Aortic Diseases: Dilation and dissections

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Objectives:
1. Describe the echocardiographic characteristics and classification of aortic aneurysms and dissections.
2. Compare the diagnostic utility of various imaging modes, including TTE and TEE, in the evaluation of suspected aortic dissection.
3. Describe the echocardiographic characteristics and prognostic significance of an intramural hematoma or a penetrating ulcer.

Introduction:
Transesophageal echocardiography is an excellent tool for evaluating the intrathoracic and proximal intra-abdominal aorta. It provides comprehensive and real time evaluation of a variety of disease processes, which may affect this segment of the aorta. TEE imaging provides high resolution views of the three layers of the aortic wall and lumen. It is a time efficient procedure and coupled with the convenience of bedside portability, TEE is an excellent tool to evaluate patients presenting with signs and symptoms of acute aortic pathology.

Evaluation of the aorta by TEE should include the aortic root, ascending aorta, transverse aorta (arch), intra-thoracic, and the upper intra-abdominal descending aorta. The aortic root is composed of the AV annulus, aortic valve cusps, sinuses of Valsalva, coronary artery ostia, sinotubular junction. The ascending aorta originates at the level of the sinotubular junction and extends to the aortic arch. Eight standard views of the aorta are part of a comprehensive exam.

A short segment of ascending aorta where it joins the aortic arch is not visible because of the air filled trachea being interposed between the aorta and the esophagus. Epiaortic scanning with a sterile sleeve covering a high frequency transducer provides detailed examination of the entire ascending aorta and proximal arch and can overcome this limitation with TEE.

Aortic Aneurysmal Disease:
Aneurysmal disease is defined as a permanent localized dilation of an artery by at least 1.5 times the normal diameter. Enlargement between 1 and 1.5 times the normal diameter is defined as ectasia. Aneurysms can be classified by location, shape, or etiology. The Crawford Classification defines 4 types of thoracoabdominal aneurysms. Type I originate in the proximal descending thoracic aorta and terminate above the renal arteries. Type II originate in the proximal descending thoracic aorta and terminate below the renal arteries. Type II aneurysms may also involve the aortic root. Type III originate in the distal
descending aorta below the 6th intercostal space. Type IV are limited to the abdominal aorta. Aneurysms may be either saccular or fusiform. Etiologies for aneurysms include congenital as well as acquired conditions. The most common connective tissue diseases that affect the aorta are Marfan’s syndrome and Ehlers Danlos syndrome. Acquired diseases account for most aneurysms and include atherosclerosis and medial degeneration associated with aging. In 2005, Loeys-Dietz syndrome was identified and consists of a genetic mutation within the TGFBR1 and TGFBR2 genes. Patients demonstrate arterial tortuosity and are risk for aortic aneurysms and dissections, which may present clinically during pregnancy. A high prevalence of aortic root dilation is also seen in bicuspid aortic valve patients, irrespective of valvular hemodynamics or age, suggesting an intrinsic aortic defect. Studies have demonstrated that patients with bicuspid aortic valves have increased activity of matrix metalloproteinases which are responsible for cell matrix turnover. Patients with bicuspid aortic valves should have their thoracic aorta evaluated for signs of aneurysm or dissection formation and should undergo routine ongoing surveillance for progression of aortic disease. First-degree relatives of patients with a bicuspid aortic valve should be referred for evaluation for the presence of a bicuspid valve and asymptomatic thoracic aortic disease.

Hypertension, smoking and aging are also associated with aortic disease. Other less common causes include infection, inflammation, trauma and iatrogenic injuries during surgery or procedures. Aneurysms larger than 5 cm or two times the normal artery size should be considered for treatment due to the risk of rupture. Patients with aneurysms secondary to congenital causes may be candidates for treatment at a smaller size. Analysis by Elefteriades demonstrated that ascending aortic aneurysms grow at a rate of 0.07 cm/yr and 0.19 cm/yr for descending aneurysms. A study of 133 patients demonstrated an equal representation in men and women yet women presented later in life (women 75.8 ± 12.7 years vs. 62.8 ± 17.3 years for men). 79% of the aneurysms were less than 6 cm at initial diagnosis while the remaining 21% exceeded 6 cm at presentation and ruptures were much more common in women than in men (5 year cumulative rupture rate 33% for women and 9% for men). In the 133 patients, 24% elected to have operative management with an 8% 30 day mortality for elective procedures and a 57% 30 day mortality for emergent operations. Management recommendations include aggressive control of hypertension, urgent evaluation for repair in patients with symptomatic aneurysms or aneurysms associated with dissections. The size of the aneurysm and the decision regarding urgent versus elective repair is under debate, with current guidelines recommending surgical replacement at smaller internal diameters than previously published.
Timing of elective repair has been outlined in recent guidelines and suggests the following:

- Idiopathic ascending aneurysm > 5.5 cm
- Patients with Marfan syndrome > 5.0 cm
- Female patient with Marfan syndrome contemplating pregnancy > 4.0 cm
- Patients with Loeys-Dietz syndrome > 4.2 cm

Specific criteria for patients with BAV and dilated ascending aorta are in the table that follows:

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<thead>
<tr>
<th>Indications for Elective Surgical Repair of Dilated Ascending Aorta Associated with BAV</th>
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<tbody>
<tr>
<td>• Aortic Diameter &gt; 5.0 cm</td>
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<tr>
<td>• Aortic Diameter &gt; 4.5 cm with any of the following:</td>
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<tr>
<td>• Expansion rate &gt; 0.5 cm/y in an adult</td>
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<tr>
<td>• Aortic coarctation, corrected or uncorrected</td>
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<tr>
<td>• First degree relative with ascending aortic dissection or rupture</td>
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<td>• Long history of smoking, especially with COPD</td>
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<td>• Small adult body size, indicated by either of the following:</td>
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<td>• Ratio of aortic area to body height &gt; 10 cm²/m</td>
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<tr>
<td>• Ratio of aortic diameter to body surface area &gt; 4.25 cm²/m²</td>
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<tr>
<td>• Aortic diameter &gt; 4.0 cm with concomitant indication for elective AVR</td>
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(adapted from 5,30)

Echocardiographic images will demonstrate aortic enlargement. The size may be measured in the 2D mode using the caliper function. Internal diameter measurements should be made perpendicular to the axis of blood flow. Areas of atherosclerosis, calcium deposition and thrombus may be seen within the aneurysmal segment. Spontaneous echo contrast may also be noted and is a reflection of slow moving blood cells. Size, length, location and associated echocardiographic findings should always be documented. Aortic root dilation may lead to aortic insufficiency and can be documented using 2D, Pulse wave and color flow Doppler techniques.

**Aortic Dissections:**
Aortic dissections are part of a disease process entitled the acute aortic syndrome. Other pathologic processes represented in this syndrome include aortic intramural hematoma (AIH) and penetrating atherosclerotic ulcer (PAU). Two separate classification systems have been developed for aortic dissections based upon anatomic location. The Stanford Classification consists of Type A dissections that involve the ascending aorta and Type B which are limited to the
descending aorta. The Debakey Classification has three subcategories. Type I dissections originate in the ascending aorta and extend into the arch and descending thoracic aorta. Type II dissections originate and are confined to only the ascending aorta. Type III originate and are limited to the descending aorta and are subclassified with IIIa limited to the thoracic aorta and IIIb extending into the abdominal aorta.\textsuperscript{10}

Aortic dissections are associated with bicuspid aortic valves, hypertension, preexisting aortic disease, pregnancy, cocaine use, trauma and iatrogenic surgical injury. Patients typically present with the acute onset of chest pain and back pain. Secondary symptoms may become manifest as end organs become ischemic either from direct extension or luminal occlusion by the dissection flap. Malperfusion of the branch vessels arising from the aorta can be compromised either by direct extension of the flap down the ostia (static obstruction) or by prolapse of the flap over the ostia (dynamic obstruction)\textsuperscript{11}.

The aortic wall is comprised of three layers, the intima, the media and the adventia. In an aortic dissection, blood penetrates the intimal layer and enters the media layer and may propagate both anterograde and retrograde along the aortic wall.

Echocardiographic assessment includes documentation of an undulating linear echodensity within the aortic lumen. This echodensity is the intimal flap which has separated from the outer two layers of the aortic wall. This intimal flap separates the true from the false lumen. Different Doppler color flow patterns can be demonstrated when comparing the true and false lumen with a predominance of systolic flow in the true lumen. Other associated aortic findings include central displacement of intimal calcification, and the presence of aortic wall thrombus.

A variety of other associated findings with dissections can complicate the patient's clinical course and lead to further surgical or medical management. The TEE is useful for diagnosis of these issues both preoperatively and intraoperatively and can be used intraoperatively to guide resuscitation and alter surgical management. Acute aortic valve insufficiency may occur if the dissection alters the anatomic relationship of the aortic valve cusps at their attachment to the aortic wall, or if the intimal flap prolapses across the aortic valve leaflets. This acute aortic insufficiency may need to be treated with resuspension of the aortic valve or by aortic valve replacement. Regional wall motion abnormalities secondary to coronary ischemia may be visualized secondary to dissection down the coronary artery or flap occlusion of the coronary ostia. The dissection may also lead to extravasation of blood into the pericardial or pleural cavities which can be detected by TEE as abnormal fluid collections. An important intraoperative use of TEE is interrogation of flow in both the true and false lumen. Flow should be assessed both prebypass, on bypass, and post bypass to ensure that malperfusion situations are recognized and addressed.\textsuperscript{12,13} TEE should be utilized on bypass to monitor for LV distension during cooling and post bypass TEE evaluation should focus upon assessment of biventricular function, aortic
valve competence and patency of coronary ostia. Presence of significant aortic insufficiency or new segmental wall motion abnormality following ascending aortic repair may lead to reinstitution of cardio-pulmonary bypass and aortic valve repair/replacement or coronary artery bypass grafting to the affected coronary wall segment.14

Type A dissections require urgent surgical management to prevent further morbidity and mortality. Type B dissections are typically treated with aggressive medical management. Type A dissections treated medically had 58% mortality in contrast to 26% mortality rate if treated surgically. Type B dissections treated medically had a 10.7% mortality compared to 31.4 % mortality if treated surgically. Most common cause of death in patients with Type A dissections was aortic rupture and tamponade (41.6%). Mortality rates approximate 1% to 2% per hour early after symptom onset in Type A dissections making early diagnosis and urgent treatment imperative.10 Similar to aortic aneurysms, females with aortic dissections demonstrated an increased mortality rate when compared to men.15

BAV disease carries a 6.14% lifetime risk of aortic dissection and this risk is 9 times higher than the general public. In comparison, Marfan syndrome has a 40% lifetime likelihood of aortic dissection. Given the much higher prevalence of BAV in comparison to Marfan syndrome, BAV is responsible for an equal or greater number of aortic dissections.31 In a retrospective review, patients with an unreplaced BAV, were followed to determine growth rates of the ascending aorta. All patients had unrepaired ascending aortic aneurysms which were greater than 3.5 cm. 13.4% of the patients followed had BAV. Ascending aorta growth rates for all patients was 0.14 cm/yr but patients with BAV had increased growth rates of 0.19 cm/yr. The growth rate for patients with BAV and associated AS was slightly elevated at 0.20 cm/yr. Based upon this study, the presence of BAV with AS warrants frequent assessment of the aorta and valve.32 Other studies have demonstrated that AVR alone for BAV does not prevent further aorta dilatation.33

Aortic Intramural Hematoma (AIH):
Since the advent of high resolution diagnostic imaging both aortic intramural hematoma and penetrating atherosclerotic ulcer are now recognized as other causes of acute aortic pathology. Aortic dissection, aortic intramural hematoma and penetrating atherosclerotic ulcer collectively comprise “the acute aortic syndrome.”12 Similarities exist between these three clinical conditions in regards to their presentation and management. All three conditions need to be treated aggressively as they share common rapidly developing life-threatening complications. AIH is considered a precursor to classic dissection and should be treated in a similar fashion.5,10
The definition of AIH is localized separation of the layers of the aortic wall by partially or totally clotted blood in the absence of an intimal tear. The proposed mechanism is rupture of the vasa vasorum in the aorta media or from a penetrating atherosclerotic ulcer.\textsuperscript{16}

Echocardiographic criteria include a 7-mm circular or crescentic thickening of the aortic wall extending 1-20 cm longitudinally along the aortic wall. In addition, no flow should be demonstrated in the thickened wall and no intimal laceration or flap should be visualized. Other features include a layered appearing segment of aorta with echolucent areas in a widened aortic wall as well as central displacement of intimal calcifications.\textsuperscript{16}

Disease progression in AIH is very similar to aortic dissection. 5 out of 15 patients progressed to a typical dissection and 4 out of 15 developed aortic rupture.\textsuperscript{17} Management is similar to patients with an aortic dissection favoring surgical repair for AIH in the ascending aorta and medical management for AIH limited to descending thoracic aorta.\textsuperscript{18} Ascending AIH can progress to rupture, tamponade or compression of coronary ostia.\textsuperscript{10}

A meta-analysis of the literature demonstrated that surgical management was superior to medical management in Stanford Type A AIH (14% vs. 36% mortality rates respectively). Both medically and surgically managed Type B AIH had a mortality rate of 14%.\textsuperscript{19}

In 2003, van der Loo and Jenni recommended the inclusion of intraluminal thrombus (primary mural thrombus) and aortitis into the acute aortic syndrome.\textsuperscript{20} TEE is one diagnostic modality to identify intraluminal thrombus. A mobile pedunculated echodensity is visualized in the thoracic aorta. In some patients the thrombus is associated with aortic sites without underlying atherosclerotic disease.\textsuperscript{21} Other literature reports indicate that thrombus originates from sites of underlying aortic pathology including atherosclerotic lesions or previous aortic procedural sites. Hypercoagulability is also a risk factor. Patients typically present with systemic embolization. Optimal management techniques for this rare condition is still debated but has included thrombolysis, long-term
Penetrating Atherosclerotic Ulcer of the Aorta (PAU):
This entity occurs when an atherosclerotic plaque erodes into the media of the aorta. Sequelae include aneurysm formation, aortic intramural hematoma, dissection or rupture. Most PAU are located in the descending thoracic aorta where atherosclerotic lesions are more frequently located. Long term outcome studies are lacking and standard management strategies have not been adopted. Presentation is similar to the other acute aortic syndromes, with acute excruciating back pain being the most commonly reported symptom. These patients may rapidly deteriorate with impending aortic rupture. Some patients with PAU will be evaluated by TEE to rule out an aortic dissection, and AIH and PAU should be considered part of the differential diagnosis for patients presenting with chest and back pain.

Echocardiographic features include a crater like defect with jagged edges located within a complex atherosclerotic plaque. It has been associated with AIH, and like AIH may be a precursor to aortic dissection. PAU disease progression may be related to maximum initial diameter and depth. Patients with PAU with an initial depth > 10 mm or maximal diameter > 20 mm have been shown to demonstrate a high risk for disease progression. These patients should be considered as early surgery or endovascular repair candidates. In addition patient's with PAU in the ascending, arch and proximal descending aorta had a more malignant course than patients with disease in their middle and distal descending thoracic aorta. These patients should also be considered for surgical or endovascular repair. In contrast, many patients with PAU in the descending thoracic aorta can be safely managed medically.
**Imaging Modalities:**

In multiple studies, TEE has been evaluated in comparison to TTE, CT, aortography, CT + aortography, and MRI. Newer multiple slice CT scanners can best identify thoracic aortic disease as well as other conditions which present similarly to aortic disease, including coronary artery disease and pulmonary embolism. Based upon this benefit, 64-slice CT is becoming the diagnostic procedure of choice in the work up of acute chest pain.\(^5\)

TEE has an overall sensitivity of 98%, which correlates well with the sensitivity of MRI. Unfortunately, the specificity of TEE in comparison to MRI is inferior secondary to artifacts, most commonly a mirror image or reverberation artifact. Based upon the sensitivity and specificity, recommendations by Nienabern and colleagues are MRI should be the initial diagnostic procedure for aortic dissections in stable patients and TEE the procedure of choice for unstable patients.\(^27\) Concerns with this recommendation include patient transport, limited monitoring in the MRI suite, length of the MRI study, and cost. The TEE provides a quick and highly sensitive procedure in a safe and cost effective fashion. The addition of M-mode echocardiography can help increase the specificity and sensitivity by helping to rule out artifacts. Application of 3D TEE imaging may also limit errors secondary to artifacts.

Comparison of TTE to TEE clearly demonstrates TEE’s superiority in the diagnosis of thoracic aortic dissections. TTE had a sensitivity of only 59.3% in comparison to 97.7% for TEE.\(^27\) Recent studies proved Live 3D TTE to be superior to standard TTE in demonstrating an aortic dissection in 10/10 patients while standard 2D TTE only isolated the defect in 5 of the same 10 patients.\(^28\) Small 3D TEE studies have demonstrated enhanced identification of true lumens, false lumens and dissection flaps.\(^29\) As 3D TEE becomes more widely utilized and accepted the specificity and sensitivity for aortic pathology is expected to increase.

References:


