

2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients with Thoracic Aortic Disease: Executive Summary

A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines, American Association for Thoracic Surgery, American College of Radiology, American Stroke Association, Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, Society of Interventional Radiology, Society of Thoracic Surgeons, and Society for Vascular Medicine

Endorsed by the North American Society for Cardiovascular Imaging

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PREAMBLE

It is essential that the medical profession play a central role in critically evaluating the evidence related to drugs, devices, and procedures for the detection, management, or prevention of disease. Properly applied, rigorous, expert analysis of the available data documenting absolute and relative benefits and risks of these therapies and procedures can improve outcomes and reduce costs of care by focusing resources on the most effective strategies. One important use of such data is the production of clinical practice guidelines which, in turn, can provide a foundation for a variety of other applications such as performance measures, appropriate use criteria, clinical decision support tools, and quality improvement tools.

The American College of Cardiology Foundation (ACCF) and the American Heart Association (AHA) have jointly engaged in the production of guidelines in the area of cardiovascular disease since 1980. The ACCF/AHA Task Force on Practice Guidelines is charged with developing, updating, and revising practice guidelines for cardiovascular diseases and procedures, and the Task Force directs and oversees this effort. Writing committees are charged with assessing the evidence as an independent group of authors to develop, update, or revise recommendations for clinical practice.

Experts in the subject under consideration have been selected from both organizations to examine subject-specific data and

write guidelines in partnership with representatives from other medical practitioner and specialty groups. Writing committees are specifically charged to perform a formal literature review, weigh the strength of evidence for or against particular treatments or procedures, and include estimates of expected health outcomes where data exist. Patient-specific modifiers, comorbidities, and issues of patient preference that may influence the choice of tests or therapies are considered. When available, information from studies on cost is considered, but data on efficacy and clinical outcomes constitute the primary basis for recommendations in these guidelines.

The ACCF/AHA Task Force on Practice Guidelines makes every effort to avoid actual, potential, or perceived conflicts of interest that may arise as a result of industry relationships or personal interests among the writing committee. Specifically, all members of the writing committee, as well as peer reviewers of the document, are asked to disclose all current relationships and those 24 months prior to initiation of the writing effort that may be perceived as relevant. All guideline recommendations require a confidential vote by the writing committee and must be approved by a consensus of the members voting. Members who were recused from voting are noted on the title page of this document. Members must recuse themselves from voting on any recommendation where their relationships with industry (RWI) and other entities apply. If a writing committee member develops a new relationship with industry during his/her tenure, he/she is required to notify guideline staff in writing. These statements are reviewed by the Task Force on Practice Guidelines and all members during each conference call and/or meeting of the writing committee, updated as changes occur, and ultimately published as an appendix to the document. For detailed information regarding guideline policies and procedures, please refer to the methodology manual for ACCF/AHA Guideline Writing Committees.¹ RWI and other entities pertinent to this guideline for authors and peer reviewers are disclosed in Appendixes 1 and 2, respectively. Disclosure information for the ACCF/AHA Task Force on Practice Guidelines is also available online at <http://www.acc.org/about/overview/ClinicalDocumentsTaskForces.cfm>.

These practice guidelines are intended to assist healthcare providers in clinical decision making by describing a range of generally acceptable approaches for diagnosis, management, and prevention of specific diseases or conditions. Clinicians should consider the quality and availability of expertise in the area where care is provided. These guidelines attempt to define practices that meet the needs of most patients in most circumstances. The recommendations reflect a consensus after a thorough review of the available current scientific evidence and are intended to improve patient care. The Task Force recognizes that situations arise where additional data are needed to better inform patient care; these areas will be identified within each respective guideline when appropriate.

Patient adherence to prescribed and agreed upon medical regimens and lifestyles is an important aspect of treatment. Prescribed courses of treatment in accordance with these recommendations are effective only if they are followed. Because lack of patient understanding and adherence may adversely affect outcomes, physicians and other healthcare providers should make every effort to engage the patient's

active participation in prescribed medical regimens and lifestyles.

If these guidelines are used as the basis for regulatory or payer decisions, the goal should be improvement in quality of care and aligned with the patient's best interest. The ultimate judgment regarding care of a particular patient must be made by the healthcare provider and the patient in light of all of the circumstances presented by that patient. Consequently, there are circumstances in which deviations from these guidelines are appropriate.

The guidelines will be reviewed annually by the ACCF/AHA Task Force on Practice Guidelines and considered current unless they are updated, revised, or withdrawn from distribution. The full-text guidelines are e-published in the April 6, 2010, issues of the *Journal of the American College of Cardiology* and *Circulation*.^{1a}

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1. INTRODUCTION

1.1. Methodology and Evidence Review

The writing committee conducted a comprehensive search of the medical and scientific literature through the use of PubMed/MEDLINE. Searches were limited to publications written in the English language. Compiled reports were reviewed and additional articles were provided by committee members. Specifically targeted searches were conducted on the following subtopics: acute aortic dissection, ankylosing spondylitis, aortic dissection and litigation, aortic neoplasm, aortic tumors, Behçet disease, bicuspid aortic valve, calcified aorta, chronic dissection, coarctation of the aorta, D-dimer, dissecting aneurysm, Ehlers-Danlos syndrome, endovascular and aortic aneurysms, medial degeneration, porcelain aorta, giant cell arteritis, imaging and thoracic aortic disease, inflammatory disease, intramural hematoma, Loeys-Dietz syndrome, Marfan syndrome, Noonan syndrome, penetrating aortic ulcer, polycystic kidney disease, thoracic and aortic aneurysms, thoracic aortic disease and patient care, thoracic aortic disease and surgery, thoracic aorta and Kawasaki disease, Takayasu arteritis, thoracoabdominal and aorta or aortic disease, and Turner syndrome. More than 850 references were reviewed, with 830 used as the primary evidence base for the final guideline. The ACCF/AHA Task Force on Practice Guidelines methodology processes were followed to write the text and recommendations. In general, published manuscripts appearing in journals listed in Index Medicus were used as the evidence base. Published abstracts were used only for emerging information but were not used in the formulation of recommendations.

The committee reviewed and ranked evidence supporting current recommendations with the weight of evidence ranked as Level A if the data were derived from multiple randomized clinical trials or meta-analyses. The committee ranked available evidence as Level B when data were derived from a single randomized trial or nonrandomized studies. Evidence was ranked as Level C when the primary source of the

Table 1. Applying Classification of Recommendations and Level of Evidence

		SIZE OF TREATMENT EFFECT →			
ESTIMATE OF CERTAINTY (PRECISION) OF TREATMENT EFFECT		CLASS I <i>Benefit >>> Risk</i> Procedure/Treatment SHOULD be performed/administered	CLASS IIa <i>Benefit >> Risk</i> Additional studies with <i>focused objectives needed</i> IT IS REASONABLE to perform procedure/administer treatment	CLASS IIb <i>Benefit ≥ Risk</i> Additional studies with <i>broad objectives needed</i> ; additional registry data would be helpful Procedure/Treatment MAY BE CONSIDERED	CLASS III <i>Risk ≥ Benefit</i> Procedure/Treatment should NOT be performed/administered SINCE IT IS NOT HELPFUL AND MAY BE HARMFUL
	LEVEL A Multiple populations evaluated* Data derived from multiple 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 	<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
	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 ■ 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 	<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 ■ 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 	<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
	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	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.

recommendation was consensus opinion, case studies, or standard of care. In the narrative portions of these guidelines, evidence is generally presented in chronologic order of development. Studies are identified as observational, retrospective, prospective, or randomized. For certain conditions for which inadequate data are available, recommendations are based on expert consensus and clinical experience and are ranked as Level C. An analogous example is the use of penicillin for pneumococcal pneumonia, where there are no randomized trials and treatment is based on clinical experience. When recommendations at Level C are supported by historical clinical data, appropriate references (including clinical reviews) are cited if available. For issues where sparse data are available, a survey of current practice among the clinicians on the writing committee formed the basis for Level C recommendations and no references are cited. The schema for classification of recommendations and level of evidence is

summarized in Table 1, which also illustrates how the grading system provides an estimate of the size of the treatment effect and an estimate of the certainty of the treatment effect.

To provide clinicians with a comprehensive set of data, whenever possible, the exact event rates in various treatment arms of clinical trials are presented to permit calculation of the absolute risk difference (ARD), number needed to harm (NNH); the relative treatment effects are described either as odds ratio (OR), relative risk (RR), or hazard ratio (HR) depending on the format in the original publication. Along with all other point statistics, confidence intervals (CIs) for those statistics are added when available.

The writing committee recognized that the evidence base for this guideline is less robust in terms of randomized clinical trials than prior ACCF/AHA guidelines, particularly those focused on coronary artery disease (CAD) and

heart failure. As the reader will discern, much of the evidence base for this topic consists of cohort studies and retrospective reviews, which largely emanate from centers with a specialized interest in specific types of thoracic aortic disease. The writing committee attempted to focus on providing the practitioner with recommendations for evaluation and treatment wherever possible and where controversy exists, identified as such in the text.

The writing committee acknowledges the expertise of the highly experienced and effective practice guidelines staff of the ACCF and AHA. The writing committee chair also acknowledges the commitment and dedication of the diverse writing committee members who were able to put aside issues of specialty “turf” and focus on providing the medical community with a guideline aimed at optimal patient care.

1.2. Organization of the Writing Committee

The guideline was written by a committee comprised of experts in cardiovascular medicine, surgery, radiology, and nursing. For many of the previous ACCF/AHA practice guidelines, writing expertise has been available within these 2 organizations. Because of the broad scope and diversity of thoracic aortic diseases, as well as the specialists who treat such patients, the ACCF and AHA sought greater involvement from many specialty organizations. Most, but not all, specialty organizations that represent the major stakeholders caring for patients with thoracic aortic diseases provided writing committee members and financial support of the project, and they are recognized as marquee level partners with the ACCF and AHA. These organizations included the American Association for Thoracic Surgery (AATS), American College of Radiology (ACR), American Stroke Association (ASA), Society of Cardiovascular Anesthesiologists (SCA), Society for Cardiovascular Angiography and Interventions (SCAI), Society of Interventional Radiology (SIR), Society of Thoracic Surgeons (STS), and Society for Vascular Medicine (SVM). The American College of Emergency Physicians (ACEP) and the American College of Physicians (ACP) were also represented on the writing committee. Where additional expertise was needed, the scientific councils of the AHA were contacted for writing committee representatives. Representation was provided or facilitated by the Councils on Cardiovascular Nursing, Cardiovascular Surgery and Anesthesia, Cardiovascular Radiology and Intervention, and Clinical Cardiology, Council for High Blood Pressure Research, and Stroke Council.

1.3. Document Review and Approval

This document was reviewed by 3 outside reviewers nominated by the ACCF and 2 outside reviewers nominated by the AHA, as well as 1 or 2 reviewers from each of the following organizations: the AATS, ACP, ACEP, ACR, ASA, SCA, SCAI, SIR, STS, and the SVM. It was also reviewed by 6 individual content reviewers—2 content reviewers from the ACCF Catheterization Committee and 1 content reviewer from the ACCF Interventional Council. All reviewer RWI information was collected and distributed to the writing committee and is published in this document (see Appendix 2).

This document was approved for publication by the governing bodies of the ACCF and the AHA; and the AATS, ACEP, ACR, ASA, SCA, SCAI, SIR, STS, and SVM and was endorsed by the North American Society for Cardiovascular Imaging.

1.4. Scope of the Guideline

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.² 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).³ The diagnosis of acute thoracic AoD or rupture is often difficult and delayed, and errors in diagnosis may account for deaths otherwise attributed to cardiac arrhythmia, myocardial infarction (MI), pulmonary embolism, or mesenteric ischemia.

Most patients with significant thoracic aortic disease will be directed to specialized practitioners and institutions. However, the importance of early recognition and prompt treatment and/or referral for a variety of thoracic aortic diseases by all healthcare professionals provides the rationale for this document. This guideline will provide the practitioner with a sufficient description of background information, diagnostic modalities, and treatment strategies so that appropriate care of these patients can be facilitated and better understood. The goal of this guideline is to improve the health outcomes and quality of life for all patients with thoracic aortic disease.

This guideline includes diseases involving any or all parts of the thoracic aorta with the exception of aortic valve diseases⁴ and includes the abdominal aorta when contiguous thoracic aortic diseases are present. Specific disease states are described in the following sections and the reader is referred to the glossary of terminology in Section 1.5 for abbreviations used throughout the guideline.

1.4.1. 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 and determine risk for future complications.
- Radiologic imaging technologies have improved in terms of accuracy of detection of thoracic aortic disease. However, as the use of these technologies has increased, so also has the potential risk associated with repeated radiation exposure, as well as contrast medium-related toxicity. Whether these technologies should be used repeatedly as a widespread screening

tool is discussed in the full-text document. In addition, the writing committee formulated recommendations on a standard reporting format for thoracic aortic findings as discussed in Section 4.

- Imaging for asymptomatic patients at high risk based on history or associated diseases 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.
- A subset of 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. This issue has come under greater medical-legal scrutiny, and specific cases have been widely discussed in the public domain. Widespread 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 9. The collaboration and cosponsorship of multiple medical specialties in the writing of this guideline will provide unique opportunities for widespread dissemination of knowledge 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. 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.^{5,6} 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.

1.5. Glossary of Terms and Abbreviations Used Throughout the Guideline

Aneurysm (or true aneurysm): a permanent localized dilatation of an artery, having at least a 50% increase in diameter compared to the expected normal diameter of the artery in question. Although all 3 layers (intima, media, and adventitia) may be present, the intima and media in large aneurysms may be so attenuated that in some sections of the wall they are undetectable.

Pseudoaneurysm (or false aneurysm): contains blood resulting from disruption of the arterial wall with

extravasation of blood contained by periarterial connective tissue and not by the arterial wall layers. Such an extravascular hematoma that freely communicates with the intravascular space is also known as a pulsating hematoma.⁷⁻⁹

Ectasia: arterial dilatation less than 150% of normal arterial diameter.

Arteriomegaly: diffuse arterial dilatation involving several arterial segments with an increase in diameter greater than 50% by comparison to the expected normal arterial diameter.

Thoracoabdominal aneurysm (TAA): aneurysm involving the thoracic and abdominal aorta.

Abdominal aortic aneurysm (AAA): aneurysm involving the infradiaphragmatic abdominal aorta.

Aortic dissection (AoD): disruption of the media layer of the aorta with bleeding within and along the wall of the aorta. Dissection may, and often does, occur without an aneurysm being present. An aneurysm may, and often does, occur without dissection. The term dissecting aortic aneurysm is often used incorrectly and should be reserved only for those cases where a dissection occurs in an aneurysmal aorta.

2. THE THORACIC AORTA

The thoracic aorta is divided into 4 parts: the aortic root (which includes the aortic valve annulus, the aortic valve cusps, and the sinuses of Valsalva); the ascending aorta (which includes the tubular portion of the ascending aorta beginning at the sinotubular junction and extending to the brachiocephalic artery origin); the aortic arch (which begins at the origin of the brachiocephalic artery, and is the origin of the head and neck arteries, coursing in front of the trachea and to the left of the esophagus and the trachea); and the descending aorta (which begins at the isthmus between the origin of the left subclavian artery and the ligamentum arteriosum and courses anterior to the vertebral column, and then through the diaphragm into the abdomen) (see Figure 1).

The normal human adult aortic wall is composed of 3 layers, listed from the blood flow surface outward:

Intima: Endothelial layer on a basement membrane with minimal ground substance and connective tissue.

Media: Bounded by an internal elastic lamina, a fenestrated sheet of elastic fibers; layers of elastic fibers arranged concentrically with interposed smooth muscle cells; bounded by an external elastic lamina, another fenestrated sheet of elastic fibers.

Adventitia: A resilient layer of collagen containing the vasa vasorum and nerves. Some of the vasa vasorum can penetrate into the outer third of the media.

3. THORACIC AORTIC HISTOPATHOLOGY

3.1. Atherosclerosis

A 1995 consensus document from the AHA defines the types and histological classes of atherosclerosis¹⁰ (Figure 2).

3.2. Aneurysms and Dissections

Aortic aneurysm histopathology, more accurately termed medial degeneration, is characterized by disruption and loss of elastic fibers and increased deposition of proteoglycans.

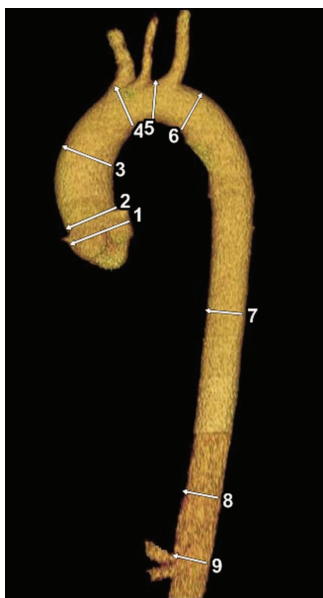


Figure 1. Normal anatomy of the thoracoabdominal aorta with standard anatomic landmarks for reporting aortic diameter as illustrated on a volume-rendered CT image of the thoracic aorta. CT indicates computed tomographic imaging. Anatomic locations: 1, Aortic sinuses of Valsalva; 2, Sinotubular junction; 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. CT indicates computed tomographic imaging.

Typically, there are areas of loss of smooth muscle cells in the aortic media, but whether there is a total loss of smooth muscle cells in the aortic wall is not clear. Recent literature supports the presence of inflammatory cell infiltration in this disease.^{11,12} Aortic pathology associated with myosin heavy chain 11, smooth muscle (*MYH11*) and actin, alpha 2, smooth muscle aorta (*ACTA2*) mutations leading to ascending aortic aneurysms demonstrates a hyperplastic response by smooth muscle cells in the aortic media. The aortic media in aneurysm tissue taken from patients harboring mutations in these genes demonstrated focal hyperplasia associated with smooth muscle cells that were remarkable for a lack of structured orientation parallel to the lumen of the aorta, but instead, the smooth muscle cells were oriented randomly with respect to one another.^{13,14}

Increased immunostaining for a subset of matrix metalloproteinases (MMPs) has been described in the media of thoracic aortic aneurysms, particularly MMP-2 and MMP-9.^{15–18} Immunostaining of aortic media from patients with Marfan syndrome has demonstrated increases of MMP-2 and MMP-9, which was associated with smooth muscle cells at the borders of areas of medial degeneration and on the surface of disrupted elastic fibers. Elevated MMP-2 and MMP-9 immunostaining has been demonstrated in ascending aneurysms from patients with either tricuspid or bicuspid aortic valves^{16,18} and inconsistently in ascending aortic tissue from patients with tricuspid aortic valves.¹⁷ These 2

MMPs are known to have elastolytic activity. Variable expression of MMPs and tissue inhibitors of MMPs has also been demonstrated in aortic tissue of patients with Marfan syndrome versus patients without Marfan syndrome.¹⁹ Although accumulation of proteoglycans in the aortic media is another consistent finding in thoracic aortic aneurysms, no studies have determined why this accumulation occurs or whether these are causative in nature.

3.3. Vasculitis and Inflammatory Diseases

Giant cell arteritis and Takayasu arteritis share important features^{19a} with T-cell clonal expansion suggesting an antigenic response. An adventitial inflammatory response is marked by augmented cytokine and MMP production causing granuloma formation, which causes vessel destruction.^{19b} Behçet disease affects both arteries and veins of all sizes.

4. 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 (see Table 2). (*Level of Evidence: C*)
2. For measurements taken by computed tomography 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 midsinus level, should be used. (*Level of Evidence: 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 midsinus level, should be used. (*Level of Evidence: C*)
4. Abnormalities of aortic morphology should be recognized and reported separately even when aortic diameters are within normal limits. (*Level of Evidence: C*)
5. The finding of aortic dissection, aneurysm, traumatic injury and/or aortic rupture should be immediately communicated to the referring physician. (*Level of Evidence: C*)
6. Techniques to minimize episodic and cumulative radiation exposure should be utilized whenever possible.^{20,21} (*Level of Evidence: 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 (see Tables 3 and 4). (*Level of Evidence: C*)

Definitive identification or exclusion of thoracic aortic disease or one of its anatomic variants requires dedicated aortic imaging. Selection of the most appropriate imaging study may depend on patient related factors (ie, hemodynamic stability, renal function, contrast allergy) and institutional

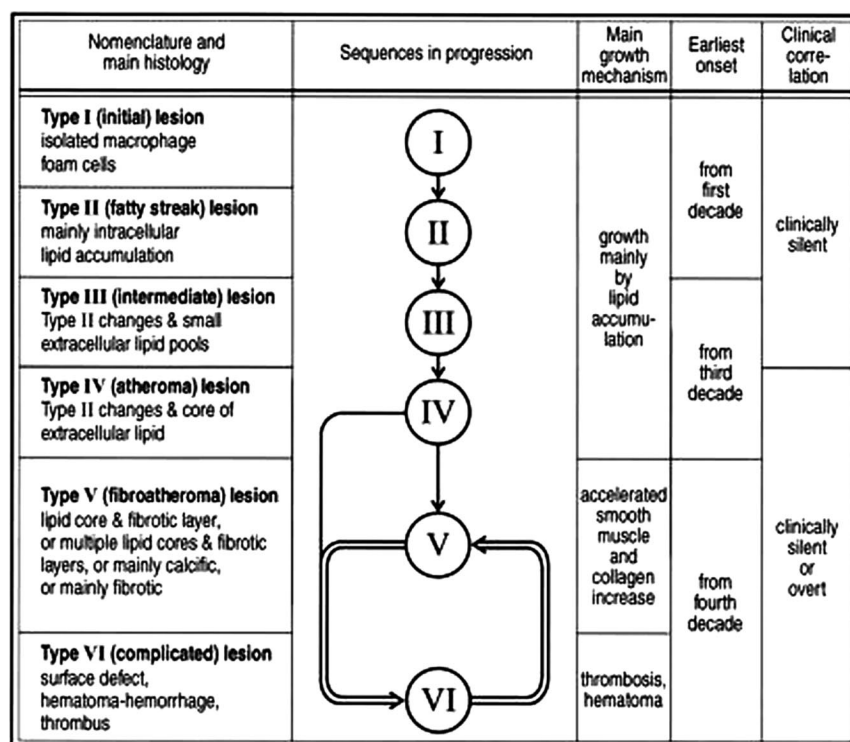


Figure 2. Atherosclerotic lesions. Flow diagram in center column indicates pathways in evolution and progression of human atherosclerotic lesions. Roman numerals indicate histologically characteristic types of lesions defined at the left of the flow diagram. The direction of the arrows indicates the sequence in which characteristic morphologies may change. From Type I to Type IV, changes in lesion morphology occur primarily because of increasing accumulation of lipid. The loop between Types V and VI illustrates how lesions increase in thickness when thrombotic deposits form on their surfaces. Thrombotic deposits may form repeatedly over varied time spans in the same location and may be the principal mechanism for gradual occlusion of medium-sized arteries. Adapted from Stary et al.¹⁰

capabilities (ie, rapid availability of individual imaging modalities, state of the technology, and imaging specialist expertise). Consideration should be given to patients with borderline abnormal renal function (serum creatinine greater than 1.8 to 2.0 mg/dL)—specifically, the tradeoffs between the use of iodinated intravenous contrast for CT and the possibility of contrast-induced nephropathy, and gadolinium agents used with MR and the risk of nephrogenic systemic fibrosis.²²

Radiation exposure should be minimized.^{21,23–26} The risk of radiation-induced malignancy is the greatest in neonates, children, and young adults.²¹ Generally, above the age of 30 to 35 years, the probability of radiation-induced

malignancy decreases substantially.^{20,21} For patients who require repeated imaging to follow an aortic abnormality, MR may be preferred to CT. MR may require sedation due to longer examination times and tendency for claustrophobia.

CT as opposed to echocardiography can best identify thoracic aortic disease, as well as other disease processes that can mimic aortic disease, including pulmonary embolism, pericardial disease, and hiatal hernia. After intervention or open surgery, CT is preferred to detect asymptomatic postprocedural leaks or pseudoaneurysms because of the presence of metallic closure devices and clips.

CT and MR measure external aortic diameter, whereas echocardiography measures internal aortic diameter. Lumen size may not accurately reflect external diameter due to intraluminal clot, wall inflammation, or AoD. A recent refinement in the CT measurement of aortic size examines the vessel size using a centerline of flow, which reduces the error of tangential measurement and allows true short-axis measurement of aortic diameter. Essential element of aortic imaging reports are listed in Table 2.

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 et al.²⁷

Table 4. Sex Differences in Aortic Root Dimensions in Adults

Aortic root	Absolute values (cm)			Indexed values (cm/m ²)		
	Men	P Value	Women	Men	P Value	Women
Annulus	2.6 ± 0.3	<0.001	2.3 ± 0.2	1.3 ± 0.1	NS	1.3 ± 0.1
Sinuses of Valsalva	3.4 ± 0.3	<0.001	3.0 ± 0.3	1.7 ± 0.2	NS	1.8 ± 0.2
Sinotubular junction	2.9 ± 0.3	<0.001	2.6 ± 0.3	1.5 ± 0.2	NS	1.5 ± 0.2
Proximal ascending aorta	3.0 ± 0.4	<0.001	2.7 ± 0.4	1.5 ± 0.2	NS	1.6 ± 0.3

NS indicates not significant. Adapted from Roman et al.²⁸

4.1. Chest X-Ray

Routine chest x-ray may occasionally detect abnormalities of aortic contour or size that require definitive aortic imaging. Chest x-ray often serves as a part of the evaluation of patients with potential acute AoD, primarily to identify other causes of patient's symptoms, but also as a screening test to identify findings due to a dilated aorta or bleeding.

4.2. Computed Tomographic Imaging

CT scanning has several advantages, including near-universal availability; the ability to image the entire aorta, including lumen, wall, and periaortic regions; to identify anatomic variants and branch vessel involvement; to distinguish among types of acute aortic syndromes (ie, intramural hematoma [IMH], penetrating atherosclerotic ulcer [PAU], and acute AoD); and the short time required to complete the imaging process and the 3-dimensional data. Electrocardiogram-gated techniques have made it possible to generate motion-free images of the aortic root and coronary arteries, similar to coronary CT angiographic imaging. Reports of newer-generation multidetector helical CT scanners show sensitivities of up to 100% and specificities of 98% to 99%.^{29–32}

The sequence for a CT performed in the potential setting of acute AoD generally would include a noncontrast study to detect subtle changes of IMH, followed by a contrast study to delineate the presence and extent of the dissection flap, identify regions of potential malperfusion, and demonstrate contrast leak indicating rupture. Imaging of the

vascular tree from the thoracic inlet to the pelvis, including the iliac and femoral arteries, provides sufficient information to plan surgical or endovascular treatment, if needed. Prompt interpretation and communication of findings to the appropriate treating physicians are essential in the acute setting. (For further information on technique parameters and anatomic coverage, see the online-only Data Supplement.)

4.3. Magnetic Resonance Imaging

MR has been shown to be very accurate in the diagnosis of thoracic aortic disease, with sensitivities and specificities that are equivalent to or may exceed those of CT and transesophageal echocardiogram (TEE).^{30,35–39} Advantages of MR include the ability to identify anatomic variants of AoD (IMH and PAU), assess branch artery involvement, and diagnose aortic valve pathology and left ventricular dysfunction without exposing the patient to either radiation or iodinated contrast. Disadvantages include prolonged duration of imaging acquisition during which the patient is inaccessible to care providers; inability to use gadolinium contrast in patients with renal insufficiency; contraindication in patients with claustrophobia, metallic implants or pacemakers, and lack of widespread availability on an emergency basis.

4.4. Echocardiography

Echocardiography can detect the presence of aortic enlargement and associated cardiac pathology that suggests the underlying etiology of the aortic disease (eg, bicuspid aortic valve). For AoD (Figure 3), one of the major limitations of both transthoracic echocardiogram (TTE) and TEE is the frequent appearance of artifacts that mimic a dissection flap (Figure 4). These usually arise from a mirror image or reverberation artifact that appears as a mobile linear echodensity overlying the aortic lumen. It is therefore essential that the echocardiographer make certain to distinguish true dissection flaps from such artifacts.

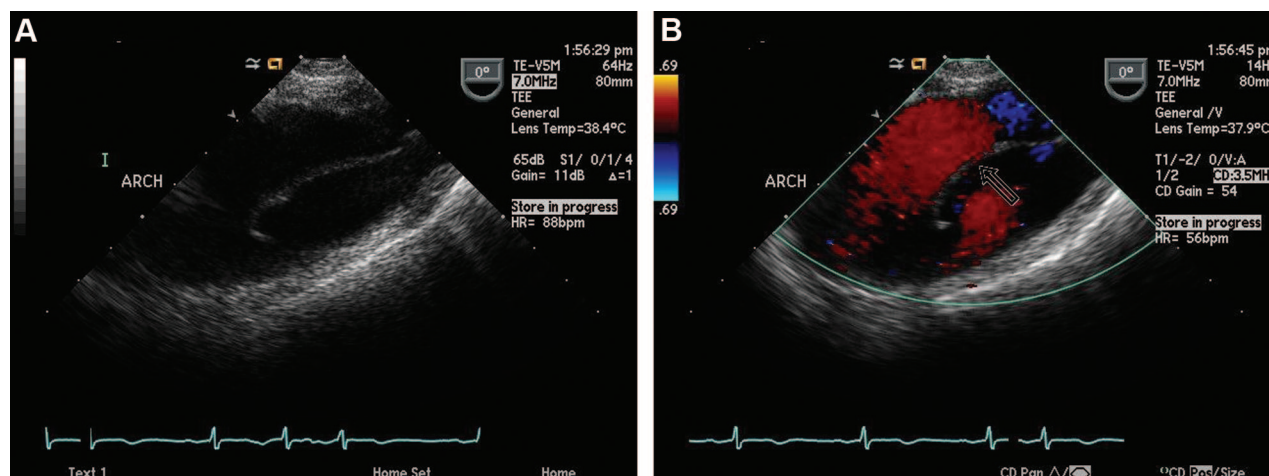


Figure 3. Arch aneurysm with dissection flap. Panel A, Arch dissection, 2-dimensional view. Panel B, Arch dissection (arrow) with color flow Doppler margination.

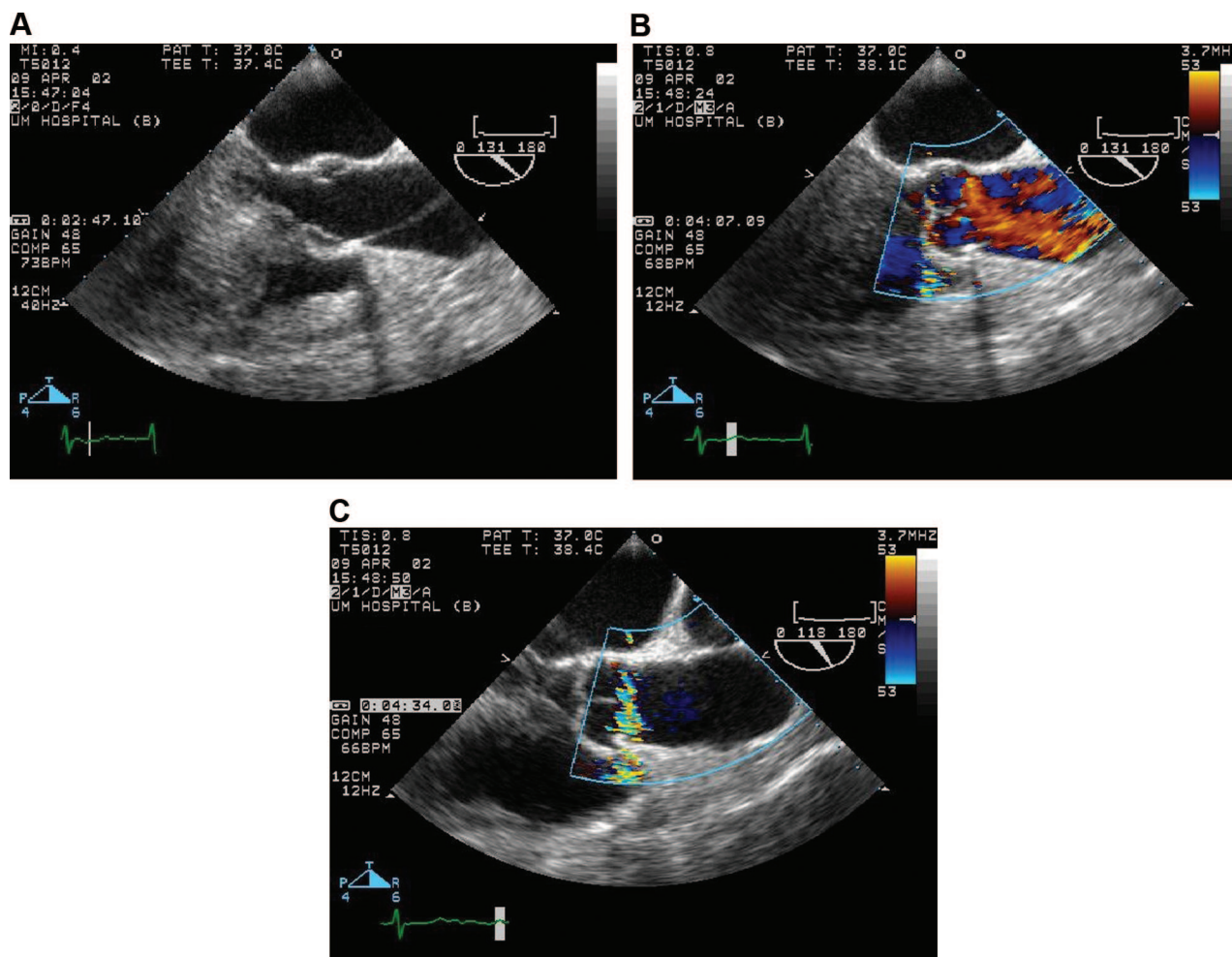


Figure 4. Artifact mimicking dissection. Panel A, 2-dimensional view. Panel B, Color flow Doppler without margination. Panel C, Artifact not seen in this view.

5. 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 (see Figure 5) and 6 months thereafter to determine the rate of enlargement of the aorta. (*Level of Evidence: 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. (*Level of Evidence: 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.^{40–43} (*Level of Evidence: C*)
4. Loeys-Dietz patients should have yearly magnetic resonance imaging from the cerebrovascular circulation to the pelvis.^{18,44,45} (*Level of Evidence: 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.⁴⁶ If initial imaging is normal and there are no risk factors for aortic dissection, repeat

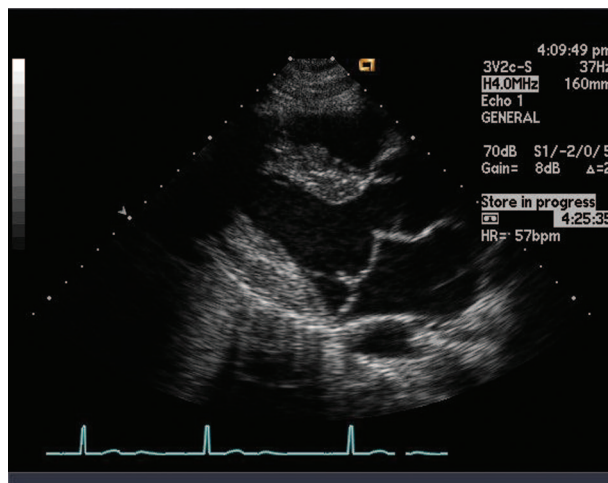


Figure 5. Transthoracic echocardiogram of a patient with Marfan syndrome with mitral valve prolapse and 4-cm ascending aortic aneurysm.

Table 5. 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 Patent ductus arteriosus	Multiple aortic dissections documented at aortic diameters <5.0 cm
<i>MYH11</i> mutations	1%		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.

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. (Level of Evidence: 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 tomography scanning and/or magnetic resonance imaging (external diameter).⁴⁴ (Level of Evidence: 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.⁴⁰ (Level of Evidence: 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.^{42,47,48} (Level of Evidence: 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. (Level of Evidence: C)

There are several syndromic and nonsyndromic genetic conditions that are associated with the development of thoracic aortic aneurysms and present with dissections at smaller diameters than usual. The following recommendations focus on these specific conditions, including Marfan syndrome, Loeys-Dietz syndrome, Turner syndrome, bicuspid aortic valve, and other genetic mutations (*TGFBR1*, *TGFBR2*, *FBN1*, *ACTA2*, *COL3A1*, *MYH11*) (see Tables 5 and 6).

A substantial proportion of Ehlers-Danlos syndrome patients who do not have the vascular form also have aortic root dilatation but the progression of this dilatation to AoD is rare.^{42,49} Similarly, patients with congenital contractural

arachnodactyly or Beals syndrome due to mutations in *FBN2* have had aortic root enlargement without documented progression to dissection.^{50,51}

There are other genetic syndromes that have multiple reports or documentation of thoracic aortic aneurysms leading to Type A dissections. There are multiple case reports of AoD in patients with autosomal dominant polycystic kidney disease.^{52,53} Although AoD is a complication of autosomal dominant polycystic kidney disease, it is less common than cerebral aneurysms leading to subarachnoid hemorrhage in this population. There is insufficient information to gauge the value of routine or screening imaging for these patients.

Similar to autosomal dominant polycystic kidney disease, there are multiple reports in the literature of patients with Noonan syndrome experiencing AoDs.^{54–56} The value of imaging or routine monitoring of these patients is unknown. A review of 200 patients with Alagille syndrome also identified thoracic aortic disease in a small subset of these patients.⁵⁷

6. 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.^{58,59} (Level of Evidence: B)
2. If the mutant gene (*FBN1*, *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. (Level of Evidence: 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.⁵⁸ (Level of Evidence: 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.^{13,14,43,44,60,61} (Level of Evidence: B)

Table 6. Genetic Syndromes Associated with Thoracic Aortic Aneurysm and Dissection

Genetic syndrome	Common clinical features	Genetic defect	Diagnostic test	Comments on aortic disease
Marfan syndrome	Skeletal features (see text) Ectopia lentis Dural ectasia	<i>FBN1</i> mutations*	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
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	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)
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	DNA for sequencing Dermal fibroblasts for analysis of type III collagen	Surgical repair is complicated by friable tissues Noninvasive imaging recommended
Turner syndrome	Short stature Primary amenorrhea Bicuspid aortic valve Aortic coarctation Webbed neck, low-set ears, low hairline, broad chest	45,X karyotype	Blood (cells) for karyotype analysis	AoD risk is increased in patients with bicuspid aortic valve, aortic coarctation, hypertension, or pregnancy

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.

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

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.^{13,14,43,44,60,61} (Level of Evidence: 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. (Level of Evidence: C)

7. 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. (Level of Evidence: 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.^{62–65} (Level of Evidence: B)

8. RECOMMENDATIONS FOR TAKAYASU ARTERITIS AND GIANT CELL ARTERITIS

See Table 7 and Figure 6.

Class I

1. Initial therapy for active Takayasu arteritis and active giant cell arteritis should be corticosteroids at a high dose (prednisone 40 to 60 mg daily at initiation or its equivalent) to reduce the active inflammatory state.^{66,67} (Level of Evidence: B)
2. The success of treatment of patients with Takayasu arteritis and giant cell arteritis should be periodically evaluated to determine disease activity by repeated physical examination and either an erythrocyte sedimentation rate or C-reactive protein level.^{68,69} (Level of Evidence: B)
3. Elective revascularization of patients with Takayasu arteritis and giant cell arteritis should be delayed until the acute inflammatory state is treated and quiescent.⁷⁰ (Level of Evidence: B)
4. The initial evaluation of Takayasu arteritis or giant cell arteritis should include thoracic aorta and branch vessel computed tomography scanning or magnetic resonance imaging to investigate the possibility of aneurysm or occlusive disease in these vessels. (Level of Evidence: C)

Table 7. Inflammatory Diseases Associated with Thoracic Aortic Aneurysm and Dissection

Names	Criteria used in diagnosis/source	When is diagnosis established?
Takayasu arteritis ⁷¹	Age of onset <40 y Intermittent claudication Diminished brachial artery pulse Subclavian artery or aortic bruit Systolic BP variation of >10 mm Hg between arms Aortographic evidence of aorta or aortic branch stenosis	≥3 criteria are present (sensitivity 90.5%; specificity 97.8%)
Giant cell arteritis ⁷²	Age >50 y Recent-onset localized headache Temporary artery tenderness or pulse attenuation Elevated erythrocyte sedimentation >50 mm/h Arterial biopsy shows necrotizing vasculitis	≥3 criteria are present (sensitivity >90%; specificity >90%)
Behçet disease ⁷³	Oral ulceration Recurrent genital ulceration Uveitis or retinal vasculitis Skin lesions—erythema nodosum, pseudo-folliculitis, or pathergy	Oral ulceration plus 2 of the other 3 criteria
Ankylosing spondylitis ⁷⁴	Onset of pain <40 y Back pain for >3 months Morning stiffness Subtle symptom onset Improvement with exercise	4 of the diagnostic criteria are present

BP indicates blood pressure.

Class IIa

1. It is reasonable to treat patients with Takayasu arteritis receiving corticosteroids with an additional anti-inflammatory agent if there is evidence of progression of vascular disease, recurrence of constitutional symptoms, or re-elevation of inflammatory marker.⁶⁶ (Level of Evidence: C)

9. 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 (see Figure 7). 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 (see Table 8).^{59,75–77} (Level of Evidence: 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 *FBN1*, *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^{75–81} (Level of Evidence: 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^{75,77,81–84} (Level of Evidence: B):
 - Pulse deficit.
 - Systolic blood pressure limb differential greater than 20 mm Hg.
 - Focal neurological 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 disorders associated with thoracic aortic disease.⁷⁶ (Level of Evidence: 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.⁷⁶ (Level of Evidence: 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. (Level of Evidence: 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 neurological injury or the presence of pericardial tamponade. (Level of Evidence: C)
6. All patients presenting with acute neurological 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 neurological pathology are less likely to report thoracic pain than the typical aortic dissection patient.⁸³ (Level of Evidence: C)

These recommendations provide guidance to improve more prompt diagnosis of acute AoD (Figure 7). The true incidence of acute AoD is difficult to define as AoD can be rapidly fatal and when patients expire prior to hospitalization, death may be erroneously attributed to another cause. Acute AoD is frequently missed on initial presentation and early mortality among this group may be misclassified as nondissection related. Classes of intimal tears are described in Figure 8. The DeBaKey and Stanford Classifications of



Figure 6. Takayasu arteritis with involvement of the thoracoabdominal aorta and great vessels as shown on contrast-enhanced CT and MR examinations. Note narrowing of the arterial lumen and circumferential soft tissue thickening of the walls of the great vessels and thoracic and abdominal aorta. Panel A, Image through the great vessels with narrowing of the left common carotid and left subclavian arteries; Panel B, Mid descending thoracic aorta (arrowheads); Panel C, Aorta just above the diaphragm (arrowheads); Panel D, Infrarenal aorta; Panel E, Volume-rendered image from CT demonstrates the extent of involvement; Panel F, Oblique sagittal MR of the thoracic aorta; Panel G, Coronal MR of the abdominal aorta. CT indicates computed tomographic imaging; and MR, magnetic resonance imaging.

AoD are pictured in Figure 9. There is no unanimity as to which classification system should be universally used.

10. INITIAL EVALUATION AND MANAGEMENT OF ACUTE THORACIC AORTIC DISEASE

10.1. 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.^{75,81,88} (*Level of Evidence: 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. (*Level of Evidence: 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. (*Level of Evidence: C*)
3. Urgent and definitive imaging of the aorta using transesophageal echocardiogram, computed tomography scanning, or magnetic resonance imaging is recommended to identify or exclude thoracic aortic

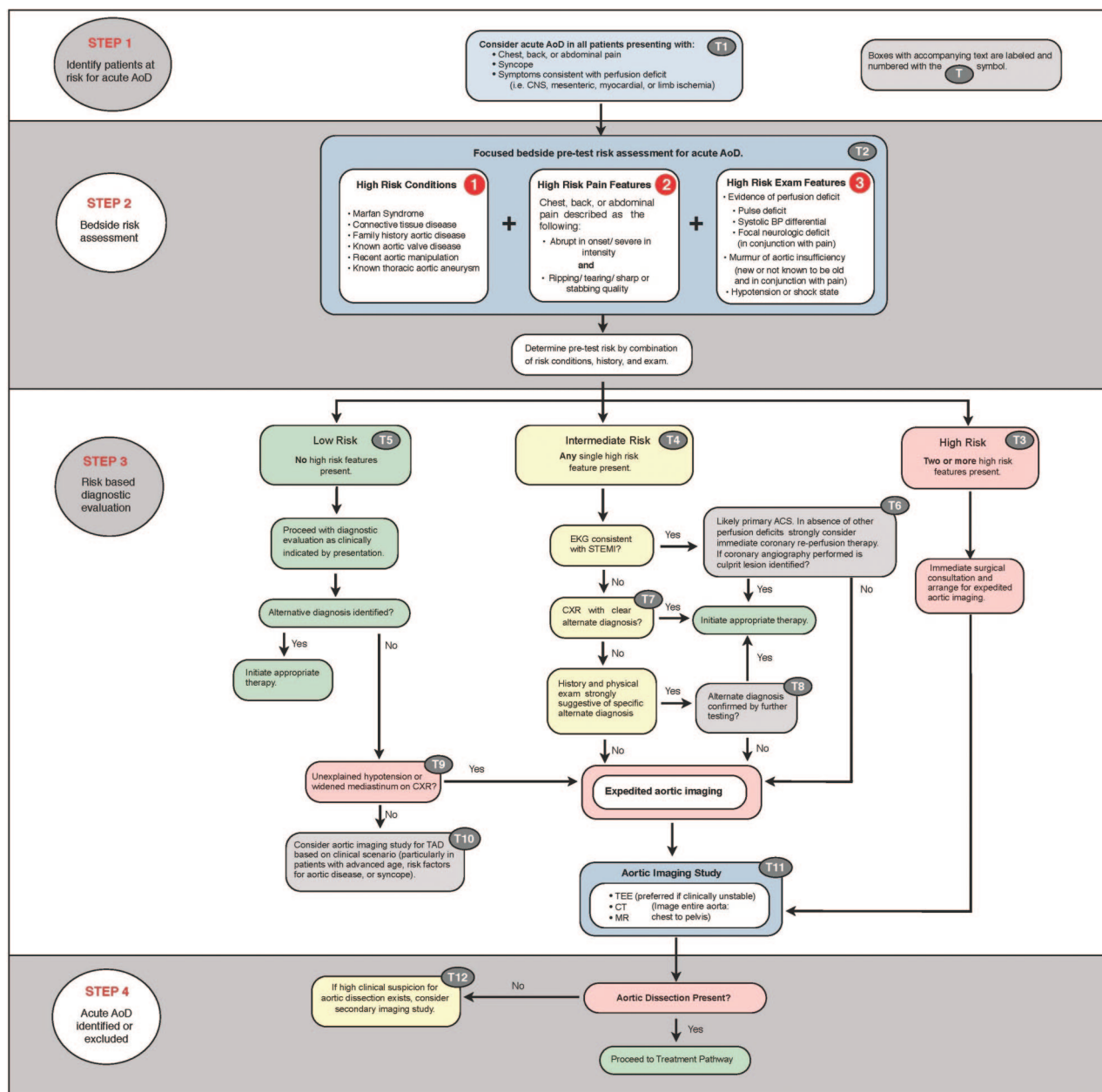


Figure 7. AoD evaluation pathway. ACS indicates acute coronary syndrome; AoD, aortic dissection; BP, blood pressure; CNS, central nervous system; CT, computed tomographic imaging; CXR, chest x-ray; EKG, electrocardiogram; MR, magnetic resonance imaging; STEMI, ST-elevated myocardial infarction; TAD; thoracic aortic disease; and TEE, transesophageal echocardiogram.

dissection in patients at high risk for the disease by initial screening.^{29–32,37,89,90} (Level of Evidence: 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. (Level of Evidence: C)

10.2. 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. (Level of Evidence: 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.⁸⁵ (Level of Evidence: C)

10.3. Recommendations for Initial Management

See Figure 10.

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:

Table 8. 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

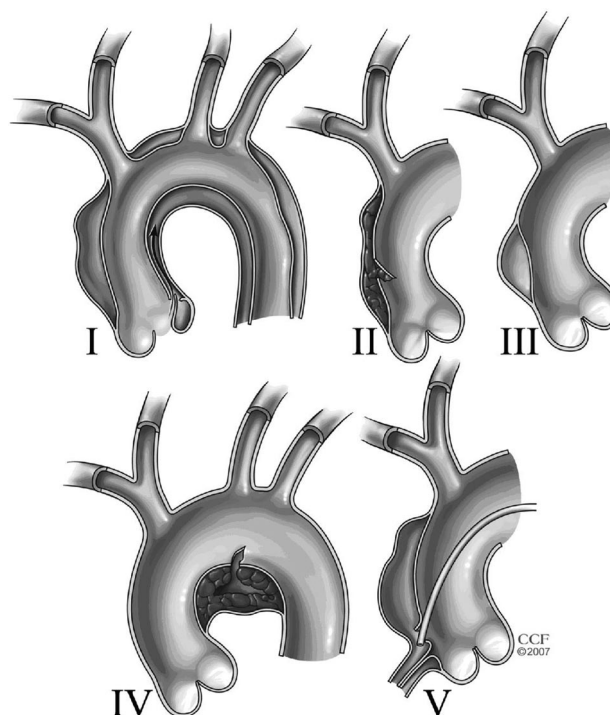


Figure 8. Classes of intimal tears. I. Classic dissection with intimal tear and double lumen separated by septum. Communication between lumens is typically in descending aorta at sheared-off intercostal arteries or distal reentry site. II. IMH. No intimal tear or septum is imaged but is usually found at surgery or autopsy. DeBakey Types II and IIIa are common extent of this lesion. III. Intimal tear without medial hematoma (limited dissection) and eccentric aortic wall bulge. Rare and difficult to detect by TEE or CT. Patients with Marfan syndrome prone to this type. May result in aortic rupture or extravasation. IV. PAU usually to the adventitia with localized hematoma or saccular aneurysm. May propagate to Class I dissection, particularly when involving ascending aorta or aortic arch. V. Iatrogenic (catheter angiography or intervention)/traumatic (deceleration) dissection. CT indicates computed tomographic imaging; IMH, intramural hematoma; PAU, penetrating atherosclerotic ulcer; and TEE, transesophageal echocardiography. Figure reprinted with permission from the Cleveland Clinic Foundation. Legend adapted from Svensson et al,⁸⁵ Chirillo et al,⁸⁶ and Murray et al.⁸⁷

- 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. (*Level of Evidence: C*)
- b. In patients with clear contraindications to beta blockade, nondihydropyridine calcium channel-blocking agents should be utilized as an alternative for rate control. (*Level of Evidence: 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. (*Level of Evidence: C*)
- d. Beta blockers should be used cautiously in the setting of acute aortic regurgitation because they will block the compensatory tachycardia.⁴ (*Level of Evidence: C*)

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. (*Level of Evidence: C*)

10.4. Recommendations for Definitive Management

See Figures 9 and 11.

Class I

1. Urgent surgical consultation should be obtained for all patients diagnosed with thoracic aortic dissection regardless of the anatomic location (ascending

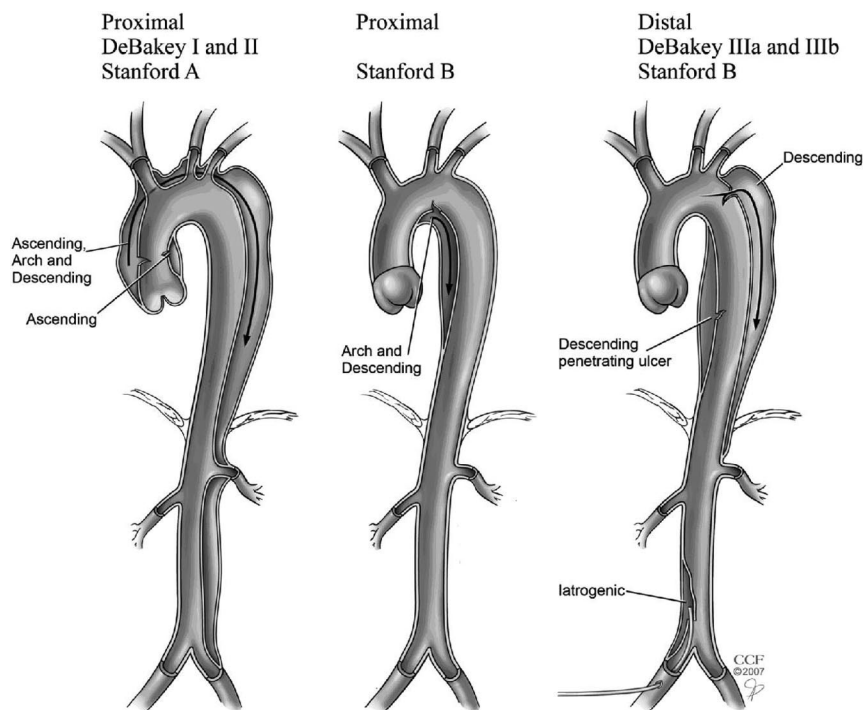
versus descending) as soon as the diagnosis is made or highly suspected. (*Level of Evidence: C*)

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.⁷⁵ (*Level of Evidence: 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).^{80,92-96} (*Level of Evidence: B*)

11. 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

Figure 9. Aortic dissection classification: DeBakey and Stanford classifications. Reprinted with permission from the Cleveland Clinic Foundation.



may be repaired with aortic valve resuspension. 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. (*Level of Evidence: C*)

12. 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. (*Level of Evidence: C*)

13. 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 neurological deficits, a murmur of aortic regurgitation, bruits, and findings compatible with possible cardiac tamponade.^{97–99} (*Level of Evidence: C*)

14. 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 (see Table 9). (*Level of Evidence: C*)

14.1. 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.^{107–111} (*Level of Evidence: 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.¹⁰¹ (*Level of Evidence: B*)

Class IIa

1. For patients with thoracic aortic aneurysm, it is reasonable to reduce blood pressure with beta

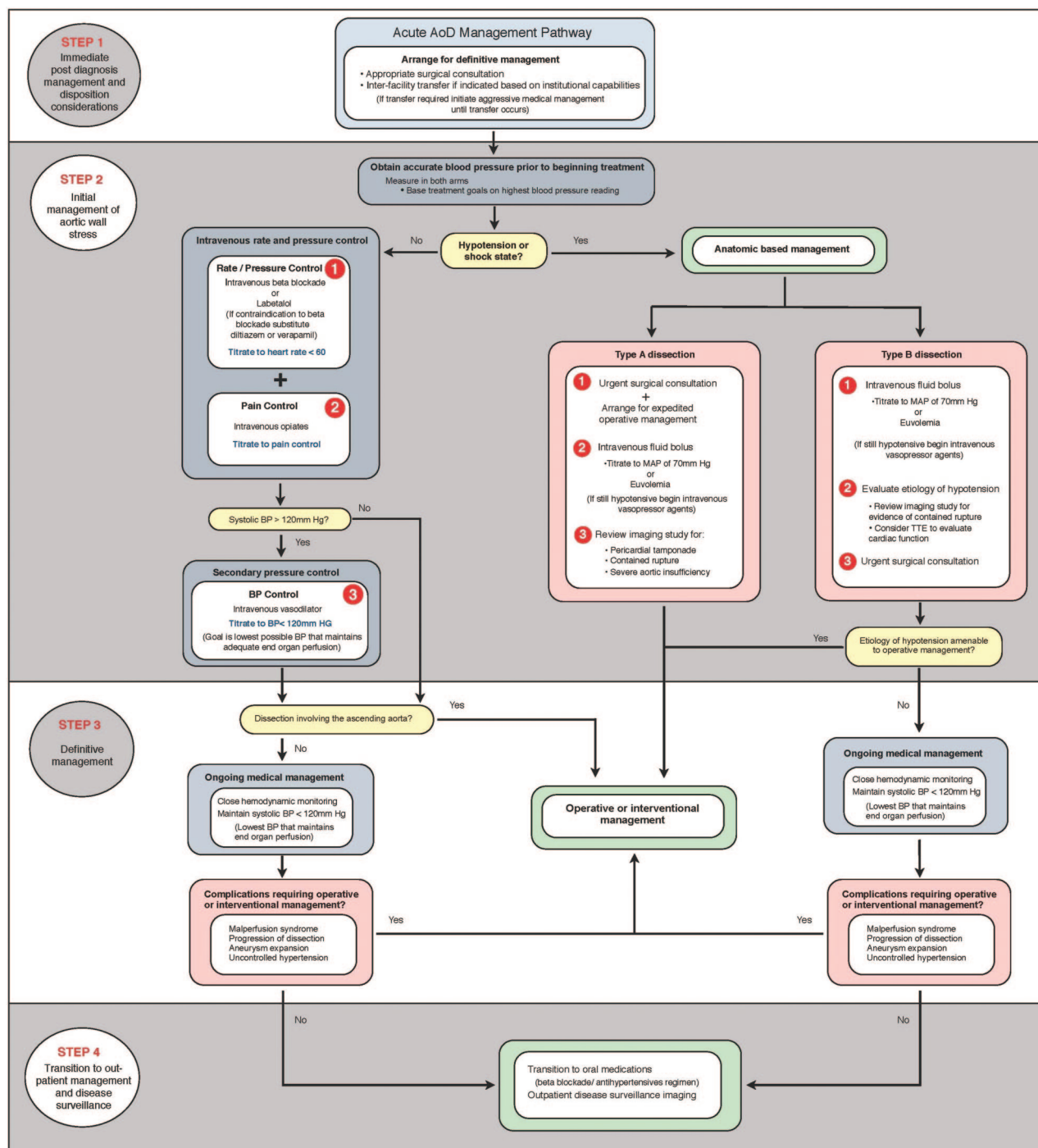


Figure 10. Acute AoD management pathway. AoD indicates aortic dissection; BP, blood pressure; MAP, mean arterial pressure; and TTE, transthoracic echocardiogram.

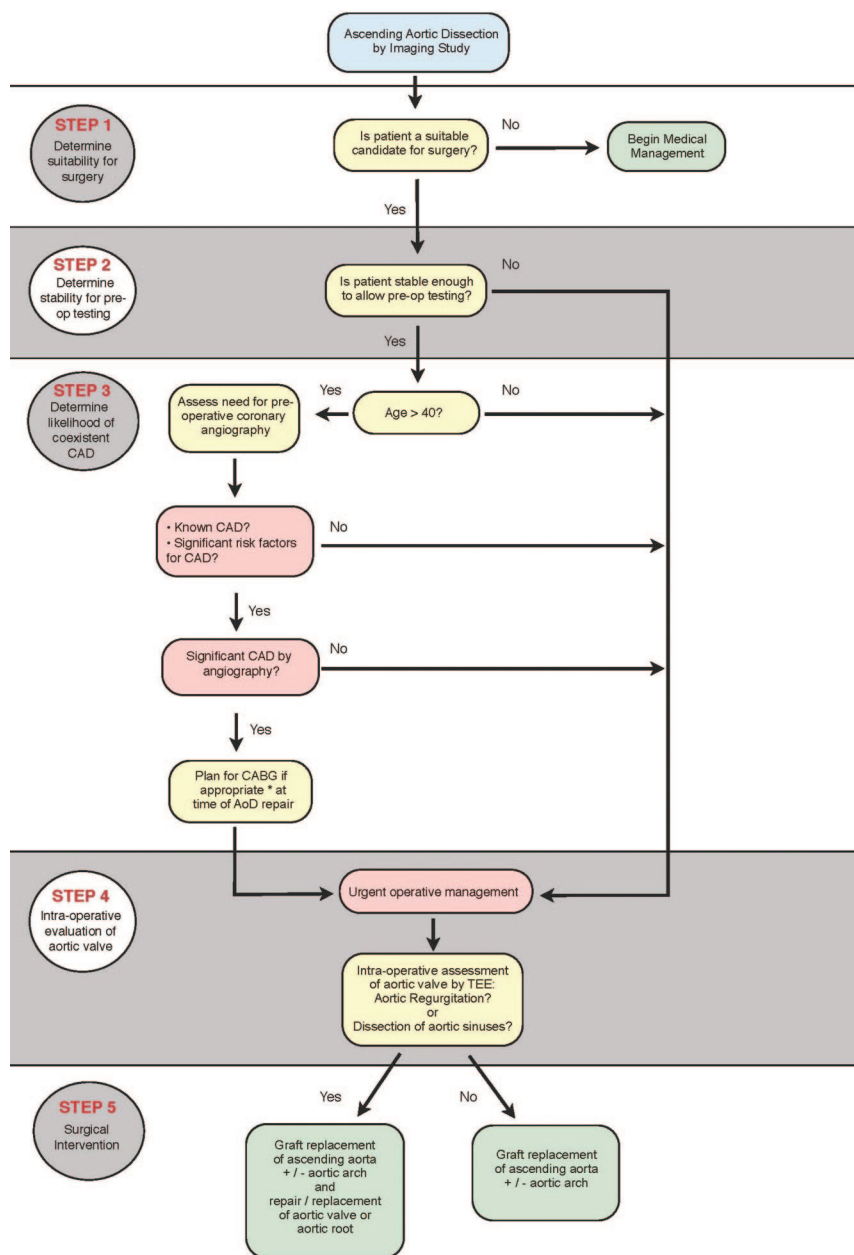
blockers and angiotensin-converting enzyme inhibitors¹⁰³ or angiotensin receptor blockers^{104,105} to the lowest point patients can tolerate without adverse effects.^{100–102} (Level of Evidence: B)

2. An angiotensin receptor blocker (losartan) is reasonable for patients with Marfan syndrome, to reduce the rate of aortic dilatation unless contraindicated.^{105,112} (Level of Evidence: B)

14.2. Recommendation for Dyslipidemia Class IIa

1. Treatment with a statin to achieve a target LDL cholesterol of less than 70 mg/dL is reasonable for patients with a coronary heart disease risk equivalent such as noncoronary atherosclerotic disease, atherosclerotic aortic aneurysm, and coexistent

Figure 11. Acute surgical management pathway for AoD. AoD indicates aortic dissection; CABG, coronary artery bypass graft surgery; CAD, coronary artery disease; TAD, thoracic aortic disease; and TEE, transesophageal echocardiogram. *Addition of 'if appropriate' based on Patel et al.^{96a}



coronary heart disease at high risk for coronary ischemic events.^{113–116} (Level of Evidence: A)

14.3. Recommendation for Smoking Cessation Class I

1. Smoking cessation and avoidance of exposure to environmental tobacco smoke at work and home are recommended. Follow-up, referral to special programs, and/or pharmacotherapy (including nicotine replacement, bupropion, or varenicline) is useful, as is adopting a stepwise strategy aimed at smoking cessation (the 5 As are Ask, Advise, Assess, Assist, and Arrange).^{117–118b} (Level of Evidence: B)

15. RECOMMENDATIONS FOR ASYMPTOMATIC PATIENTS WITH ASCENDING AORTIC ANEURYSM

See Figures 12 and 13.

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.¹¹⁹ (Level of Evidence: C)

Table 9. Studies of Medical Treatment of Thoracic Aortic Aneurysm

Treatment	Studies	Results
Beta blockers	Genoni M, Paul M, Jenni R, et al ¹⁰⁰	Retrospective, case-record review of 78 patients with chronic Type B dissection who received medical treatment. 51 of 71 received beta-blocker treatment, 20 of 71 were treated with other antihypertensive drugs. 10 of 51 (20%) of the beta-blocker-treated patients and 9 of 20 (45%) from the other treatment group needed dissection-related surgery ($P=0.002$). The incidence of increasing aortic diameter was 12% (6 of 51) in the beta-blocker group and 40% (8 of 20) in the other treatment group ($P=0.002$).
	Shores J, Berger KR, Murphy EA, et al ¹⁰¹	Open-label, randomized, control study of propranolol in 70 patients with Marfan syndrome. The treated group received a mean daily propranolol dose of 212 ± 68 mg/d. Propranolol therapy slowed aortic root dilation (0.023 vs 0.084 per year, $P<0.001$).
Angiotensin-converting enzyme inhibitors	Ladouceur M, Fermanian C, Lupoglazoff JM, et al ¹⁰²	Retrospective evaluation of aortic dilation in children with Marfan syndrome. Aortic dilatation was slowed by 0.2 mm/y in children treated with beta blockers.
	Ahimastos AA, Aggarwal A, D'Orsa KM, et al ¹⁰³	Randomized, double-blind, placebo-controlled trial of 17 patients with Marfan syndrome taking beta-blocker therapy to perindopril or placebo. After 24 weeks of therapy, the perindopril-treated subjects compared with placebo-treated subjects had smaller growth in the ascending aortic diameter during systole (1.2 vs 0.3 mm/m ² , $P=0.01$) and a significant reduction in ascending aortic diameter during diastole (0.4 vs -1.2 mm/m ² , $P<0.001$), respectively.
Angiotensin receptor blockers	Mochizuki S, Dahlof B, Shimizu M, et al ¹⁰⁴	3081 Japanese patients with hypertension, coronary heart disease, heart failure, or a combination were randomly assigned either to open-label valsartan (40 to 160 mg/d) or to other treatment without angiotension receptor blockers. Patients randomized to valsartan had reduction in composite cardiovascular outcome (OR 0.61 , 95% CI 0.47 to 0.79) and reduction in aortic dissection (OR 0.18 , 95% CI 0.04 to 0.88). Open-label, randomized.
	Brooke BS, Habashi JP, Judge DP, et al ¹⁰⁵	The clinical response to angiotension receptor blockers (losartan in 17 patients and irbesartan in 1 patient) were evaluated in pediatric patients with Marfan syndrome with severe aortic root enlargement. The mean (\pm SD) rate of change in aortic root diameter decreased significantly from 3.54 ± 2.87 mm/y during previous medical therapy to 0.46 ± 0.62 mm/y during angiotension receptor blocker therapy ($P<0.001$). The deviation of aortic root enlargement from normal, as expressed by the rate of change in z scores, was reduced by a mean difference of 1.47 z scores/y (95% CI 0.70 to 2.24 , $P<0.001$) after the initiation of angiotension receptor blocker therapy. The sinotubular junction showed a reduced rate of change in diameter during angiotension receptor blocker therapy ($P<0.05$), whereas the distal ascending aorta was not affected by angiotension receptor blocker therapy.
Statins	Diehm N, Decker G, Katzen B, et al ¹⁰⁶	A nonrandomized propensity-score-adjusted study of statin use effect on long-term mortality of patients after endovascular repair of AAA (731 patients) or TAA (59 patients) was done. Statin use was associated with decreased long-term mortality in patients with AAA (adjusted HR 0.613 , 95% CI 0.379 to 0.993 , $P=0.047$), but not for patients with TAA (adjusted HR 1.795 , 95% CI 0.147 to 21.942 , $P=0.647$).

AAA indicates abdominal aortic aneurysm; CI, confidence interval; SD, standard deviation; and TAA, thoracic aortic aneurysm.

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 5) to avoid acute dissection or rupture.^{47,119–125} (Level of Evidence: 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. (Level of Evidence: 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. (Level of Evidence: 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

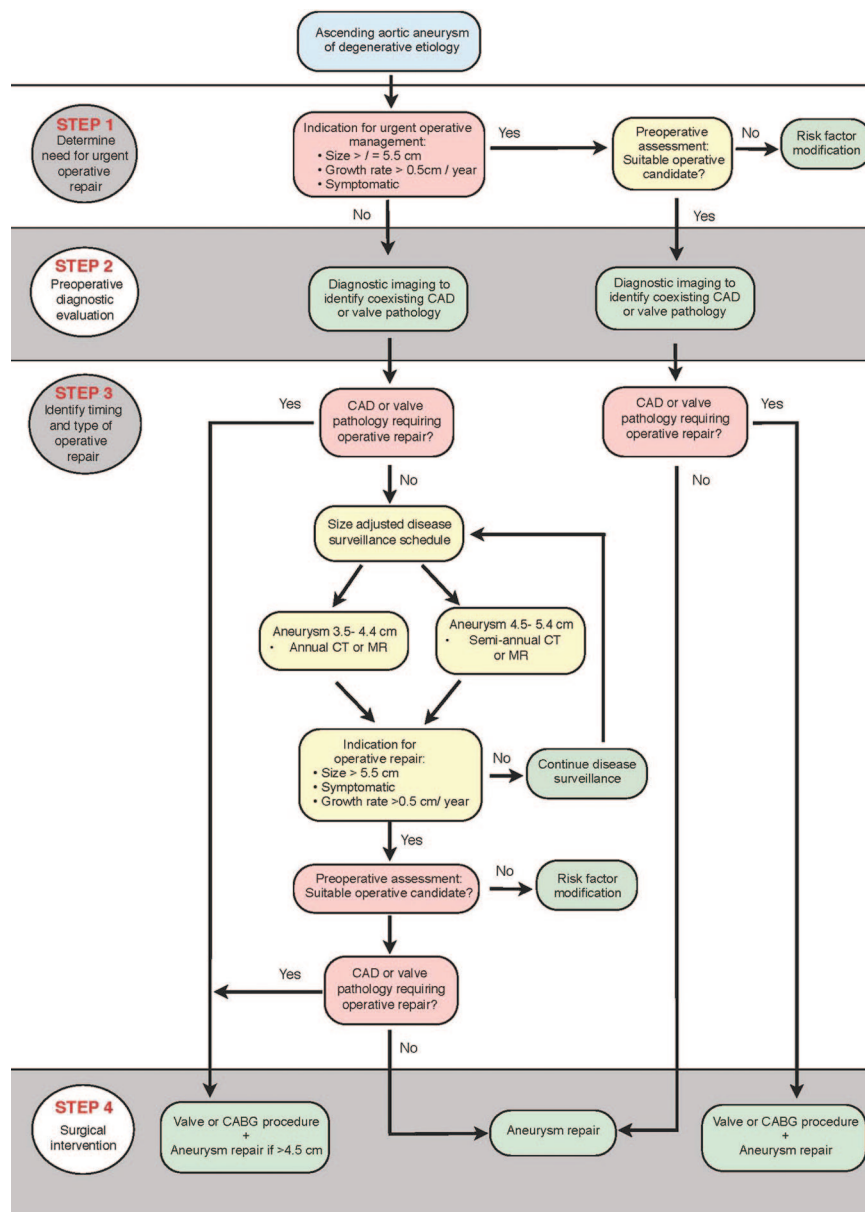
maximal ascending or aortic root area (πr^2) in cm² divided by the patient's height in meters exceeds 10 .^{48,123} (Level of Evidence: C)

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 tomography scanning and/or magnetic resonance imaging (external diameter).⁴⁴ (Level of Evidence: C)

16. 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. (Level of Evidence: C)

Figure 12. 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.



17. 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. (Level of Evidence: 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.^{72,126-129} (Level of Evidence: B)

18. 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.¹³⁰⁻¹³² (Level of Evidence: 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.^{131,132} (Level of Evidence: 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

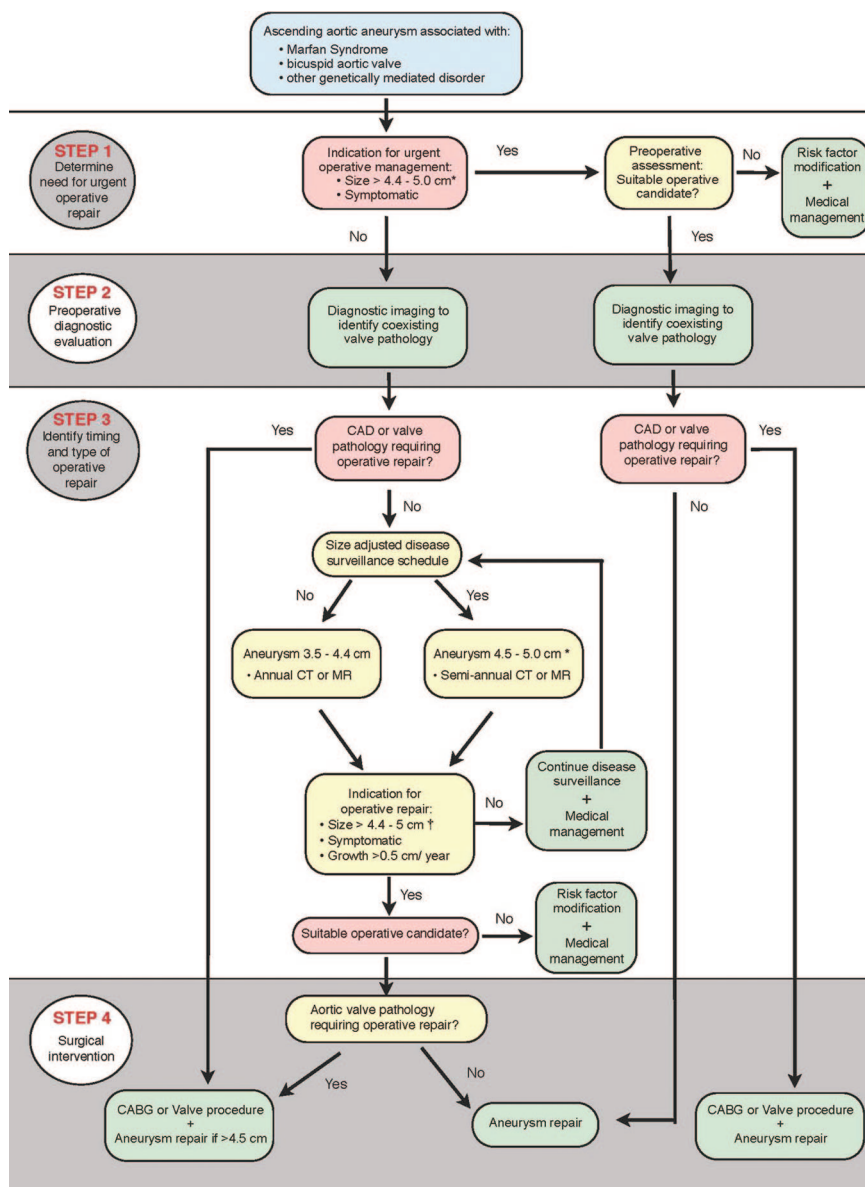


Figure 13. Ascending aortic aneurysms associated with genetic disorder. CABG indicates coronary artery bypass graft surgery; CAD, coronary artery disease; CT, computed tomographic imaging; and MR, magnetic resonance imaging. *Depends on specific genetic condition. †See Recommendations for Asymptomatic Patients With Ascending Aortic Aneurysm (Section 15), and Recommendations for Bicuspid Aortic Valve and Associated Congenital Variants in Adults (Section 7).

thoracic aorta, usually with the elephant trunk procedure (see Figure 14).^{133–135} (Level of Evidence: B)

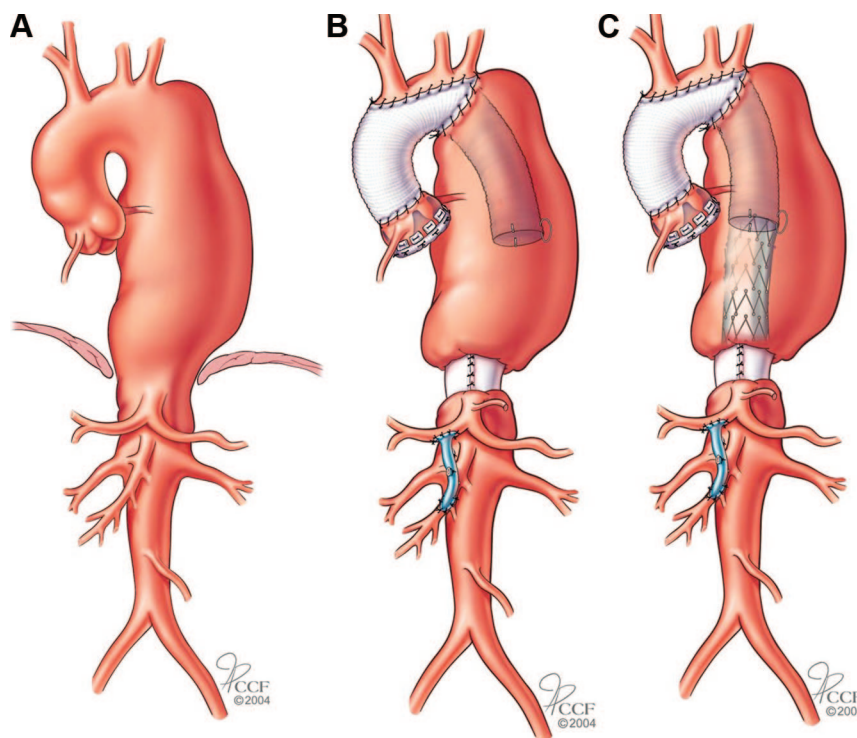
- For patients with low operative risk in whom an isolated degenerative or atherosclerotic aneurysm of the aortic arch is present, operative treatment is reasonable for asymptomatic patients when the diameter of the arch exceeds 5.5 cm.¹³⁶ (Level of Evidence: B)
- For patients with isolated aortic arch aneurysms less than 4.0 cm in diameter, it is reasonable to reimaging using computed tomography scanning or magnetic resonance imaging, at 12-month intervals, to detect enlargement of the aneurysm. (Level of Evidence: C)
- For patients with isolated aortic arch aneurysms 4.0 cm or greater in diameter, it is reasonable to reimaging using computed tomography scanning or magnetic resonance imaging, at 6-month intervals, to detect enlargement of the aneurysm. (Level of Evidence: C)

19. RECOMMENDATIONS FOR DESCENDING THORACIC AORTA AND THORACOABDOMINAL AORTIC ANEURYSMS

Class I

- 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.^{119,137,138} (Level of Evidence: B)
- 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^{119,139} (see Table 10). (Level of Evidence: B)
- For patients with thoracoabdominal aneurysms, in whom endovascular stent graft options are limited

Figure 14. Elephant trunk procedure. Panel A, Preoperative disease. Panel B, Stage I with replacement of the ascending aorta and arch with a Dacron graft, with the distal graft sutured circumferentially to the aorta distal to the left subclavian artery and the free end of the graft (“elephant trunk”) within the descending aneurysm. Panel C, Completion of the procedure using an endovascular stent graft attached proximally to the “elephant trunk” and the distal end secured to a Dacron graft cuff. Images reprinted with permission from the Cleveland Clinic Foundation.



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.¹¹⁹ (*Level of Evidence: 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.¹⁴⁰ (*Level of Evidence: B*)

Table 10. 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 et al.¹¹⁹

20. 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.^{40,141} (*Level of Evidence: 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. (*Level of Evidence: C*)
3. For all pregnant women with known aortic root or ascending aortic dilatation, monthly or bi-monthly echocardiographic measurements of the ascending aortic dimensions are recommended to detect aortic expansion until birth. (*Level of Evidence: C*)
4. For imaging of pregnant women with aortic arch, descending, or abdominal aortic dilatation, magnetic resonance imaging (without gadolinium) is recommended over computed tomography scanning to avoid exposing both the mother and fetus to ionizing radiation. Transesophageal echocardiogram is an option for imaging of the thoracic aorta. (*Level of Evidence: C*)
5. Pregnant women with aortic aneurysms should be delivered where cardiothoracic surgery is available. (*Level of Evidence: C*)

Class IIa

1. Fetal delivery via cesarean section is reasonable for patients with significant aortic enlargement, dissection, or severe aortic valve regurgitation.¹⁴¹ (*Level of Evidence: C*)

Class IIb

1. If progressive aortic dilatation and/or advancing aortic valve regurgitation are documented, prophylactic surgery may be considered.¹⁴² (*Level of Evidence: C*)

21. 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.¹⁴³ (*Level of Evidence: 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. (*Level of Evidence: C*)

22. PERIPROCEDURAL AND PERIOPERATIVE MANAGEMENT

Sections 22.1 to 22.6 list recommendations regarding the periprocedural and perioperative management of patients undergoing open surgical or thoracic aortic endograft procedures including strategies to preserve end-organ function. More detailed discussions are available in the full-text document.

22.1 Recommendations for Preoperative Evaluation

Class I

1. In preparation for surgery, imaging studies adequate to establish the extent of disease and the potential limits of the planned procedure are recommended. (*Level of Evidence: C*)
2. Patients with thoracic aortic disease requiring a surgical or catheter-based intervention who have symptoms or other findings of myocardial ischemia should undergo additional studies to determine the presence of significant coronary artery disease. (*Level of Evidence: C*)
3. Patients with unstable coronary syndromes and significant coronary artery disease should undergo revascularization prior to or at the time of thoracic aortic surgery or endovascular intervention with percutaneous coronary intervention or concomitant coronary artery bypass graft surgery. (*Level of Evidence: C*)

Class IIa

1. Additional testing is reasonable to quantitate the patient's comorbid states and develop a risk profile. These may include pulmonary function tests,

cardiac catheterization, aortography, 24-hour Holter monitoring, noninvasive carotid artery screening, brain imaging, echocardiography, and neurocognitive testing. (*Level of Evidence: C*)

2. For patients who are to undergo surgery for ascending or arch aortic disease, and who have clinically stable, but significant (flow limiting), coronary artery disease, it is reasonable to perform concomitant coronary artery bypass graft surgery. (*Level of Evidence: C*)

Class IIb

1. For patients who are to undergo surgery or endovascular intervention for descending thoracic aortic disease, and who have clinically stable, but significant (flow limiting), coronary artery disease, the benefits of coronary revascularization are not well established.^{144–146} (*Level of Evidence: B*)

22.2. Recommendations for Choice of Anesthetic and Monitoring Techniques

Class I

1. The choice of anesthetic techniques and agents and patient monitoring techniques should be tailored to individual patient needs to facilitate surgical and perfusion techniques and the monitoring of hemodynamics and organ function. (*Level of Evidence: C*)

Class IIa

1. Transesophageal echocardiography is reasonable in all open surgical repairs of the thoracic aorta, unless there are specific contraindications to its use. Transesophageal echocardiography is reasonable in endovascular thoracic aortic procedures for monitoring, procedural guidance, and/or endovascular graft leak detection.^{147–149} (*Level of Evidence: B*)
2. Motor or somatosensory evoked potential monitoring can be useful when the data will help to guide therapy. It is reasonable to base the decision to use neurophysiologic monitoring on individual patient needs, institutional resources, the urgency of the procedure, and the surgical and perfusion techniques to be employed in the open or endovascular thoracic aortic repair.^{150,151} (*Level of Evidence: B*)

Class III

1. Regional anesthetic techniques are not recommended in patients at risk of neuraxial hematoma formation due to thienopyridine antiplatelet therapy, low-molecular-weight heparins, or clinically significant anticoagulation.¹⁵² (*Level of Evidence: C*)
2. Routinely changing double-lumen endotracheal (endobronchial) tubes to single-lumen tubes at the end of surgical procedures complicated by significant upper airway edema or hemorrhage is not recommended. (*Level of Evidence: C*)

22.3. Recommendation for Transfusion Management and Anticoagulation in Thoracic Aortic Surgery

Class IIa

1. An algorithmic approach to transfusion, antifibrinolytic, and anticoagulation management is reasonable to use in both open and endovascular thoracic aortic repairs during the perioperative period. Institutional variations in coagulation testing capability and availability of transfusion products and other prothrombotic and antithrombotic agents are important considerations in defining such an approach.¹⁵³ (*Level of Evidence: C*)

22.4. 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.^{154–160} (*Level of Evidence: 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.^{161–184} (*Level of Evidence: 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.^{185–187} (*Level of Evidence: B*)

22.5. 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.^{188–190} (*Level of Evidence: 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.^{138,191–193} (*Level of Evidence: B*)

2. Moderate systemic hypothermia is reasonable for protection of the spinal cord during open repairs of the descending thoracic aorta.¹⁹⁴ (*Level of Evidence: 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.^{193,195–197} (*Level of Evidence: 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.^{150,198–200} (*Level of Evidence: B*)

22.6. Recommendations for Renal Protection During Descending Aortic Open Surgical and Endovascular Repairs

Class IIb

1. Preoperative hydration and intraoperative mannitol administration may be reasonable strategies for preservation of renal function in open repairs of the descending aorta. (*Level of Evidence: C*)
2. During thoracoabdominal or descending aortic repairs with exposure of the renal arteries, renal protection by either cold crystalloid or blood perfusion may be considered.^{201–203} (*Level of Evidence: B*)

Class III

1. Furosemide, mannitol, or dopamine should not be given solely for the purpose of renal protection in descending aortic repairs.^{204,205} (*Level of Evidence: B*)

23. RECOMMENDATIONS FOR SURVEILLANCE OF THORACIC AORTIC DISEASE OR PREVIOUSLY REPAIRED PATIENTS

Class IIa

1. Computed tomography 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.⁴⁰ (*Level of Evidence: C*)
2. Computed tomography 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. (*Level of Evidence: C*)

Table 11. Suggested Follow-Up of Aortic Pathologies After Repair or Treatment

Pathology	Interval	Study
Acute dissection	Before discharge, 1 month, 6 months, 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 aortic arch	Before discharge, yearly	TTE
Thoracic aortic stent	Before discharge, 1 month, 2 months, 6 months, yearly Or 30 days*	CT or MR, chest plus abdomen CXR, CT, chest plus abdomen
Acute IMH/PAU	Before discharge, 1 month, 3 months, 6 months, yearly	CT or MR, chest plus abdomen

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.

*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 pre-discharge 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 et al.²⁰⁸

- 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. (Level of Evidence: C)
- If a thoracic aortic aneurysm is only moderate in size and remains relatively stable over time, magnetic resonance imaging instead of computed tomography scanning is reasonable to minimize the patient's radiation exposure. (Level of Evidence: C)
- Surveillance imaging similar to classic aortic dissection is reasonable in patients with intramural hematoma. (Level of Evidence: C)

The mean rate of growth for all thoracic aortic aneurysms is approximately 1 mm/y, but that growth rate increases with increasing aneurysm diameter. Growth rates tend to be faster for aneurysms involving the descending versus the ascending aorta, for dissected versus nondissected aortas, for those with Marfan syndrome versus those without,²⁰⁶ and for those with bicuspid versus tricuspid aortic valves.²⁰⁷ Table 11 notes suggested intervals for follow up.

24. RECOMMENDATION FOR EMPLOYMENT AND LIFESTYLE IN PATIENTS WITH THORACIC AORTIC DISEASE

Class IIa

- 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. (Level of Evidence: C)

Establishing clear lifestyle goals for patients with thoracic aortic disease is important in improving long-term health and reducing the risk of complications.

There are no outcomes data, and scant data of any variety for that matter, to indicate how much exercise is safe or beneficial for patients with thoracic aortic disease. However, aerobic exercise, sometimes referred to as dynamic exercise, is associated with only a modest increase in mean arterial pressure,²⁰⁹ and AoD rarely occurs during aerobic exercise. Consequently, most experts believe that aerobic exercise, particularly when heart rate and blood pressure are well controlled with medications, is beneficial overall. Nevertheless, if patients wish to engage in vigorous aerobic exercise, such as running or basketball, one might consider performing a symptom limited stress test to ensure that the patient does not have a hypertensive response to exercise.

Conversely, with isometric exercise, there is a significant increase in mean arterial pressure. When the Valsalva maneuver is used for the lifting of heavy weights, there is a superimposed increase in intrathoracic pressure, followed by a dramatic increase in systemic arterial pressure,²⁰⁹ with systolic pressures reaching 300 mm Hg or more.²¹⁰ As a result, most experts believe that heavy weight lifting or competitive athletics involving isometric exercise may trigger AoD and/or rupture and that such activities should be avoided.²¹¹ Working with patients on an individualized basis to streamline these goals based on insufficient data can be challenging. For patients who are very much interested in maintaining some sort of weight lifting program, choosing sets of repetitive light weights appears to make more sense than permitting heavy weight lifting.²⁰⁹

25. TUMORS OF THE THORACIC AORTA

Neoplasms of the thoracic aorta are usually secondary and related to contiguous spread of adjacent primary malignancies, particularly lung and adjacent primary malignancies or subsequent metastases, particularly lung and esophagus.^{212–215} Primary neoplasms of the thoracic aorta are rare.²¹³ Metastatic disease is often demonstrated at the time of diagnosis of primary aortic neoplasms. Symptoms may include malaise, fatigue, weight loss and nausea or the occurrence of distal arterial embolization (with histopathologic examination showing neoplasm, or identified by imaging techniques during a search for an embolic source).^{216–218} AoD may originate in the area of the neoplasm or the aortic occlusion.²¹⁹ Resection and reconstruction of the segment of aorta containing the neoplasm have been described, but because most patients present with metastatic disease, overall prognosis is poor.²²⁰

26. RECOMMENDATIONS FOR QUALITY ASSESSMENT AND IMPROVEMENT FOR THORACIC AORTIC DISEASE

Class I

- Hospitals that provide regional care for patients with acute sequelae of thoracic aortic disease (eg, procedures for thoracic aortic dissection and rupture) should participate in standardized quality assessment and improvement activities, including thoracic aortic disease registries. Such activities should include periodic measurement and regional/national

interfacility comparisons of thoracic aortic disease-related procedural volumes, complications and risk-adjusted mortality rates. (*Level of Evidence: C*)

2. Hospitals that provide regional care for patients with acute sequelae of thoracic aortic disease (eg, procedures for thoracic aortic dissection and rupture) should facilitate and coordinate standardized quality assessment and improvement activities with transferring facilities and emergency medical services teams. Such activities might include:
 - a. cooperative joint facility meetings to discuss opportunities for quality improvement and
 - b. interfacility and emergency medical services team comparisons of pretransfer care based on available outcome data and future performance measures developed in accordance with this guideline. (*Level of Evidence: C*)

Patients with acute aortic syndromes may require transfer to specialized institutions. Ideally, the communications between institutions will completely and accurately portray the condition of the patient including items listed in Table 12.

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Table 12. Standardized Transferring Facility Assessment, Communication, and Documentation for the Following Domains

- Blood pressure control for hypertension
- Heart rate control for tachycardia
- Hemodynamic instability
- Blood volume
- Cardiac ischemia
- Neurologic ischemia
- Renal function
- Mesenteric ischemia
- Peripheral arterial pulses and perfusion
- Activation of receiving team
- Imaging expectations and communications
- Timeliness and efficiency
- EMS characteristics of transferring facility, including requisite personnel, requisite in-transport equipment, including catastrophic resuscitation capabilities, in-transfer contingency planning, weather conditions, estimated transfer time, etc.

EMS indicates emergency medical services.

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DISCLOSURES

Appendix 1. Author Relationships With Industry and Other Entities—2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients with Thoracic Aortic Disease

Committee member	Employment	Consultant	Speaker	Ownership/ partnership/ principal	Research	Institutional, organizational or other financial benefit	Expert witness
Loren F. Hiratzka, <i>Chair</i>	Cardiac, Vascular & Thoracic Surgeons Inc. and TriHealth Inc.—Medical Director, Cardiac Surgery	None	None	None	None	None	● 2007; Defense; Aortic Dissection
George L. Bakris	University of Chicago Medical Center—Professor of Medicine; Director, Hypertension Center	● Abbott ● Boehringer Ingelheim ● Bristol-Myers Squibb/Sanofi-aventis ● Forest Laboratories ● GlaxoSmithKline ● Merck	● Forest Laboratories ● GlaxoSmithKline ● Merck ● Novartis	None	● Forest Laboratories ● GlaxoSmithKline ● Myogen ● National Institutes of Health (NIDDK/NHLBI)	None	None
Joshua A. Beckman	Brigham & Women’s Hospital—Director, Cardiovascular Fellows Program	● Bristol-Myers Squibb ● Sanofi-aventis	● Bristol-Myers Squibb ● GlaxoSmithKline ● Merck ● Sanofi-aventis	None	None	None	None
Robert M. Bersin	Seattle Cardiology—Director, Endovascular Services & Clinical Research	● Abbott Vascular ● Boston Scientific ● Bristol-Myers Squibb ● Cordis Endovascular ● Eli Lilly ● EV3 ● ReVascular Therapeutics ● Sanofi-aventis ● Vascular Solutions ● W.L. Gore	● Boston Scientific ● Bristol-Myers Squibb ● Daiichi Sankyo ● Eli Lilly ● Sanofi-aventis ● The Medicines Company	● Vascular Solutions	● Boston Scientific	● Boston Scientific ● Cordis Endovascular ● Vascular Solutions	● Expert witness in a case involving iatrogenic type B dissection
Vincent F. Carr	Uniformed Services University of Health Science—Professor of Medicine	None	None	None	None	None	None
Donald E. Casey, Jr	Atlantic Health—Vice President of Quality & Chief Medical Officer; Associate Professor of Medicine, Mount Sinai School of Medicine	None	None	None	None	None	None
Kim A. Eagle	University of Michigan Health System—Albion Walter Professor of Internal Medicine; Clinical Director, Cardiovascular Center	● NHLBI ● Robert Wood Johnson Foundation ● Sanofi-aventis	None	None	● Blue Cross/Blue Shield ● Bristol-Myers Squibb ● National Institutes of Health ● Pfizer	None	None
Luke K. Hermann	Mount Sinai Medical Center—Assistant Professor of Emergency Medicine; Director, Chest Pain Unit	None	None	None	None	None	None
Eric M. Isselbacher	Massachusetts General Hospital—Associate Professor of Medicine, Harvard Medical School; Co-Director, Thoracic Aortic Center	None	None	None	None	None	● 2007; Plaintiff; Aortic Dissection
Ella A. Kazerooni	University of Michigan Health System—Professor of Medicine; Director, Cardiothoracic Radiology	● GE Healthcare ● Vital Images	None	None	None	● GERRAF (GE Radiology Research Fellowship)	None

(Continued)

Appendix 1. Continued

Committee member	Employment	Consultant	Speaker	Ownership/ partnership/ principal	Research	Institutional, organizational or other financial benefit	Expert witness
Nicholas T. Kouchoukos	Missouri Baptist Medical Center—Cardiovascular Surgeon	• Edwards Lifesciences	None	None	None	None	• 2006; Defense; Aortic Dissection
Bruce W. Lytle	The Cleveland Clinic—Chair, Heart and Vascular Institute	None	None	None	None	None	None
Dianna M. Milewicz	University of Texas Southwestern Medical Center—President George H.W. Bush Chair in Cardiovascular Medicine; Professor & Director, Division of Medical Genetics	None	None	None	• Doris Duke Foundation • Genetech • National Institutes of Health • Vivian Smith Foundation	None	None
David L. Reich	Mount Sinai Medical Center—Professor & Chair, Department of Anesthesiology	None	None	None	None	None	None
Souvik Sen	University of South Carolina School of Medicine—Professor and Chair, Department of Neurology	• Coaxia	• Boehringer Ingelheim • Bristol-Myers Squibb • Pfizer • Sanofi-aventis	None	• American Heart Association • Genetech • Sanofi-aventis	None	None
Julie A. Shinn	Stanford University School of Medicine—Cardiovascular Clinical Nurse Specialist	None	None	None	None	None	None
Lars G. Svensson	The Cleveland Clinic—Director, The Center for Aortic Surgery; Director, Marfan Syndrome and Collective Tissue Disorder Clinic	None	None	None	• Edwards Lifesciences • Evalve	None	None
David M. Williams	University of Michigan Health System—Professor, Department of Radiology; Director, Interventional Radiology	• W.L. Gore	None	None	• W.L. Gore • Medtronic	None	• 2000; Defense; Failure to diagnose and treat mesenteric ischemia with aortic dissection • 2009; Defense; Failure to diagnose and treat mesenteric ischemia with aortic dissection

NHLBI indicates National Heart, Lung, and Blood Institute; NIDDK, National Institute of Diabetes and Digestive and Kidney Diseases.

This table represents the relevant relationships of committee members with industry and other entities that were reported orally at the initial writing committee meeting and updated in conjunction with all meetings and conference calls of the writing committee during the document development process. It does not necessarily reflect relationships with industry at the time of publication. A person is deemed to have a significant interest in a business if the interest represents ownership of 5% or more of the voting stock or share of the business entity, or ownership of \$10 000 or more of the fair market value of the business entity; or if funds received by the person from the business entity exceed 5% of the person's gross income for the previous year. A relationship is considered to be modest if it is less than significant under the preceding definition. Relationships noted in this table are modest unless otherwise noted.

*Significant (greater than \$10 000) relationship.

Appendix 2. Reviewer Relationships With Industry and Other Entities—2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients with Thoracic Aortic Disease

Peer reviewer	Representation	Consultant	Speaker	Ownership/ partnership/ principal	Research	Institutional, organizational or other financial benefit	Expert witness
Amjad Almahameed	Official Reviewer—Society for Vascular Medicine	None	None	None	None	None	None
Richard A. Bernstein	Official Reviewer—American Stroke Association	None	None	None	None	None	None
Christopher E. Buller	Official Reviewer—ACCF/AHA Task Force Lead Reviewer	None	None	None	None	None	None
Albert T. Cheung	Official Reviewer—Society of Cardiovascular Anesthesiologists	<ul style="list-style-type: none"> • EKR Therapeutics • The Medicines Company • Neuralstem • Schering Plough 	<ul style="list-style-type: none"> • EKR Therapeutics • The Medicines Company • PDL Biopharm* 	None	<ul style="list-style-type: none"> • The Medicines Company* • Neuralstem* 	None	None
Michael D. Dake	Official Reviewer—Society of Interventional Radiologists	<ul style="list-style-type: none"> • W.L. Gore* • Medtronic 	<ul style="list-style-type: none"> • Cook 	None	<ul style="list-style-type: none"> • Cook • W.L. Gore* • Medtronic 	None	None
Antionette S. Gomes	Official Reviewer—AHA Cardiovascular Surgery and Anesthesia Committee	None	None	None	None	None	None
Robert A. Guyton	Official Reviewer—ACCF Board of Trustees	<ul style="list-style-type: none"> • Medtronic 	None	None	None	None	None
Clifford J. Kavinsky	Official Reviewer—Society for Cardiovascular Angiography and Interventions	None	None	None	<ul style="list-style-type: none"> • Possis Corp. 	None	None
Scott Kinlay	Official Reviewer—Society for Vascular Medicine	None	<ul style="list-style-type: none"> • Merck • Pfizer 	None	<ul style="list-style-type: none"> • Pfizer 	None	None
Richard J. Kovacs	Official Reviewer—ACCF Board of Governors	None	None	None	None	None	None
Christine Mora Mangano	Official Reviewer—Society of Cardiovascular Anesthesiologists	None	None	None	None	None	None
Steven R. Messé	Official Reviewer—American Stroke Association	None	<ul style="list-style-type: none"> • Boehringer Ingelheim 	None	<ul style="list-style-type: none"> • American Heart Association* 	None	None
Eric Roselli	Official Reviewer—Society of Thoracic Surgeons	<ul style="list-style-type: none"> • Medtronic • Vascutek 	None	None	<ul style="list-style-type: none"> • Cook 	None	None
Geoff D. Rubin	Official Reviewer—American College of Radiology	<ul style="list-style-type: none"> • Fovia 	<ul style="list-style-type: none"> • Bracco 	<ul style="list-style-type: none"> • TeraRecon 	<ul style="list-style-type: none"> • Biosense-Webster* 	None	None
Frank J. Rybicki	Official Reviewer—American College of Radiology	<ul style="list-style-type: none"> • Bracco • Siemens Medical • Toshiba Medical Systems* • Vital Images 	<ul style="list-style-type: none"> • Bracco • Siemens Medical • Toshiba Medical Systems* • Vital Images 	<ul style="list-style-type: none"> • Siemens Medical 	<ul style="list-style-type: none"> • Bracco • Toshiba Medical Systems* 	None	None
Thoralf M. Sundt	Official Reviewer—American Association for Thoracic Surgery	None	None	None	<ul style="list-style-type: none"> • Bolton Medical 	<ul style="list-style-type: none"> • Atricure • Bolton Medical 	None
Richard D. White	Official Reviewer—AHA Peripheral Vascular Disease Council	None	None	None	None	<ul style="list-style-type: none"> • Jarvik Heart • Medtronic • Sorin Group/Carbomedics • St. Jude Medical • Thoratec Corporation • Ventracor • W.L. Gore 	None
James P. Zidar	Official Reviewer—Society for Cardiovascular Angiography and Interventions	<ul style="list-style-type: none"> • Abbott Vascular • Cordis* • Medtronic Vascular 	<ul style="list-style-type: none"> • Abbott Vascular • Cordis* • Medtronic Vascular 	None	<ul style="list-style-type: none"> • Abbott Vascular • Cordis* • Medtronic Vascular 	None	None

(Continued)

Appendix 2. Continued

Peer reviewer	Representation	Consultant	Speaker	Ownership/ partnership/ principal	Research	Institutional, organizational or other financial benefit	Expert witness
Wyatt Decker	Organizational Reviewer—American College of Emergency Physicians	None	None	None	None	None	None
Josh M. Kosowsky	Organizational Reviewer—American College of Emergency Physicians	None	None	None	None	None	None
Emile Mohler	Organizational Reviewer—American College of Physicians	None	None	None	None	None	None
James Burke	Content Reviewer—ACCF Catherization Committee	None	None	None	None	None	None
Edward P. Chen	Content Reviewer	None	None	None	None	None	None
Mark A. Creager	Content Reviewer— ACCF/AHA Task Force on Practice Guidelines	None	None	None	None	None	None
Jose G. Diez	Content Reviewer—ACCF Catherization Committee	● Sanofi-aventis	None	None	None	None	None
John A. Eleftheriades	Content Reviewer	● Baxter	None	● Coolspine	● Celera Diagnostics	None	● 2006; Plaintiff; Aortic Dissection*
D. Craig Miller	Content Reviewer	● Medtronic	● St. Jude Medical	None	● NHLBI ● Stanford PARTNER Trial	None	None
Rick Nishimura	Content Reviewer— ACCF/AHA Task Force on Practice Guidelines	None	None	None	None	None	None
Patrick T. O'Gara	Content Reviewer	None	None	None	None	None	None
Carlos Ruiz	Content Reviewer—ACCF Interventional Council	None	None	None	None	None	None

ACCF indicates American College of Cardiology Foundation; AHA, American Heart Association; and NHLBI, National Heart, Lung, and Blood Institute.

This table represents the relevant relationships with industry and other entities that were disclosed at the time of peer review. It does not necessarily reflect relationships with industry at the time of publication. A person is deemed to have a significant interest in a business if the interest represents ownership of 5% or more of the voting stock or share of the business entity, or ownership of \$10 000 or more of the fair market value of the business entity; or if funds received by the person from the business entity exceed 5% of the person's gross income for the previous year. A relationship is considered to be modest if it is less than significant under the preceding definition. Relationships in this table are modest unless otherwise noted. Names are listed in alphabetical order within each category of review.

*Significant (greater than \$10 000) relationship. ■■

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An Evolving Role of Anesthesiologists in the Management of Thoracic Aortic Diseases

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Thoracic aortic diseases are a spectrum of medical conditions for which surgery is the definitive treatment. For that reason, anesthesiologists are likely to encounter patients with thoracic aortic diseases and have an important role in their care. The importance of the anesthesiologist was recognized in the 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM guidelines for the diagnosis and management of patients with thoracic aortic disease appearing in this issue of the journal and originally published in *Circulation*.¹ In this first iteration, The Society of Cardiovascular Anesthesiologists (SCA) and 9 other medical societies worked together to create a multidisciplinary set of guidelines for screening, diagnosis, and treatment of patients with thoracic aortic diseases. Together, we believe that guidelines are most valuable to clinicians when they are disease focused rather than specialty focused or intervention focused.

By providing detailed, accessible guidelines for the detection, diagnosis, and management of thoracic aortic diseases, the American College of Cardiology Foundation (ACCF) and American Heart Association (AHA) also addressed the publicity generated by the death of John Ritter, a well known actor who died on September 11, 2003 of an acute aortic dissection. After his death, John Ritter's widow, also an actress, filed a wrongful death lawsuit against the physicians who had failed to diagnose and immediately treat her husband's condition. Although she did not win the lawsuit, she stated that the case was important because it brought together experts and raised public awareness about aortic disease. "I don't know what you call it in medicine, but in acting or art, it would be called a master class. The most brilliant minds in the aorta business were there."^{*}

In the same year, beginning in January 2003, *The Wall Street Journal* published a series of articles to bring attention to the incidence of aortic disease and the absence of any organized effort on a national level to identify and treat

patients with these conditions.[†] These articles, by Kevin Helliker and Thomas Burton, had provocative titles such as, "A Death Sentence You Can Avoid," "Ordering an Autopsy Could Save Your Life," and "Knowledge Gap. Medical Ignorance Contributes to the Toll from Aortic Illness. Many Doctors Don't Realize Aneurysms Are Treatable; a Paucity of Experts." From a physician's perspective, the articles were especially haunting. Kevin Helliker, an avid triathlete, described vividly in first hand what it was like to be diagnosed with a thoracic aortic aneurysm at the age of 43 years and the inevitable lifestyle changes and decisions that he will have to face. In 2004, the Pulitzer Prize for Explanatory Reporting was awarded to Kevin Helliker and Thomas Burton "for their groundbreaking examination of aneurysms, an often overlooked medical condition that kills thousands of Americans each year." One such American, Michael DeBakey, the cardiac surgeon who pioneered the use of the Dacron vascular graft for aortic repair and whose name is used in the nomenclature for classifying aortic dissections, almost died from an aortic dissection in 2005. This publicity, together with the publication of the guidelines from the ACCF and AHA, will increase the public's expectations and the responsibilities of health care providers for diagnosing and managing patients with thoracic aortic diseases.

Improved screening and early diagnosis of thoracic aortic diseases would provide little benefit if the prospects for surgical repair were only marginally better than the consequences of the disease itself. Undertaking operations on the thoracic aorta, the conduit for blood to the entire body, is a serious commitment with inherent risks. Strategies to protect the heart, brain, spinal cord, and mesenteric organs from ischemic injury are necessary to ensure successful operations. Fortunately, cardiothoracic surgeons together with anesthesiologists have accomplished much to improve the safety and outcome of these operations. Some examples of specialized organ-protection strategies advocated by the guidelines include: (a) deep hypothermic circulatory arrest, antegrade cerebral perfusion, and retrograde cerebral perfusion alone or in combination to minimize brain injury (section 14.5.1); (b) cerebrospinal fluid drainage and spinal cord perfusion pressure optimization

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[†]Pulitzer Prize-Winning Articles for Explanatory Journalism. April 5, 2004. Available at: <http://online.wsj.com/public/resources/documents/PulitzerKH04052004.htm>. Accessed April 20, 2010.

to decrease the risk of spinal cord ischemic injury in patients at risk (section 14.5.2); and (c) motor or somatosensory evoked potential monitoring to detect spinal cord ischemia, guide reimplantation of intercostal arteries, and guide hemodynamic optimization to prevent or treat spinal cord ischemia (sections 14.2 and 14.5.2). In addition to organ-protection strategies, new approaches, such as thoracic endovascular aortic repair and hybrid repairs using both open and endovascular techniques, have shown promise in decreasing the risk of operations.

Despite many advances in the management of thoracic aortic diseases, authors of the guidelines were careful to point out that many of the recommendations are supported by only limited studies, expert opinion, or specific institutional experience (Level of Evidence B or C). Limited institutional case experiences, the heterogeneous patient population that is affected by these diseases, and regional variability in surgical and anesthetic practices all pose serious challenges to investigators trying to accumulate the necessary data to generate evidenced-based guidelines. Nevertheless, the guidelines' contributors attempted to include descriptions of all techniques used in contemporary practice that are of potential benefit. The contributors acknowledge the limitations of the recommendations in the guidelines by rating both the strength of the recommendations and the strength of the evidence supporting them. For this reason, many of the recommendations constitute a range of acceptable approaches rather than a "standard of care." Many of the recommendations, particularly those involving periprocedural and perioperative care, are qualified by the statement, "Institutional experience is an important factor in selecting these techniques." The field of thoracic aortic surgery and anesthesia continues to evolve with ample opportunity for further progress.

The importance of imaging in thoracic aortic disease is a major emphasis of the guidelines. Imaging is important, because thoracic aortic diseases are manifested by structural changes that make imaging essential for diagnosis and management. The widespread availability of transesophageal echocardiography (TEE) and increased expertise with intraoperative TEE has affected the diagnosis and management of acute aortic syndromes. For this reason, TEE, together with computed tomography and magnetic resonance imaging, were assigned a Class I recommendation, meaning that the procedures *should* be performed for urgent and definitive imaging of the aorta to identify or exclude thoracic aortic dissection in patients at high risk for the disease by initial screening (section 8.6.1.3). TEE also received a Class IIa recommendation for all open thoracic aortic repairs, meaning that its benefits exceed its risks. TEE was also judged reasonable for use in endovascular procedures (section 14.2). The guidelines provided criteria for

elective repair based on aortic diameters for Loeys-Dietz syndrome (section 5.1.2), Marfan syndrome (section 5.1.1), ascending aortic aneurysms (section 9.2.2.1.1), bicuspid aortic valve with dilated ascending aorta (section 9.2.2.1.1), and descending thoracic or thoracoabdominal aortic aneurysms (section 9.2.2.3.1). These criteria will serve as valuable references for training programs, referring physicians, surgeons, and anesthesiologists using intraoperative TEE for surgical decision-making. In addition, the guidelines specify precisely how and where measurements should be made in the thoracic aorta. For example, measurements performed using TEE measure the internal diameter of the aorta whereas computed tomography measures the external diameter of the aorta (section 4.1). Establishing uniform reporting standards based on precise definitions combined with disease registries and systematic quality-improvement efforts will increase the prognostic value of these tests in the future.

Publication of the thoracic aortic disease guidelines is the latest in a series of multidisciplinary guidelines on which the SCA has collaborated with the American College of Cardiology, AHA, the American Society of Anesthesiologists, the Society of Thoracic Surgeons, and the American Society of Echocardiography.† The SCA leadership believes that fostering professional dialog and continued collaboration among medical societies in areas of common interest is an important direction for the subspecialty. The SCA applauds the ACCF and the AHA for finding common ground among participating subspecialties in the creation of this set of guidelines to advance the care of patients with thoracic aortic diseases. ■

DISCLOSURE

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