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SOCIETAL STATEMENT

2024 Hypertrophic Cardiomyopathy Guideline-at-a-Glance

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INTRODUCTION

The 2024 AHA/ACC/AMSSM/HRS/PACES/SCMR Guideline for the Management of Hypertrophic Cardiomyopathy (AHA/ACC/Multisociety HCM Guideline) provides guidance for clinicians on the management of patients with hypertrophic cardiomyopathy (HCM).[1] The guideline contains updated, evidence-based recommendations that build on those from the 2020 HCM Guideline[2] in the following areas: risk assessment of sudden cardiac death (SCD), exercise recommendation nuances, and new pharmacological options. This Guideline-at-a-Glance highlights practice-changing recommendations from the guideline to accelerate adoption into clinical practice.

ACC guideline dissemination is an organization-wide effort overseen by the Solution Set Oversight Committee to ensure the integration of guideline content throughout ACC's clinical policy, education, registry, membership, and advocacy efforts. For each guideline, an individual ACC Guideline Dissemination Workgroup is created to influence dissemination strategy and to create tools to facilitate the implementation of key changes in practice. These tools include the *JACC* **Central Illustration**, as well as tables highlighting updates in the 2024 AHA/ACC/Multisociety HCM Guideline and comparisons to the 2023 European Society of Cardiology (ESC) Cardiomyopathy Guideline.[3]

TOP 10 TAKE-HOME MESSAGES

The following Top 10 Take-Home Messages are taken directly from the AHA/ACC/Multisociety HCM Guideline. The ACC HCM Guideline Dissemination Workgroup selected 3 of the Top 10 Take-Home Messages (in bold) as key themes for this Guideline-at-a-Glance as they represent the most impactful recommendation changes compared to previous guidelines and address known gaps in clinical practice.

1. Shared decision-making is essential to provide the best clinical care. This involves thoughtful dialogue among patients, families, and their care team in which health care professionals present all available testing and treatment options; discuss the risks, benefits, and applicability of those options to the individual patient; and ensure the patient expresses their personal preferences and goals to develop their treatment plan.
2. Although the primary cardiology team can initiate evaluation, treatment, and longitudinal care, referral to multidisciplinary HCM centers with appropriate expertise can be important to optimizing care for patients with HCM. Challenging treatment decisions—where reasonable alternatives exist, where the strength of recommendation is weak (eg, any decision relying on a Class of Recommendation 2b) or is particularly nuanced (eg, interpretation of genetic testing; primary prevention implantable cardioverter-defibrillator decision-making), and for HCM-specific invasive procedures—may critically benefit from involving specialized HCM centers.
3. Careful ascertainment of family history, counseling patients with HCM about the potential for genetic transmission of HCM, and options for genetic testing are cornerstones of care. Screening first-degree family members of patients with HCM, using either genetic testing, serial imaging, or electrocardiographic surveillance as appropriate, can begin at any age and can be influenced by specifics of the patient and family history and family preference. Because screening

*On behalf of the ACC Solution Set Oversight Committee.

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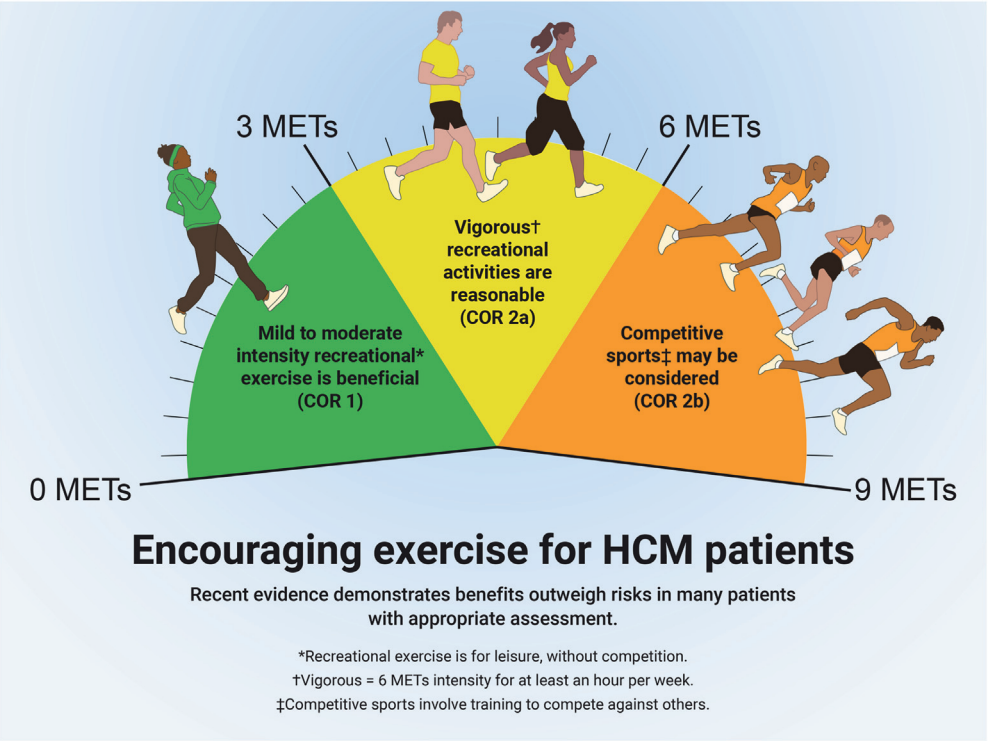
CENTRAL ILLUSTRATION **2024 AHA/ACC/Multisociety HCM Guideline**



Coylewright M, et al. J Am Coll Cardiol. 2024;83(23):2406–2410.

COR = class of recommendation; HCM = hypertrophic cardiomyopathy; METs = metabolic equivalents.

recommendations for family members hinge on the pathogenicity of any detected variants, the reported pathogenicity should be reconfirmed every 2 to 3 years, and input from specialized HCM centers with genetics expertise may be valuable.

4. Assessing a patient's risk for sudden cardiac death is an important component of management. Integrating the presence or absence of established risk markers with tools to estimate individual risk score will facilitate the patient's ability to participate in decision-making regarding implantable cardioverter defibrillator placement. These discussions should incorporate a patient's personal level of risk tolerance and their specific treatment goals.
5. **The risk factors for sudden cardiac death in children with HCM carry different weights and components than those used in adult patients. Pediatric risk stratification also varies with age and must account for different body sizes. Coupled with the complexity of placing implantable cardioverter-defibrillators in young patients with anticipated growth and a higher risk of device complications, the threshold for implantable cardioverter-defibrillator implantation in children often differs from adults. These differences are best addressed at comprehensive HCM centers with expertise in caring for children with HCM. New risk calculators, specific to children and adolescents, have been validated and can help young patients and their families contextualize their estimated risk of sudden cardiac death.**
6. **Cardiac myosin inhibitors are now available to treat patients with symptomatic obstructive HCM. This new class of medication inhibits actin-myosin interaction, thus decreasing cardiac contractility and reducing left ventricular outflow tract obstruction. Mavacamten is currently the only U.S. Food and**

Drug Administration-approved agent. These agents can be beneficial for patients with obstructive HCM who do not derive adequate symptomatic relief from first-line drug therapy.

7. Invasive septal reduction therapies (surgical septal myectomy and alcohol septal ablation), when performed by experienced HCM teams at dedicated centers, can provide safe and effective symptomatic relief for patients with drug-refractory or severe outflow tract obstruction. Given the data on the significantly improved outcomes at comprehensive HCM centers, these decisions represent an optimal opportunity for referral.
8. Patients with HCM and persistent or paroxysmal atrial fibrillation have a sufficiently increased risk of stroke such that oral anticoagulation with direct-acting oral anticoagulants (or alternatively warfarin) should be considered the default treatment option irrespective of the CHA_2DS_2-VASc score. New tools to stratify risk for incident atrial fibrillation have been developed and may assist in determining the frequency of screening patients with ambulatory telemetry. Because rapid atrial fibrillation is often poorly tolerated in patients with HCM, maintenance of sinus rhythm and rate control are key treatment goals.
9. Exercise stress testing is particularly helpful in determining overall exercise tolerance and for latent exercise provoked left ventricular outflow tract obstruction. Because children may not describe symptoms readily, routine exercise testing can be particularly important for young patients.
10. **Increasingly, data affirm that the beneficial effects of exercise on general health are extended to patients with HCM. Healthy recreational exercise (light [<3 metabolic equivalents], moderate [3-6 metabolic equivalents], and vigorous [>6 metabolic equivalents] intensity levels) has not been associated with increased risk of ventricular arrhythmia events in short-term studies. If patients pursue rigorous exercise training for the purpose of performance or competition, it is important to engage in a comprehensive discussion and seek input from expert HCM professionals regarding the potential risks and benefits, to develop an individualized training plan, and to establish a regular schedule for reevaluation.**

JACC ILLUSTRATION

Central Illustration: Encouraging Exercise for HCM Patients

A key message in the 2024 guideline is the growing evidence regarding the beneficial effects of exercise for patients with HCM. There has been a long-standing belief among clinicians that patients with HCM should not exercise, but recent evidence demonstrates that benefits outweigh the risks in many patients with appropriate assessment.[4,5]

The *JACC* **Central Illustration** for the AHA/ACC/Multisociety HCM Guideline focuses on the changing norms surrounding recreational exercise in patients with HCM. The image is meant to encourage clinicians to advise their HCM patients that recreational exercise can improve their health. Tools for discussing exercise with patients can be found at CardioSmart.org/HCM.

This clinician tool focuses on Top 10 Take-Home Message 10. For additional information, see Section 9.1. "Recreational Physical Activity and Competitive Sports."[1]

COMPARISON TO PREVIOUS ACC/AHA GUIDELINE

The AHA/ACC/Multisociety HCM Guideline includes and updates content previously covered in the 2020 HCM guideline.[2] **Table 1** outlines changes in SCD risk assessment, use of cardiac myosin inhibitors, and beneficial effects of exercise in patients with HCM between the 2020 and the 2024 version of the guideline. The comparison focuses on Top 10 Take-Home Messages 5, 6, and 10.

For further details, refer to the corresponding sections of the 2024 AHA/ACC/Multisociety HCM Guideline:[1]

- Section 7.1.2. "SCD Risk Assessment in Children and Adolescents With HCM";
- Section 8.1.1. "Pharmacological Management of Symptomatic Patients With Obstructive HCM";
- Section 9.1. "Recreational Physical Activity and Competitive Sports."

COMPARISON OF AHA/ACC/MULTISOCIETY HCM GUIDELINE TO ESC CARDIOMYOPATHIES GUIDELINE

The ESC published a guideline on the management of cardiomyopathies in 2023. **Table 2** compares the recommendations assessing the risk of SCD in children and adolescents, the use of cardiac myosin inhibitors, and recommendations regarding exercise between the 2024 AHA/ACC/Multisociety HCM Guideline[1] and the 2023 ESC Guidelines for the Management of Cardiomyopathies.[3] The comparison focuses on Top 10 Take-Home Messages 5, 6, and 10.

For further details, refer to the corresponding sections of the 2023 ESC Cardiomyopathies Guideline:[3]

- Section 7.1.5. "Sudden Cardiac Death Prevention in Hypertrophic Cardiomyopathy";
- Section 7.1.4. "Management of Symptoms and Complications";

TABLE 1 Select Differences Between the 2020 and the 2024 AHA/ACC/Multisociety HCM Guidelines

	2020[2]	2024[1]
SCD risk assessment in children and adolescents (Top 10 Take-Home Message 5)	No corresponding guideline recommendations; Risk Stratification Considerations in Pediatric Patients was discussed in text.	For children and adolescents with HCM, a comprehensive, systematic noninvasive SCD risk assessment at initial evaluation and every 1 to 2 years thereafter is recommended and should include evaluation of these risk factors (Figures 1 and 3, Table 8): • Personal history of cardiac arrest or sustained ventricular arrhythmias; • Personal history of syncope suspected by clinical history to be arrhythmic; • Family history in close relative of premature HCM-related sudden death, cardiac arrest, or sustained ventricular arrhythmias; • Maximal LV wall thickness, EF, LV apical aneurysm; • NSVT episodes on continuous ambulatory electrocardiographic monitoring (COR 1). For children and adolescents with HCM who have a borderline risk for SCD, or in whom a decision to proceed with ICD placement remains uncertain after clinical assessment that includes personal and family history, echocardiography, and ambulatory electrocardiographic monitoring, CMR imaging is beneficial to assess for extent of myocardial fibrosis with LGE (Table 8) (COR 1).
		For patients <16 years of age with HCM, it is reasonable to calculate an estimated 5-year sudden death risk that includes echocardiographic parameters (interventricular septal thickness in diastole, LV posterior wall thickness in end-diastole, left atrial diameter, maximal LVOT gradient) and genotype, which may be useful during shared decision-making for ICD placement (Table 8) (COR 2a).
Cardiac myosin inhibitors (Top 10 Take-Home Message 6)	For patients with obstructive HCM who have persistent severe symptoms attributable to LVOTO despite beta-blockers or non-dihydropyridine calcium channel blockers, **either adding disopyramide in combination with 1 of the other drugs, or SRT performed at experienced centers**, is recommended (COR 1).	For patients with obstructive HCM who have persistent symptoms attributable to LVOTO despite beta blockers or nondihydropyridine calcium-channel blockers, **adding a myosin inhibitor (adult patients only), or disopyramide (in combination with an atrioventricular nodal blocking agent), or SRT performed at experienced centers**, is recommended (COR 1).
Beneficial effects of exercise on patients with HCM (Top 10 Take-Home Message 10)	For patients with HCM, participation in **high-intensity recreational activities or moderate- to high-intensity competitive sports** activities may be considered after a comprehensive evaluation and shared discussion, repeated annually with an expert provider who conveys that the risk of sudden death and ICD shocks may be increased, and with the understanding that eligibility decisions for competitive sports participation often involve third parties (eg, team physicians, consultants, and other institutional leadership) acting on behalf of the schools or teams (COR 2b).	For patients with HCM, participation in **vigorous recreational activities** is reasonable after an annual comprehensive evaluation and shared decision-making with an expert professional who balances potential benefits and risks (COR 2a).
		For patients with HCM who are capable of a high level of physical performance, participation in **competitive sports** may be considered after review by an expert provider with experience managing athletes with HCM who conducts an annual comprehensive evaluation and shared decision-making that balances potential benefits and risks (COR 2b).
	No corresponding guideline recommendation.	For most patients with HCM, universal restriction from vigorous physical activity or competitive sports is not indicated (COR 3: No benefit).

Colors in the table align with the classification system found in Table 3, "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 AHA/ACC/Multisociety HCM Guideline.[1]

ACC = American College of Cardiology; AHA = American Heart Association; COR = Class of Recommendation; CMR = cardiovascular magnetic resonance; EF = ejection fraction; HCM = hypertrophic cardiomyopathy; ICD = implantable cardioverter-defibrillator; LGE = late gadolinium enhancement; LV = left ventricular; LVOT = left ventricular outflow tract; LVOTO = left ventricular outflow tract obstruction; NSVT = nonsustained ventricular tachycardia; SCD = sudden cardiac death; SRT = septal reduction therapy.

- Section 8.1.3. "Exercise Recommendations in Hypertrophic Cardiomyopathy."

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TABLE 2 Select Comparison of AHA/ACC/Multisociety HCM Guideline and ESC Cardiomyopathies Guideline

	ESC Cardiomyopathies Guideline[3]	AHA/ACC/Multisociety HCM Guideline[1]
SCD risk assessment in children and adolescents (Top 10 Take-Home Message 5)	Validated pediatric-specific risk prediction models (eg, HCM Risk-Kids) are recommended as a method of estimating risk of sudden death at 5 years in patients aged <16 years for primary prevention (COR 1).	For patients <16 years of age with HCM, it is reasonable to calculate an estimated 5-year sudden death risk that includes echocardiographic parameters (interventricular septal thickness in diastole, LV posterior wall thickness in end-diastole, left atrial diameter, maximal LVOT gradient) and genotype, which may be useful during shared decision-making for ICD placement (Table 8) (COR 2a).
Cardiac myosin inhibitors (Top 10 Take-Home Message 6)	Cardiac myosin ATPase inhibitor (mavacamten), titrated to maximum tolerated dose with echocardiographic surveillance of LVEF, should be considered in addition to a beta-blocker (or, if this is not possible, with verapamil or diltiazem) to improve symptoms in adult patients with resting or provoked LVOTO (COR 2a). Cardiac myosin ATPase inhibitor (mavacamten), titrated to maximum tolerated dose with echocardiographic surveillance of LVEF, should be considered as monotherapy in symptomatic adult patients with resting or provoked LVOTO (exercise or Valsalva maneuver) who are intolerant or have contraindications to beta-blockers, verapamil/diltiazem, or disopyramide (COR 2a).	For patients with obstructive HCM who have persistent symptoms attributable to LVOTO despite beta blockers or nondihydropyridine calcium-channel blockers, then adding a myosin inhibitor (adult patients only), or disopyramide (in combination with an atrioventricular nodal blocking agent), or SRT performed at experienced centers, is recommended (COR 1).
Beneficial effects of exercise on patients with HCM (Top 10 Take-Home Message 10)	High-intensity exercise and competitive sport may be considered in asymptomatic low-risk individuals with morphologically mild hypertrophic cardiomyopathy in the absence of resting or inducible left ventricular outflow obstruction and exercise-induced complex ventricular arrhythmias (COR 2b).	For patients with HCM, participation in vigorous recreational activities is reasonable after an annual comprehensive evaluation and shared decision-making with an expert professional who balances potential benefits and risks (COR 2a). For patients with HCM who are capable of a high level of physical performance, participation in competitive sports may be considered after review by an expert provider with experience managing athletes with HCM who conducts an annual comprehensive evaluation and shared decision-making that balances potential benefits and risks (COR 2b).
	No corresponding guideline recommendation.	For most patients with HCM, universal restriction from vigorous physical activity or competitive sports is not indicated (COR 3: No benefit).

Colors in the table align with the classification system found in Table 3, "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 AHA/ACC/Multisociety HCM Guideline.[1]

ACC = American College of Cardiology; AHA = American Heart Association; ATPase = adenosine triphosphatase; COR = Class of Recommendation; ESC = European Society of Cardiology; HCM = hypertrophic cardiomyopathy; ICD = implantable cardioverter-defibrillator; LV = left ventricular; LVEF = left ventricular ejection fraction; LVOT = left ventricular outflow tract; LVOTO = left ventricular outflow tract obstruction; SRT = septal reduction therapy.

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