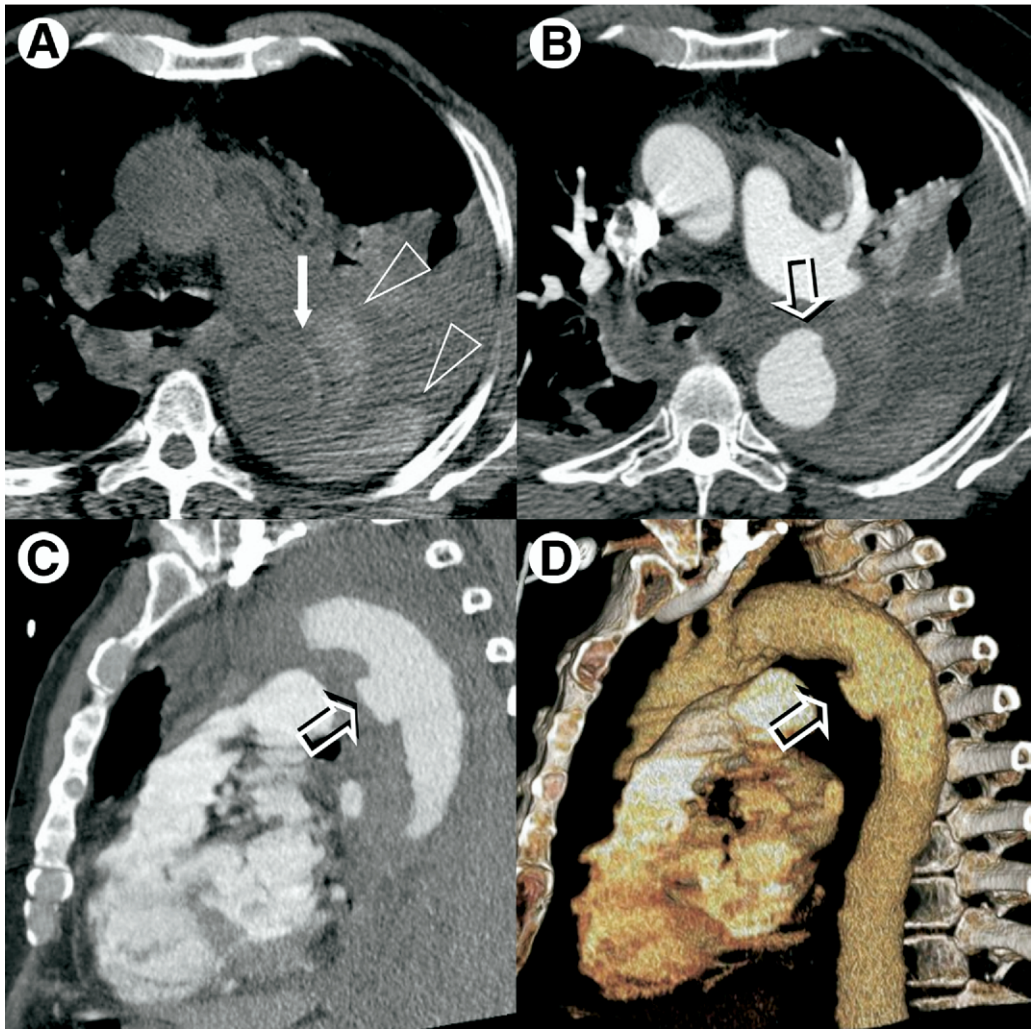

Figure 7 Atherosclerotic penetrating ulcer (APU). Nonenhanced CT image (A) in an 83-year-old man with acute chest pain shows small hyperdense crescent shaped abnormality (arrow) in the descending thoracic aorta, consistent with intramural hematoma. Note a large amount of hyperdense periaortic and pleural fluid (arrowheads) indicating mediastinal hematoma and hemothorax. (B) Corresponding postcontrast medium CTA image at the same anatomic level shows large ulcer (open arrow) in the descending thoracic aorta, which is better characterized on a sagittal reformation (C) and on a volume rendered image (D).

on involvement or no involvement of the ascending aorta (Fig. 6). This is clinically meaningful because it predicts the expected biological behavior of the process if untreated and, moreover, dictates proper management, e.g., ascending aortic involvement (acute type A dissection) mandates emergency surgical repair. While most type B dissections commonly involve the descending aorta distal to the subclavian artery and frequently the abdominal aorta, this is not how type B dissections are strictly defined (despite notorious propagation of misinterpretation in the literature). It is important clinically to emphasize that type B dissections commonly have some limited retrograde involvement of the transverse aorta (aortic arch), and the false lumen in the arch may either be thrombosed or patent. Regardless of arch involvement, most patients with acute type B dissections are treated conservatively unless complications such as rupture, leak, distal body malperfusion due to true lumen collapse, rapid false lumen expansion, refractory pain, malignant hy-

Table 2 Diagnostic Information Sought in Patients with Acute Aortic Syndrome

1. Lesion characterization
Dissection and its variants, IMH, APU
For dissections, identify location of PIT
2. Lesion location and extent
Involvement of the ascending aorta (Type A vs. Type B)
Distal extent
Location of distal re-entry points and flap fenestrations
3. Complications
Type A lesions
Presence of pericardial fluid
Sinus of Valsalva and aortic valve involvement
Coronary artery involvement
Distal complications
Side branch involvement and organ perfusion status
4. Signs of leakage and rupture

IMH, intramural hematoma; APU, atherosclerotic penetrating ulcer; PIT, primary intimal tear.

pertension, or branch vessel ischemia require urgent surgical or stent-graft intervention. The terms type A and type B are also used to describe the location of aortic intramural hematoma (IMH). More detailed anatomic descriptions of the location of IMH have recently been proposed in the literature.[6]

Atherosclerotic Penetrating Ulcer

Atherosclerotic penetrating ulcers (APU) are a rather distinct entity within the acute aortic syndromes. Pathologically, these lesions are defined as ulcers of the inner lining of the aorta (commonly thickened intima with chronic atherosclerotic changes) that penetrate through the internal elastic lamina into the aortic media. This can result in a local aortic wall hematoma, or it can penetrate through the entire aortic wall and result in a (contained) rupture (Fig. 7).

The radiologic diagnosis of an APU is based on the presence of an ulcer-like aortic wall lesion (the internal elastic lamina is not visible on imaging studies), which typically protrudes beyond the aortic circumference, and is associated with a local intramural hematoma or signs of rupture. Indeed, the term ulcer-like projection (ULP) has been used in the Japanese literature. Clinical correlation is important, since ulcer-like lesions can be an incidental finding in asymptomatic patients, representing either an ulcerated plaque (not penetrating into the media), or a chronic, healed (re-endothelialized) ulcer. There is a continuum between the latter lesions and atherosclerotic aneurysms, as some believe most degenerative atherosclerotic aneurysms originate from an APU.

Penetrating aortic ulcers can be considered a consequence of a diseased intima (i.e., atherosclerosis), in contradistinction to aortic dissection and its variants, where intimal disease is not a prerequisite, but in which degenerative changes of the elastic fibers and smooth muscle cells of the aortic media are paramount. This is also in keeping with the observation that APUs tend to occur older patients with significant atherosclerotic plaque burden and a history of hypertension and the predilection for involvement of the descending thoracic aorta. More than one APU is not uncommon in a given patient. The fact that both APU and the dissection complex can result in the accumulation of blood in the aortic wall as an intramural hematoma can be considered a mere coincidence. The IMH seen in patients with APU is usually more focal than those seen in IMH without an intimal ulcer. If "intimal flaps" are seen in patients with an APU, these are usually deep overhanging edges of an ulcer, rather than "true" dissections, (which would require entry- and exit-tears, and a double barrel flow channel). Some morphologic overlap and ambiguity on imaging studies, however, does occur. The therapeutic consequences in the appropriate clinical setting are the same for any morphology, independent of the semantics.

CTA Image Evaluation in Acute Aortic Syndromes

CT image analysis always includes a thorough review of the transverse CT source images for any extra-vascular abnormality within the chest or abdominal wall, lung, airways, and abdominal organs. Following the lines of the above discussion and description of pathology and imaging findings, specific detailed information is sought in patients with acute aortic syndromes as listed in Table 2.

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