Perspectives on the 2022 Joint Guidelines on the Diagnosis and Management of Aortic Disease

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Central Picture Legend: Multidisciplinary collaboration is key to consensus guidelines and care of aortic patients

Central Message: The 2022 AHA/ACC aortic guidelines provide the first update to the 2010 guidelines with balanced representation across disciplines and stronger evidence incorporating advances in surgical techniques.
Perspective Statement: Developments in open and endovascular aortic surgery over the past decade have led to improvements in the treatment of aortic diseases. A critical evaluation of the evidence requires input from all stakeholders, including patients. Short-term improvements in care must be weighed against the implication for long-term outcomes, and evolving technologies require ongoing evaluation.

Keywords: Aorta, aortic surgery, aortic aneurysm, aortic dissection
Introduction

While pathologic conditions of the aorta were recognized as early as 1760, with King George II’s private physician Frank Nicholls identifying an acute aortic dissection on post-mortem examination, it was not until the 1950s that definitive surgery for aortic diseases was reported by Denton Cooley and Michael DeBakey. In the last half-decade, developments in imaging technology, surgical techniques, genetic testing, and endovascular devices have all led to improvements in the management of aortic diseases. Despite a broader understanding of pathophysiology and the role of medical management, death rates globally from aortic diseases continue to rise.

The first multi-society guidelines document on the management of aortic diseases was published in 2010 and limited to the thoracic aorta. Since then, writing groups have addressed controversies and provided expert consensus documents and clinical practice guidelines on selected topics in aortic disease management. A broad multisociety guideline update of the evidence-based recommendations from 2010, however, had not occurred in over a decade. In this issue of the Journal, the 2022 American College of Cardiology (ACC)/American Heart Association (AHA) guidelines for the diagnosis and management of aortic disease are presented with representation from the American Association for Thoracic Surgery, Society of Thoracic Surgeons (STS), Society of Vascular Surgery (SVS), Society of Cardiovascular Anesthesiologists, Society of Cardiovascular Angiography and Interventions, and American College of Radiology.

Guideline Development - Key Standards and Principles
Unique to this current aortic guidelines document is the breadth of disciplines involved: representatives from radiology, cardiovascular medicine, cardiothoracic surgery, vascular surgery, anesthesiology/critical care, internal medicine, medical genetics, emergency medicine, and a lay/patient representative all of whom contributed to its development. In contrast to the 2010 guidelines, the committee was designed to achieve an intentional balanced representation of specialties among cardiology, vascular surgery, and cardiothoracic surgery. Inclusion of the lay/patient representative is a recognition of the importance of shared decision-making and patient-reported outcomes in guiding care.

The keystone of guideline development is a critical, methodologic appraisal of the evidence via a systematic, rigorous analysis, focusing on risks, benefits, and patient outcomes. With the recognition that aortopathy is a lifelong and systemic disease, the evaluation of risks and benefits should account for not only short-term outcomes but also long-term risks of mortality, morbidity, and reintervention. Focus on the quality of evidence and open communication about disagreements facilitated the writing committee to achieve consensus amongst the many specialty representatives and to overcome any potential biases.

Comment on Quality of Evidence

Compared to the 2010 guidelines, the writing committee had the benefit of a more robust body of evidence. The International Registry of Acute Aortic Dissection (IRAD), formed in 1996, has expanded from 24 centers in 2010 to 58 currently, and has in the intervening years contributed a number of important studies that contributed to the guideline development, with 38 references in the 2022 guidelines. Another crucial registry is the German Registry for Acute Aortic Dissection Type A (GERAADA), founded in 2006 and now with over 50 centers from
Germany, Austria, and Switzerland contributing. Other sources of national data are the STS Adult Cardiac Surgery Database and the Canadian Thoracic Aortic Collaborative. Importantly, as the guidelines highlight, center expertise and experience are important, and a number of high-volume centers have set the standard for excellence of care, as well as pushing the field forward. Numerous citations in the guidelines are based on these centers’ contributions to the literature. Literature from these registries and high-volume centers represent the best available evidence, as large-scale prospective randomized controlled trials are lacking and unlikely to be conducted.

Endovascular registries have lagged behind surgical ones, likely due to the more recent development of commercial devices which restricted registries to device-specific studies in the initial phases. Comparative studies are relatively lacking. The SVS’s Vascular Quality Initiative (VQI), which began in 2010 as an expansion of the Vascular Study Group of New England, now includes a TEVAR/Complex EVAR Registry encompassing the whole of aortic pathologies beyond dissections. As the management of chronic type B aortic dissections matures, there is not yet consensus on the important radiologic parameters to predict long-term aortic reintervention and aortic mortality risk. Expert radiologic measurement techniques and high-quality longitudinal data will be critical. Standardized reporting across registries will be necessary for meta-analytic studies. In the growth and evolution of these registries, including web-based data entry systems, critical assessment of data validity is paramount and may necessitate system redesigns.\textsuperscript{11}

In particular, the aortic components of the STS Adult Cardiac Surgery Database, have changed dynamically over the last 15 years in response to technologic advances, to reflect the anatomic and complexity of aortic operations, and to quantify the use of adjuncts such as cerebral perfusion and spinal cord protection as highlighted by Bavaria.\textsuperscript{12} The STS Aortic
Database, established in V2.9 in 2017 has allowed collection of high-quality, granular data. This in turn has made feasible the development of an aortic-specific risk model through the work of the Aortic Task Force, which is currently in validation and expected to roll-out imminently.\textsuperscript{13} Importantly, surgeons should actively participate in the data input to ensure ongoing high-quality and accurate data.

\textit{Comment on scope and granularity}

The 2010 guidelines, along with many others, provided an excellent foundation on which to write the 2022 document. In the last 12 years, major open surgical developments have included valve-sparing root replacement for aneurysmal disease\textsuperscript{14} and extended arch techniques for aortic dissections. Pioneered by high-volume centers, these techniques are meant to extend long-term benefit, particularly for younger patients, without imposing additional operative risk. Accordingly, surgeon experience in a multidisciplinary aortic team (MAT) is of great importance to risk mitigation for these complex operations. A similar sentiment underlies the granularity of recommendations for bicuspid and sporadic tricuspid valve aortopathy, which now factors in not only absolute size but also surgeon and team experience, as well as the recognition of patient factors such as body size and personalized risk. Notably, these new 2a and 2b recommendations underscore the importance of surgical expertise in MAT. The STS database demonstrated that the observed to expected ratio for mortality after proximal thoracic aortic operations did not decrease below 1 until center volume exceeded 24 cases/year; however, only 116 centers performed \( \geq 20 \) cases/year. Of 1045 total centers, only 24 centers performed \( \geq 50 \) cases/year with a 2.1\% operative mortality.\textsuperscript{15} This operative risk at high-volume centers must be weighed against the risk of aortic events, which can be challenging to determine. In the most recent and largest natural history study, the risk of dissection for 5.0-5.4 cm ascending thoracic aortic aneurysms
was only 0.43(0.20-0.97)/100 person-years (95% confidence interval).\textsuperscript{16} As such, lower thresholds for aneurysm repair apply to experienced centers. Nonetheless, while adverse aortic events are not entirely captured by such low rates of dissection, all-cause mortality overestimates aortic-related events; thus better prospective data is required.

The interrelationship of the aortic valve and ascending aorta has implications for transcatheter aortic valve therapy offerings. As bicuspid aortic valve (BAV) patients often present at a younger age, interventions should aim for the strategy that provides the best long-term benefit. Experienced surgeons faced with patients having BAV dysfunction and an ascending aortic aneurysm of 4.5-5.0 cm are supported in an open approach for low-risk patients, rather than transcatheter aortic valve replacement (TAVR) alone. This is especially true if aortic dissection risk factors (family history, root phenotype, growth $\geq 0.3$ cm/year) are present. As TAVR technology expands to include treatment of pure aortic insufficiency (AI), the possibility of aortic valve replacement for AI in a patient with aortopathy demands that surgeons remain involved in shared decision-making. If such patients undergo TAVR, then followup of aortopathy is even more critical than in patients with predominant stenosis, as the risk of late aortic dissection is higher in AI patients (odds ratio: 10.0; 95% confidence interval: 6.2 to 16.2; $p < 0.001$).\textsuperscript{17}

Despite advances in endovascular therapies, open surgery remains the standard of care for patients with connective tissue disorders, with excellent outcomes in the short and long-term.\textsuperscript{18} The guidelines do recognize the role of endovascular therapies for acutely life-threatening complications in such patients, but primarily as a temporizing measure to definitive management. There has been marked expansion of specific recommendations on the various
genetic and heritable aortopathies which will serve as a useful guide for surgeons without access to a geneticist locally.

Limitations and Future Directions

The intent of these guidelines is to be a leading document for all practitioners worldwide. There has been a paradigm shift in the development and update of guideline recommendations. While plans for the update process are in evolution, the current intent is to annually reaffirm, update, and add new information so that the guidelines become a living document. This is in recognition of the dynamic rate of technical and technologic development in all aspects of care and the need to provide guidance to physicians in a timely manner when practice-changing evidence emerges. Certain topics were not included in the 2022 guidelines on which we wish to comment. First, it is increasingly recognized that gender and social disparities exist broadly in medicine. Literature has demonstrated such disparities in aortic surgery, both in diagnosis and treatment. The linkage to different biologic behavior of the aorta along gender lines remain unknown. Further, the untangling of socioeconomic status from baseline health and from access to quality care similarly remain largely unaddressed. A second limitation of the guidelines is the absence of recommendations for the treatment of chronic TBAD. For these, we refer the reader to the clinical practice guidelines published jointly by the STS and AATS. This remains an area where new endovascular techniques are rapidly developing and being studied. High-quality registries, validated data, and uniformity of reporting and endpoints will lead to a stronger evidence base to provide future recommendations.

Lastly, future guidelines may amend the current size thresholds for intervention based on accumulating evidence. The limitations of using absolute aortic diameter and the value of body
size indexing have been recognized for some time and are incorporated into the current
guidelines. Elefteriades, re-examining the “hinge points” that he pioneered two decades ago,
suggests for a multitude of reasons that a leftward shift for elective aneurysmectomy may be
prudent to further reduce population dissection mortality. Whether this alters the balance of
risk and benefit may be addressed by the ongoing Treatment in Thoracic Aortic Aneurysm:
Surgery versus Surveillance (TITAN; SvS) trial, one of the rare randomized controlled trials in
open aortic surgery; patients with ascending aortic aneurysms of 5.0-5.5 cm will be randomized
to surgical intervention or surveillance.

With regards to more concrete recommendations on imaging surveillance of sporadic and
BAV aneurysms, there is limited prospective high- quality population data regarding aneurysm
growth rates. As incidental detection of aneurysms by computed tomography increases, such data
will be necessary to develop evidence-based cost-effective surveillance strategies that safely
capture growth, and risk of adverse aortic events while reducing overall patient life-time
radiation exposure.

Conclusion

Given the continuous changing nature of the landscape of aortic management, the
guidelines should be a living document, updated to include emerging evidence while maintaining
the principles of improving patient outcomes through a multidisciplinary approach and
teamwork. The guidelines are not meant to be prescriptive but rather to provide direction to all
colleagues in the aortic community towards better patient care and long-term outcomes.
References


5. 2010 ACCF/AHA/AATS/ACR/ASA/SCA/SCAI/SIR/STS/SVM Guidelines for the Diagnosis and Management of Patients With Thoracic Aortic Disease Representative Members; Hiratzka LF, Creager MA, Isselbacher EM, Svensson LG; 2014

6. AHA/ACC Guideline for the Management of Patients With Valvular Heart Disease Representative Members; Nishimura RA, Bonow RO, Guyton RA, Sundt TM 3rd.


8. Citation TBD pending JTCVS info


Descending aortic diameter of 5.5 cm or greater is not an accurate predictor of acute type B aortic dissection. J Thorac Cardiovasc Surg. 2011 Sep;142:e101-7.


Table. Top 12 Highlights of the 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease for Cardiothoracic Surgeons

<table>
<thead>
<tr>
<th>Guideline Section</th>
<th>Recommendation</th>
</tr>
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<tbody>
<tr>
<td>4</td>
<td>Multidisciplinary Aortic Teams are those with expertise in cardiac, vascular, or endovascular surgery; radiology; anesthesiology; and critical care of aortic patients. For patients with extensive disease, requiring complex procedure, or high-risk profiles, referral to high-volume centers (30-40 cases/year) is reasonable to optimize outcome (COR IIa).</td>
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<tr>
<td>5,6</td>
<td>Shared decision-making is particularly recommended to discuss thresholds for intervention, choice between open and endovascular repairs, in women who are pregnant or contemplating pregnancy, and in non-syndromic heritable thoracic aortic disease (COR I).</td>
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<tr>
<td>6.1.2.2.3</td>
<td>In addition to a threshold of 5.0 cm, elective replacement of the root or ascending aorta for aortopathy in Marfan syndrome is reasonable at lower thresholds when performed by experienced surgeons in a Multidisciplinary Aortic Team (COR IIa); valve-sparing root replacement may be reasonable in low-risk Marfan patients when performed by experienced surgeons in a Multidisciplinary Aortic Team (COR IIb).</td>
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<tr>
<td>6.1.2.3.3</td>
<td>In patients with Loeys-Dietz syndrome and aortic dilation, the threshold for surgical prophylactic replacement should be informed by the genetic variant, aortic diameter, growth rate, and extra-aortic features, amongst other factors (COR I).</td>
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<tr>
<td>6.1.3.2</td>
<td>In addition to a threshold of 5.5 cm (COR I), elective replacement of the root or ascending aorta for asymptomatic bicuspid aortopathy is reasonable at lower thresholds when performed by experienced surgeons in a Multidisciplinary Aortic Team (COR IIa).</td>
</tr>
<tr>
<td>6.5.1</td>
<td>In patients with asymptomatic aortic root or ascending aneurysms &lt; 5.5 cm, a growth rate of $&gt;0.3$ cm/yr in 2 consecutive years, or $&gt;0.5$ cm in 1 year is an indication for surgery (COR I).</td>
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<tr>
<td>6.5.1</td>
<td>Replacement of sporadic asymptomatic aortic aneurysm is recommended or reasonable in the following situations:</td>
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<td>6.5.4.2</td>
<td>In patients with Marfan syndrome, Loeys-Dietz syndrome, or vascular Ehlers-Danlos syndrome and intact thoracoabdominal aortic aneurysm requiring intervention, open repair is recommended over endovascular repair (COR I).</td>
</tr>
<tr>
<td>7.4.1.1</td>
<td>Transfer of stable patients with acute type A aortic dissection from a low to a high-volume aortic center is reasonable to improve survival (COR IIa).</td>
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<td>7.4.1.3</td>
<td>Contemporary management of acute type A aortic dissection includes, when deemed safe and feasible:</td>
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<tr>
<td>7.5</td>
<td>Urgent repair is recommended for patients with complicated acute type A or type B aortic intramural hematoma (COR I); prompt open repair is recommended for uncomplicated acute type A intramural hematoma (COR I).</td>
</tr>
<tr>
<td>10</td>
<td>For patients with aortic disease, lifestyle guidance should include the following:</td>
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</table>

BP, blood pressure; COR, Class of Recommendation; green shading indicates COR I; yellow shading indicates COR II