

THE JOHNS HOPKINS
**Hypertrophic
Cardiomyopathy
Center**



JOHNS HOPKINS
MEDICINE

Welcome to the Johns Hopkins Hypertrophic Cardiomyopathy Center

If you or a family member have been diagnosed recently with hypertrophic cardiomyopathy (HCM), chances are that you have a lot of questions. This guide was designed with you in mind! We want you to get the most out of your visit to the Johns Hopkins HCM Center.

The purpose of this guide is to familiarize you with some of the basics of HCM, introduce you to the expert members of our team, give you an idea of the cardiac testing we may perform, and tell you about some of the advanced treatment options that are available through the Johns Hopkins HCM Center.

Rest assured: You are in good hands!

Jose Madrazo, M.D.
HCM Center Director

Richard Carrick, M.D., Ph.D.
HCM Center Co-Director



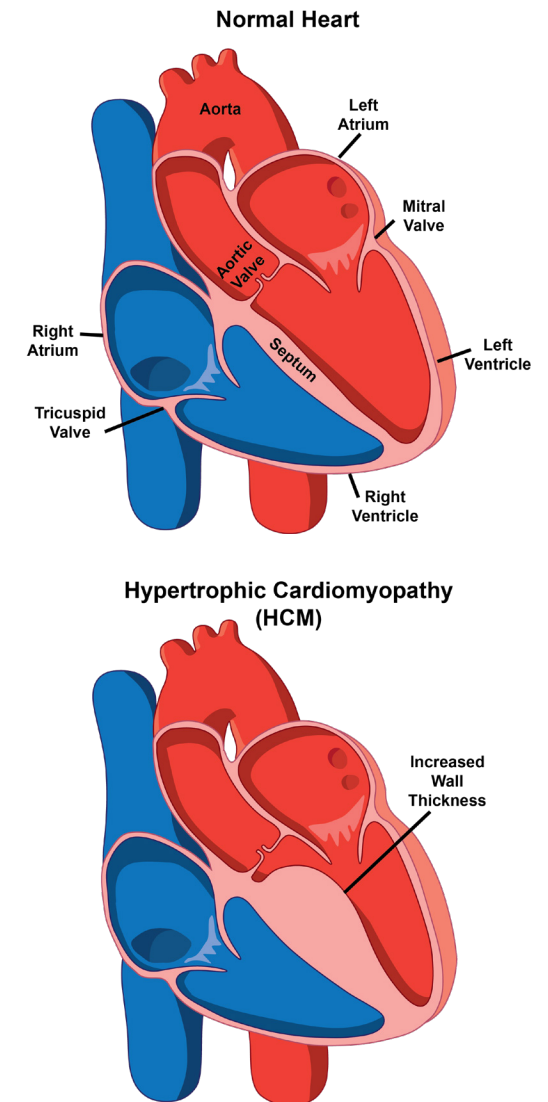
What Is Hypertrophic Cardiomyopathy (HCM)?

Hypertrophic = increased thickness
Cardiomyopathy = disease of the heart muscle

HCM is the most common form of inherited cardiomyopathy, affecting approximately 1 in 500 people. Fortunately, in the era of modern HCM treatment, the prognosis is excellent for the majority of patients.

Patients with HCM have increased thickness of the heart muscle, typically greater than 1.5 centimeters (normal is less than 1.0 centimeters). For most patients with HCM, heart function is normal.

Patients with HCM are at increased risk for abnormal heart rhythms (arrhythmias), and may develop symptoms such as shortness of breath, dizziness, or chest discomfort.



Preparing for Your Visit

At the Johns Hopkins HCM Center, we want you to feel informed and supported from the very start.

These tips will help you get the most out of your visit.

What to bring:

- A list of your current medications (including dosages and frequency)
- Copies of any recent cardiac test results (echocardiograms, exercise testing, cardiac MRIs, heart rhythm monitoring, heart catheterization)
- Your family history, particularly related to heart disease or sudden death
- Your insurance card and a valid photo ID
- A written list of questions or concerns you'd like to discuss
- Any medical paperwork related to your condition that needs to be completed by a member of our HCM Center

Before your appointment:

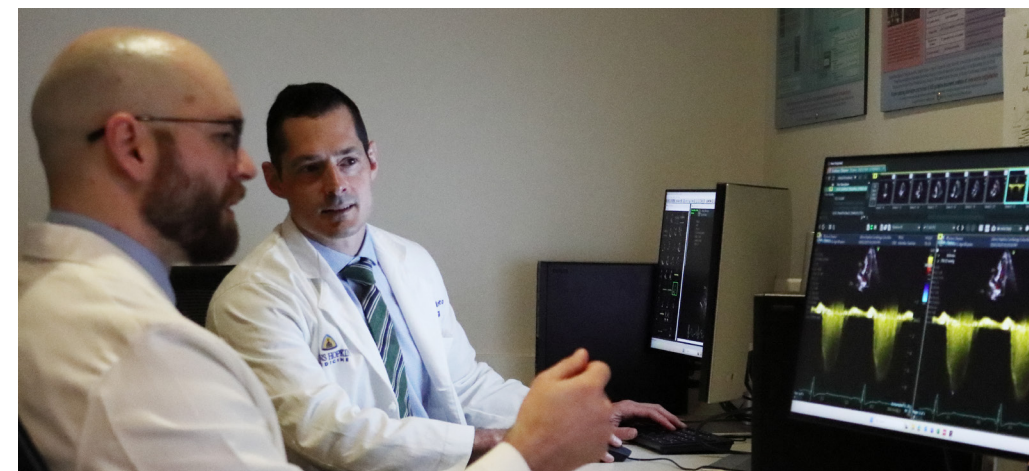
- Complete any pre-visit paperwork through MyChart or as directed.
- Arrange for a family member or support person to accompany you, if possible—especially for your first visit.

Plan to arrive at least 15 minutes before your appointment — there will be a lot to talk about!



Cardiac Testing in HCM

Cardiac testing is an essential part of the medical care of a patient with HCM. At Johns Hopkins, we offer a full spectrum of cardiac testing, including echocardiography, exercise testing, cardiac MRI, genetic testing, heart rhythm monitoring, and heart catheterization. These tests help us determine your risk for abnormal heart rhythms and guide HCM-specific treatments.



Genetics and Family Screening



HCM is an inherited heart condition, which means it runs in families. If one member of a family is affected by HCM, other family members may also be at risk for developing disease. All first-degree relatives (parents, siblings and children) should undergo cardiac testing to screen for HCM.

In some patients, genetic testing may reveal abnormalities (variants) that cause HCM by interfering with the normal function of proteins of the heart muscle. Identifying these genetic variants can be helpful for several reasons. It can help us rule out alternative diagnoses, determine which family members are or are not at risk for developing HCM, identify high-risk forms of HCM that require closer monitoring, and to guide preconception planning. We work closely with our pediatric cardiology team to care for our younger HCM patients and at-risk family members.



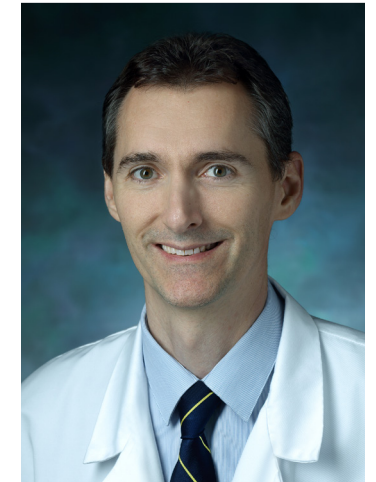
Bryana Rivers, CGC
Cardiac Genetics

Arrhythmias in HCM

Patients with HCM are at increased risk for abnormal heart rhythms (arrhythmias). The two most common types of arrhythmia are:

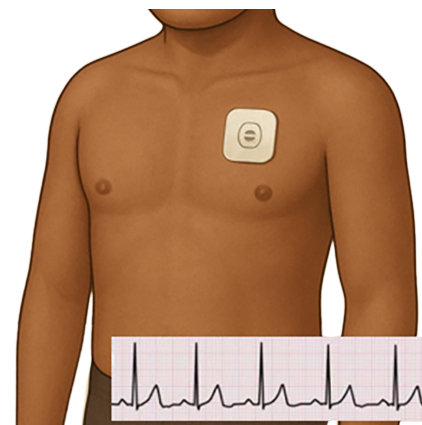
1) Atrial fibrillation (AF): an arrhythmia coming from the top chamber of the heart. Patients with HCM and AF are at increased risk for blood clots, and they may benefit from blood thinners to prevent stroke.

2) Ventricular arrhythmias (VA): abnormal heart rhythms coming from the bottom chambers of the heart. VA can be life-threatening, and some patients with HCM who are found to be at high risk for VA after cardiac testing may benefit from an implantable cardioverter defibrillator (ICD) to prevent sudden cardiac death. Heart rhythm monitoring and cardiac MRI help guide this decision-making.

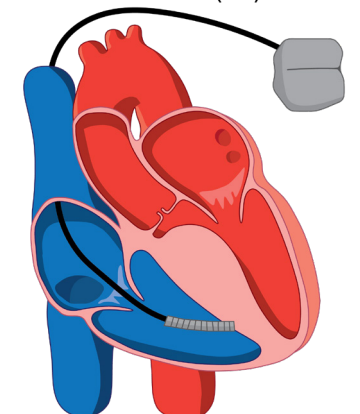


Dr. Andreas Barth
Cardiac Electrophysiology
Director, Inherited Heart
Diseases

Ambulatory Rhythm Monitor

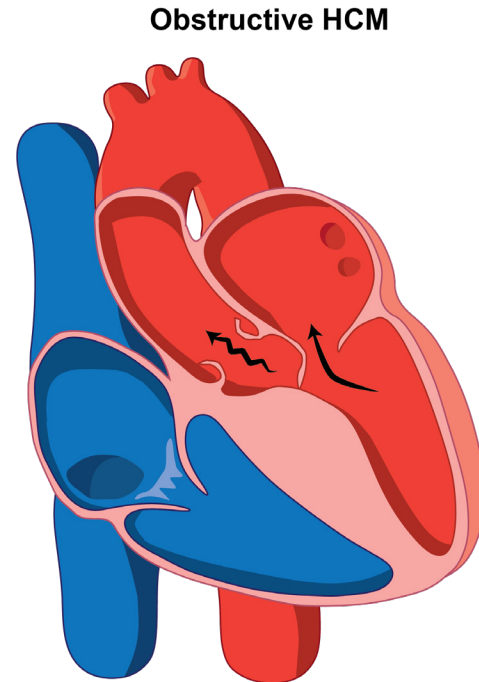


Implantable Cardioverter
Defibrillator (ICD)



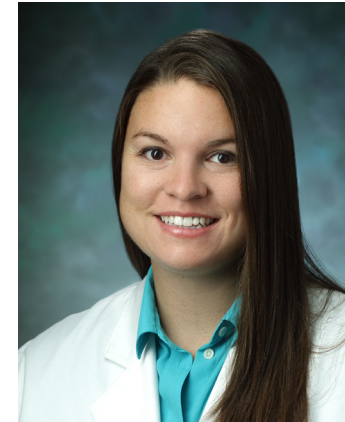
Obstruction in HCM

The most common cause of symptoms in patients with HCM is a phenomenon known as obstruction. In HCM with obstruction, the thickened muscle of the heart leads to narrowing in the path that blood flows through as it leaves the heart. This is just like when you put your thumb over the end of a garden hose – the heart has to work harder to pump blood past the area of narrowing. Obstruction often produces a heart murmur, and it often causes patients to experience shortness of breath, chest pain, or dizziness, particularly during exercise.



We measure the severity of obstruction using heart ultrasound (echocardiography). The degree of obstruction may change from day to day – things like being dehydrated, eating a large meal, or hot weather can make obstruction worse. Some patients only have obstruction with exercise, and we need to perform a treadmill stress echocardiogram.

Treating Obstruction with Medicines



Sarah Riley, CRNP
JHH HCM Center

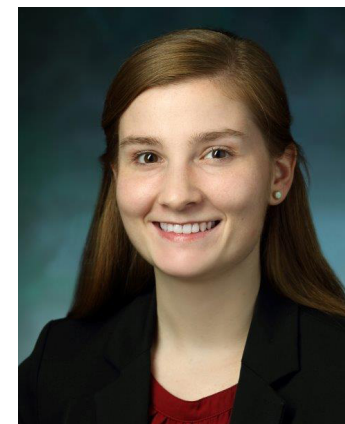
Medicines that slow the heart rate or cause the heart to squeeze less vigorously can reduce obstruction and improve symptoms.

First-Line Medical Therapy

1. Beta blocker (BB) medications (e.g., metoprolol, carvedilol or bisoprolol)
2. Calcium channel blocker (CCB) medications (diltiazem and verapamil)

Cardiac Myosin Inhibitors

For many HCM patients with obstruction, treatment with first-line medications does not get rid of their symptoms completely. Cardiac myosin inhibitors (mavacamten or aficamten) are more potent medications to treat obstruction. These medications work by purposely reducing the squeeze function of the heart. Close monitoring with repeat echocardiography and clinical visits is required for patients on these medications.



Victoria Blake, PharmD
JHH Specialty Pharmacy

Advanced Treatments of Obstruction

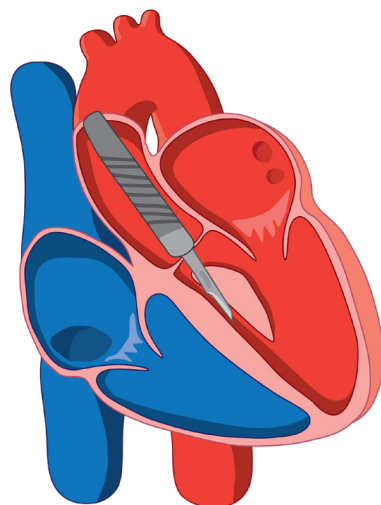
Surgical Treatment

In high-volume centers with experienced cardiac surgeons, an operation called a septal myectomy can be a safe and effective method for permanently eliminating an HCM-related obstruction and improving patient symptoms. During this surgery, the thickened part of the heart muscle that causes obstruction is removed.

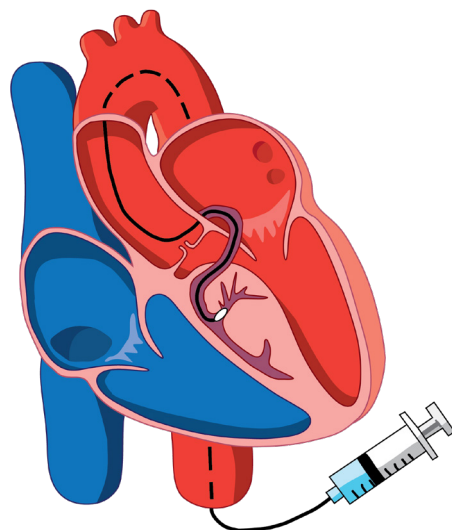
Catheter-Based Treatment

For patients in whom surgery is high risk due to their age or other medical conditions, a catheter-based procedure called an alcohol septal ablation may be an effective way to improve obstruction. During this procedure, alcohol is injected into the small coronary artery that supplies the thickened part of the heart muscle with blood, causing it to shrink. Not all patients are candidates for this procedure due to the individual anatomy of their coronary arteries.



Surgical Myectomy



Alcohol Septal Ablation



Frequently Asked Questions

- **Can I lead a normal life with HCM?**
With appropriate management, many individuals with HCM live full, active lives.
- **Is HCM curable?**
While there's no cure, treatments can effectively manage symptoms and reduce risks.
- **Is it safe to exercise?**
Yes, but the type and intensity should be discussed with your physician.
- **What are the signs of worsening HCM?**
Increased shortness of breath, chest pain, fainting or palpitations warrant medical attention.
- **What treatment options are available for patients with advanced HCM disease?**
At Johns Hopkins, we offer advanced heart failure/transplant services to patients with advanced disease.
- **Is the Johns Hopkins HCM Center a center of excellence?**
Yes, the Johns Hopkins HCM Center was designated as a center of excellence by the HCMA, a national patient advocacy group.
- **How I can help support the Johns Hopkins HCM program?**
 We are extremely grateful for philanthropic support; learn more at [Our Giving Page](https://hopkins.org/SupportHCMCenter).
hopkins.org/SupportHCMCenter
- **Do you have patient education events?**
 We hold an annual HCM patient symposium directed towards HCM patients, family members, and friends. Learn more about the [HCM Patient Symposium](https://hopkins.org/HCMPatientSymposium).
hopkins.org/HCMPatientSymposium

Schedule an Appointment at the Johns Hopkins HCM Center

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Learn more:
hopkinsmedicine.org/hcm