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

Developed in partnership with 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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Citation

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The full-text guidelines are also available on the following Web sites:
ACC (www.acc.org) and,
AHA (www.americanheart.org)

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Classification of Recommendations

Class I	Class IIa	Class IIb	Class III
Benefit >>> Risk Procedure/ Treatment **SHOULD** be performed/ administered	*Benefit >> Risk* *Additional studies with focused objectives needed* **IT IS REASONABLE** to perform procedure/administer treatment	*Benefit ≥ Risk* *Additional studies with broad objectives needed; Additional registry data would be helpful* Procedure/Treatment **MAY BE CONSIDERED**	*Risk ≥ Benefit* *No additional studies needed* Procedure/Treatment should **NOT** be performed/administered **SINCE IT IS NOT HELPFUL AND MAY BE HARMFUL**

Alternative Phrasing:

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

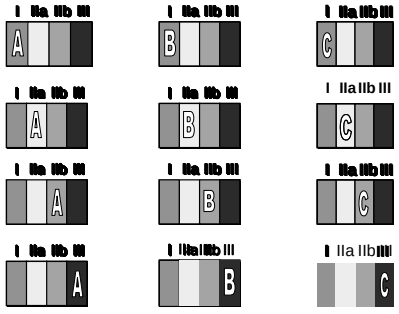
Applying Classification of Recommendations and Level of Evidence

Class I	Class IIa	Class IIb	Class III
Benefit >>> Risk Procedure/ Treatment **SHOULD** be performed/ administered	*Benefit >> Risk* *Additional studies with focused objectives needed* **IT IS REASONABLE** to perform procedure/administer treatment	*Benefit ≥ Risk* *Additional studies with broad objectives needed; Additional registry data would be helpful* Procedure/Treatment **MAY BE CONSIDERED**	*Risk ≥ Benefit* *No additional studies needed* Procedure/Treatment should **NOT** be performed/administered **SINCE IT IS NOT HELPFUL AND MAY BE HARMFUL**

Level of Evidence:

Level A:	Data derived from multiple randomized clinical trials or meta-analyses Multiple populations evaluated;
Level B:	Data derived from a single randomized trial or nonrandomized studies Limited populations evaluated
Level C:	Only consensus of experts opinion, case studies, or standard of care Very limited populations evaluated

Icons representing the Classification and Evidence Levels for Recommendations



Critical Issues for Thoracic Aortic Diseases

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

- Thoracic aortic diseases (TADs) are usually asymptomatic and not easily detectable until an acute and often catastrophic complication occurs.
- The identification and treatment of stable patients at risk for acute and catastrophic disease presentations (eg, thoracic aortic dissection (AoD) and thoracic aneurysm rupture) prior to such an occurrence are paramount to eliminating the high morbidity and mortality associated with acute presentations.
- Imaging of the thoracic aorta 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 TAD. However, increased use of these technologies increases the potential risk associated with repeated radiation exposure, as well as contrast medium–related toxicity.
  - Imaging for asymptomatic patients at high risk based on history or associated diseases is expensive and not always covered by payers.

Critical Issues for TADs (continued)

- 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.
  - Widespread awareness of the varied and complex nature of TAD presentations has been lacking, especially for acute AoD.
- There is rapidly accumulating evidence that genetic alterations or mutations predispose some individuals to aortic diseases.
  - Identification of the genetic alterations leading to these aortic diseases has potential for early detection among at-risk individuals.
  - 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.

Guidelines for Thoracic Aortic Disease

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

Recommendations for Aortic Imaging Techniques to Determine the Presence and Progression of TAD

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 entitled “Essential Elements of Aortic Imaging Reports”).

For measurements taken by computed tomographic imaging or magnetic resonance imaging, the external diameter should be measured perpendicular to the axis of blood flow. For aortic root measurements, the widest diameter, typically at the mid-sinus level, should be used.

Recommendations for Imaging Techniques to Determine the Presence and Progression of TAD

For measurements taken by echocardiography, the internal diameter should be measured perpendicular to the axis of blood flow. For aortic root measurements, the widest diameter, typically at the mid-sinus level, should be used.

Abnormalities of aortic morphology should be recognized and reported separately even when aortic diameters are within normal limits.

Recommendations for Imaging Techniques to Determine the Presence and Progression of TAD

The finding of aortic dissection, aneurysm, traumatic injury and/or aortic rupture should be immediately communicated to the referring physician.

Techniques to minimize episodic and cumulative radiation exposure should be utilized whenever possible.

If clinical information is available, it can be useful to relate aortic diameter to the patient’s age and body size.

Technical Parameters for Computed Tomographic Imaging

Recommended details of CT technique includes the following:

CT angiographic acquisition using intravenous contrast delivered at rate of 3-5 mL/s using a power injector, followed by a saline chaser bolus.

Total contrast volume should be as low as possible (no more than 150 mL)

Recommended technical parameters for image acquisition:

- Slices thickness of 3 mm or less with a reconstruction interval of 50% or smaller than the slice thickness
- Tube rotation of 1 second or less
- 120-140 kVp; mA adjusted to patient size
- ECG gating particularly useful for AoD (Note: prospective triggering has lower radiation exposure than retrospective gating)
- Coverage: thoracic inlet to groin

2- and 3- dimensional reconstructions (e.g., multiplanar and curved multiplanar reformations) and volume rendering *may* augment interpretation and improve communication of findings, and are likely to play an important role in planning surgical or endovascular treatment approaches.

Essential Elements of Aortic Imaging Reports

The following table outlines specific qualitative and quantitative elements that are important to include in CT and MR 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 intramural hematoma (IMH), penetrating atherosclerotic ulcer (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.

Note: This is Table 5 in the full-text version of the TAD Guideline

Guidelines for Thoracic Aortic Disease

Recommendations for Genetic Syndromes Associated with Thoracic Aortic Aneurysms and Dissections

Recommendations for Genetic Syndromes Associated with Thoracic Aortic Aneurysms and Dissections

An echocardiogram is recommended at the time of diagnosis of Marfan syndrome to determine the aortic root and ascending aortic diameters and 6 months thereafter to determine the rate of enlargement of the aorta.

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.

Recommendations for Genetic Syndromes Associated with Thoracic Aortic Aneurysms and Dissections

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.

Loeys-Dietz patients should have yearly magnetic resonance imaging from the cerebrovascular circulation to the pelvis.

Loeys-Dietz Syndrome

- Autosominal dominant
- TGFBR1 and TGFBR2 gene mutation
- The main clinical characteristics include:
  - Widely spaced eyes (orbital hypertelorism)
  - Cleft palate or bifid uvula (a split in the tissue that hangs down in the back of the throat)
  - Aortic and arterial aneurysms/dissections with tortuosity (corkscrew structure) of the arteries.

Loeys-Dietz Syndrome

- Other findings can include :
  - Scoliosis
  - Indented or protruding chest wall (pectus excavatum or pectus carinatum)
  - Contractures of fingers and toes (camptodactyly)
  - Long fingers and lax joints
  - Club foot
  - Premature fusion of the skull bones (craniosynostosis)
  - Joint hypermobility
  - Congenital heart problems including patent ductus arteriosus (connection between the aorta and the lung circulation) and atrial septal defect (connection between heart chambers)
  - Translucency of the skin with velvety texture
  - Abnormal junction of the brain and medulla (Arnold-Chiari malformation)
  - Bicuspid aortic valves

Recommendations for Genetic Syndromes Associated with Thoracic Aortic Aneurysms and Dissections

I IIa IIb III

Patients with Turner syndrome should undergo imaging of the heart and aorta for evidence of bicuspid aortic valve, coarctation of the aorta, or dilatation of the ascending thoracic aorta. If initial imaging is normal and there are no risk factors for aortic dissection, repeat imaging should be performed every 5 to 10 years or if otherwise clinically indicated. If abnormalities exist, annual imaging or follow-up imaging should be done.

Recommendations for Genetic Syndromes Associated with Thoracic Aortic Aneurysms and Dissections

I IIa IIb III

It is reasonable to consider surgical repair of the aorta in all adult patients with Loeys-Dietz syndrome or a confirmed *TGFBR1* or *TGFBR2* mutation and an aortic diameter of 4.2 cm or greater by transesophageal echocardiogram (internal diameter) or 4.4 to 4.6 cm or greater by computed tomographic imaging and/or magnetic resonance imaging (external diameter).

I IIa IIb III

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.

Recommendations for Genetic Syndromes Associated with Thoracic Aortic Aneurysms and Dissections

I IIa IIb III

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.

I IIa IIb III

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.

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
TGFBR2 (transforming growth factor-beta receptor type 2) mutations	4%	•Thin, translucent skin •Arterial or aortic tortuosity •Aneurysm of arteries	Multiple aortic dissections documented at aortic diameters <5.0 cm
MYH11 (smooth muscle specific beta-myosin heavy chain) mutations	1%	•Patent ductus arteriosus	Patient with documented dissection at 4.5 cm
ACTA2 (actin, alpha 2, smooth muscle aorta) mutations	14%	•Livedo reticularis •Iris flocculi •Patent ductus arteriosus •Bicuspid aortic valve	Two of 13 patients with documented dissections <5.0 cm

Note: Table 6 in full-text version of TAD Guidelines

Genetic Syndromes Associated With Thoracic Aortic Aneurysm and Dissection

Genetic Syndrome Genetic	Common Clinical Features	Defect	Diagnostic Test	Comments on Aortic Disease
Marfan syndrome	Skeletal features (see text); Ectopia Lentis; Dural ectasia	*FBN1* 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	*TGFBR2* or *TGFBR1* 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	*COL3A1* mutations	DNA for sequencing Dermal fibroblasts for analysis of type 3 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

* The defective gene at a second locus for MFS is *TGFBR2* but the clinical phenotype as MFS is debated.
AoD = aortic dissection; *COL3A1*, type III collagen; *FBN1*, fibrillin 1; MFS, Marfan syndrome; *TGFBR1*, transforming growth factor-beta receptor type 1; and *TGFBR2*, transforming growth factor beta receptor type 2.

Note: Table 7 in full-text version of TAD Guidelines

Guidelines for Thoracic Aortic Disease

Recommendations for Familial Thoracic Aortic Aneurysm and Dissections

Recommendations for Familial Thoracic Aortic Aneurysm and Dissections

Aortic imaging is recommended for first degree relatives of patients with thoracic aortic aneurysm and/or dissection to identify those with asymptomatic disease.

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.

Recommendations for Familial Thoracic Aortic Aneurysm and Dissections

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.

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.

Recommendations for Familial Thoracic Aortic Aneurysm and Dissections

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.

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.

Guidelines for Thoracic Aortic Disease

Recommendations for Bicuspid Aortic Valve and Associated Congenital Variants in Adults

Recommendations for Bicuspid Aortic Valve and Associated Congenital Variants in Adults

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.

All patients with a bicuspid aortic valve should have both the aortic root and ascending thoracic aorta evaluated for evidence of aortic dilatation.

Guidelines for Thoracic Aortic Disease

Recommendations for Takayasu Arteritis and Giant Cell Arteritis

Recommendations for Takayasu Arteritis and Giant Cell Arteritis

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.

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.

Elective revascularization of patients with Takayasu arteritis and giant cell arteritis should be delayed until the acute inflammatory state is treated and quiescent.

Recommendations for Takayasu Arteritis and Giant Cell Arteritis

The initial evaluation of Takayasu arteritis or giant cell arteritis should include thoracic aorta and branch vessel computed tomographic imaging or magnetic resonance imaging to investigate the possibility of aneurysm or occlusive disease in these vessels.

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.

Guidelines for Thoracic Aortic Disease

Recommendations for Initial Evaluation and Management of Acute Thoracic Aortic Disease

Recommendations for Estimation of Pretest Risk of Thoracic Aortic Dissection

Providers should routinely evaluate any patient presenting with complaints that may represent acute thoracic aortic dissection to establish a pretest risk of disease that can then be used to guide diagnostic decisions. This process should include specific questions about:

- medical history,
- family history, and
- pain features.

This process should also include a focused examination to identify findings that are associated with aortic dissection (outlined in the next 3 slides).

Estimation of Pretest Risk of Thoracic Aortic Dissection

High Risk Conditions 1

- Marfan Syndrome
- Connective tissue disease*
- Family history of aortic disease
- Known aortic valve disease
- Recent aortic manipulation (surgical or catheter-based)
- Known thoracic aortic aneurysm
- Genetic conditions that predispose to AoD†

* 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*.

Estimation of Pretest Risk of Thoracic Aortic Dissection

High Risk Pain Features 2

Chest, back, or abdominal pain features described as pain that:

- is abrupt or instantaneous in onset.
- is severe in intensity.
- has a ripping, tearing, stabbing, or sharp quality.

Estimation of Pretest Risk of Thoracic Aortic Dissection

High Risk Examination Features 3

- Pulse deficit
- Systolic BP limb differential > 20mm Hg
- Focal neurologic deficit
- Murmur of aortic regurgitation (new or not known to be old and in conjunction with pain)

Recommendations for Estimation of Pretest Risk of Thoracic Aortic Dissection

Patients presenting with sudden onset of severe chest, back, and/or abdominal pain, particularly those less than 40 years of age, should be questioned about a history and examined for physical features of Marfan syndrome, Loeys-Dietz syndrome, vascular Ehlers-Danlos syndrome, Turner syndrome, or other connective tissue disorder associated with thoracic aortic disease.

Recommendations for Estimation of Pretest Risk of Thoracic Aortic Dissection

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.

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.

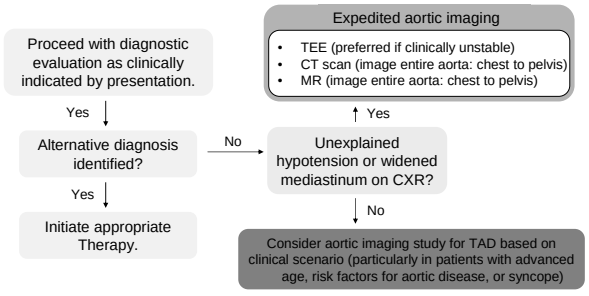
Recommendations for Estimation of Pretest Risk of Thoracic Aortic Dissection

In patients with suspected or confirmed aortic dissection who have experienced a syncopal episode, a focused examination should be performed to identify associated neurologic injury or the presence of pericardial tamponade.

All patients presenting with acute neurologic complaints should be questioned about the presence of chest, back, and/or abdominal pain and checked for peripheral pulse deficits as patients with dissection-related neurologic pathology are less likely to report thoracic pain than the typical aortic dissection patient.

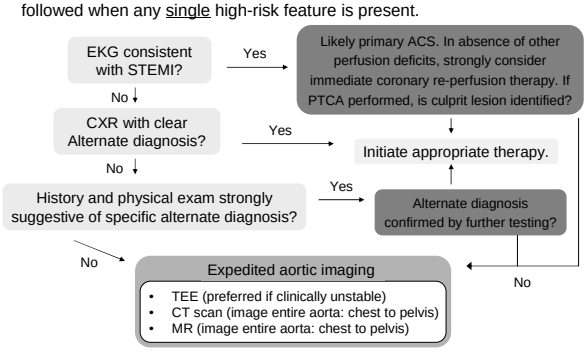
Risk-based Diagnostic Evaluation: Patients with Low Risk of TAD

Patients with no high-risk features of TAD present are considered at *low risk* for TAD. The following clinical steps are recommended for low-risk TAD patients:



Risk-based Diagnostic Evaluation: Patients with Intermediate Risk of TAD

The following steps for patients with *intermediate risk* of TAD should be followed when any single high-risk feature is present.



Risk-based Diagnostic Evaluation: Patients with High Risk of TAD

Patients at *high-risk* for TAD are those that present with at least 2 high-risk features (outlined in more detail in the following slides)

The recommended course of action for high-risk TAD patients is to seek immediate surgical consultation and arrange for expedited aortic imaging.

Expedited aortic imaging

- TEE (preferred if clinically unstable)
- CT scan (image entire aorta: chest to pelvis)
- MR (image entire aorta: chest to pelvis)

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

Note: Information on this slide is adapted from Table 9 in full-text version of TAD Guidelines

Risk Factors for Development of Thoracic Aortic Dissection (continued)

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

Note: Information on this slide is adapted from Table 9 in full-text version of TAD Guidelines

Risk Factors for Development of Thoracic Aortic Dissection (continued)

Conditions Associated With Aortic Media Abnormalities ***(continued)***

Inflammatory vasculitides

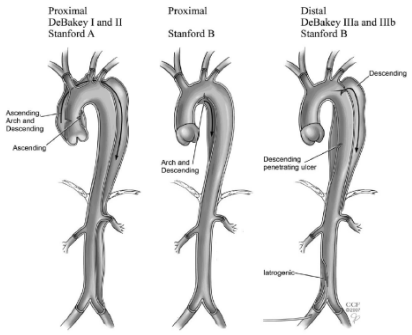
- Takayasu arteritis
- Giant cell arteritis
- Behçet arteritis

Other

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

Note: Information on this slide is adapted from Table 9 in full-text version of TAD Guidelines

Aortic Dissection Classification: DeBakey and Stanford Classifications



Note: Figure 20 in full-text version of TAD Guidelines. Reprinted with permission from The Cleveland Clinic Foundation.

Recommendations for Screening Tests

An electrocardiogram should be obtained on all patients who present with symptoms that may represent acute thoracic aortic dissection.

- 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.

Recommendations for Screening Tests (continued)

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.

- 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.
- 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.

Recommendations for Screening Tests (continued)

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

A negative chest x-ray should not delay definitive aortic imaging in patients determined to be high risk for aortic dissection by initial screening.

Recommendations for Diagnostic Imaging Studies

Selection of a specific imaging modality to identify or exclude aortic dissection should be based on patient variables and institutional capabilities, including immediate availability.

If a high clinical suspicion exists for acute aortic dissection but initial aortic imaging is negative, a second imaging study should be obtained.

Recommendations for Initial Management

Initial management of thoracic aortic dissection should be directed at decreasing aortic wall stress by controlling heart rate and blood pressure as follows:

a. In the absence of contraindications, intravenous beta blockade should be initiated and titrated to a target heart rate of 60 beats per minute or less.

b. In patients with clear contraindications to beta blockade, nondihydropyridine calcium channel–blocking agents should be used as an alternative for rate control.

Recommendations for Initial Management (continued)

c. If systolic blood pressures remain greater than 120mm 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.

d. Beta blockers should be used cautiously in the setting of acute aortic regurgitation because they will block the compensatory tachycardia.

Recommendations for Initial Management (continued)

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.

Acute AoD Management Pathway

STEP 1: Immediate post-diagnosis management and disposition considerations

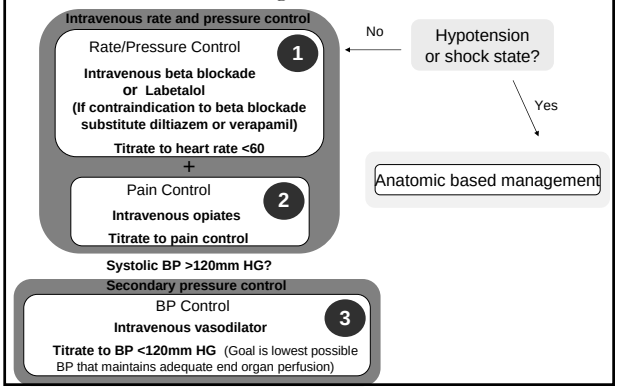
- Arrange for definitive management:
  - Appropriate surgical consultation
  - Inter-facility transfer if indicated based on institutional capabilities
- If transfer required, initiate aggressive medical management until transfer occurs.

Acute AoD Management Pathway

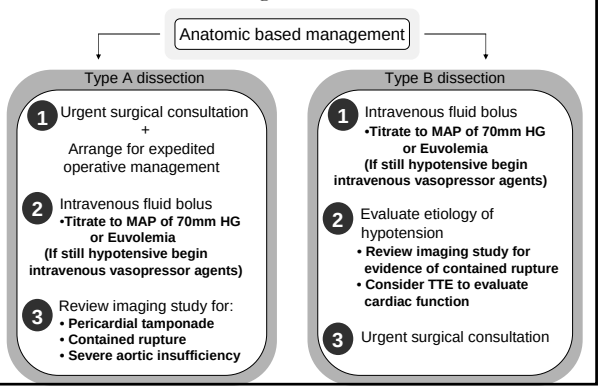
STEP 2: Initial management of aortic wall stress

- Obtain accurate blood pressure prior to beginning treatment.
- Measure in both arms.
- Base treatment goals on highest blood pressure reading.

Acute AoD Management Pathway
STEP 2: Initial management of aortic wall stress



Acute AoD Management Pathway
STEP 2: Initial management of aortic wall stress



Acute AoD Management Pathway

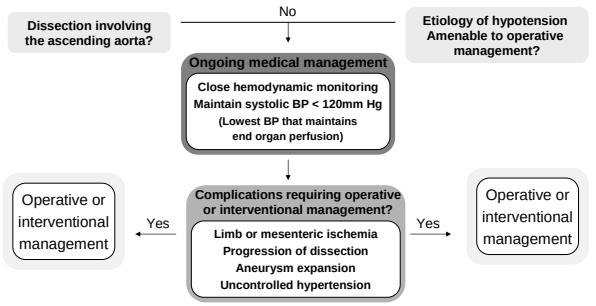
STEP 3: Definitive management

- Depending on the results from the pressure control or anatomic based management, continued treatment will involve either:
  - ongoing medical management, or
  - operative or interventional management.

Acute AoD Management Pathway
STEP 3: Definitive management

Based on results from intravenous rate and pressure control:

Based on results from anatomic based management:



Acute AoD Management Pathway

STEP 4: Transition to outpatient management and disease surveillance

- If no complications present requiring operative or interventional management, transition to:
  - Oral medications (beta blockade/ antihypertensives regimen)
  - Outpatient disease surveillance imaging

Note: For full algorithm, see Figure 26 in full-text version of TAD Guidelines

Recommendations for Definitive Management

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.

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.

Recommendations for Definitive Management (continued)

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).

Guidelines for Thoracic Aortic Disease

Recommendation for Surgical Intervention for Acute Thoracic Aortic Dissection

Recommendation for Surgical Intervention for Acute Thoracic Aortic Dissection

For patients with ascending thoracic aortic dissection, all aneurysmal aorta and the proximal extent of the dissection should be resected. A partially dissected aortic root may be repaired with aortic valve 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.

Acute Surgical Management Pathway for AoD

The following steps outline ascending TAD by imaging study.

STEP 1: Determine patient suitability for surgery

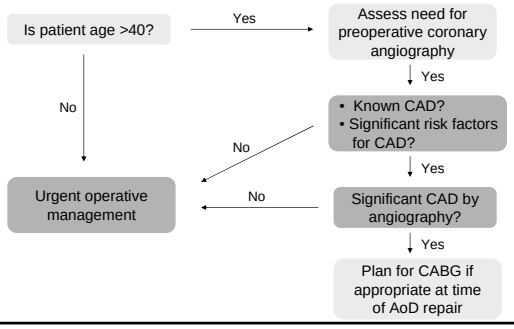
- If not suitable, begin medical management.

STEP 2: Determine stability for pre-op testing

- If not sufficiently stable, proceed with urgent operative management.

Acute Surgical Management Pathway for AoD

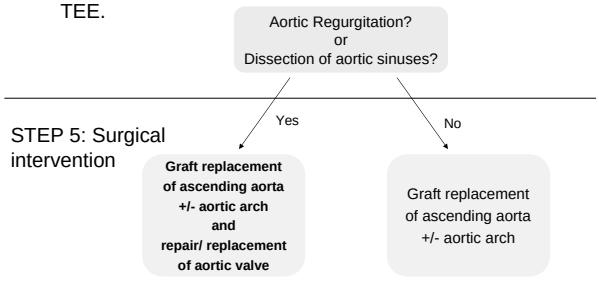
STEP 3: Determine likelihood of coexistent CAD



Acute Surgical Management Pathway for AoD

STEP 4: Intra-operative evaluation of aortic valve

- Perform intra-operative assessment of aortic valve by TEE.



STEP 5: Surgical intervention

Note: For full algorithm, see Figure 22 in full-text version of TAD Guidelines.

Guidelines for Thoracic Aortic Disease

Recommendation for Intramural Hematoma Without Intimal Defect

Recommendation for Intramural Hematoma Without Intimal Defect

It is reasonable to treat intramural hematoma similar to aortic dissection in the corresponding segment of the aorta.

Guidelines for Thoracic Aortic Disease

Recommendation for History and Physical Examination for Thoracic Aortic Disease

Recommendation for History and Physical Exam for TAD

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

Guidelines for Thoracic Aortic Disease

Recommendations for General Medical Treatment and Risk Factor Management for Patients with Thoracic Aortic Diseases

Recommendation for Medical Treatment of patients with TAD

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 to be surgical or stent graft candidates.

(See list of studies outlined in next 4 slides for related evidence base.)

Studies of Medical Treatment of Thoracic Aortic Aneurysm

Treatment: Beta blockers

Study	Results
Genoni M, Paul M, Jenni R, et al., 2001	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.· 1994	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).
Ladouceur M, Fermanian C, Lupoglazoff JM, et al., 2007	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.

Note: Adapted from Table 13 in full-text version of TAD Guidelines

Studies of Medical Treatment of Thoracic Aortic Aneurysm

Treatment: Angiotensin-converting enzyme inhibitors

Study	Results
Ahimastos AA, Aggarwal A, D'Orsa KM, et al., 2007	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/m2, *P*=0.01) and a significant reduction in ascending aortic diameter during diastole (0.4 vs –1.2 mm/m2, *P*<0.001), respectively.

Note: Adapted from Table 13 in full-text version of TAD Guidelines

Studies of Medical Treatment of Thoracic Aortic Aneurysm

Treatment: Angiotensin receptor blockers

Study	Results
Mochizuki S, Dahlof B, Shimizu M, et al., 2007	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., 2008	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 (±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.

Note: Adapted from Table 13 in full-text version of TAD Guidelines

Studies of Medical Treatment of Thoracic Aortic Aneurysm

Treatment: Statins

Study	Results
Diehm N, Decker G, Katzen B, et al., 2008	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).

Note: Adapted from Table 13 in full-text version of TAD Guidelines

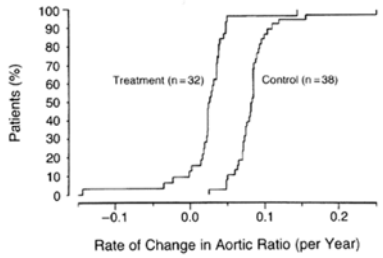
Recommendations for Blood Pressure Control

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.

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.

Beta Adrenergic Blockade Slows Aorta Growth in Marfan's

Randomized trial of propranolol in 70 adolescent and adult patients with classic Marfan's syndrome



SOURCE: Shores, J. *New England Journal of Medicine,* 1994; 330(19):1335-1341. Copyright © *1994 Massachusetts Medical Society. All rights reserved.*

Recommendations for Blood Pressure Control

For patients with thoracic aortic aneurysm, it is reasonable to reduce blood pressure with beta blockers and angiotensin-converting enzyme inhibitors or angiotensin receptor blockers to the lowest point patients can tolerate without adverse effects.

An angiotensin receptor blocker (losartan) is reasonable for patients with Marfan syndrome, to reduce the rate of aortic dilatation unless contraindicated.

Recommendation for Dyslipidemia

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 coronary heart disease at high risk for coronary ischemic events.

Recommendation for Smoking Cessation

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, buproprion, or varenicline) is useful, as is adopting a stepwise strategy aimed at smoking cessation (the 5 As are Ask, Advise, Assess, Assist, and Arrange).

Guidelines for Thoracic Aortic Disease

Recommendations for Asymptomatic Patients with Ascending Aortic Aneurysm

Recommendations for Asymptomatic Patients with Ascending Aortic Aneurysm

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.

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) to avoid acute dissection or rupture.

Recommendations for Asymptomatic Patients with Ascending Aortic Aneurysm

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.

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.

Recommendations for Asymptomatic Patients with Ascending Aortic Aneurysm

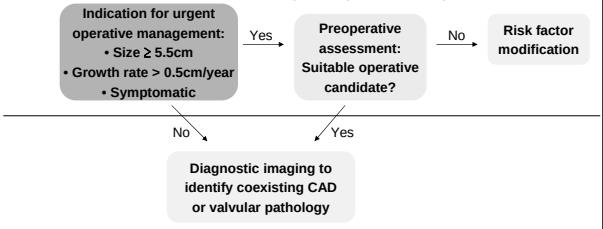
Elective aortic replacement is reasonable for patients with Marfan syndrome, other genetic diseases, or bicuspid aortic valves, when the ratio of maximal ascending or aortic root area (π r^2) in cm^2 divided by the patient's height in meters exceeds 10.

It is reasonable for patients with Loeys-Dietz syndrome or a confirmed *TGFBR1* or *TGFBR2* mutation to undergo aortic repair when the aortic diameter reaches 4.2 cm or greater by transesophageal echocardiogram (internal diameter) or 4.4 to 4.6 cm or greater by computed tomographic imaging and/or magnetic resonance imaging (external diameter).

Ascending Aortic Aneurysm of Degenerative Etiology

The following are recommended pathway steps for the diagnosis and management of ascending aortic aneurysm of degenerative etiology.

STEP 1: Determine need for urgent operative repair



STEP 2: Preoperative diagnostic evaluation

Ascending Aortic Aneurysm of Degenerative Etiology

STEP 3: Identify timing and type of operative repair

For patients that indicated urgent operative management in Step 1: Is there CAD or valve pathology requiring operative repair?

Yes: Proceed with CABG or valve procedure and aneurysm repair.

No: Proceed with aneurysm repair.

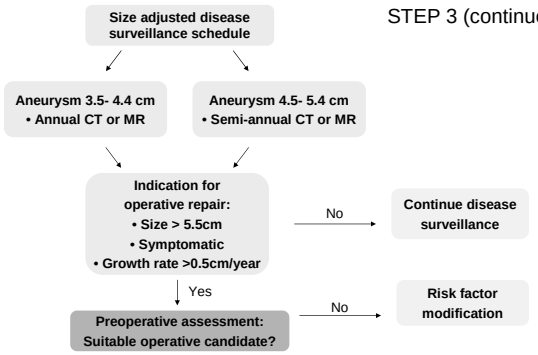
For patients that did <u>not</u> indicate a need for urgent operative management in Step 1: Is there CAD or valve pathology requiring operative repair?

Yes: Proceed with CABG or valve procedure and aneurysm repair if > 4.5cm.

No: Proceed with the following pathway steps.

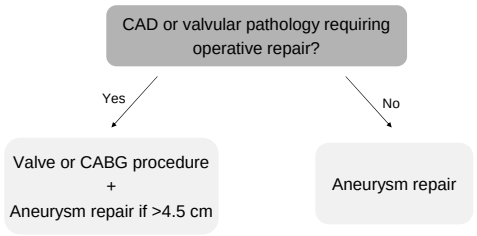
Ascending Aortic Aneurysm of Degenerative Etiology

STEP 3 (continued)



Ascending Aortic Aneurysm of Degenerative Etiology

STEP 4: Surgical intervention



Note: For full algorithm, see Figure 31 in full-text version of TAD Guidelines.

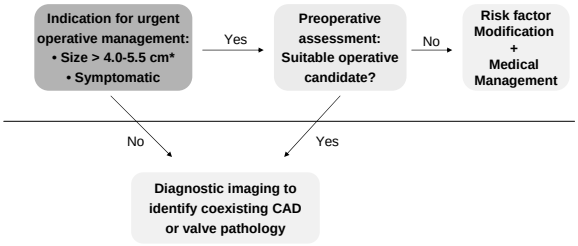
Ascending Aortic Aneurysms Associated with Genetic Disorder

The following are recommended pathway steps for the diagnosis and management of ascending aortic aneurysm associated with:

- Marfan Syndrome
- bicuspid aortic valve
- other genetically mediated disorder

Ascending Aortic Aneurysms Associated with Genetic Disorder

STEP 1: Determine need for urgent operative repair



STEP 2: Preoperative diagnostic evaluation

* Depends on specific condition

Ascending Aortic Aneurysms Associated with Genetic Disorder

STEP 3: Identify timing and type of operative repair

For patients that indicated urgent operative management in Step 1: Is there CAD or valve pathology requiring operative repair?

Yes: Proceed with CABG or valve procedure and aneurysm repair

No: Proceed with aneurysm repair

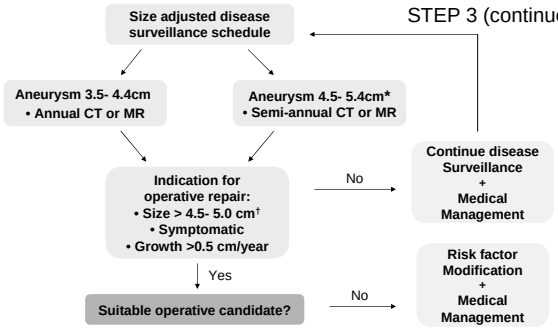
For patients that did <u>not</u> indicate a need for urgent operative management in Step 1: Is there aortic valve pathology requiring operative repair?

Yes: Proceed with CABG or valve procedure and aneurysm repair if > 4.5cm.

No: Proceed with the following pathway steps.

Ascending Aortic Aneurysms Associated with Genetic Disorder

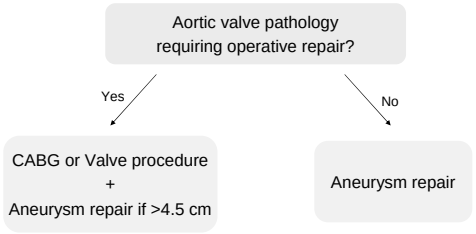
STEP 3 (continued)



* Depends on specific condition; † See Recommendations for Asymptomatic Patients With Ascending Aortic Aneurysm and for Bicuspid Aortic Valve and Associated Congenital Variants in Adults in full-text version of TAD Guidelines

Ascending Aortic Aneurysms Associated with Genetic Disorder

STEP 4: Surgical intervention



Note: For full algorithm, see Figure 32 in full-text version of TAD Guidelines.

Guidelines for Thoracic Aortic Disease

Recommendation for Symptomatic Patients With Thoracic Aortic Aneurysm

Recommendation for Symptomatic Patients With thoracic Aortic Aneurysm

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.

Guidelines for Thoracic Aortic Disease

Recommendations for Open Surgery for Ascending Aortic Aneurysm

Recommendations for Open Surgery for Ascending Aortic Aneurysm

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.

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.

Guidelines for Thoracic Aortic Disease

Recommendations for Aortic Arch Aneurysms

Recommendations for Aortic Arch Aneurysms

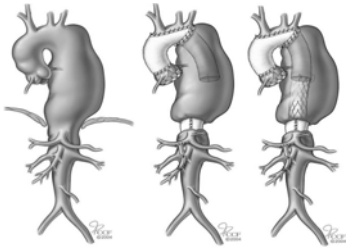
For thoracic aortic aneurysms also involving the proximal aortic arch, partial arch replacement together with ascending aorta repair using right subclavian/axillary artery inflow and hypothermic circulatory arrest is reasonable.

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.

Replacement of the entire aortic arch is reasonable for aneurysms of the entire arch, for chronic dissection when the arch is enlarged, and for distal arch aneurysms that also involve the proximal descending thoracic aorta, usually with the elephant trunk procedure.

"Elephant Trunk" Procedure

Left, Preoperative disease. **Middle,** Stage I with replacement of 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. **Right,** Completion of procedure using an endovascular stent graft attached proximally to the "elephant trunk" and the distal end secured to a Dacron graftcuff.



SOURCE: Images reprinted with permission from the Cleveland Clinic Foundation.

Recommendations for Aortic Arch Aneurysms

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.

For patients with isolated aortic arch aneurysms less than 4.0 cm in diameter, it is reasonable to reimage using computed tomographic imaging or magnetic resonance imaging, at 12-month intervals, to detect enlargement of the aneurysm.

For patients with isolated aortic arch aneurysms 4.0 cm or greater in diameter, it is reasonable to reimage using computed tomographic imaging or magnetic resonance imaging, at 6-month intervals, to detect enlargement of the aneurysm.

Guidelines for Thoracic Aortic Disease

Recommendations for Descending Thoracic Aorta and Thoracoabdominal Aortic Aneurysms

Recommendations for Descending Thoracic Aorta and Thoracoabdominal Aortic Aneurysms

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.

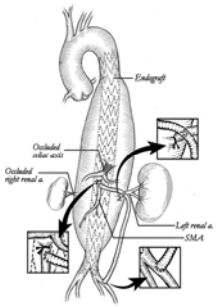
For patients with degenerative or traumatic aneurysms of the descending thoracic aorta exceeding 5.5 cm, saccular aneurysms, or postoperative pseudoaneurysms, endovascular stent grafting should be strongly considered when feasible.

Recommendations for Descending Thoracic Aorta and Thoracoabdominal Aortic Aneurysms

For patients with thoracoabdominal aneurysms, in whom endovascular stent graft options are limited and surgical morbidity is elevated, elective surgery is recommended if the aortic diameter exceeds 6.0 cm, or less if a connective tissue disorder such as Marfan or Loeys-Dietz syndrome is present.

For patients with thoracoabdominal aneurysms and with end-organ ischemia or significant stenosis from atherosclerotic visceral artery disease, an additional revascularization procedure is recommended.

Hybrid Open Visceral Revascularization and Endograft Aneurysm Exclusion



Schema of TAA treated with initial left iliac artery–to–left renal artery–to–superior mesenteric artery bypass graft and subsequent placement of a thoracoabdominal endograft.

Proximal superior mesenteric artery and left renal arteries were ligated.

SMA indicates superior mesenteric artery; and TAA, thoracoabdominal aneurysm.

SOURCE: Adapted from Flye, et al. *J Vasc Surg*. 2004;39:454–8.

Society of Thoracic Surgeons Recommendations for Thoracic Stent Graft Insertion (summary)

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

Note: Table 15 in full-text version of TAD Guidelines. Reprinted from Svensson et al. Expert consensus document on the treatment of descending thoracic aortic disease using endovascular stent grafts. Ann Thorac Surg. 2008;85:S1–41.

Guidelines for Thoracic Aortic Disease

Recommendations for Counseling and Management of Chronic Aortic Diseases in Pregnancy

Recommendations for Counseling and Management of Chronic Aortic Diseases in Pregnancy

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.

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.

For all pregnant women with known aortic root or ascending aortic dilatation, monthly or bimonthly echocardiographic measurements of the ascending aortic dimensions are recommended to detect aortic expansion until birth.

Recommendations for Counseling and Management of Chronic Aortic Diseases in Pregnancy

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

Pregnant women with aortic aneurysms should be delivered where cardiothoracic surgery is available.

Recommendations for Counseling and Management of Chronic Aortic Diseases in Pregnancy

Fetal delivery via cesarean section is reasonable for patients with significant aortic enlargement, dissection, or severe aortic valve regurgitation.

If progressive aortic dilatation and/or advancing aortic valve regurgitation are documented, prophylactic surgery may be considered.

Guidelines for Thoracic Aortic Disease

Recommendations for Aortic Arch and Thoracic Aortic Atheroma and Atheroembolic Disease

Recommendations for Aortic Arch and Thoracic Aortic Atheroma and Atheroembolic Disease

Treatment with a statin is a reasonable option for patients with aortic arch atheroma to reduce the risk of stroke.

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.

Guidelines for Thoracic Aortic Disease

Recommendations for Preoperative Evaluation

Recommendations for Preoperative Evaluation

In preparation for surgery, imaging studies adequate to establish the extent of disease and the potential limits of the planned procedure are recommended.

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.

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.

Recommendations for Preoperative Evaluation

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.

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.

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.

Guidelines for Thoracic Aortic Disease

Recommendations for Choice of Anesthetic and Monitoring Techniques

Recommendations for Choice of Anesthetic and Monitoring Techniques

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.

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.

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.

Recommendations for Choice of Anesthetic and Monitoring Techniques

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.

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.

Guidelines for Thoracic Aortic Disease

Recommendation for Transfusion Management and Anticoagulation in Thoracic Aortic Surgery

Recommendation for Transfusion Management and Anticoagulation in Thoracic Aortic Surgery

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.

Guidelines for Thoracic Aortic Disease

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

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

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.

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.

Perioperative brain hyperthermia is not recommended in repairs of the ascending aortic and transverse aortic arch as it is probably injurious to the brain.

Guidelines for Thoracic Aortic Disease

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

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

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.

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.

Moderate systemic hypothermia is reasonable for protection of the spinal cord during open repairs of the descending thoracic aorta.

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

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.

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.

Guidelines for Thoracic Aortic Disease

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

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

Preoperative hydration and intraoperative mannitol administration may be reasonable strategies for preservation of renal function in open repairs of the descending aorta.

During thoracoabdominal or descending aortic repairs with exposure of the renal arteries, renal protection by either cold crystalloid or blood perfusion may be considered.

Furosemide, mannitol, or dopamine should not be given solely for the purpose of renal protection in descending aortic repairs.

Guidelines for Thoracic Aortic Disease

Recommendations for Surveillance of Thoracic Aortic Disease or Previously Repaired Patients

Recommendations for Surveillance of Thoracic Aortic Disease or Previously Repaired Patients

Computed tomographic imaging or magnetic resonance imaging of the thoracic aorta is reasonable after a Type A or B aortic dissection or after prophylactic repair of the aortic root/ascending aorta.

Computed tomographic imaging or magnetic resonance imaging of the aorta is reasonable at 1, 3, 6, and 12 months postdissection and, if stable, annually thereafter so that any threatening enlargement can be detected in a timely fashion.

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.

Recommendations for Surveillance of Thoracic Aortic Disease or Previously Repaired Patients

If a thoracic aortic aneurysm is only moderate in size and remains relatively stable over time, magnetic resonance imaging instead of computed tomographic imaging is reasonable to minimize the patient's radiation exposure.

Surveillance imaging similar to classic aortic dissection is reasonable in patients with intramural hematoma.

Suggested Follow-up of Aortic Pathologies after Repair or Treatment

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

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

Note: CT, computed tomographic imaging; CXR, chest X-ray; IMH, intramural hematoma; MR, magnetic resonance imaging; PAU, penetrating atherosclerotic ulcer; and TTE, transthoracic echocardiography.

Table 17 in full-text version of TAD Guidelines.

Guidelines for Thoracic Aortic Disease

Recommendation for Employment and Lifestyle in Patients With Thoracic Aortic Disease

Recommendation for Employment and Lifestyle in Patients With TAD

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.

Guidelines for Thoracic Aortic Disease

Recommendations for Quality Assessment and Improvement for Thoracic Aortic Disease

Recommendations for Quality Assessment and Improvement for TAD

Hospitals that provide regional care for patients with acute sequelae of thoracic aortic disease (e.g., 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.

Recommendations for Quality Assessment and Improvement for TAD

Hospitals that provide regional care for patients with acute sequelae of thoracic aortic disease (e.g., 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 upon available outcome data and future performance measures developed in accordance with this guideline.

Recommendations for Quality Assessment and Improvement for TAD

Standardized Transferring Facility Assessment, Communication, and Documentation should occur for the following domains:

- **BP Control for hypertension**
- **HR 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 & 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.**

Note: BP = blood pressure; EMS = emergency medical service; HR =heart rate.
This information was adapted from Table 20 in the full-text version of the TAD guidelines

Quiz 1

- Which one of the following is NOT high risk for thoracic aortic dissection?
  1. Marfan Syndrome
  2. Connective tissue disease (eg. LDS)
  3. Systematic lupus erythematosus
  4. Recent aortic manipulation
  5. Known thoracic aortic aneurysm

Quiz 2

- Which one of the following is NOT high risk examination feature for thoracic aortic dissection?
  1. Pulse deficit
  2. Systolic BP limb differential > 20mm Hg
  3. Focal neurologic deficit
  4. Differential cyanosis
  5. Murmur of aortic regurgitation (new or not known to be old and in conjunction with pain)

Quiz 3

- Which one of the following is NOT high risk for increased aortic wall stress?
  1. Hypertension, uncontrolled
  2. Deceleration or torsional injury (eg, motor vehicle crash, fall)
  3. Pheochromocytoma
  4. Valsalva maneuver
  5. Bicuspid aortic stenosis

Quiz 4

- Which one of the following is the first priority for treating aortic dissection if not contraindicated?
  1. Beta blockers or labetalol
  2. Diltiazem
  3. ACEI
  4. ARB
  5. Clonidine

Quiz 5

- Which one of the following is CORRECT for aorta surgical or endovascular repairs?
  1. Endovascular stenting for saccular aneurysm
  2. CSF drainage for brain protection
  3. Regional anesthetic techniques
  4. Furosemide for renal protection
  5. Routinely changing double-lumen endotracheal (endobronchial) tubes to single-lumen tubes at the end of surgical procedures