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Diagnosis and Management of Acute Aortic Syndromes: Dissection, Penetrating Aortic Ulcer, and Intramural Hematoma

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Abstract

Purpose of Review—Acute aortic syndromes, including aortic dissection, intramural hematoma, and penetrating aortic ulcer, are a group of highly morbid, related pathologies that are defined by compromised aortic wall integrity. The purpose of this review is to summarize current management strategies for acute aortic syndromes.

Recent Findings—All acute aortic syndromes have potential for high morbidity and mortality and must be quickly identified and managed with the appropriate algorithm to prevent suboptimal outcomes. Recent trials suggest that TEVAR is increasingly useful in stabilizing pathology of the descending thoracic aorta but when possible should be applied in a delayed fashion and with limited coverage to minimize neurologic complications.

Summary—Treatment for acute aortic syndrome is frequently dictated by the anatomic location and extent of the wall compromise as well as patient comorbidities. Therapy is often individualized and often includes some combination of medical, procedural, and surgical intervention.

Keywords

Aortic dissection; Intramural hematoma; Penetrating aortic ulcer; Acute aortic syndrome; TEVAR; Type B dissection

Introduction

An acute aortic syndrome occurs when the integrity of the aortic wall is compromised or disrupted in some way, resulting in increased potential for rupture or malperfusion associated with end organ ischemia [1]. This may either be caused by direct disruption of the intima, in the form of a dissection tear or penetrating aortic ulcer (PAU), or by bleeding

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within the aortic media leading to an intramural hematoma (IMH). Because of their high potential for morbidity and mortality, all types of acute aortic syndrome should be treated as emergencies when suspected clinically or identified on high-quality imaging studies [2]. It is well established that the risk of morbidity and mortality increases from the time of onset of an acute aortic syndrome, making prompt diagnosis and rapid implementation of therapy, either medical or procedural, of utmost importance in optimizing outcomes [3]. The care and management of patients with acute aortic syndromes involves multiple specialties, including emergency medicine, radiology, critical care, cardiothoracic surgery, and vascular surgery, and communication between providers is key to providing patients with timely access to appropriate therapy.

Epidemiology

The incidence of acute aortic syndromes is estimated at approximately 10.2 and 5.7 cases per 100,000 person-years for males and females, respectively [4, 5]. Patients at higher risk include those with increased age (> 60 years), males, and those with uncontrolled hypertension; cocaine use resulting in uncontrolled hypertension may also play a role [6, 7]. Disease processes that affect the integrity of the underlying aortic wall are associated with the presentation of acute aortic syndromes in younger, non-hypertensive patients, including connective tissue disease, vascular inflammatory disease, or trauma [8, 9]. Dissection and related syndromes may also be iatrogenic, resulting from percutaneous vascular procedures or recent cardiac or aortic surgery [10].

Clinical Presentation

Acute aortic syndromes have variable presentation and can generate a wide variety of symptoms that are dependent on the extent of aortic wall compromise. Pain is common, with pathology of the ascending aorta (type A) classically results in severe anterior chest pain, while pathology of the descending aortic pathology (type B) classically presents with severe back pain [11]. If pain is present, it may be described as “tearing,” “ripping,” or “stabbing,” but acute aortic syndromes may also be painless or present with atypical pain. Additional symptoms may indicate the anatomic extent of dissection —involvement of the brachiocephalic vessels may lead to neurologic or upper extremity symptoms, visceral dissection may generate abdominal pain, and lower extremity pain signifies iliac involvement [12]. Type A aortic dissection (TAAD) may result in coronary dissection, tamponade, or acute valvular dysfunction that can masquerade as a primary acute coronary syndrome. Syncope is a particularly sinister symptom and may portend neurologic involvement or major cardiac dysfunction [1].

Diagnosis

The history of a patient presenting with the aforementioned symptoms should include family history of aortic aneurysm, dissection, genetic aortopathy, or sudden unexplained death. Physical exam should quickly assess for the physical manifestations of malperfusion, auscultating for aortic valve murmurs and any asymmetry in breath sounds, palpating any differences in extremity pulses, assessing for abdominal pain, and performing an efficient neurologic exam. One can also obtain blood pressures in both extremities if a pulse difference is noted, or if the initial blood pressure is not consistent with the clinical scenario.

Electrocardiogram (ECG) changes and chest radiograph abnormalities such as mediastinal widening and displacement of aortic calcifications are present in over two-thirds of patients who present with acute aortic dissection; however, these findings are frequently non-specific [6]. While important, the investigations listed here cannot absolutely rule out acute aortic syndrome and should not delay or preclude obtaining axial imaging for definitive diagnosis [3, 12, 13].

Imaging Studies

The mainstay of diagnosis of acute aortic syndromes is high quality axial imaging, ideally in the form of computed tomographic (CT) angiography, due to its wide accessibility and short accession time [14]. Contrast bolus optimization with electrocardiographic gating to minimize motion artifact should be considered to maximize CTA yield, but any CT with contrast is usually sufficient to make the diagnosis. The entire aorta (chest, abdomen, and pelvis) should be imaged in anyone with suspected acute aortic syndrome, as complete visualization of the extent of disease is imperative for risk assessment and therapeutic planning. Thus, concern for acute aortic syndrome should be communicated to the radiology staff performing the study in order to optimize the quality of the study [1]. Magnetic resonance angiography (MRA) is an alternative option to CTA, but its utility is limited by long acquisition times. Transesophageal echo (TEE) is useful to make a diagnosis of TAAD, but is technician-dependent and may not reliably visualize more distal pathology [15]. CT without contrast is occasionally obtained in patients with chronic kidney disease, but is suboptimal particularly for visualizing visceral involvement. CTA can identify acute aortic pathology with a sensitivity and specificity of over 95%; however, if high clinical suspicion exists in the setting of a negative CTA, other imaging modalities should be employed as a secondary study [16].

Classification

The management of aortic dissection as well as related acute aortic syndromes such as PAU and IMH is largely dictated by the location of the pathology [17]. This is most frequently and simply dictated by the Stanford classification [18], where type A is any extent of pathology that involves the ascending aorta and type B is any extent of pathology that excludes the ascending aorta. Type A pathologies are treated as surgical emergencies, whereas type B pathologies are generally managed with aggressive medical therapy, with surgical and procedural interventions to manage complicated cases including those with malperfusion syndromes, persistent pain despite appropriate blood pressure management, and rapid progression of disease on interval imaging [19]. In the past decade, thoracic endovascular aortic repair (TEVAR) has become the treatment of choice for stabilizing wall abnormalities of the descending thoracic aorta [20]. One area that remains challenging in terms of management is the “non-A non-B” dissection, which originates in the aortic arch and descends distally, as this type of dissection has a poorly characterized natural history. More recently the Society for Vascular Surgery/Society for Thoracic Surgery (SVS/STS) classification systems was developed, which defines dissections with a greater degree of anatomic detail, including defining both the proximal and distal extent of the dissection [21••].

Aortic Dissection

Aortic dissection makes up 85–95% of all acute aortic syndromes [2]. Dissection occurs when a tear is formed in the intima of the aortic lumen. Intimal tears can be generated either by undue hemodynamic shear stress, frequently in the form of uncontrolled hypertension, or by compromised quality of connective tissues, such as secondary to a collagen vascular disease such as Marfan, Loeys-Dietz, or vascular Ehlers-Danlos syndrome, or from traumatic or inflammatory processes [9, 14]. Once an intimal tear is generated, forward, pulsatile blood flow generates a shearing force that forces blood through the intimal tear and into the aortic media rather than the true lumen, thereby propagating the dissection and forming a false lumen [12]. As the dissection propagates, it may do so in either an antegrade or retrograde fashion, with the potential for the extending false lumen to occlude true lumen flow, causing malperfusion of the coronary vessels or the cerebrovascular vessels (type A) or the visceral vessels (type B). The false lumen is, by definition, fragile, with no full thickness wall surrounding it; it is thus prone to tearing further, either rupturing through the intima to rejoin the true lumen, forming a so-called re-entry tear, or rupturing through the adventitia, leading to free hemorrhage into a body cavity or pericardium [12].

Natural History

Aortic dissection is recognized as a highly morbid process, with overall 30-day mortality for patients presenting to the hospital with TAAD exceeding 50% [6]. Autopsy-based community studies suggest that roughly 50% of patients with a TAAD die before ever reaching a hospital, suggesting that less than 1 in 4 patients who experience an ascending aortic syndrome will be alive one month later [6, 7]. Delayed recognition of aortic dissection is associated with worse outcomes, with a mortality rate of 1–2% per hour over the first 24 h and exceeding 75% in the first 2 weeks [6].

Type B dissection (TBAD) is less common than TAAD; however, patients with type B aortic syndromes do better than type A. Rather than the 50% mortality rate associated with TAAD at 30 days, TBAD is associated with a 30-day mortality rate of only 13% [22, 23]. TBAD, however, can be complex and present with increased 30-day mortality risk (20.0%) if they are complicated by a malperfusion syndrome [23]. It has been estimated that 30–50% of patients presenting with TBAD develop a complicated dissection, defined as being accompanied by a malperfusion syndrome, persistent pain or refractory hypertension, or rapid progression of disease on interval imaging [22, 23]. One study estimated that 27% of patients presenting with a TBAD required a procedural intervention during their index hospitalization [22].

Treatment

TAAD (Fig. 1a) is a cardiothoracic surgical emergency and should prompt expeditious evaluation by a cardiac surgeon regardless of a patient’s symptoms or clinical stability. Surgical treatment is dictated by the involvement of the coronary vessels and valve compromise, with valve sparing techniques and coronary reimplantation employed where possible [19]. Broadly, repair involves replacement of the ascending aorta with an interposition graft via a median sternotomy under hypothermic circulatory arrest [1].

The mainstay of treatment for uncomplicated TBAD (Fig. 1b) is medical management in the form of blood pressure and heart rate control to decrease aortic wall stress, usually with an intravenous, easily titratable beta-blocker [24, 25] and, secondarily, vasodilators [26, 27]. The lowest blood pressure that does not compromise end organ function should be targeted [14], typically at least systolic blood pressure < 120 mmHg and diastolic blood pressure < 80 mmHg, with target heart rate < 80 beats per minute. Patients should be admitted to the intensive care unit with close blood pressure monitoring and frequent assessments in the form of neurovascular and laboratory checks for the development of a malperfusion syndrome [14]. Regardless of whether or not TBAD is deemed uncomplicated, early evaluation by a vascular surgeon is critical in the event that the dissection propagates, requiring thoracic or complex visceral endografting or surgery.

The seminal trials applying TEVAR in TBAD applied it in complicated dissection and demonstrated 8% perioperative mortality and a survival rate of 84–88% at 1–2 years [28-30]. Roughly 20% of patients in these early trials required a reintervention at 30 days, mostly due to false lumen growth on imaging, development of a malperfusion syndrome, or retrograde dissection [28]. More recently, data from the STABLE II trial demonstrated favorable 30-day and 1-year clinical and anatomical outcomes for the treatment of rupture and malperfusion in the setting of acute, complicated TBAD using a composite device design (covered stent graft and bare metal stent) with partial or complete false lumen thrombosis seen in 100% of the stent grafted segments and 97.7% of the bare mental stented segments at 1 year [31••]. From a durability standpoint, it remains unclear if TEVAR is superior to open surgery in the modern era; however, the morbidity reduction associated with TEVAR has been demonstrated for emergent operations, as well as for older, more frail patients [32].

Recently, interest around the role of TEVAR in uncomplicated acute TBAD has generated several trials investigating its application. However, as the mortality rate associated with medical management is significantly lower for uncomplicated TBAD, it is more challenging to demonstrate short-term mortality benefits of TEVAR in this setting, causing investigators to look at remodeling endpoints instead. The Investigation of Stent Grafts in Aortic Dissection with Extended Length of Follow-up (INSTEAD-XL) trial investigated the role of TEVAR in stable, chronic (> 14 days) TBAD, and demonstrated improved aorta-specific mortality at 5 years (6.9% vs. 19.3%, $p = 0.04$) but not all-cause mortality (11.1% vs. 19.3%, $p = 0.13$) for TEVAR compared with medical therapy alone [33]. More recently, the ADSORB (Acute Dissection: Stent Graft or Best Medical Treatment) trial enrolled 61 patients presenting with acute TBAD < 14 days from onset and demonstrated improved aortic remodeling as measured by false lumen thrombosis and freedom from aortic dilation for patients receiving TEVAR over medical therapy (97% vs 43%, $p < 0.001$) [34].

As a result of these data, many physicians are treating acute TBAD earlier in the disease process than previously recommended in an effort to promote aortic remodeling and reduce long-term aortic degeneration and related complications. Currently, upfront TEVAR is recommended only for TBAD that are complicated by a malperfusion syndrome, persistent pain or refractory hypertension, or rapid progression of disease on interval imaging [14]. For those patients presenting with uncomplicated TBAD but with high-risk features (Table 1), the risks and benefits of applying TEVAR early to prevent complications must be

weighed against the risks of inducing retrograde dissection by deploying a stent graft in a freshly dissected aorta. If possible, many surgeons opt to wait until 4 weeks after the acute event before placing a TEVAR, consistent with the results of the INSTEAD trial [33]. There currently remain no recommendations for the application of TEVAR in low-risk uncomplicated TBAD; all of these patients should be initially managed medically and followed with serial imaging as the risk of early TEVAR likely outweighs the benefits in this patient population.

Regardless of the time period in which TEVAR is performed, there are several best practices which should be followed [35••]. Percutaneous femoral artery access under ultrasound guidance is now considered the standard of care for TEVAR deployment. Intravascular ultrasound (IVUS) should be used to confirm true lumen positioning and allows assessment of dissection flap dynamics both before and after stent deployment. The aortic coverage achieved with stent graft deployment should cover the entry tear but should otherwise be minimized in an effort to reduce the risk of spinal cord injury; the hybrid covered and bare metal dissection stents investigated in the STABLE II trial allow for stabilization of the true lumen through the visceral segment without extended coverage of the aorta [31••]. Coverage of the left subclavian artery in urgent and emergent setting is acceptable, with delayed revascularization performed for patients with left arm ischemia, claudication, or signs of spinal cord ischemia or vertebrobasilar insufficiency. This is in contrast to the practice for elective TEVAR for dissection, where revascularization of the left subclavian should be performed prior to stent graft placement to reduce the risk of adverse neurologic outcomes.

Intramural Hematoma

Intramural hematoma (IMH) (Fig. 1c) is a similar entity to acute aortic dissection but lacks two criteria seen in frank dissection: an evident intimal tear and a perfused false lumen [26]. IMH is thought to occur either from spontaneous rupture of the vasa vasorum within the aortic media, leading to medial bleeding that propagates in a manner similar to dissection and eventually tamponades, or from an intimal tear too small to be discerned on imaging studies that generates a thrombosed false lumen [2]. As a result, IMH is generally thought of as being on the spectrum of aortic dissection pathology. Diagnosis of IMH is defined by circumferential or crescent shaped thickening of the aortic wall > 5 mm. The imaging study of choice to identify IMH is generally a CT scan with and without contrast, as the hematoma enhances at a higher density than unenhanced blood but does not perfuse on the administration of contrast, signifying the presence of fresh thrombus without active flow [1].

Isolated IMH is present in 5–15% of in patients presenting with acute aortic syndromes [24, 36-38]. It more frequently affects the descending thoracic aorta, but can also involve the ascending aorta (58% type B vs. 42% type A, $p < 0.001$) [24]. While IMH is considered to be a more stable acute aortic syndrome than frank dissection due to the lack of pulsatile flow and overt intimal tear, it nonetheless leads to weakening of the aortic wall and carries a high rate of in-hospital mortality nearly 30% for type A IMH [24]. The intramural hematoma can rupture through the intima, allowing an acute dissection to propagate secondarily, a phenomenon that has been estimated to occur in 16–47% of cases [2]. Progression to dissection is much more common in the presence of concomitant compromise of the intima,

such as in the presence of penetrating aortic ulcer (PAU) (48% chance of progression with PAU versus 8% without, $p = 0.002$) [36].

It is estimated that as little as 10% of identified IMH resolve spontaneously, with as many as 47% evolving into dissection or aneurysmal degradation without treatment [39]. Current American Heart Association [19] as well as SVS [35••] guidelines suggest that management of IMH should reflect that of an acute dissection of a similar anatomic segment, with type A IMH patients undergoing surgery when possible and type B IMH patients receiving medical management and regular surveillance, utilizing TEVAR where appropriate for patients who have persistent symptoms or complications or show evidence of disease progression on follow-up imaging after a period of hypertension control [40]. TEVAR is effective in stabilizing IMH and inducing regression in the setting of appropriate anatomy [41•] and is appropriate for patients with complicated IMH or uncomplicated IMH with high-risk imaging features, such as PAU-like findings (ulcer-like projection and focal intimal disruption), hematoma > 10 mm, or aortic diameter >45 mm [36, 42-44]. Medical management of uncomplicated type B IMH is generally effective in resolving 50–90% of lesions [2, 14, 45]. Medical management of type A IMH is not recommended, with mortality as high as 40% noted in one retrospective study, versus 24% mortality of those treated surgically [46].

Penetrating Aortic Ulcer

Penetrating aortic ulcer (PAU) is a phenomenon in which atherosclerosis progresses to the point that the inflammatory plaque ulcerates, creating a tear in the aortic intima that can lead to focal IMH, dissection, or rupture [19, 47]. The appearance of PAU on cross-sectional imaging is a focal outpouching of the aortic lumen within the aortic wall itself (Fig. 1d) [48]. PAU generally results from long-standing atherosclerosis in the setting of poorly controlled hypertension. They are frequently identified in older, more frail patients with extensive atherosclerotic disease burden and a high rate of comorbidities such as tobacco use, chronic obstructive pulmonary disease, coronary artery disease, and chronic renal insufficiency [26]. PAU account for as little as 2–7% of all cases of acute aortic syndromes [1] and most frequently occur in the descending thoracic aorta, followed by the ascending and abdominal aorta [49].

The natural history of PAU is poorly defined and can either be stable or progress to IMH, dissection, or rupture. There is substantial variation in the reported natural history of PAU, with studies reporting rupture risk ranging from 3% to 40%, depending on the presence or absence of symptomatology or high-risk characteristics [47,50••]. Symptomatic PAU is associated with a high risk of early disease progression, and depth and diameter of PAU on imaging can be used to delineate which lesions may be of highest risk; specifically PAU diameters over 13–20 mm and deeper than 10 mm are associated with worse outcomes [48]. In contrast, asymptomatic PAU without high-risk features on imaging is associated with minimal growth and infrequent complications including rupture [50••].

Management of PAU is often dictated by the comorbidity burden of the affected patient—often there is opportunity to initiate antihypertensives and statins to stabilize

the atherosclerotic plaque and minimize hemodynamic stress while initiating imaging surveillance [1]. If the patient demonstrates signs of rapid disease progression, concerning imaging features including ulcer growth or presence of concomitant IMH, or has poorly controlled hypertension or ongoing pain, TEVAR can be considered as an option to stabilize and exclude acute type B PAU [51, 52]. For type A PAU, this is more challenging, as few patients presenting with type A PAU are good candidates for surgical intervention. Often care for PAU is individualized and involves close follow-up with a multidisciplinary team as these patients are at high risk for future hospitalizations even if the course of the PAU is stable and benign.

Follow-up

Once stabilized, patients who present with acute aortic syndrome must be surveilled with serial imaging and close outpatient follow-up regardless of management strategy. Patients receiving medical management (i.e., those who do not receive upfront surgical or endovascular intervention, mostly those with type B pathology) must be followed closely by a vascular surgeon with serial cross-sectional (CTA) imaging at 1, 3, 6, and 12 months post-pathology and yearly thereafter [1]. It is also critical these patients regularly see a primary care provider or cardiologist to maintain an outpatient medication regimen that controls their blood pressure and heart rate within predefined parameters [25, 53]. Recent studies have demonstrated that ACE inhibitors and ARBs are beneficial in the long-term medical management of aortic dissection [26]. Roughly one-third of type B aortic dissection patients who are initially managed medically will ultimately go on to require surgery, usually to repair aneurysmal degeneration of the dissected segment. Patients with partially thrombosed or large (> 22 mm) false lumens are at higher risk for late aneurysmal degeneration [54], as are patients with connective tissue disease [55].

Receiving surgical or procedural intervention does not preclude the need for imaging surveillance. For those patients undergoing surgery for type A aortic pathology, surveillance imaging is recommended at 1, 3, 6, and 12 months with no recommendations on continuing surveillance if repair is stable at 1 year [19]. Surveillance imaging post-TEVAR is more involved due to the risk of endoleak, aneurysmal degeneration, or disease progression. Among patients undergoing upfront TEVAR for complicated acute TBAD, 5-year mortality following the initial hospitalization has been estimated to be as high as 40% [56]. It is therefore recommended that those patients receiving TEVAR should be followed with CTA at 1 month, 6 months, 12 months, then yearly thereafter; however, when applied for acute aortic syndrome earlier evaluation may be warranted, usually prior to discharge from the hospital or within 1 week of stent graft placement [35••].

Conclusions

Acute aortic syndromes are life-threatening disease processes that threaten the aortic wall, requiring prompt diagnosis and multidisciplinary care both upfront and longitudinally. While aortic dissection is the most common and perhaps most feared of these syndromes, PAU and IMH are related, malignant arterial pathologies that are increasingly identified on imaging studies and must be managed appropriately. Communication between specialties

and individualized care are critical to improve outcomes and select the appropriate combination of medical and procedural therapy, follow-up, and imaging surveillance for patients presenting with acute aortic syndromes.

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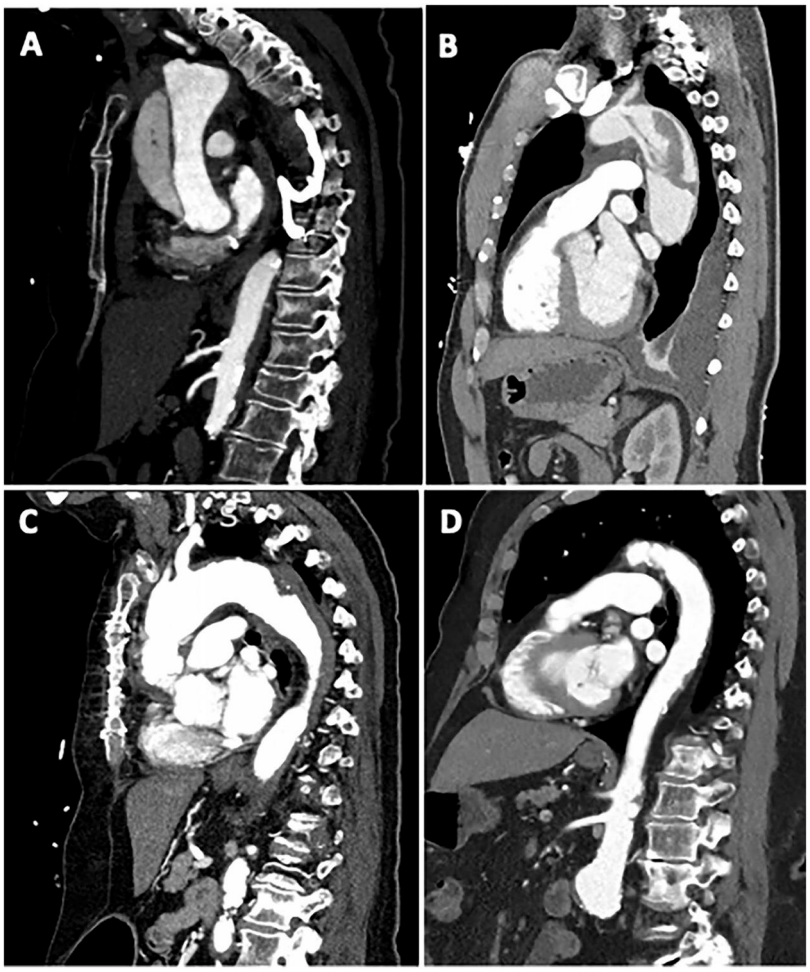

Fig. 1.

a 84F with acute TAAD requiring surgical repair. **b** 42 M with hypertensive crisis and TBAD. **c** 84F with thickening and intramural hematoma of the descending thoracic aorta. **d** 78F with penetrating aortic ulcer of the abdominal aorta adjacent to the celiac axis

Table 1

Criteria for complicated versus high risk Type B acute aortic syndromes

Complicated	High Risk
• Rupture (contained or free)	• Refractory pain >12 h despite maximal doses of pain medication
• Extension or enlargement of false lumen on interval imaging	• Refractory hypertension despite > 3 classes of antihypertensive at maximal doses
• Aortic diameter > 5 cm	• Aortic diameter > 4 cm
• Visceral malperfusion, clinically evident	• Radiographic only malperfusion
• Peripheral malperfusion, clinically evident	• Bloody pleural effusion
• Retrograde type A dissection	• False lumen diameter > 22 mm
	• Entry tear on lesser curvature