JAMA Clinical Guidelines Synopsis

Diagnosis and Management of Aortic Diseases

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GUIDELINE TITLE 2022 ACC/AHA Guideline for the Diagnosis and Management of Aortic Disease

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DEVELOPER American College of Cardiology (ACC) and American Heart Association (AHA)

FUNDING SOURCE ACC and AHA

TARGET POPULATION Adults with or at risk of aortic disease

SELECTED RECOMMENDATIONS

- A multidisciplinary team should determine the most appropriate type of intervention for patients with acute aortic disease requiring urgent repair (strong recommendation [SR; benefit much greater than risk]; expert opinion [EO]). Patients with asymptomatic extensive disease, multiple comorbidities, or who may require complex repairs may be referred to centers with higher case volumes (≥30-40 cases/y) and a multidisciplinary aortic team (moderate recommendation [MR; benefit greater than risk]; limited data [LD]).
- All patients with dilated thoracic aorta or thoracic aortic aneurysm (TAA) should have transthoracic echocardiography (TTE) at the time of diagnosis to assess aortic valve anatomy and function (SR; LD).
- Repair of sporadic aortic root and ascending aortic aneurysms when they reach a maximum diameter of 5.5 cm is strongly recommended (SR; nonrandomized studies [NR]); it is reasonable to repair at a diameter of 5.0 cm when care can occur at a high-volume center with management by a multidisciplinary aortic team (MR; NR). It is reasonable to repair the aorta in patients at increased risk of adverse aortic events, and some genetic aortopathies at even lower thresholds (MR; LD).
- First-degree relatives of patients with aortic root aneurysms, ascending aortic aneurysms, or history of aortic dissection should have screening for aortic disease (SR; LD).
- Preconception counseling on risks of pregnancy-associated aortic dissection is recommended for patients with bicuspid aortic valve and aortic dilatation, syndromic and nonsyndromic thoracic aortic disease, and Turner syndrome (SR; LD).

Summary of the Clinical Problem

Type A aortic dissection has a mortality rate of 57% without emergency surgery and up to 25% with emergency surgery.[1] This guideline addresses thoracic and abdominal aortic disease, genetic and hereditable aortopathies, peripheral artery disease, bicuspid aortic valves, and Turner syndrome.[2] Herein, we focus mainly on recommendations for aneurysms that involve the aortic root and/or the ascending aorta.

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Characteristics of the Guidelines Source

These guidelines (**Table**) were developed and funded by the ACC and AHA. The writing committee consisted of experts in cardiology, internal medicine, interventional medicine, surgery, radiology, anesthesiology, and nursing, as well as a patient representative. All committee members disclosed conflicts of interest.

Evidence Base

Patients with acute type A aortic dissection benefit from multidisciplinary teams, with improved outcomes sustained for 5 years after team formation ($P < .002$; absolute rates not given).[2] Teams typically include cardiac and vascular surgeons, endovascular and imaging specialists, anesthesiologists, and intensivists experienced in managing aortic disease. Centers with high case volume and experienced operators have improved outcomes for patients with TAA. An analysis of 13 358 elective aortic root and aortic valve–ascending aortic procedures at 741 North American hospitals found an overall unadjusted mortality rate of 4.48%. After adjustment for risk, in centers performing 30 to 100 cases per year, the odds ratio for mortality was 0.42 (95% CI, 0.31-0.58) vs 0.55 (95% CI, 0.42-0.73) for centers with 13 to 30 cases per year and 0.79 (95% CI, 0.62-1.00) for those with 6 to 13 cases annually.[2]

Although all patients diagnosed with aneurysms of the aortic sinus or TAA should have TTE to assess aortic valve structure and function, the frequency of follow-up imaging (echocardiogram, computed tomography, or magnetic resonance imaging) should be based on aneurysm size, risk factors, and rate of growth. Follow-up can vary between 6 and 24 months.

In a major change, the guideline suggests that it is reasonable to repair sporadic aneurysms of the aortic root or ascending aorta in select patients with low surgical risk when the aneurysm reaches 5.0 cm, provided management is at a high-volume center by experienced surgeons as part of a multidisciplinary aortic team. This suggestion is based on observational data from the Multi-Ethnic Study of Atherosclerosis database, which modeled risk of dissection at various aortic diameters relative to 3.4 cm or smaller: aorta diameters were less than 3.5 cm in 79.2%, 3.5 to 3.9 cm in 18.0%, 4.0 to 4.4 cm in 2.6%, and 4.5 cm or greater in 0.22%. Individuals with an aorta larger than

Table. Guideline Rating[10]

Standard	Rating
Establishing transparency	Good
Management of conflict of interest in the guideline development group	Good
Guideline development group composition	Good
Clinical practice guideline–systematic review intersection	Good
Establishing evidence foundations and rating strength for each of the guideline recommendations	Good
Articulation of recommendations	Good
External review	Fair
Updating	Good
Implementation issues	Fair

4.5 cm were 6305 times more likely to have aortic dissection than those with an aorta smaller than 3.5 cm.[3] The prior cutoff of 5.5 cm for surgery was based on natural history studies that examined aortic diameter at the time of an adverse event[4,5] and supported a strong recommendation to repair aortic root aneurysms or TAAs greater than 5.5 cm in asymptomatic patients.

Cross-sectional imaging or echocardiography should be done to screen first-degree relatives of all patients with aortic root aneurysms, ascending aortic aneurysms, or history of aortic dissection. Nonrandomized data demonstrate that screening of relatives of patients with nonsyndromic TAA identifies affected relatives in up to half of those screened, many with significant aortic dilatation.[6] No upper age limit in an index patient precludes screening first-degree relatives when screening would otherwise be appropriate.

Pregnant patients with aortic disease should receive a screening echocardiogram at least every trimester and several weeks postpartum based on limited observational studies.[2] It is reasonable to have maternal-fetal medicine and cardiology specialists in a center that can perform emergent aortic repair manage the care of pregnant patients with aortic aneurysms or who are at risk of aortic disease.

Patients with significant aortic disease should avoid exercise involving intense isometric contraction or prolonged Valsalva maneuvers, such as heavy weight lifting, in favor of light weight lifting and low-intensity aerobic exercise. These recommendations are based on consensus expert opinion founded in case series and animal models examining the hemodynamic effects of various types of exercise, which have shown transient increase of systolic BP to greater than 300 mm Hg during heavy lifting with Valsalva maneuvers.

Benefits and Harms

The benefits and harms of these recommendations balance operative risk with risk of spontaneous aortic dissection or rupture. Lower aortic dimension thresholds for surgery will increase the number of and cost associated with surgeries and may raise the risk of additional complications, including coronary artery bypass graft surgery, pacemaker implantation, mechanical circulatory support, and death. The guideline's focus on higher risk types for screening and management is a potentially significant advance. For example, most genetic aortopathies are currently underdiagnosed and are recognized only after rupture.

Discussion

There are few randomized clinical trials that inform recommendations in this guideline. The most notable changes from previous guidelines (consideration of a lower surgical threshold for TAA repair in select scenarios and increasing family screening) are significant practice changes backed by suggestive but lower-quality evidence. A number of recommendations address risk factor management, including medication and lifestyle changes. These include BP management goals (medication is advised for BP ≥130/80 mm Hg), use of statins in abdominal aortic aneurysm, and smoking cessation. While aortic-specific data are limited, low-dose aspirin may be considered, unless contraindicated, if there are concomitant atheroma and/or penetrating aortic ulcers.

Areas for Future Investigation

Knowledge gaps remain in management and recognition of aortic disease. One ongoing randomized clinical trial compares medical surveillance with early intervention in patients with TAAs measuring 5.0 cm to 5.4 cm.[7] Advances in endovascular technology will require research across a spectrum of aortic anatomy and to define optimal patient selection and operator training requirements.

The recommendation for exercise limits deserves additional supportive evidence, as it is not clear to what degree aortic dissections are precipitated by increased wall stress or decreased wall strength.[8] The recommendation of referrals to high-volume centers for certain complex asymptomatic patients may face insurance and travel barriers. Studies are needed to better stratify which patients merit travel to high-volume centers. Similarly, prospective studies show that despite a lower overall incidence, women appear to have a 2- to 3-fold increased risk of fatal aortic rupture relative to men (hazard ratio, 2.60; 95% CI, 1.58-4.29; $P < .001$).[9] Determining sex-specific surgical thresholds for aneurysms of the aortic root and TAA are a high priority for future investigation.

ARTICLE INFORMATION

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