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INTERMEDIATE

CASE REPORT: CLINICAL CASE

# Applying Shared Decision Making to Sports Participation for a Patient With Hypertrophic Cardiomyopathy

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## ABSTRACT

A patient with hypertrophic cardiomyopathy and a cardio-defibrillator implanted for primary prevention would like to compete on the ski team at his school. This case illustrates how a shared decision-making approach can be applied when counseling patients with hypertrophic cardiomyopathy about exercise and sports participation. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:6–9) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 15-year-old male with a history of familial hypertrophic cardiomyopathy (HCM) presented to discuss participation in exercise and competitive sports. He was diagnosed with HCM at age 2 and subsequent echocardiograms showed marked septal hypertrophy and a resting left ventricular outflow tract (LVOT) gradient of 36 mm Hg. He was started on beta-blockade therapy and has remained on monotherapy since. At age 9 years, cardiac magnetic resonance imaging showed a septal wall thickness of 32 mm with late gadolinium enhancement. His resting LVOT gradients have been stable over the years in the moderate range, and he has remained asymptomatic. An implantable cardioverter-defibrillator (ICD) was inserted due to his hypertrophy burden and a family history of sudden cardiac death.

## LEARNING OBJECTIVES

- To summarize the historical context of guideline-based exercise restrictions in patients with HCM.
- To recognize the nuances and complexities of counseling patients with HCM on participation in competitive sports.
- To restate the elements of a shared decision making approach to sports participation in athletes with HCM.

The patient participated in gym class but not in competitive sports in middle school. In high school, he joined the golf team but has not participated in gym class because of uncertainty around the intensity of exercise that would be considered “safe.” He engages in low to moderate-intensity recreational activity, including weightlifting, jogging, skiing, and pick-up basketball games. He does not have any physical limitations or symptoms with exercise. He

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has no other medical history. His examination was notable for a 3/6 grade systolic ejection murmur that increases with squat-to-stand.

## FAMILY HISTORY

The patient's paternal uncle died suddenly in his 50's while jogging. The patient's father has a primary prevention ICD, but it has never fired. The patient's 2 siblings each have primary prevention subcutaneous ICDs. Genetic testing identified a pathogenic variant in the *MYH7* gene that was also present in his father and 2 siblings.

## DIAGNOSTIC TESTING

Electrocardiogram showed left ventricular hypertrophy with repolarization abnormalities (**Figure 1**). Transthoracic echocardiography showed left atrial dilation, interventricular septum measuring 30 mm, systolic anterior motion of the mitral valve with peak gradient of 48 mm Hg at rest, and moderate to severe mitral regurgitation with an ejection fraction of 70% (Videos 1, 2, 3, and 4).

## MANAGEMENT

Because he was currently asymptomatic, beta-blockade was continued for treatment of outflow tract obstruction with plans to perform regular cardiopulmonary exercise testing with echocardiography.

A shared decision-making discussion was initiated with the patient and his parents about participation in physical education class and extracurricular sports (**Table 1**). The patient and his parents expressed a strong desire that he participate fully in gym class, the golf team, and ski club. He and his parents were fully cognizant of the potential risks of exercise-induced episodes of arrhythmia or defibrillator shocks, which were estimated to be very low but not zero. The following precautions were advised:

- It is preferable not to train alone.
- It should be ensured that the teams and school have at least 1 automated external defibrillator on site and accessible, with working batteries and an emergency action plan.
- All coaches, teammates, and physical education teachers should know about his heart condition.
- He should build up his conditioning slowly.
- He should be permitted to rest as needed and not be graded based on performance or timed events in physical education class.
- He should be able to take frequent water breaks.

### ABBREVIATIONS AND ACRONYMS

**HCM** = hypertrophic cardiomyopathy

**ICD** = implantable cardioverter-defibrillator

**LVOT** = left ventricular outflow tract

**SAM** = systolic anterior motion

**FIGURE 1** Electrocardiogram at Initial Visit

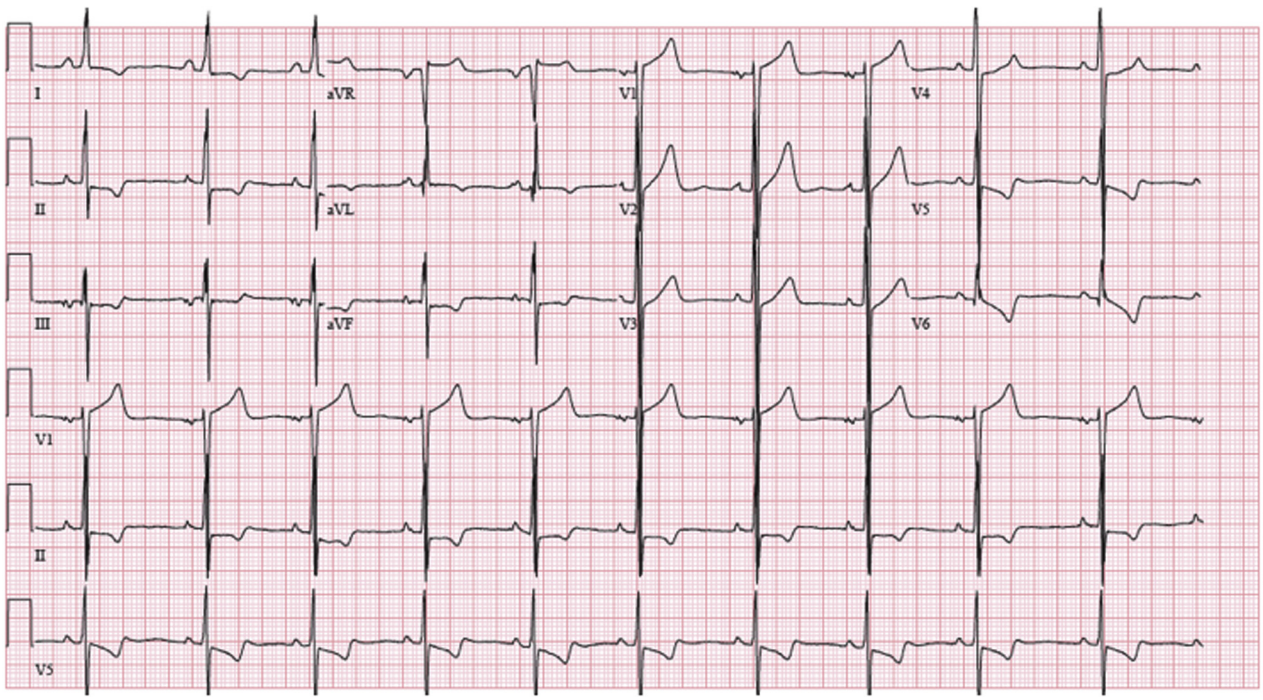

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**TABLE 1 Steps in Shared Decision Making for Sports Participation in Athletes With HCM**

| Steps of Shared Decision Making | Details |
|---|---|
| 1. Confirmation of Diagnosis | Often uses advanced imaging and genetic testing |
| 2. Individualized risk stratification and treatment plans | Testing: ECG, ambulatory monitoring, stress testing, echocardiography, CMR, family history, genetic testing.<br>Treatment: role for pharmacotherapy, intervention, and risk stratification for SCD |
| 3. Patient and family education | Discuss potential risks/benefits, review relevant research studies, acknowledge uncertainty, provide your own expert opinion |
| 4. Assessment of the patient's preferences and values | Discuss the role that athletics plays in their lives and athletic aspirations; gauge their risk tolerance and comfort with uncertainty |
| 5. Synthesis of information and arrival at a shared decision | Review the treatment plan, summarize key points, discuss the balance of risks and benefits |
| 6. Stakeholder engagement | Engage third parties, including school athletic departments, coaches, and team physicians |
| 7. Longitudinal care and follow-up | Regular surveillance (at least yearly): monitoring for arrhythmia, stress testing, and imaging<br>Reinforce precautions and best practices for athletes choosing to continue or returning to play and for recreational exercise |

CMR = cardiac magnetic resonance; ECG = electrocardiography; HCM = hypertrophic cardiomyopathy; SCD = sudden cardiac death.

## DISCUSSION

Whether exercise at any level is associated with an increased risk of ventricular arrhythmia and sudden cardiac death in patients with HCM has been a subject of much debate and controversy. The RESET-HCM (Randomized Exploratory Study of Exercise Training in Hypertrophic Cardiomyopathy) study, in which patients were randomized to moderate intensity exercise or usual activity, showed a significant improvement in exercise capacity with exercise training without a signal for harm, including episodes of ventricular arrhythmia (1). That study provided the evidence base for a new Class 1 recommendation in the 2020 American Heart Association/American College of Cardiology (AHA/ACC) guidelines for the diagnosis and management of patients with HCM that supports recreational, moderate intensity exercise to improve exercise capacity, physical functioning, and quality of life (2).

Uncertainty remains for potential arrhythmic risks associated with more vigorous or competitive physical activities. In 2005, the 36th Bethesda Guidelines recommended that all patients with HCM be excluded from vigorous exercise and competitive sports (3), a recommendation that was upheld by the AHA/ACC guideline committee for the clinical management of HCM in 2011 (4). Since those guidelines were published, a number of studies have demonstrated that the absolute risk of sudden cardiac death associated with HCM in adults was low and suggested that the risk may not be increased with engagement in vigorous exercise or competitive sports (4). In a study of ~400 competitive athletes (65 with HCM) with implantable defibrillators, there were no deaths, device malfunctions, or damage associated with participation in competitive athletics. The proportion of shocks in most participants, including those individuals with HCM, was not greater during competition or practice than during other leisure activities (5,6).

The writing committee for the 2020 AHA/ACC guidelines carefully weighed the existing evidence and uncertainties surrounding risks of engagement in vigorous and competitive athletics and balanced these risks against the desire to support patient autonomy in making informed decisions around their lifestyle choices. Ultimately, the committee settled on a Class 2b recommendation that participation in such activities "may be considered, after a comprehensive evaluation and shared discussion, repeated annually, with an expert provider." The nuances around such discussions are significant, particularly in children with HCM. Incorporating input from both the child and the caregivers, acknowledging the paucity of pediatric-specific data to inform the team about risk, understanding what level of risk a school or athletic program is willing to take on, and tackling potential legal ramifications of making decisions with and for a minor are among the many challenges that pediatric HCM specialists face in adopting a shared decision making approach to children's participation in sports. The committee fully acknowledged these challenges, and all were in agreement that more longitudinal data are needed.

## FOLLOW-UP

Several months after his initial visit, the patient had a syncopal episode while jogging ~one-quarter of a mile at a very leisurely pace. Interrogation of his ICD showed an episode of ventricular fibrillation that was appropriately terminated with 1 shock. He was unaware he had been shocked and, apart from a few bruises from the fall, had no memory of the event. He and his family are comfortable with him continuing to exercise, and the importance of exercising with a partner was stressed. He has been doing moderate weight training at home without any more issues and

plans to compete on the ski team in the coming winter.

## CONCLUSIONS

Although there has been a long-standing belief that high-intensity exercise in subjects with HCM is associated with increased risk of arrhythmia or sudden death, mounting evidence suggests that the risks are considerably lower than initially estimated more than a decade ago (7,8). A wealth of data show that cardiorespiratory fitness reduces all-cause mortality in the general population and that randomized clinical trial data in adults with HCM confirm the benefits of aerobic exercise (1,9). In recommending significant restrictions for all patients with HCM, the previous guidelines may have inadvertently led to adverse outcomes associated with sedentary lifestyles (10). The substantial revisions to this section of the US guidelines in 2020 will hopefully lead to improvements in both the physical and the mental wellbeing of patients living with HCM.

## AUTHOR DISCLOSURES

Dr. Day has received a research grant and honoraria from MyoKardia and Tenaya Therapeutics. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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**KEY WORDS** echocardiography, exercise, hypertrophic cardiomyopathy, shared decision making, sports cardiology, sudden cardiac death

**APPENDIX** For supplemental videos, please see the online version of this paper.


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