

SOCIETAL STATEMENT

# 2022 Aortic Disease Guideline-at-a-Glance



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## INTRODUCTION

The 2022 American College of Cardiology/American Heart Association Guideline for the Diagnosis and Management of Aortic Disease (ACC/AHA Aortic Disease Guideline) provides guidance for clinicians on the “diagnosis, genetic evaluation and family screening, medical therapy, endovascular and surgical treatment, and long-term surveillance of patients with aortic disease.”<sup>1</sup> In this Guideline-at-a-Glance, practice-changing recommendations from the guideline are highlighted to accelerate dissemination.

American College of Cardiology (ACC) Guideline dissemination is an organization-wide effort overseen by the ACC Solution Set Oversight Committee. A goal of the Solution Set Oversight Committee is to ensure guideline content is integrated throughout ACC clinical policy, education, registry, membership, and advocacy efforts. The clinical tools presented here are part of a larger ACC dissemination strategy to facilitate the implementation of key changes in practice.

Three of the Top 10 Take-Home Messages from the ACC/AHA Aortic Disease Guideline are selected here to provide a framework for initial dissemination. The 3 selected messages are focused on imaging (#3), surgical thresholds (#4), and family screening (#10). These messages are emphasized in various ACC clinician tools: the JACC Illustrations, an old/new comparison table, and a table comparing ACC/AHA guidelines with those of the European Society of Cardiology (ESC).

## TOP 10 TAKE-HOME MESSAGES

The following Top 10 Take-Home Messages are taken directly from the ACC/AHA Aortic Disease Guideline.<sup>1</sup> Messages in **bold** were selected as key themes for this Guideline-at-a-Glance because they are changes in

recommendations from previous guidelines and address known practice gaps.

1. Because outcomes for patients with aortic disease are enhanced at programs with higher volumes, experienced practitioners, and extensive management capabilities, Multidisciplinary Aortic Team care is considered in determining the appropriate timing of intervention.
2. Shared decision-making involving the patient and a multidisciplinary team is highly encouraged to determine the optimal medical, endovascular, and open surgical therapies. In patients with aortic disease who are contemplating pregnancy or who are pregnant, shared decision-making is especially important when considering the cardiovascular risks of pregnancy, the diameter thresholds for prophylactic aortic surgery, and the mode of delivery.
3. **Computed tomography, magnetic resonance imaging, and echocardiographic imaging of patients with aortic disease should follow recommended approaches for image acquisition, measurement and reporting of relevant aortic dimensions, and the frequency of surveillance before and after intervention.**
4. **At centers with Multidisciplinary Aortic Teams and experienced surgeons, the threshold for surgical intervention for sporadic aortic root and ascending aortic aneurysms has been lowered from 5.5 to 5.0 cm in selected patients, and even lower in specific scenarios among patients with heritable thoracic aortic aneurysms.**
5. In patients who are significantly smaller or taller than average, surgical thresholds may incorporate indexing of the aortic root or ascending aortic diameter to either patient body surface area or height, or aortic cross-sectional area to patient height.
6. Rapid aortic root growth or ascending aortic aneurysm growth, an indication for intervention,

is defined as  $\geq 0.5$  cm in 1 year or  $\geq 0.3$  cm/y in 2 consecutive years for those with sporadic aneurysms and  $\geq 0.3$  cm in 1 year for those with heritable thoracic aortic disease or bicuspid aortic valve.

7. In patients undergoing aortic root replacement surgery, valve-sparing aortic root replacement is reasonable if the valve is suitable for repair and when performed by experienced surgeons in a Multidisciplinary Aortic Team.
8. Patients with acute type A aortic dissection, if clinically stable, should be considered for transfer to a high-volume aortic center to improve survival. The operative repair of type A aortic dissection should entail at least an open distal anastomosis rather than just a simple supracoarotary interposition graft.

9. There is an increasing role for thoracic endovascular aortic repair in the management of uncomplicated type B aortic dissection. Clinical trials of repair of thoracoabdominal aortic aneurysms with endografts are reporting results that suggest endovascular repair is an option for patients with suitable anatomy.
10. **In patients with aneurysms of the aortic root or ascending aorta, or those with aortic dissection, screening of first-degree relatives with aortic imaging is recommended.**

### JACC ILLUSTRATIONS

#### Central Illustration: Updated Surgical Thresholds for Aneurysms

The JACC **Central Illustration** for the ACC/AHA Aortic Disease Guideline identifies the updated surgical

## CENTRAL ILLUSTRATION 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease

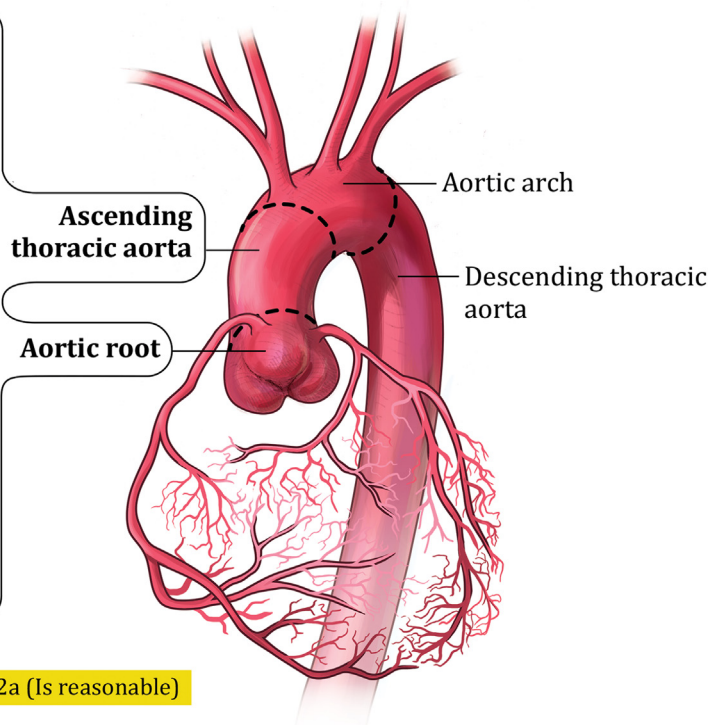
### Surgical intervention thresholds for aortic root & ascending aorta in patients with...

#### Sporadic and BAV aneurysms\*:

**5.5 cm** (COR 1)  
**5.0 cm** by experienced surgeons in a Multidisciplinary Aortic Team (COR 2a)

#### Marfan syndrome#:

**5.0 cm** (COR 1)  
 **$\geq 4.5$  cm** in those with an increased risk of aortic dissection when performed by experienced surgeons in a Multidisciplinary Aortic Team (COR 2a)



**COR 1** (Is recommended) **COR 2a** (Is reasonable)

\*Surgical thresholds may be adjusted based on patient genetics, rapid aortic growth rate, cross-sectional aortic area/height ratio  $\geq 10$  cm<sup>2</sup>/m, aortic size index of  $\geq 3.08$  cm/m<sup>2</sup>, or aortic height index of  $\geq 3.21$  cm/m.

#For more on rapid aortic growth rate and patients with nonsyndromic heritable thoracic aortic aneurysms or with genetic aortopathies other than Marfan syndrome (e.g., Loeys-Dietz syndrome), please see the 2022 ACC/AHA Guideline for the Diagnosis & Management of Aortic Disease.

intervention thresholds for sporadic aortic root and ascending aortic aneurysms. This clinician tool focuses on Top 10 Take-Home Message #4 and represents a major change in clinical practice. The tool is intended to quickly and memorably communicate new information from the guideline.

For additional information, see Section 6.5.1, “Surgery for Sporadic Aneurysms of the Aortic Root and Ascending Aorta,” in the guideline.<sup>1</sup>

#### Interactive Illustration: Family Screening for Aortic Disease

The [JACC Interactive Illustration](#) for the ACC/AHA Aortic Disease Guideline outlines screening of first-degree relatives with aortic imaging and genetic screening in patients with aortic aneurysms or dissections. The tool is accessible at <https://www.jacc.org/AorticDiseaseInteractive>.

This clinician tool focuses on Top 10 Take-Home Message #10 and aims to close a current clinical practice gap. A user of the interactive tool can select factors that best match the clinical situation and review the corresponding guideline recommendations to decide

how to best screen family members of patients with aortic disease. As an interactive tool, a user can make different selections to understand their impact on recommendations for screening. The interactive tool can also be used with patients as part of a clinician-patient conversation.

For additional information, see Section. 6.1.2.1, “Heritable Thoracic Aortic Disease: Genetic Testing and Screening of Family Members for Thoracic Aortic Disease,” in the guideline.<sup>1</sup>

#### COMPARISON TO PREVIOUS ACC/AHA GUIDELINES

The scope of the 2022 ACC/AHA Aortic Disease Guideline includes both the abdominal and thoracic aorta, topics previously covered in 3 other guidelines.<sup>2-4</sup> **Table 1** outlines changes in surgical thresholds, imaging, and family screening between the 2010 guidelines and the 2022 ACC/AHA Aortic Disease Guideline.

**TABLE 1** Select Differences Between the 2010 and 2022 ACC/AHA Aortic Disease Guidelines

	2010 <sup>3</sup>	2022 <sup>1</sup>
Surgical thresholds (Top 10 #4)	<p>≥5.5-cm threshold for surgical intervention for sporadic aortic root and ascending aortic aneurysms (COR 1).</p> <p>Surgical thresholds adjusted by patient sex.</p>	<p>≥5.0 cm threshold for surgical intervention is reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team for sporadic aortic root and ascending aortic aneurysms (COR 2).</p> <p>Surgical thresholds adjusted by patient genetics, sex, and height (maximal cross-sectional aortic area/height ratio ≥10 cm<sup>2</sup>/m, aortic size index ≥3.08 cm/m<sup>2</sup>, or aortic height index ≥3.21 cm/m).</p>
Imaging (Top 10 #3)	<p>Basic guidance on accurate reporting of dimensions by echocardiography, CT, and MRI.</p>	<p>Much more specific guidance on:</p> <ul style="list-style-type: none"> <li>Recommended approaches for image acquisition;</li> <li>Measurement and reporting of relevant aortic dimensions;</li> <li>The frequency of surveillance before and after intervention by CT, MRI, and echocardiographic imaging of patients with aortic disease.</li> </ul> <p>Measurement methods should be reported in a clear and consistent fashion (COR1).</p> <p>Measuring from sinus to sinus and from inner-edge to inner-edge on CT and MRI has shown good correlation with TTE for measurements of the root (leading-edge to leading-edge at end-diastole) and ascending segments (COR 1).</p> <p>The use of ECG-gated images decreases motion artifact and improves edge depiction in aortic root imaging, with diminished measurement variability (COR 1).</p> <p>In patients with known or suspected aortic disease, when performing echocardiography, it is reasonable to measure the aorta from leading-edge to leading-edge, perpendicular to the axis of blood flow (COR 2a).</p> <p>Using inner-edge to inner-edge measurements may also be considered, particularly on short-axis imaging (COR 2b).</p>
Family screening (Top 10 #10)	<p>Only relatives with genetic mutations should undergo aortic imaging (COR 1).</p> <p>If 1 or more first-degree relatives of a patient are found to have aortic disease, then referral to a geneticist may be considered (COR 2b).</p> <p>Sequencing of the <i>ACTA2</i> gene is reasonable (COR 2a).</p> <p>Sequencing of other genes (<i>TGFBR1</i>, <i>TGFBR2</i>, <i>MYH11</i>) may be considered (COR 2b).</p>	<p>Screening aortic imaging for relatives is recommended, even if a disease-causing variant is not identified with genetic testing (COR 1).</p> <p>In patients with an established pathogenic or likely pathogenic variant in a gene predisposing to HTAD, it is recommended that genetic counseling be provided (COR 1).</p> <p>Genetic testing to identify variants is recommended in the proband. Cascade testing for family members would then be indicated if the mutation is present in the proband. Testing panels include <i>FBN1</i>, <i>LOX</i>, <i>COL3A1</i>, <i>TGFBR1</i>, <i>TGFBR2</i>, <i>SMAD3</i>, <i>TGFBR2</i>, <i>ACTA2</i>, <i>MYH11</i>, <i>MYLK</i>, and <i>PRKG1</i> (COR 1).</p>

Colors in the table align with the classification system found in Table 2, “Applying American College of Cardiology/American Heart Association Class of Recommendation and Level of Evidence to Clinical Strategies, Interventions, Treatments, or Diagnostic Testing in Patient Care” in the 2022 ACC/AHA Aortic Disease Guideline.<sup>1</sup>

ACC = American College of Cardiology; AHA = American Heart Association; COR = class of recommendation; CT = computed tomography; ECG = electrocardiogram; HTAD = heritable thoracic aortic disease; MRI = magnetic resonance imaging; TTE = transesophageal echocardiography.

**TABLE 2 Surgical Intervention Thresholds for Aortic Aneurysms: Select Comparison of ACC/AHA Guideline and ESC Guideline**

	ESC Guideline <sup>5</sup>	ACC/AHA Guideline <sup>1</sup>
Intervention for aortic root aneurysm (Top 10 #4)	Surgery should be considered in patients who have aortic root aneurysm, with a maximal ascending aortic diameter of 55 mm for other patients with no elastopathy (COR 2a).	In asymptomatic patients with aneurysms of the aortic root or ascending aorta with a maximum diameter $\geq 5.5$ cm, surgery is indicated (COR 1).
	No comparative recommendation.	In asymptomatic patients with aneurysms of the aortic root or ascending aorta with a maximum diameter of $\geq 5.0$ cm, surgery is reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team (COR 2a).
	Surgery should be considered in patients who have aortic root aneurysm, with maximal ascending aortic diameter of 45 mm for patients with Marfan syndrome with risk factors (COR 2a).	In patients with Marfan syndrome and an aortic root diameter $\geq 5.0$ cm, surgery to replace the aortic root and ascending aorta is recommended (COR 1).
	Surgery should be considered in patients who have aortic root aneurysm, with maximal ascending aortic diameter of 50 mm for patients with bicuspid valve with risk factors (COR 2a).	In patients with Marfan syndrome, an aortic root diameter $\geq 4.5$ cm and features associated with an increased risk of AoD, surgery to replace the aortic root and ascending aorta is reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team (COR 2a).
	Surgery should be considered in patients who have aortic root aneurysm, with maximal ascending aortic diameter of 50 mm for patients with bicuspid valve with risk factors (COR 2a).	In patients with a BAV, a diameter of the aortic root or ascending aorta of 5.0 to 5.4 cm and an additional risk factor for AoD, surgery to replace the aortic root and/or ascending aorta is reasonable when performed by experienced surgeons in a Multidisciplinary Aortic Team (COR 2a).
Intervention for descending aortic aneurysms	TEVAR should be considered in patients who have descending aortic aneurysm with maximal diameter 55 mm (COR 2a).	In patients with intact descending TAAs, repair is recommended when the diameter is $\geq 5.5$ cm (COR 1).

Colors in the table align with the classification system found in Table 2, "Applying American College of Cardiology/American Heart Association Class of Recommendation and Level of Evidence to Clinical Strategies, Interventions, Treatments, or Diagnostic Testing in Patient Care" in the 2022 ACC/AHA Aortic Disease Guideline.<sup>1</sup>

ACC = American College of Cardiology; AHA = American Heart Association; AoD = aortic dissection; BAV = bicuspid aortic valve; COR = class of recommendation; TAA = thoracic aortic aneurysm; TEVAR = thoracic endovascular aortic repair.

Additional information for **Table 1** can be found in the following 2022 ACC/AHA Aortic Disease Guideline sections:

- Imaging (Top 10 Take-Home Message #3): Section 3.1, "Aortic Imaging Techniques to Determine Presence and Progression of Aortic Disease," and Section 3.2, "Conventions of Measurements"
- Surgical thresholds (Top 10 Take-Home Message #4): Section 6.5.1, "Surgery for Sporadic Aneurysms of the Aortic Root and Ascending Aorta"
- Family screening (Top 10 Take-Home Message #10): Section 6.1.2.1, "Heritable Thoracic Aortic Disease: Genetic Testing and Screening of Family Members for Thoracic Aortic Disease."<sup>1</sup>

**ACC/AHA GUIDELINE COMPARISON TO ESC GUIDELINE**

The ESC has also published an aortic disease clinical practice guideline. **Table 2** highlights the subtle differences in surgical interventions for

aneurysms between the 2022 ACC/AHA Aortic Disease Guideline and the 2014 ESC Aortic Diseases Guideline.<sup>5</sup> The comparison table is focused on Top 10 Take-Home Message #4.

Additional information to **Table 2** can be found in the 2022 ACC/AHA Aortic Disease Guideline Section 6.5.1, "Surgery for Sporadic Aneurysms of the Aortic Root and Ascending Aorta,"<sup>1</sup> and the 2014 ESC Aortic Diseases Guideline Section 7.1, "Thoracic Aortic Aneurysms."<sup>5</sup>

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