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Guidelines for the Diagnosis and Management of Patients With Thoracic Aortic Disease

March 2010

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Contents

1. Introduction	6
2. Critical Issues	10
3. Recommendations for Aortic Imaging Techniques to Determine the Presence and Progression of Thoracic Aortic Disease	14
4. Recommendations for Genetic Syndromes	18
5. Recommendations for Familial Thoracic Aortic Aneurysms and Dissections	24
6. Recommendations for Bicuspid Aortic Valve and Associated Congenital Variants in Adults	26
7. Recommendations for Estimation of Pretest Risk of Thoracic Aortic Dissection	27
8. Initial Evaluation and Management of Acute Thoracic Aortic Disease	34
<i>8.1. Recommendations for Screening Tests</i>	34
<i>8.2. Recommendations for Diagnostic Imaging Studies</i>	35
<i>8.3. Recommendations for Initial Management</i>	36
<i>8.4. Recommendations for Definitive Management</i>	37
9. Recommendation for Surgical Intervention for Acute Thoracic Aortic Dissection	41

10. Recommendation for Intramural Hematoma Without Intimal Defect	42
11. Recommendation for History and Physical Examination for Thoracic Aortic Disease	43
12. Recommendation for Medical Treatment of Patients With Thoracic Aortic Diseases	44
<i>12.1. Recommendations for Blood Pressure Control</i>	44
13. Recommendations for Asymptomatic Patients With Ascending Aortic Aneurysm	46
14. Recommendation for Symptomatic Patients With Thoracic Aortic Aneurysm	50
15. Recommendations for Open Surgery for Ascending Aortic Aneurysm	51
16. Recommendations for Aortic Arch Aneurysms	52
17. Recommendations for Descending Thoracic Aorta and Thoracoabdominal Aortic Aneurysms	54
18. Recommendations for Counseling and Management of Chronic Aortic Diseases in Pregnancy	56
19. Recommendations for Aortic Arch and Thoracic Aortic Atheroma and Atheroembolic Disease	58

20. Periprocedural and Perioperative Management	59
20.1. <i>Recommendations for Brain Protection During Ascending Aortic and Transverse Aortic Arch Surgery</i>	59
20.2. <i>Recommendations for Spinal Cord Protection During Descending Aortic Open Surgical and Endovascular Repairs</i>	60

21. Recommendations for Surveillance of Thoracic Aortic Disease or Previously Repaired Patients	62
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22. Recommendation for Employment and Lifestyle in Patients With Thoracic Aortic Disease	64
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1. Introduction

The term “thoracic aortic disease” encompasses a broad range of degenerative, structural, acquired, genetic-based, and traumatic disease states and presentations. According to the Centers for Disease Control and Prevention death certificate data, diseases of the aorta and its branches account for 43 000 to 47 000 deaths annually in the United States. The precise number of deaths attributable to thoracic aortic diseases is unclear. However, autopsy studies suggest that the presentation of thoracic aortic disease is often death due to aortic dissection (AoD) and rupture, and these deaths account for twice as





many deaths as attributed to ruptured abdominal aortic aneurysms (AAAs). This guideline includes diseases involving any or all parts of the thoracic aorta with the exception of aortic valve diseases and includes the abdominal aorta when contiguous thoracic aortic diseases are present.

Table 1. Applying Classification of Recommendations and Level of Evidence

		SIZE OF TREATMENT EFFECT	
		CLASS I <i>Benefit >>> Risk</i> Procedure/Treatment SHOULD be performed/ administered	CLASS IIA <i>Benefit >> Risk</i> <i>Additional studies with focused objectives needed</i> IT IS REASONABLE to per- form procedure/administer treatment
ESTIMATE OF CERTAINTY (PRECISION) OF TREATMENT EFFECT	LEVEL A Multiple populations evaluated* Data derived from multi- ple randomized clinical trials or meta-analyses	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is useful/effective ■ Sufficient evidence from multiple randomized trials or meta-analyses 	<ul style="list-style-type: none"> ■ Recommendation in favor of treatment or procedure being useful/effective ■ Some conflicting evidence from multiple randomized trials or meta-analyses
	LEVEL B Limited populations evaluated* Data derived from a single randomized trial or nonrandomized studies	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is useful/effective ■ Limited evidence from single randomized trial or nonrandomized studies 	<ul style="list-style-type: none"> ■ Recommendation in favor of treatment or procedure being useful/effective ■ Some conflicting evidence from single randomized trial or nonrandomized studies
	LEVEL C Very limited populations evaluated* Only consensus opinion of experts, case studies, or standard of care	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is useful/effective ■ Only expert opinion, case studies, or standard of care 	<ul style="list-style-type: none"> ■ Recommendation in favor of treatment or procedure being useful/effective ■ Only diverging expert opinion, case studies, or standard of care
Suggested phrases for writing recommendations†		should is recommended is indicated is useful/effective/beneficial	is reasonable can be useful/effective/beneficial is probably recommended or indicated



<p>Class IIb <i>Benefit ≥ Risk</i> <i>Additional studies with broad objectives needed; additional registry data would be helpful</i> Procedure/Treatment MAY BE CONSIDERED</p>	<p>Class III <i>Risk ≥ Benefit</i> Procedure/Treatment should NOT be performed/administered SINCE IT IS NOT HELPFUL AND MAY BE HARMFUL</p>
<ul style="list-style-type: none"> ■ Recommendation's usefulness/efficacy less well established ■ Greater conflicting evidence from multiple randomized trials or meta-analyses 	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Sufficient evidence from multiple randomized trials or meta-analyses
<ul style="list-style-type: none"> ■ Recommendation's usefulness/efficacy less well established ■ Greater conflicting evidence from single randomized trial or nonrandomized studies 	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Limited evidence from single randomized trial or nonrandomized studies
<ul style="list-style-type: none"> ■ Recommendation's usefulness/efficacy less well established ■ Only diverging expert opinion, case studies, or standard of care 	<ul style="list-style-type: none"> ■ Recommendation that procedure or treatment is not useful/effective and may be harmful ■ Only expert opinion, case studies, or standard of care

may/might be considered
 may/might be reasonable
 usefulness/effectiveness is
 unknown/unclear/uncertain
 or not well established

is not recommended
 is not indicated
 should not
 is not useful/effective/beneficial
 may be harmful

* Data available from clinical trials or registries about the usefulness/efficacy in different subpopulations, such as sex, age, history of diabetes, history of prior myocardial infarction, history of heart failure, and prior aspirin use. A recommendation with Level of Evidence B or C does not imply that the recommendation is weak. Many important clinical questions addressed in the guidelines do not lend themselves to clinical trials. Even though randomized trials are not available, there may be a very clear clinical consensus that a particular test or therapy is useful or effective.

† In 2003, the ACCF/AHA Task Force on Practice Guidelines developed a list of suggested phrases to use when writing recommendations. All guideline recommendations have been written in full sentences that express a complete thought, such that a recommendation, even if separated and presented apart from the rest of the document (including headings above sets of recommendations), would still convey the full intent of the recommendation. It is hoped that this will increase readers' comprehension of the guidelines and will allow queries at the individual recommendation level.

2. Critical Issues

As the writing committee developed this guideline, several critical issues emerged:

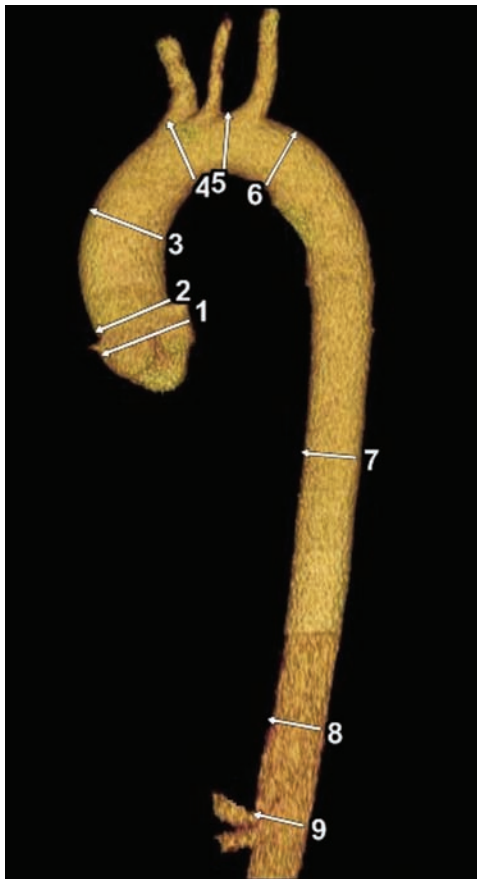
- Thoracic aortic diseases are usually asymptomatic and not easily detectable until an acute and often catastrophic complication occurs. Imaging of the thoracic aorta with computed tomographic imaging (CT), magnetic resonance imaging (MR), or in some cases, echocardiographic examination is the only method to detect thoracic aortic diseases
- Radiologic imaging technologies have improved in terms of accuracy of detection of thoracic aortic disease. However risks associated with repeated radiation exposure, as well as contrast medium–related toxicity have also been recognized. The writing committee therefore formulated recommendations on a standard reporting format (Section 3) as well as surveillance schedules (Section 21).
- Imaging for asymptomatic patients at high risk based on history or associated disease is expensive and not always covered by payers.
- For many thoracic aortic diseases, results of treatment for stable, often asymptomatic, but high-risk conditions are far better than the results of treatment required for acute and often catastrophic disease presentations. Thus, the identification and treatment of patients at risk for acute and

catastrophic disease presentations (eg, thoracic AoD and thoracic aneurysm rupture) prior to such an occurrence are paramount to eliminating the high morbidity and mortality associated with acute presentations.

■ Patients with acute AoD are subject to missed or delayed detection of this catastrophic disease state. Many present with atypical symptoms and findings, making diagnosis even more difficult. Awareness of the varied and complex nature of thoracic aortic disease presentations has been lacking, especially for acute AoD. Risk factors and clinical presentation clues are noted in Section 7. The collaboration of multiple medical specialties for this guideline will provide opportunities to raise the level of awareness among all medical specialties.

■ There is rapidly accumulating evidence that genetic alterations or mutations predispose some individuals to aortic diseases (see Sections 4–6). Therefore, identification of the genetic alterations leading to these aortic diseases has the potential for early identification of individuals at risk. In addition, biochemical abnormalities involved in the progression of aortic disease are being identified through studies of patients' aortic samples and animal models of the disease. The biochemical alterations identified in the aortic tissue have the potential to serve as biomarkers for aortic disease. Understanding the molecular pathogenesis may lead to targeted therapy to prevent aortic disease. Medical and gene-based treatments are beginning to show promise for reducing or delaying catastrophic complications of thoracic aortic diseases.

Figure 1. Normal Anatomy of the Thoracoabdominal Aorta.



Anatomic Location

1. Aortic sinuses of Valsalva
2. Sinotubular junction
3. Mid ascending aorta (midpoint in length between Nos. 2 and 4)
4. Proximal aortic arch (aorta at the origin of the innominate artery)
5. Mid aortic arch (between left common carotid and subclavian arteries)
6. Proximal descending thoracic aorta (begins at the isthmus, approximately 2 cm distal to left subclavian artery)
7. Mid descending aorta (midpoint in length between Nos. 6 and 8)
8. Aorta at diaphragm (2 cm above the celiac axis origin)
9. Abdominal aorta at the celiac axis origin

Normal anatomy of the thoracoabdominal aorta with standard anatomic landmarks for reporting aortic diameter as illustrated on a volume-rendered CT image of the thoracic aorta. CT indicates computed tomographic imaging.

3. Recommendations for Aortic Imaging Techniques to Determine the Presence and Progression of Thoracic Aortic Disease

- Class I**
1. Measurements of aortic diameter should be taken at reproducible anatomic landmarks, perpendicular to the axis of blood flow, and reported in a clear and consistent format. (LOE: C)
 2. For measurements taken by computed tomographic imaging or magnetic resonance imaging, the external diameter should be measured perpendicular to the axis of blood flow. For aortic root measurements, the widest diameter, typically at the mid-sinus level, should be used. (LOE: C)
 3. For measurements taken by echocardiography, the internal diameter should be measured perpendicular to the axis of blood flow. For aortic root measurements the widest diameter, typically at the mid-sinus level, should be used. (LOE: C)
 4. Abnormalities of aortic morphology should be recognized and reported separately even when aortic diameters are within normal limits. (LOE: C)

5. The finding of aortic dissection, aneurysm, traumatic injury and/or aortic rupture should be immediately communicated to the referring physician. (LOE: C)

6. Techniques to minimize episodic and cumulative radiation exposure should be utilized whenever possible. (LOE: B)

Class IIa 1. If clinical information is available, it can be useful to relate aortic diameter to the patient's age and body size. (LOE: C)

Table 2. Essential Elements of Aortic Imaging Reports

1. The location at which the aorta is abnormal.
2. The maximum diameter of any dilatation, measured from the external wall of the aorta, perpendicular to the axis of flow, and the length of the aorta that is abnormal.
3. For patients with presumed or documented genetic syndromes at risk for aortic root disease measurements of aortic valve, sinuses of Valsalva, sinotubular junction, and ascending aorta.
4. The presence of internal filling defects consistent with thrombus or atheroma.
5. The presence of IMH, PAU, and calcification.
6. Extension of aortic abnormality into branch vessels, including dissection and aneurysm, and secondary evidence of end-organ injury (eg, renal or bowel hypoperfusion)
7. Evidence of aortic rupture, including periaortic and mediastinal hematoma, pericardial and pleural fluid, and contrast extravasation from the aortic lumen.
8. When a prior examination is available, direct image to image comparison to determine if there has been any increase in diameter.

IMH indicates intramural hematoma; and PAU, penetrating atherosclerotic ulcer.

Table 3. Normal Adult Thoracic Aortic Diameters

Thoracic Aorta	Range of Reported Mean (cm)	Reported SD (cm)	Assessment Method
Root (female)	3.50 to 3.72	0.38	CT
Root (male)	3.63 to 3.91	0.38	CT
Ascending (female, male)	2.86	NA	CXR
Mid-descending (female)	2.45 to 2.64	0.31	CT
Mid-descending (male)	2.39 to 2.98	0.31	CT
Diaphragmatic (female)	2.40 to 2.44	0.32	CT
Diaphragmatic (male)	2.43 to 2.69	0.27 to 0.40	CT, arteriography

CT indicates computed tomographic imaging; CXR, chest x-ray; and NA, not applicable.

Reprinted with permission from Johnston KW, Rutherford RB, Tilson MD, et al. Suggested standards for reporting on arterial aneurysms. Subcommittee on Reporting Standards for Arterial Aneurysms, Ad Hoc Committee on Reporting Standards, Society for Vascular Surgery and North American Chapter, International Society for Cardiovascular Surgery. *J Vasc Surg.* 1991;13:452–8.

4. Recommendations for Genetic Syndromes

- Class I**
- 1.** An echocardiogram is recommended at the time of diagnosis of Marfan syndrome to determine the aortic root and ascending aortic diameters and 6 months thereafter to determine the rate of enlargement of the aorta. (LOE: C)
 - 2.** Annual imaging is recommended for patients with Marfan syndrome if stability of the aortic diameter is documented. If the maximal aortic diameter is 4.5 cm or greater, or if the aortic diameter shows significant growth from baseline, more frequent imaging should be considered. (LOE: C)



- 3.** Patients with Loeys-Dietz syndrome or a confirmed genetic mutation known to predispose to aortic aneurysms and aortic dissections (*TGFBR1*, *TGFBR2*, *FBN1*, *ACTA2*, or *MYH11*) should undergo complete aortic imaging at initial diagnosis and 6 months thereafter to establish if enlargement is occurring. (LOE: C)
- 4.** Patients with Loeys-Dietz syndrome should have yearly magnetic resonance imaging from the cerebrovascular circulation to the pelvis. (LOE: B)
- 5.** Patients with Turner syndrome should undergo imaging of the heart and aorta for evidence of bicuspid aortic valve, coarctation of the aorta, or dilatation of the ascending thoracic aorta. If initial imaging is normal and there are no risk factors for aortic dissection, repeat imaging should be performed every 5 to 10 years or if otherwise clinically indicated. If abnormalities exist, annual imaging or follow-up imaging should be done. (LOE: C)

-
- Class IIa**
1. It is reasonable to consider surgical repair of the aorta in all adult patients with Loeys-Dietz syndrome or a confirmed *TGFBR1* or *TGFBR2* mutation and an aortic diameter of 4.2 cm or greater by transesophageal echocardiogram (internal diameter) or 4.4 to 4.6 cm or greater by computed tomographic imaging and/or magnetic resonance imaging (external diameter). (LOE: C)
 2. For women with Marfan syndrome contemplating pregnancy, it is reasonable to prophylactically replace the aortic root and ascending aorta if the diameter exceeds 4.0 cm. (LOE: C)
 3. If the maximal cross-sectional area in square centimeters of the ascending aorta or root divided by the patient's height in meters exceeds a ratio of 10, surgical repair is reasonable because shorter patients have dissection at a smaller size and 15% of patients with Marfan syndrome have dissection at a size smaller than 5.0 cm. (LOE: C)

-
- Class IIb**
1. In patients with Turner syndrome with additional risk factors, including bicuspid aortic valve, coarctation of the aorta, and/or hypertension, and in patients who attempt to become pregnant or who become pregnant, it may be reasonable to perform imaging of the heart and aorta to help determine the risk of aortic dissection. (LOE: C)

Table 4. Gene Defects Associated With Familial Thoracic Aortic Aneurysm and Dissection

Defective Gene Leading to Familial Thoracic Aortic Aneurysms and Dissection	Contribution to Familial Thoracic Aortic Aneurysms and Dissection	Associated Clinical Features	Comments on Aortic Disease
<i>TGFBR2</i> mutations	4%	Thin, translucent skin Arterial or aortic tortuosity Aneurysm of arteries	Multiple aortic dissections documented at aortic diameters <5.0 cm
<i>MYH11</i> mutations	1%	Patent ductus arteriosus	Patient with documented dissection at 4.5 cm
<i>ACTA2</i> mutations	14%	Livedo reticularis Iris flocculi Patent ductus arteriosus Bicuspid aortic valve	Two of 13 patients with documented dissections <5.0 cm

ACTA2 indicates actin, alpha 2, smooth muscle aorta; *MYH11*, smooth muscle specific beta-myosin heavy chain; and *TGFBR2*, transforming growth factor-beta receptor type II.

Table 5. Genetic Syndromes Associated With Thoracic Aortic Aneurysm

Genetic Syndrome	Common Clinical Features	Genetic Defect
Marfan syndrome	Skeletal features (see text) Ectopia lentis Dural ectasia	<i>FBN1</i> mutations*
Loeys-Dietz syndrome	Bifid uvula or cleft palate Arterial tortuosity Hypertelorism Skeletal features similar to MFS Craniosynostosis Aneurysms and dissections of other arteries	<i>TGFBR2</i> or <i>TGFBR1</i> mutations
Ehlers-Danlos syndrome, vascular form	Thin, translucent skin Gastrointestinal rupture Rupture of the gravid uterus Rupture of medium-sized to large arteries	<i>COL3A1</i> mutations
Turner syndrome	Short stature Primary amenorrhea Bicuspid aortic valve Aortic coarctation Webbed neck, low-set ears, low hairline, broad chest	45,X karyotype

* The defective gene at a second locus for MFS is *TGFBR2* but the clinical phenotype as MFS is debated.

AoD indicates aortic dissection; *COL3A1*, type III collagen; CT, computed tomographic imaging; *FBN1*, fibrillin 1; MFS, Marfan syndrome; MR, magnetic resonance imaging; TEE, transesophageal echocardiogram; *TGFBR1*, transforming growth factor-beta receptor type I; and *TGFBR2*, transforming growth factor-beta receptor type II.

and Dissection

Diagnostic Test	Comments on Aortic Disease
Ghent diagnostic criteria DNA for sequencing	Surgical repair when the aorta reaches 5.0 cm unless there is a family history of AoD at <5.0 cm, a rapidly expanding aneurysm or presence or significant aortic valve regurgitation
DNA for sequencing	Surgical repair recommended at an aortic diameter of ≥ 4.2 cm by TEE (internal diameter) or 4.4 to ≥ 4.6 cm by CT and/or MR (external diameter)
DNA for sequencing Dermal fibroblasts for analysis of type III collagen	Surgical repair is complicated by friable tissues Noninvasive imaging recommended
Blood (cells) for karyotype analysis	AoD risk is increased in patients with bicuspid aortic valve, aortic coarctation, hypertension, or pregnancy

5. Recommendations for Familial Thoracic Aortic Aneurysms and Dissections

- Class I**
1. Aortic imaging is recommended for first-degree relatives of patients with thoracic aortic aneurysm and/or dissection to identify those with asymptomatic disease. (LOE: B)
 2. If the mutant gene (*FBNI*, *TGFBR1*, *TGFBR2*, *COL3A1*, *ACTA2*, *MYH11*) associated with aortic aneurysm and/or dissection is identified in a patient, first-degree relatives should undergo counseling and testing. Then, only the relatives with the genetic mutation should undergo aortic imaging. (LOE: C)
-

- Class IIa**
1. If one or more first-degree relatives of a patient with known thoracic aortic aneurysm and/or dissection are found to have thoracic aortic dilatation, aneurysm, or dissection, then imaging of second-degree relatives is reasonable. (LOE: B)

2. Sequencing of the *ACTA2* gene is reasonable in patients with a family history of thoracic aortic aneurysms and/or dissections to determine if *ACTA2* mutations are responsible for the inherited predisposition. (LOE: B)

Class IIb

1. Sequencing of other genes known to cause familial thoracic aortic aneurysms and/or dissection (*TGFBR1*, *TGFBR2*, *MYH11*) may be considered in patients with a family history and clinical features associated with mutations in these genes. (LOE: B)

2. If one or more first-degree relatives of a patient with known thoracic aortic aneurysm and/or dissection are found to have thoracic aortic dilatation, aneurysm, or dissection, then referral to a geneticist may be considered. (LOE: C)

6. Recommendations for Bicuspid Aortic Valve and Associated Congenital Variants in Adults

- Class I**
- 1.** First-degree relatives of patients with a bicuspid aortic valve, premature onset of thoracic aortic disease with minimal risk factors, and/or a familial form of thoracic aortic aneurysm and dissection should be evaluated for the presence of a bicuspid aortic valve and asymptomatic thoracic aortic disease. (LOE: C)
 - 2.** All patients with a bicuspid aortic valve should have both the aortic root and ascending thoracic aorta evaluated for evidence of aortic dilatation. (LOE: B)



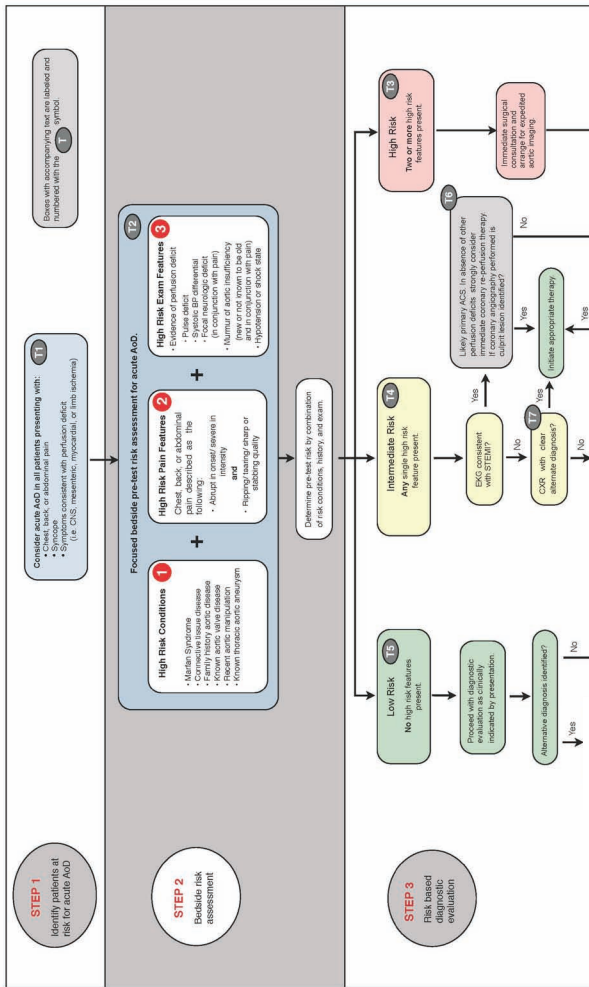
7. Recommendations for Estimation of Pretest Risk of Thoracic Aortic Dissection

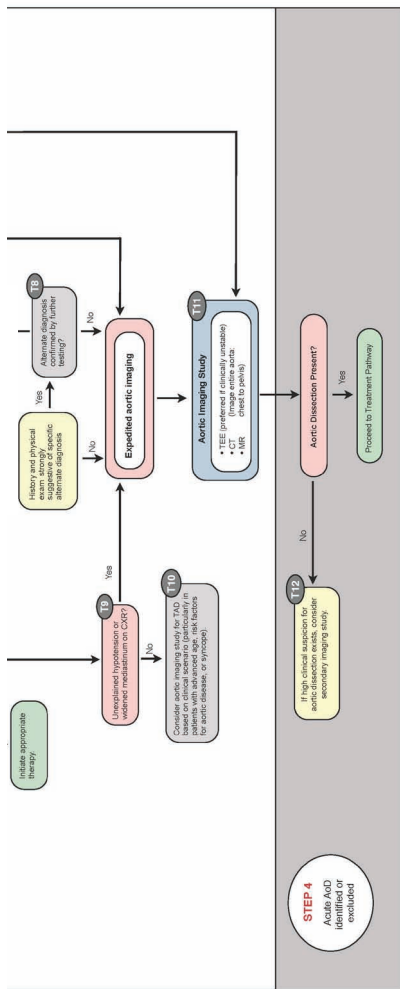
- Class I** **1.** Providers should routinely evaluate any patient presenting with complaints that may represent acute thoracic aortic dissection to establish a pretest risk of disease that can then be used to guide diagnostic decisions. This process should include specific questions about medical history, family history, and pain features as well as a focused examination to identify findings that are associated with aortic dissection, including:
- a. High-risk conditions and historical features (*LOE: B*):
 - Marfan syndrome, Loeys-Dietz syndrome, vascular Ehlers-Danlos syndrome, Turner syndrome, or other connective tissue disease.
 - Patients with mutations in genes known to predispose to thoracic aortic aneurysms and dissection, such as *FBNI*, *TGFBR1*, *TGFBR2*, *ACTA2*, and *MYH11*.
 - Family history of aortic dissection or thoracic aortic aneurysm.
 - Known aortic valve disease.
 - Recent aortic manipulation (surgical or catheter-based).
 - Known thoracic aortic aneurysm.

- b. High-risk chest, back or abdominal pain features (*LOE: B*):
- Pain that is abrupt or instantaneous in onset.
 - Pain that is severe in intensity.
 - Pain that has a ripping, tearing, stabbing, or sharp quality.
- c. High-risk examination features (*LOE: B*):
- Pulse deficit.
 - Systolic blood pressure limb differential greater than 20 mm Hg.
 - Focal neurologic deficit.
 - Murmur of aortic regurgitation (new).
- 2.** Patients presenting with sudden onset of severe chest, back and/or abdominal pain, particularly those less than 40 years of age, should be questioned about a history and examined for physical features of Marfan syndrome, Loeys-Dietz syndrome, vascular Ehlers-Danlos syndrome, Turner syndrome, or other connective tissue disorder associated with thoracic aortic disease. (*LOE: B*)
- 3.** Patients presenting with sudden onset of severe chest, back, and/or abdominal pain should be questioned about a history of aortic pathology in immediate family members as there is a strong familial component to acute thoracic aortic disease. (*LOE: B*)

4. Patients presenting with sudden onset of severe chest, back and/or abdominal pain should be questioned about recent aortic manipulation (surgical or catheter-based) or a known history of aortic valvular disease, as these factors predispose to acute aortic dissection. *(LOE: C)*
5. In patients with suspected or confirmed aortic dissection who have experienced a syncopal episode, a focused examination should be performed to identify associated neurologic injury or the presence of pericardial tamponade. *(LOE: C)*
6. All patients presenting with acute neurologic complaints should be questioned about the presence of chest, back, and/or abdominal pain and checked for peripheral pulse deficits as patients with dissection-related neurologic pathology are less likely to report thoracic pain than the typical aortic dissection patient. *(LOE: C)*

Figure 2. AoD Evaluation Pathway.





ACS indicates acute coronary syndrome; AoD, aortic dissection; BP, blood pressure; CNS, central nervous system; CT, computed tomographic imaging; CXR, chest x-ray; EKG, electrocardiogram; MR, magnetic resonance imaging; STEMI, ST-elevated myocardial infarction; TAD, thoracic aortic disease; and TEE, transesophageal echocardiogram.

Table 6. Risk Factors for Development of Thoracic Aortic Dissection

Conditions Associated With Increased Aortic Wall Stress

Hypertension, particularly if uncontrolled
Pheochromocytoma
Cocaine or other stimulant use
Weight lifting or other Valsalva maneuver
Trauma
Deceleration or torsional injury (eg, motor vehicle crash, fall)
Coarctation of the aorta

Conditions Associated With Aortic Media Abnormalities

Genetic

Marfan syndrome
Ehlers-Danlos syndrome, vascular form
Bicuspid aortic valve (including prior aortic valve replacement)
Turner syndrome
Loeys-Dietz syndrome
Familial thoracic aortic aneurysm and dissection syndrome

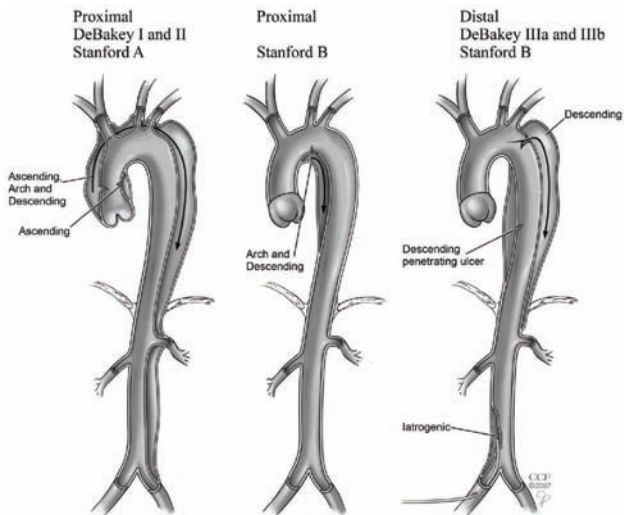
Inflammatory vasculitides

Takayasu arteritis
Giant cell arteritis
Behçet arteritis

Other

Pregnancy
Polycystic kidney disease
Chronic corticosteroid or immunosuppression agent administration
Infections involving the aortic wall either from bacteremia or extension of adjacent infection

Figure 3. Aortic Dissection Classification: DeBakey and Stanford Classifications.



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8. Initial Evaluation and Management of Acute Thoracic Aortic Disease

8.1. Recommendations for Screening Tests

- Class I**
1. An electrocardiogram should be obtained on all patients who present with symptoms that may represent acute thoracic aortic dissection.
 - a. Given the relative infrequency of dissection-related coronary artery occlusion, the presence of ST-segment elevation suggestive of myocardial infarction should be treated as a primary cardiac event without delay for definitive aortic imaging unless the patient is at high risk for aortic dissection. (LOE: B)
 2. The role of chest x-ray in the evaluation of possible thoracic aortic disease should be directed by the patient's pretest risk of disease as follows.
 - a. Intermediate risk: Chest x-ray should be performed on all intermediate-risk patients, as it may establish a clear alternate diagnosis that will obviate the need for definitive aortic imaging. (LOE: C)
 - b. Low risk: Chest x-ray should be performed on all low-risk patients, as it may either establish an alternative diagnosis or demonstrate findings that are suggestive of thoracic aortic disease, indicating the need for urgent definitive aortic imaging. (LOE: C)

3. Urgent and definitive imaging of the aorta using transesophageal echocardiogram, computed tomographic imaging, or magnetic resonance imaging is recommended to identify or exclude thoracic aortic dissection in patients at high risk for the disease by initial screening. (LOE: B)

Class III **1.** A negative chest x-ray should not delay definitive aortic imaging in patients determined to be high risk for aortic dissection by initial screening. (LOE: C)

8.2. Recommendations for Diagnostic Imaging Studies

Class I **1.** Selection of a specific imaging modality to identify or exclude aortic dissection should be based on patient variables and institutional capabilities, including immediate availability. (LOE: C)

2. If a high clinical suspicion exists for acute aortic dissection but initial aortic imaging is negative, a second imaging study should be obtained. (LOE: C)

8.3. Recommendations for Initial Management (see Figure 4)

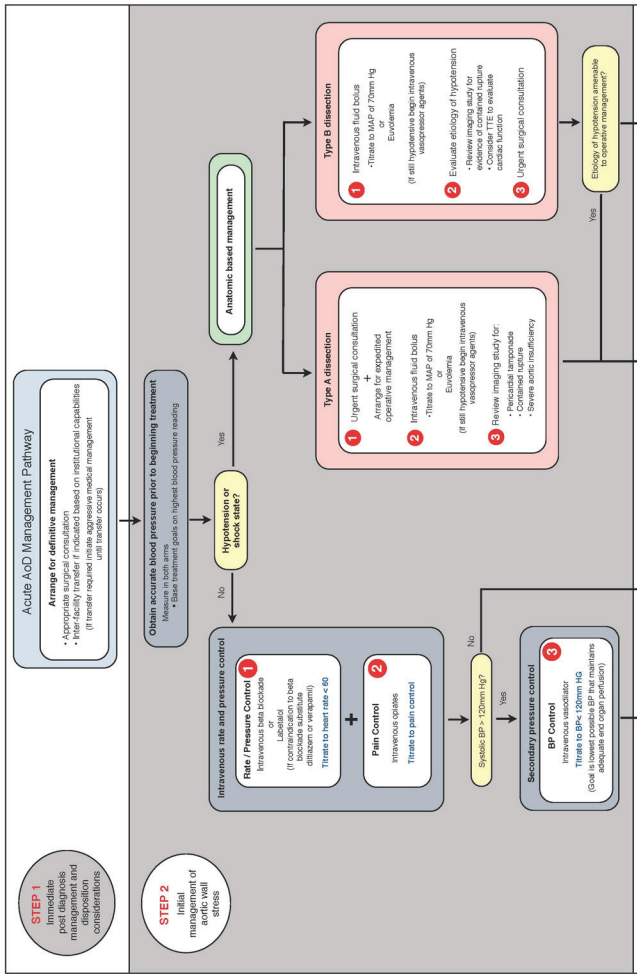
- Class I**
- 1.** Initial management of thoracic aortic dissection should be directed at decreasing aortic wall stress by controlling heart rate and blood pressure as follows:
 - a. In the absence of contraindications, intravenous beta blockade should be initiated and titrated to a target heart rate of 60 beats per minute or less. (LOE: C)
 - b. In patients with clear contraindications to beta blockade, nondihydropyridine calcium channel-blocking agents should be utilized as an alternative for rate control. (LOE: C)
 - c. If systolic blood pressures remain greater than 120 mm Hg after adequate heart rate control has been obtained, then angiotensin-converting enzyme inhibitors and/or other vasodilators should be administered intravenously to further reduce blood pressure that maintains adequate end-organ perfusion. (LOE: C)
 - d. Beta blockers should be used cautiously in the setting of acute aortic regurgitation because they will block the compensatory tachycardia. (LOE: C)

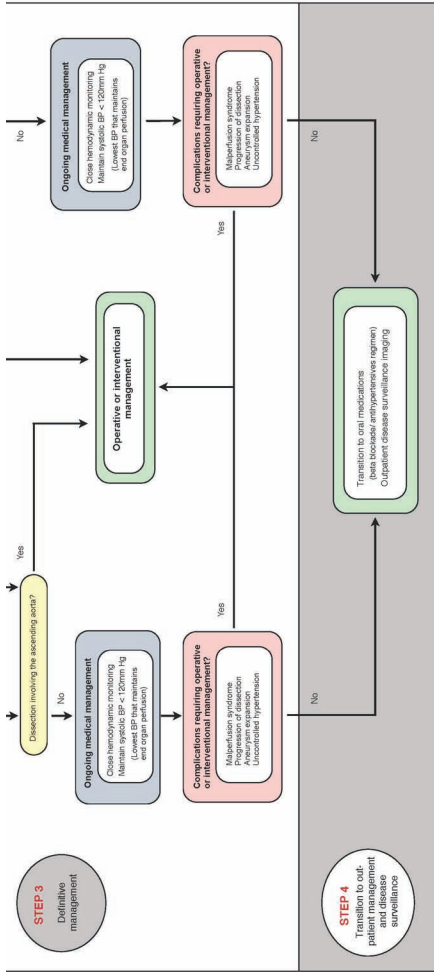
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- Class III** **1.** Vasodilator therapy should not be initiated prior to rate control so as to avoid associated reflex tachycardia that may increase aortic wall stress, leading to propagation or expansion of a thoracic aortic dissection. (LOE: C)

8.4. Recommendations for Definitive Management (see Figures 3 and 5)

- Class I**
- 1.** Urgent surgical consultation should be obtained for all patients diagnosed with thoracic aortic dissection regardless of the anatomic location (ascending versus descending) as soon as the diagnosis is made or highly suspected. (LOE: C)
 - 2.** Acute thoracic aortic dissection involving the ascending aorta should be urgently evaluated for emergent surgical repair because of the high risk of associated life-threatening complications such as rupture. (LOE: B)
 - 3.** Acute thoracic aortic dissection involving the descending aorta should be managed medically unless life-threatening complications develop (ie, malperfusion syndrome, progression of dissection, enlarging aneurysm, inability to control blood pressure or symptoms). (LOE: B)

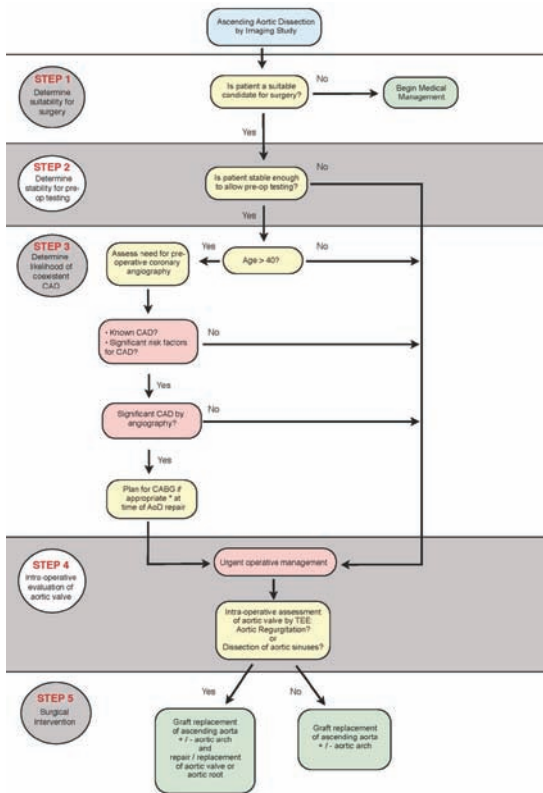
Figure 4. Acute AoD Management Pathway.





AoD indicates aortic dissection; BP, blood pressure; MAP, mean arterial pressure; and TTE, transthoracic echocardiogram.

Figure 5. Acute Surgical Management Pathway for AoD.



*Addition of 'if appropriate' based on Patel MR, Dehmer GJ, Hirshfeld JW, et al. ACCF/SCAI/STS/AATS/AHA/ASNC 2009 Appropriateness Criteria for Coronary Revascularization. J Am Coll Cardiol. 2009;53:530–53.

AoD indicates aortic dissection; CABG, coronary artery bypass graft surgery; CAD, coronary artery disease; TAD, thoracic aortic disease; and TEE, transesophageal echocardiogram.

9. Recommendation for Surgical Intervention for Acute Thoracic Aortic Dissection

- Class I** **1.** For patients with ascending thoracic aortic dissection, all aneurysmal aorta and the proximal extent of the dissection should be resected. A partially dissected aortic root may be repaired with aortic valve re-suspension. Extensive dissection of the aortic root should be treated with aortic root replacement with a composite graft or with a valve sparing root replacement. If a DeBakey Type II dissection is present, the entire dissected aorta should be replaced. (*LOE: C*)

10. Recommendation for Intramural Hematoma Without Intimal Defect

- Class IIa** **1.** It is reasonable to treat intramural hematoma similar to aortic dissection in the corresponding segment of the aorta. (*LOE: C*)



11. Recommendation for History and Physical Examination for Thoracic Aortic Disease

- Class I** **1.** For patients presenting with a history of acute cardiac and noncardiac symptoms associated with a significant likelihood of thoracic aortic disease, the clinician should perform a focused physical examination, including a careful and complete search for arterial perfusion differentials in both upper and lower extremities, evidence of visceral ischemia, focal neurologic deficits, a murmur of aortic regurgitation, bruits, and findings compatible with possible cardiac tamponade. (*LOE: C*)

12. Recommendation for Medical Treatment of Patients With Thoracic Aortic Diseases

- Class I** **1.** Stringent control of hypertension, lipid profile optimization, smoking cessation, and other atherosclerosis risk-reduction measures should be instituted for patients with small aneurysms not requiring surgery, as well as for patients who are not considered surgical or stent graft candidates. (*LOE: C*)

12.1. Recommendations for Blood Pressure Control

- Class I** **1.** Antihypertensive therapy should be administered to hypertensive patients with thoracic aortic diseases to achieve a goal of less than 140/90 mm Hg (patients without diabetes) or less than 130/80 mm Hg (patients with diabetes or chronic renal disease) to reduce the risk of stroke, myocardial infarction, heart failure, and cardiovascular death. (*LOE: B*)
- 2.** Beta adrenergic–blocking drugs should be administered to all patients with Marfan syndrome and aortic aneurysm to reduce the rate of aortic dilatation unless contraindicated. (*LOE: B*)

-
- Class IIa**
- 1.** For patients with thoracic aortic aneurysm, it is reasonable to reduce blood pressure with beta blockers and angiotensin-converting enzyme inhibitors or angiotensin receptor blockers to the lowest point patients can tolerate without adverse effects. *(LOE: B)*
 - 2.** An angiotensin receptor blocker (losartan) is reasonable for patients with Marfan syndrome, to reduce the rate of aortic dilatation unless contraindicated. *(LOE: B)*



13. Recommendations for Asymptomatic Patients With Ascending Aortic Aneurysm (see Figures 6 and 7)

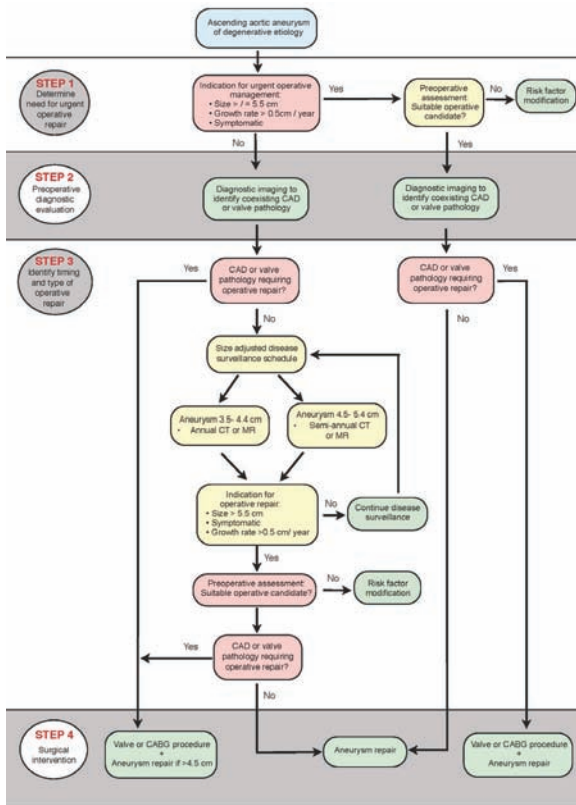
- Class I**
1. Asymptomatic patients with degenerative thoracic aneurysm, chronic aortic dissection, intramural hematoma, penetrating atherosclerotic ulcer, mycotic aneurysm, or pseudoaneurysm, who are otherwise suitable candidates and for whom the ascending aorta or aortic sinus diameter is 5.5 cm or greater should be evaluated for surgical repair. (*LOE: C*)
 2. Patients with Marfan syndrome or other genetically mediated disorders (vascular Ehlers-Danlos syndrome, Turner syndrome, bicuspid aortic valve, or familial thoracic aortic aneurysm and dissection) should undergo elective operation at smaller diameters (4.0 to 5.0 cm depending on the condition; see Section 4) to avoid acute dissection or rupture. (*LOE: C*)

3. Patients with a growth rate of more than 0.5 cm/y in an aorta that is less than 5.5 cm in diameter should be considered for operation. (LOE: C)
4. Patients undergoing aortic valve repair or replacement and who have an ascending aorta or aortic root of greater than 4.5 cm should be considered for concomitant repair of the aortic root or replacement of the ascending aorta. (LOE: C)

Class IIa

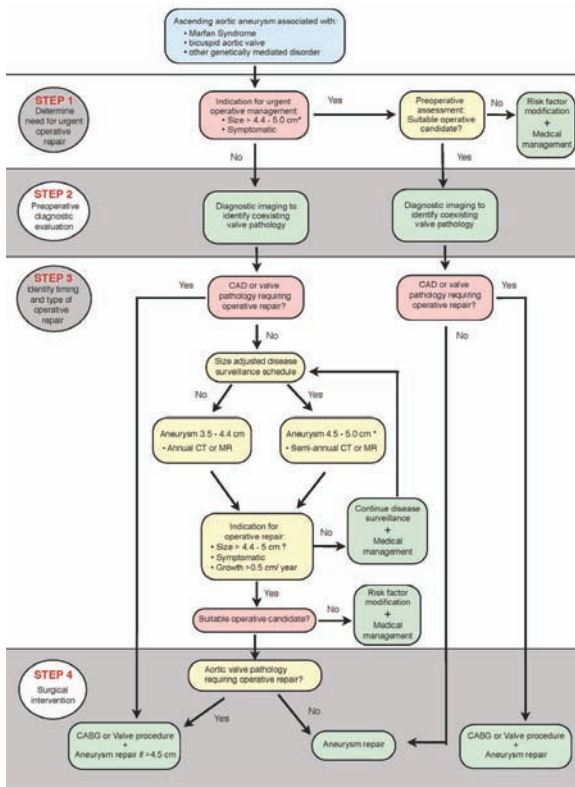
1. Elective aortic replacement is reasonable for patients with Marfan syndrome, other genetic diseases, or bicuspid aortic valves, when the ratio of maximal ascending or aortic root area (πr^2) in cm^2 divided by the patient's height in meters exceeds 10. (LOE: C)
2. It is reasonable for patients with Loeys-Dietz syndrome or a confirmed *TGFBR1* or *TGFBR2* mutation to undergo aortic repair when the aortic diameter reaches 4.2 cm or greater by transesophageal echocardiogram (internal diameter) or 4.4 to 4.6 cm or greater by computed tomographic imaging and/or magnetic resonance imaging (external diameter). (LOE: C)

Figure 6. Ascending Aortic Aneurysm of Degenerative Etiology.



CABG indicates coronary artery bypass graft surgery; CAD, coronary artery disease; CT, computed tomographic imaging; and MR, magnetic resonance imaging.

Figure 7. Ascending Aortic Aneurysms Associated With Genetic Disorder.



*Depends on specific genetic condition. †See Recommendations for Asymptomatic Patients With Ascending Aortic Aneurysm (Section 13), and Recommendations for Bicuspid Aortic Valve and Associated Congenital Variants in Adults (Section 6). CABG indicates coronary artery bypass graft surgery; CAD, coronary artery disease; CT, computed tomographic imaging; and MR, magnetic resonance imaging.

14. Recommendation for Symptomatic Patients With Thoracic Aortic Aneurysm

- Class I** **1.** Patients with symptoms suggestive of expansion of a thoracic aneurysm should be evaluated for prompt surgical intervention unless life expectancy from comorbid conditions is limited or quality of life is substantially impaired. (LOE: C)



15. Recommendations for Open Surgery for Ascending Aortic Aneurysm

- Class I**
1. Separate valve and ascending aortic replacement are recommended in patients without significant aortic root dilatation, in elderly patients, or in young patients with minimal dilatation who have aortic valve disease. (*LOE: C*)
 2. Patients with Marfan, Loeys-Dietz, and Ehlers-Danlos syndromes and other patients with dilatation of the aortic root and sinuses of Valsalva should undergo excision of the sinuses in combination with a modified David reimplantation operation if technically feasible or, if not, root replacement with valved graft conduit. (*LOE: B*)

16. Recommendations for Aortic Arch Aneurysms

- Class IIa**
1. For thoracic aortic aneurysms also involving the proximal aortic arch, partial arch replacement together with ascending aorta repair using right subclavian/axillary artery inflow and hypothermic circulatory arrest is reasonable. (*LOE: B*)
 2. Replacement of the entire aortic arch is reasonable for acute dissection when the arch is aneurysmal or there is extensive aortic arch destruction and leakage. (*LOE: B*)
 3. Replacement of the entire aortic arch is reasonable for aneurysms of the entire arch, for chronic dissection when the arch is enlarged, and for distal arch aneurysms that also involve the proximal descending thoracic aorta, usually with the elephant trunk procedure. (*LOE: B*)

4. For patients with low operative risk in whom an isolated degenerative or atherosclerotic aneurysm of the aortic arch is present, operative treatment is reasonable for asymptomatic patients when the diameter of the arch exceeds 5.5 cm. (LOE: B)
5. For patients with isolated aortic arch aneurysms less than 4.0 cm in diameter, it is reasonable to reimage using computed tomographic imaging or magnetic resonance imaging, at 12-month intervals, to detect enlargement of the aneurysm. (LOE: C)
6. For patients with isolated aortic arch aneurysms 4.0 cm or greater in diameter, it is reasonable to reimage using computed tomographic imaging or magnetic resonance imaging, at 6-month intervals, to detect enlargement of the aneurysm. (LOE: C)

17. Recommendations for Descending Thoracic Aorta and Thoracoabdominal Aortic Aneurysms

- Class I**
- 1.** For patients with chronic dissection, particularly if associated with a connective tissue disorder, but without significant comorbid disease, and a descending thoracic aortic diameter exceeding 5.5 cm, open repair is recommended. (*LOE: B*)
 - 2.** For patients with degenerative or traumatic aneurysms of the descending thoracic aorta exceeding 5.5 cm, saccular aneurysms, or postoperative pseudoaneurysms, endovascular stent grafting should be strongly considered when feasible. (*LOE: B*)
 - 3.** For patients with thoracoabdominal aneurysms, in whom endovascular stent graft options are limited and surgical morbidity is elevated, elective surgery is recommended if the aortic diameter exceeds 6.0 cm, or less if a connective tissue disorder such as Marfan or Loeys-Dietz syndrome is present. (*LOE: C*)
 - 4.** For patients with thoracoabdominal aneurysms and with end-organ ischemia or significant stenosis from atherosclerotic visceral artery disease, an additional revascularization procedure is recommended. (*LOE: B*)

Table 7. Summary of Society of Thoracic Surgeons Recommendations for Thoracic Stent Graft Insertion

Entity/Subgroup	Classification	Level of Evidence
Penetrating ulcer/intramural hematoma		
Asymptomatic	III	C
Symptomatic	IIa	C
Acute traumatic	I	B
Chronic traumatic	IIa	C
Acute Type B dissection		
Ischemia	I	A
No ischemia	IIb	C
Subacute dissection	IIb	B
Chronic dissection	IIb	B
Degenerative descending		
>5.5 cm, comorbidity	IIa	B
>5.5 cm, no comorbidity	IIb	C
<5.5 cm	III	C
Arch		
Reasonable open risk	III	A
Severe comorbidity	IIb	C
Thoracoabdominal/Severe comorbidity	IIb	C

Reprinted from Svensson LG, Kouchoukos NT, Miller DC, et al. Expert consensus document on the treatment of descending thoracic aortic disease using endovascular stent-grafts. *Ann Thorac Surg.* 2008;85:S1–41.

18. Recommendations for Counseling and Management of Chronic Aortic Diseases in Pregnancy

- Class I**
- 1.** Women with Marfan syndrome and aortic dilatation, as well as patients without Marfan syndrome who have known aortic disease, should be counseled about the risk of aortic dissection as well as the heritable nature of the disease prior to pregnancy. (LOE: C)
 - 2.** For pregnant women with known thoracic aortic dilatation or a familial or genetic predisposition for aortic dissection, strict blood pressure control, specifically to prevent Stage II hypertension, is recommended. (LOE: C)
 - 3.** For all pregnant women with known aortic root or ascending aortic dilatation, monthly or bimonthly echocardiographic measurements of the ascending aortic dimensions are recommended to detect aortic expansion until birth. (LOE: C)

4. For imaging of pregnant women with aortic arch, descending, or abdominal aortic dilatation, magnetic resonance imaging (without gadolinium) is recommended over computed tomographic imaging to avoid exposing both the mother and fetus to ionizing radiation. Transesophageal echocardiogram is an option for imaging of the thoracic aorta. (LOE: C)

5. Pregnant women with aortic aneurysms should be delivered where cardiothoracic surgery is available. (LOE: C)

Class IIa 1. Fetal delivery via cesarean section is reasonable for patients with significant aortic enlargement, dissection, or severe aortic valve regurgitation. (LOE: C)

Class IIb 1. If progressive aortic dilatation and/or advancing aortic valve regurgitation are documented, prophylactic surgery may be considered. (LOE: C)

19. Recommendations for Aortic Arch and Thoracic Aortic Atheroma and Atheroembolic Disease

Class IIa 1. Treatment with a statin is a reasonable option for patients with aortic arch atheroma to reduce the risk of stroke. (LOE: C)

Class IIb 1. Oral anticoagulation therapy with warfarin (INR 2.0 to 3.0) or antiplatelet therapy may be considered in stroke patients with aortic arch atheroma 4.0 mm or greater to prevent recurrent stroke. (LOE: C)



20. Periprocedural and Perioperative Management

20.1. Recommendations for Brain Protection During Ascending Aortic and Transverse Aortic Arch Surgery

Class I **1.** A brain protection strategy to prevent stroke and preserve cognitive function should be a key element of the surgical, anesthetic, and perfusion techniques used to accomplish repairs of the ascending aorta and transverse aortic arch. (*LOE: B*)

Class IIa **1.** Deep hypothermic circulatory arrest, selective antegrade brain perfusion, and retrograde brain perfusion are techniques that alone or in combination are reasonable to minimize brain injury during surgical repairs of the ascending aorta and transverse aortic arch. Institutional experience is an important factor in selecting these techniques. (*LOE: B*)

Class III **1.** Perioperative brain hyperthermia is not recommended in repairs of the ascending aortic and transverse aortic arch as it is probably injurious to the brain. (LOE: B)

20.2. Recommendations for Spinal Cord Protection During Descending Aortic Open Surgical and Endovascular Repairs

Class I **1.** Cerebrospinal fluid drainage is recommended as a spinal cord protective strategy in open and endovascular thoracic aortic repair for patients at high risk of spinal cord ischemic injury. (LOE: B)

Class IIa **1.** Spinal cord perfusion pressure optimization using techniques, such as proximal aortic pressure maintenance and distal aortic perfusion, is reasonable as an integral part of the surgical, anesthetic, and perfusion strategy in open and endovascular thoracic aortic repair patients at high risk of spinal cord ischemic injury. Institutional experience is an important factor in selecting these techniques. (LOE: B)

2. Moderate systemic hypothermia is reasonable for protection of the spinal cord during open repairs of the descending thoracic aorta. (LOE: B)

Class IIb

- 1.** Adjunctive techniques to increase the tolerance of the spinal cord to impaired perfusion may be considered during open and endovascular thoracic aortic repair for patients at high risk of spinal cord injury. These include distal perfusion, epidural irrigation with hypothermic solutions, high-dose systemic glucocorticoids, osmotic diuresis with mannitol, intrathecal papaverine, and cellular metabolic suppression with anesthetic agents. (*LOE: B*)
- 2.** Neurophysiological monitoring of the spinal cord (somatosensory evoked potentials or motor evoked potentials) may be considered as a strategy to detect spinal cord ischemia and to guide reimplantation of intercostal arteries and/or hemodynamic optimization to prevent or treat spinal cord ischemia. (*LOE: B*)

21. Recommendations for Surveillance of Thoracic Aortic Disease or Previously Repaired Patients

Class IIa

1. Computed tomographic imaging or magnetic resonance imaging of the thoracic aorta is reasonable after a Type A or B aortic dissection or after prophylactic repair of the aortic root/ascending aorta. (LOE: C)
2. Computed tomographic imaging or magnetic resonance imaging of the aorta is reasonable at 1, 3, 6, and 12 months postdissection and, if stable, annually thereafter so that any threatening enlargement can be detected in a timely fashion. (LOE: C)
3. When following patients with imaging, utilization of the same modality at the same institution is reasonable, so that similar images of matching anatomic segments can be compared side by side. (LOE: C)
4. If a thoracic aortic aneurysm is only moderate in size and remains relatively stable over time, magnetic resonance imaging instead of computed tomographic imaging is reasonable to minimize the patient's radiation exposure. (LOE: C)
5. Surveillance imaging similar to classic aortic dissection is reasonable in patients with intramural hematoma. (LOE: C)

Table 8. Suggested Follow-up of Aortic Pathologies After Repair or Treatment

Pathology	Interval	Study
Acute dissection	Before discharge, 1 mo, 6 mo, yearly	CT or MR, chest plus abdomen TTE
Chronic dissection	Before discharge, 1 y, 2 to 3 y	CT or MR, chest plus abdomen TTE
Aortic root repair	Before discharge, yearly	TTE
AVR plus ascending	Before discharge, yearly	TTE
Aortic arch	Before discharge, 1 y, 2 to 3 y	CT or MR, chest plus abdomen
Thoracic aortic stent	Before discharge, 1 mo, 2 mo, 6 mo, yearly Or 30 days*	CXR, CT, chest plus abdomen
Acute IMH/PAU	Before discharge, 1 mo, 3 mo, 6 mo, yearly	CT or MR, chest plus abdomen

*US Food and Drug Administration stent graft studies usually required before discharge or at 30-day CT scan to detect endovascular leaks. If there is concern about a leak, a predischarge study is recommended; however, the risk of renal injury should be borne in mind. All patients should be receiving beta blockers after surgery or medically managed aortic dissection, if tolerated. Adapted from Erbel R, Alfonso F, Boileau C, et al. Diagnosis and management of aortic dissection. *Eur Heart J.* 2001;22:1642–81.

AVR indicates aortic valve replacement; CT, computed tomographic imaging; CXR, chest X-ray; IMH, intramural hematoma; MR, magnetic resonance imaging; PAU, penetrating atherosclerotic ulcer; and TTE, transthoracic echocardiography.

22. Recommendation for Employment and Lifestyle in Patients With Thoracic Aortic Disease

- Class IIa** 1. For patients with a current thoracic aortic aneurysm or dissection, or previously repaired aortic dissection, employment and lifestyle restrictions are reasonable, including the avoidance of strenuous lifting, pushing or straining that would require a Valsalva maneuver. (LOE: C)



