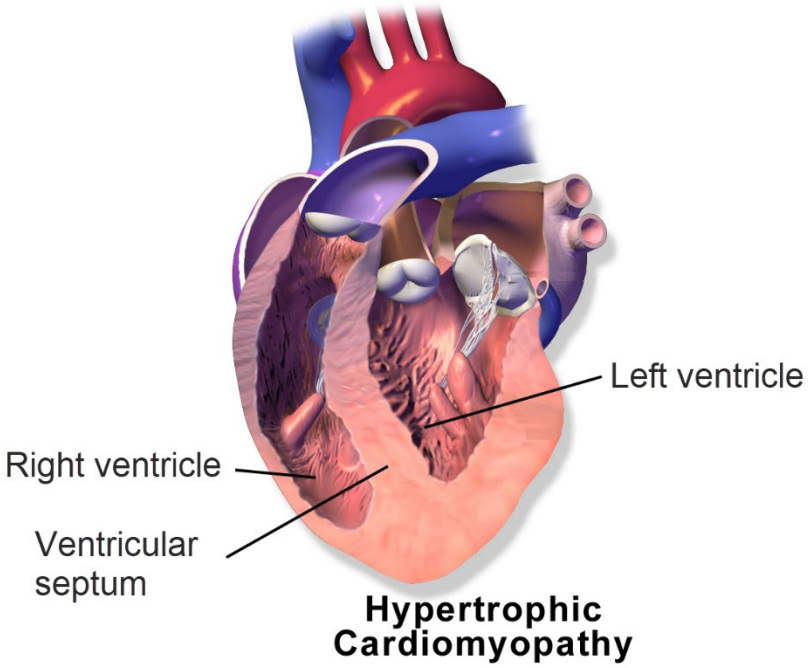
UNIVERSITY OF MICHIGAN HEALTH
MICHIGAN MEDICINE

Hypertrophic Cardiomyopathy (HCM) in Children

What is hypertrophic cardiomyopathy?

Hypertrophic cardiomyopathy (HCM) is a condition that affects the heart muscle causing it to become thickened and "muscle-bound". In HCM, the muscle cells are enlarged (the medical term for this is hypertrophy). This causes the heart walls to be thick. This effect is seen most in the **ventricular septum**, which is the wall between the heart's two lower chambers (the ventricles**)**. The **ventricles** are the primary pumping chambers of the heart with the **right ventricle** pumping blood to the lungs and the **left ventricle** pumping blood to the body.

In some cases, the wall between the two ventricles becomes so thick that it blocks blood flow out of the heart.



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What are the symptoms?

While many children with HCM have no symptoms, some may experience symptoms with exercise which may include lightheadedness, difficulty

breathing or chest pain. HCM has been linked to life-threatening abnormal heart rhythms that result in syncope (passing out or loss of consciousness) or sudden death. It is an important cause of sudden death in young athletes.

How common is it?

HCM can be inherited meaning it can be passed from one generation to the next. A diagnosis of one person in the family should lead to a family screening to determine who else might be affected. It is quite common for a genetic disease, affecting about 1 in 500 adults. Many are not diagnosed until adulthood making it far less common in childhood, though it affects an estimated 1 in 200,000 children.

How does this problem affect my child's health?

Although HCM is a serious problem, the health effects vary widely with the age it starts and how severe it is.

- Most children with HCM will not show any symptoms and will be at low risk for sudden death.
- Treatments, such as medication or surgery, can help improve symptoms for children with symptoms such as chest pain or low endurance with exercise, helping them maintain an active lifestyle.
- For children who are determined to be at high risk of sudden death, a device called an ICD (see below) can be placed to help significantly lower that risk.

HCM is a progressive condition (meaning it can get more severe with time) and your child will need to be monitored regularly as they get older.

How is this problem diagnosed?

- **Family history:** HCM is quite commonly an inherited condition and many children with HCM are identified during family screenings after another family member has been identified with the condition. Even children who have no previous family history of HCM can pass it on to their children (as the condition may be due to a new genetic mutation that can now be passed on).

- **Symptoms:** Possible symptoms include palpitations ("heart-racing" or "skipping heartbeat"), chest pain and shortness of breath with exercise, and fainting (syncope).
- **Physical findings:** The exam is usually normal other than a heart murmur.

Medical tests:

Electrocardiogram (ECG or EKG)

An ECG records the electrical signal from your heart. It is often the first test performed in any child who may have a cardiac condition. An ECG is often abnormal in people with significant HCM, but it is not a very sensitive test and may show a normal result for others, especially early in the course of the disease.

Echocardiogram (ECHO)

An ECHO uses sound waves to create pictures of your heart. It is the main test used to make the diagnosis of hypertrophic cardiomyopathy. It can detect thickening of the heart muscle which is the characteristic feature of hypertrophic cardiomyopathy.

Magnetic Resonance Imaging (MRI)

An MRI uses a strong magnet to create pictures. It is like an ECHO in that it assesses the structure and function of the heart. It can be more sensitive than an ECHO in that it can detect hypertrophy in parts of the heart that are not well seen on an ECHO. It can detect areas of scarring in the heart which may be associated with higher risk of dangerous arrhythmias and sudden death.

Holter monitor

A Holter monitor records the rhythm of the heart for a duration of 1 day to 2 weeks. It will help detect any arrhythmias that may require special treatment through medication or placing an implantable cardioverter-defibrillator (ICD).

How is the problem treated?

Medications

The primary medication used to treat HCM is a beta-blocker. **Beta-blockers** tend to slow the heart rate which can allow more time for the heart to fill with blood

between beats. This can be helpful for people with HCM who have heart stiffness that weakens the heart's ability to fill easily (diastolic dysfunction).

A heart with more blood and a slower heart rate can also be helpful in people with excess heart muscle that blocks (obstructs) the ability of blood to get out of the heart during a contraction (obstructive HCM).

Other medicines that can be used to help alleviate symptoms include calcium channel blockers (such as verapamil) and disopyramide. There are newer medicines (such as myosin inhibitors) being evaluated in research studies, but none have yet been approved for use by the FDA. Medications are usually recommended for people that have symptoms but may be considered for people without symptoms who have HCM with significant blockage.

Surgery

In some people with HCM, the excess muscle in the left ventricle can get in the way of blood getting out of the heart. This can be especially noticeable with exercise and can lead to lightheadedness or fainting with exercise or standing up too quickly. The medications mentioned above may help improve symptoms significantly, but children with muscular blockage who still have symptoms with medication may benefit from a **surgical myectomy**, a procedure to remove the muscle that is blocking blood from getting out of the heart.

Implantable Cardioverter-Defibrillator (ICD)

Patients with HCM can be at risk of sudden death. An ICD may be recommended for people with HCM who have had a cardiac arrest or who are at risk for one based on their evaluation. The ICD continuously monitors the heartbeat and if it detects a dangerous heart rhythm it will reset the heart using a small electric shock.

Exercise restrictions/safe environment

We very much encourage children with HCM to remain active. There are many benefits to physical activity and even team sports are possible if conditions can be met to maximize the safety of the child. Every child and situation is different, so your doctor will work with your child, your family and others

(including coaches, school staff, school nurses and trainers) to identify activities and environments with a good chance of safety. School staff, teachers, gym instructors and coaches should be aware of the child's medical condition and have a plan ready so they can respond in case of an emergency.

What are the long-term health issues for these children?

While HCM can be a serious problem, most children have no symptoms and are at low risk of a dangerous heart arrhythmia leading to sudden death. They should be monitored regularly depending on their age, symptoms and degree of heart thickening.

HCM is a progressive condition, meaning that it will get worse as they get older. However, the rate, timing and degree of progression can vary from one person to the next. Perhaps the greatest rate of change occurs when children become teens (adolescence). They should be followed more closely during this time so that their exercise guidelines and risk assessments can be updated appropriately.

Exercise guidelines

Personal exercise recommendations are developed in discussions involving your child, your family, and your doctor. As noted above, exercise is important, and encouraged, after discussions about safe activities. In general, children with HCM should be allowed to self-limit their exercise and they should not be graded on their performance (which could pressure them to push past their limit of comfort).

Our team

The University of Michigan Congenital Heart Center has developed a multidisciplinary team to care for patients and families affected by HCM and other inherited cardiomyopathies. The team includes cardiologists who specialize in:

- Cardiomyopathy
- Heart failure
- Heart transplantation

- Heart rhythm disorders
- Heart imaging
- Exercise

In addition, we have a genetic counselor and an advanced practice nurse and we work closely with our pediatric cardiovascular surgery team.

Mark Russell, MD
David Peng, MD
David Bradley, MD
Patty Arscott, MS (Genetic Counselor)
Kristin Evans, NP (Nurse Practitioner)
Adam Dorfman, MD ()
Jessie Hansen, MD ()
Richard Ohye, MD ()
Ming-Sing Si, MD ()

Frankel Cardiovascular Center's Inherited Cardiomyopathy program

HCM is a progressive condition that is more common in adults. For this reason, we work very closely with our colleagues in adult cardiology to create consistency for patients and families as patients transition from childhood to adulthood. We meet monthly to discuss patient care issues, to update research activities and to identify opportunities to enhance or improve care practices. In addition, both the adult and pediatric teams are assisted by the same genetic counselor who provides care for the whole family, coordinating family screenings and facilitating follow-up.

More details regarding the Inherited Cardiomyopathy Program can be found on their website:
https://www.umcvc.org/conditions-treatments/hypertrophic-cardiomyopathy

References

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Heart Association Joint Committee on Clinical Practice Guidelines. J Am Coll Cardiol. 2020 Dec 22;76(25):e159-e240. (https://pubmed.ncbi.nlm.nih.gov/33229116/)