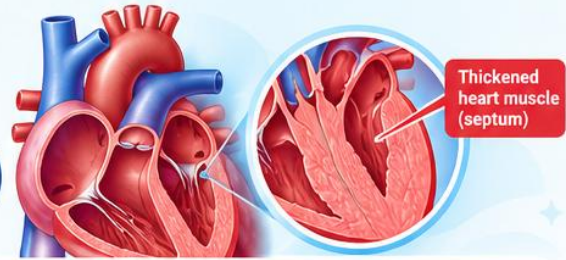


UNDERSTANDING HYPERTROPHIC CARDIOMYOPATHY (HCM) AND LONG-TERM MANAGEMENT



What Is HCM?

Hypertrophic cardiomyopathy (HCM) is a **genetic heart muscle disease** characterized by **abnormal thickening of the heart muscle**, most commonly involving the interventricular septum.

Normal Heart



Normal thickness of heart muscle

Obstructive HCM (most common)



Thickened septum blocks blood flow to the aorta

Non-Obstructive HCM



Thickened heart muscle without blockage

How Common?



Affects
1 in 200 – 500
people

One of the most common inherited heart diseases.

Causes and Genetics



- Most often caused by mutations in genes that encode cardiac sarcomere proteins (e.g., MYH7, MYBPC3, TNNT2, TNNT3).
- Usually inherited in an autosomal dominant pattern (50% chance of passing to children).
- Can also occur without a family history.

Types of HCM

- **Obstructive HCM (HOCM)**
Thickened septum obstructs blood flow.
- **Non-Obstructive HCM**
Thickening without obstruction.
- **Apical HCM**
Thickening at the apex of the heart.
- **Mid-Ventricular HCM**
Thickening in the middle portion of the ventricle.

Symptoms



Shortness of breath



Chest pain



Exercise intolerance



Dizziness or fainting



Palpitations



Fatigue



May be asymptomatic, especially in children and adolescents.

Diagnosis



Electrocardiogram (ECG)
May show LV hypertrophy, abnormalities, or arrhythmias.



Echocardiogram (Echo)
Primary test to evaluate wall thickness, obstruction, and heart function.



Cardiac MRI
Detailed anatomy and scar assessment.



Exercise Stress Testing
Evaluates exercise capacity and arrhythmias.



Genetic Testing
Helpful for diagnosis and family screening.

Potential Complications



Heart Failure
Due to impaired relaxation, obstruction, or scarring.



Arrhythmias
Atrial fibrillation or ventricular arrhythmias.



Sudden Cardiac Death (SCD)
Uncommon, but one of the leading causes in young athletes.

Treatment Options



Medications

- Beta-blockers (e.g., metoprolol, atenolol)
- Calcium channel blockers (e.g., verapamil, diltiazem)
- Cardiac myosin inhibitors (e.g., mavacamten) for selected patients with obstructive HCM.



Septal Reduction Therapies (for obstructive HCM)

- Surgical septal myectomy (gold standard)
- Alcohol septal ablation (catheter-based option)



Implantable Cardioverter Defibrillator (ICD)

Recommended for patients at high risk for sudden cardiac death.

Long-Term Follow-Up



Children and Adolescents

- Follow-up every 6–12 months
- Serial ECG and echocardiograms
- Holter monitoring when indicated
- Reassess during growth and puberty



Adults

- Annual cardiology evaluation
- Echocardiogram every 1–2 years
- Periodic cardiac MRI
- Rhythm monitoring as indicated



Family Screening

- ECG and echocardiogram for first-degree relatives
- Genetic testing when a mutation is identified
- Screening often begins at age 10–12 years (or earlier if symptoms or strong family history).

Lifestyle and Activity



Generally Encouraged

- Walking, hiking, swimming
- Recreational cycling
- Moderate aerobic activity



Individualized Evaluation Needed

- Competitive sports
- High-intensity athletics
- Endurance activities

Exercise recommendations should be personalized based on symptoms, obstruction, arrhythmia risk, and overall clinical profile.

Pregnancy and HCM



- Most women with stable HCM tolerate pregnancy well.
- Pre-pregnancy assessment is recommended.
- High-risk patients should be managed by a multidisciplinary team.

When to Seek Immediate Medical Attention



- Fainting or near-fainting
- Sustained palpitations
- Chest pain during exertion
- New shortness of breath
- ICD shocks
- Symptoms of heart failure

Key Takeaway



With regular follow-up, appropriate treatment, and lifestyle guidance, most people with HCM can enjoy a long and active life with excellent outcomes.



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