Imaging Standards for Thoracic Aortic Diseases

Although diseases of the thoracic aorta are the consequence of degenerative, acquired, genetic, or traumatic conditions, they all are ultimately manifested by structural changes in the aorta that can be diagnosed by imaging studies. For that reason, systematic and precise characterization of thoracic aortic anatomy and pathophysiologic changes by transesophageal echocardiography (TEE) in combination with angiography, computed tomography (CT), or magnetic resonance imaging (MRI) studies provide important information for determining prognosis, the timing of surgical repair, the type of surgical repair, and the risk of the planned procedures.

The 2010 ACCF/AHA Guidelines for the management of patients with thoracic aortic disease provided recommendations for standard definitions and essential elements to be included in the reporting of thoracic aortic imaging studies. Aneurysm was defined as a localized dilation > 50% the diameter of normal aorta. Pseudoaneurysm was defined as a disruption of the arterial wall with contained extravasation of blood. Ectasia was defined as arterial dilation with a diameter < 150% of the diameter of normal artery. Arteriomegaly was defined as diffuse dilation involving several segments that are > 50% the diameter of normal aorta. Thoracoabdominal aneurysm was defined as an aneurysm involving the thoracic and abdominal aorta. An abdominal aortic aneurysm was defined as an aneurysm involving the infra-diaphragmatic aorta. Aortic dissection was defined as a disruption of the media layer of the aorta with bleeding within the wall of the aorta.

Thoracic aortic imaging studies should provide the following details: 1) anatomic location of abnormality, 2) maximum diameter, perpendicular to the axis of blood flow and the extent of the abnormality, 3) diameter of aortic valve annulus (AoV annulus), sinus of Valsalva (SOV), sinotubular junction (STJ), and the ascending aorta (Asc Ao), 4) the presence of internal filling defects such as atheroma or thrombus, 5) the presence of dissection, intramural hematoma, penetrating atherosclerotic ulcer, or calcification, 6) evidence of extension into or aortic branch vessel involvement, 7) evidence of rupture, hematoma, or pericardial effusion, pleural effusion, or extravasation, and 8) Comparison to prior examinations if available to determine the magnitude and rate of progression of disease.

Thoracic Aortic Aneurysms

The ascending aorta including the aortic root is the most common site for aneurysms of the thoracic aorta. It is estimated that approximately sixty percent of thoracic aortic aneurysms involve the ascending aorta or aortic root. The objectives for surgical operation to repair a dilated ascending aorta or aortic root include correcting associated aortic valve disease, eliminating the mass effect caused by the aneurysm, relieving
refractory pain caused by the aneurysm, preventing aortic rupture, and preventing aortic dissection. In the absence of clinical symptoms, the decision to repair an ascending aortic aneurysm is based on weighing the risks associated with operation against the risks of eventual aortic rupture or dissection.

Clinical decisions important to planning the operative repair include: 1) Is there significant aortic valve disease? 2) Does the aortic valve need to be replaced? 3) Can the aortic valve be repaired? 4) Is there disease of the aortic root? 5) Does the aortic root need to be replaced? And 6) Does the ascending aorta or aortic arch need to be replaced? Many of these questions can be addressed by TEE examination and quantification of the aortic valve, aortic root, and ascending aorta to guide surgical decision making for choosing among the following options: 1) Aortic valve replacement alone, 2) Ascending aortic graft alone, 3) Combined aortic valve replacement and ascending aortic graft, 4) Composite aortic root replacement (Bentall procedure), or 5) Aortic valve-sparing aortic root replacement (David procedure).

Clinical experience and epidemiologic studies of the natural history of thoracic aortic aneurysm support the concept that aneurysm diameter is the most important factor predicting the risk of rupture or dissection. Based on the Yale University thoracic aortic aneurysm and dissection database, the cumulative lifetime risk of complications related to an thoracic aortic aneurysm increased abruptly to 31% for patients with thoracic aortic size $\geq 6.0 \text{ cm}$ in diameter. This information, in combination with an estimated surgical mortality in the range of 2.5% to 5.0%, provided support for the general recommendation that surgical repair was warranted for thoracic aortic aneurysm of at least 5.5 cm in diameter among patients with ascending aortic aneurysms, isolated descending thoracic aortic aneurysm amenable to endovascular repair or descending thoracic aortic aneurysms in patients with dissection or connective tissue diseases without significant co-morbidities. Because operative mortality is greater for open repair of descending thoracoabdominal aortic aneurysms, open surgical repair is generally delayed until the aneurysm diameter exceeds 6.0 cm.

Using aneurysm size greater than 5.5 cm as the only criteria for surgical repair to prevent complications of thoracic aortic aneurysm has certain limitations. It has been recognized that measurement of aortic diameter may be associated with an uncertainty in the range of 0.5 cm caused by differences in imaging techniques or physician interpretation. Data used in analyzing the relation between aortic diameter and the risk of rupture or dissection were based primarily on axial cross sectional images obtained by computed tomography scans that could potentially overestimate the true cross sectional diameter of the aorta if an oblique view of the vessel was used for the measurement. Multiplane transesophageal echocardiography, intravascular ultrasound, or three-dimensional reconstruction of computed tomographic angiography studies may yield smaller aortic diameters because it is more likely that the true center-line cross sectional diameter of the aorta perpendicular to the axis of blood flow can be measured even if the vessel is tortuous. Conventions applied to measure aneurysm diameter may also introduce variability among different imaging techniques. For example, the convention in CT angiography is to measure aneurysm diameter from vessel adventitia to adventitia, while the convention in
ultrasound is to measure aneurysm diameter between the leading edges of the vessel wall or the vessel intima to intima diameter. Differences in the convention and technique used to measure aneurysm diameter need to be accounted for when using these values as criteria for surgical intervention.

Aneurysm etiology has also been demonstrated to be an important risk factor predicting complication rates. For this reason, surgical repair may be justified for thoracic aneurysm diameter in the range of 4.0 cm to 5.0 cm in patients with Marfan syndrome, bicuspid aortic valve, or familial thoracic aortic aneurysm syndrome who exhibit higher rates of rupture or dissection at smaller aneurysm diameters. The natural history and rate of complications in patients with saccular aneurysms or pseudoaneurysms are less predictable in comparison to degenerative fusiform aneurysms and justify surgical repair irregardless of size whenever feasible.

It is also important to recognize that knowledge of the natural history of thoracic aortic aneurysms is incomplete. Epidemiologic studies have analyzed and followed only a selected subset or limited number of patients without intervention until aneurysm rupture or dissection. Also, outcomes may have been influenced by reasons not to operate in certain patients with aneurysms, differences in criteria applied for surgical intervention, variations in the type of operations performed for repair, or the presence of co-morbid conditions that affect mortality.

Aortic diameter also varies among individual patients based upon body size and age. For this reason, it may be advisable to refer to published values for the normal ranges for thoracic aortic dimensions according to age and body surface area. Another approach that has been advocated, especially among patients with congenital or genetic disorders is to index the ascending aortic cross sectional area to the patient height. Using this approach an ascending aortic cross sectional area in centimeters to patient height in meters ratio of greater than 10 provides surgical criteria for ascending aortic replacement.

Finally, it has been established that aortic aneurysm is a dynamic disease state and that aneurysms grow in size over time. For this reason, aneurysm growth rate is another important factor that influences the decision to repair a dilated thoracic aorta. For example, knowing the aneurysm growth rate may be important for deciding whether to repair a dilated ascending aorta detected incidentally by intraoperative transesophageal echocardiography because delaying aneurysm repair until standard aneurysm size criteria have been satisfied would be associated with the added risks of re-operation. Data from the Yale Thoracic Aortic Aneurysm Database determined that the average rate of increase in diameter for ascending aortic aneurysms was 0.07 cm per year. However, reported growth rates for thoracic aortic aneurysm diameter in smaller series have ranged from 0.1 cm per year to as high as 0.42 cm per year. Factors associated with faster aneurysm growth rates were larger initial aneurysm diameter, the presence of dissection, Marfan syndrome, or poorly controlled hypertension. Anticipating eventual growth in the size of an ascending aortic aneurysm over time may be used to justify prophylactic surgical repair of the ascending aorta even if the aneurysm diameter is less than 5.5 cm at the time of aortic valve or root replacement operations in order to avoid the need for re-operation.
within 5 to 10 years. It is also possible to justify earlier surgical intervention in individual patients exhibiting rapid, sudden, or unpredictable increases in the size of their aneurysm over time.

Considerable variability remains on the precise criteria for surgical intervention in patients with thoracic aortic aneurysm and it is likely that criteria may change and evolve based on improvements in surgical techniques and options combined with an improved understanding of the natural history and pathophysiology of thoracic aortic aneurysms. A summary of conditions and criteria for surgical repair of thoracic aortic aneurysm from the 2010 ACCF/AHA guidelines for thoracic aortic disease is listed in the table below. The full text of the guidelines should be referred to address specific issues among each of the individual disease conditions.

<table>
<thead>
<tr>
<th>CONDITIONS</th>
<th>INDICATION FOR SURGICAL REPAIR</th>
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<tr>
<td>Degenerative</td>
<td>Asc Ao ≥5.5 cm</td>
</tr>
<tr>
<td>Aneurysms</td>
<td>Asc Ao &lt;5.5 cm and growth rate &gt;0.5 cm/year</td>
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<tr>
<td></td>
<td>Desc Ao &gt;6.0 cm</td>
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<td></td>
<td>Desc Ao &gt;5.5 cm and candidate for TEVAR</td>
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<tr>
<td></td>
<td>Saccular aneurysm</td>
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<td></td>
<td>Pseudoaneurysm</td>
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<tr>
<td>Marfans</td>
<td>Asc Ao 4.0 cm - 5.0 cm</td>
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<tr>
<td>Ehlers-Danlos</td>
<td>Asc Ao 4.0 cm - 5.0 cm and family history of aortic dissection</td>
</tr>
<tr>
<td>Turners</td>
<td>Asc Ao 4.0 cm - 5.0 cm and rapidly expanding aneurysm</td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td>Asc Ao 4.0 cm - 5.0 cm and planned pregnancy</td>
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<tr>
<td>Familial TAA</td>
<td>Asc Ao 4.0 cm - 5.0 cm and significant aortic regurgitation cm</td>
</tr>
<tr>
<td>Familial Dissection</td>
<td>Desc Ao &gt;5.5 cm</td>
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<tr>
<td>Loey's-Dietz</td>
<td>Asc Ao ≥4.2 cm by TEE</td>
</tr>
<tr>
<td>Syndrome</td>
<td>Asc Ao ≥4.4 cm by CT or MRI</td>
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<tr>
<td>Aortic valve repair or replacement</td>
<td>Asc Ao &gt;4.5 cm</td>
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**Aortic Dissection**

Aortic dissection is the most common surgical emergency of the aorta. Aortic dissection is a result of intimal disruption, allowing blood to dissect through the media creating a “false” lumen that often communicates with the “true” lumen. The most common site of initiation of aortic dissection is the ascending aorta in up to 50% of cases.

TEE is an established imaging technique for the diagnosis of aortic dissection and detection of life-threatening complications of aortic dissection. The two most commonly used classification systems for aortic dissection are the DeBakey (type I, II and III) and Stanford (type A and B) classifications. The distinguishing features of the two classification schemes are that the Stanford classification is based on the site and extent of thoracic aortic involvement, regardless of the location or number of intimal tears, while the DeBakey classification is based upon the anatomic site of intimal tear together with the extent of thoracic aortic involvement. Treatment decisions for aortic dissection
are initially based on classification, with surgical repair indicated for Stanford type A or DeBakey types I and II dissections. While medical treatment is recommended for Stanford type B or DeBakey type III dissection, endovascular repair is emerging as a surgical option, especially in patients with evidence of malperfusion.

Aortic dissection can be diagnosed by TEE, CT, MRI or angiography. For acute aortic dissection, CT and TEE are the most frequent initial diagnostic tests, however, than two-thirds of patients will require at least two imaging tests to confirm the diagnosis. The advantages of TEE over other imaging modalities include its rapidity in diagnosis, the absence of need for intravenous radiographic contrast agents, portability (diagnosis can be made in the emergency room, operating room, or patient bedside), and the ability to provide functional information about ventricular and aortic valve performance. Disadvantages of TEE are that it is somewhat invasive, requires esophageal intubation, cannot reliably image the distal ascending aorta and aortic arch, the confounding effects of ultrasound imaging artifacts and that the diagnostic accuracy of TEE depends upon the experience of the echocardiographer for diagnosing aortic dissection.

Studies have examined the diagnostic accuracy of TEE for the diagnosis of aortic dissection in experienced centers. Both sensitivity and specificity of TEE ranged between 97-100%. Importantly, the negative predictive value of transesophageal echocardiography for aortic dissection approached 100%. To improve specificity it is recommended to image the intimal flap in at least two independent cross sectional views, confirm independent motion of the intimal flap within the vessel walls, and to use Doppler color flow imaging to demonstrate an intimal tear or differential blood flow on both sides of the intimal flap to decrease the chance of artifact. Specific artifacts that may mimic an intimal flap include reverberation or side-lobe artifacts, generated by a vessel wall or by an intravascular catheter or pacemaker lead. The sensitivity of TEE for detection of aortic dissection isolated to the distal ascending aorta or the proximal aortic arch is limited because the air-filled trachea obscures the acoustic window for imaging.

Echocardiography is also important for diagnosing complications associated with aortic dissection. Dissection involving the aortic root can cause aortic regurgitation that can then be quantified echocardiographic techniques. Other complications of aortic dissection include hemopericardium, cardiac tamponade, myocardial ischemia caused by dissection involving the coronary ostia, or stroke from dissection occluding flow into the aortic arch branch vessels. Cardiac tamponade can be diagnosed diagnosis by demonstrating right ventricular or right atrial diastolic collapse in combination with a pericardial effusion. Myocardial ischemia or infarction can be diagnosed by demonstrating myocardial segmental wall motion abnormalities in the distribution of the affected coronary artery. Dissection and malperfusion of the carotid arteries can also be detected using ultrasound vascular Duplex imaging.

Rupture of a descending type B aortic dissection or dissecting descending thoracic aortic aneurysm can sometimes be diagnosed by TEE as a left pleural effusion with blood flow in the pleural space demonstrated by Doppler color flow imaging. Imaging the descending aorta is also important intra-operatively during repair of ascending aortic
dissections using cardiopulmonary bypass to ensure blood flow in the true lumen of the aorta with the initiation of cardiopulmonary bypass and after cross-clamping the ascending aorta.

A variant of aortic dissection is aortic intramural hematoma (IMH). Similar to the Stanford classification for aortic dissection, intramural hematoma may be classified according to involvement of the ascending aorta or aortic arch (Type A) or confinement to the descending thoracic aorta (Type B). The characteristic feature of IMH is the presence of hematoma within the medial layer of aorta causing a crescent shaped thickening of the aortic wall. The echocardiographic definition of aortic intramural hematoma by TEE examination is a greater than 7 mm circumferential or crescentic thickening of the aortic wall imaged in short axis with central displacement of intimal calcification and a layered appearance of the aortic wall imaged in long axis extending for greater than 1 cm along the length of the vessel in the absence of an intimal flap. Intramural hematoma (IMH) is believed to be caused by hemorrhage into the aortic media without luminal continuity. From an imaging standpoint, IMH can be differentiated from its more common variant, the classic aortic dissection, by the absence of an intimal flap. Risk factors for acute mortality and thus surgical repair in patients with IMH include involvement of the ascending aorta, a maximum aortic diameter in the region of IMH of $\geq 48$ mm, or IMH thickness $\geq 11$ mm.

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