

What is Symptomatic obstructive HCM?

Symptomatic Obstructive Hypertrophic Cardiomyopathy (HCM) is a condition where the heart muscle thickens, especially in the wall between the heart's chambers. This thickening can block blood flow from the heart to the aorta, causing symptoms.

Symptomatic obstructive HCM

First line therapy

Medications: Non-vasodilating beta-blockers (e.g., metoprolol) or calcium-channel blockers (like verapamil) are used to control symptoms.



Avoid: Medications that widen blood vessels (vasodilators) and dehydration.

Persistent symptomatic obstruction

Second line therapies to consider:

Choice depends on patient preference, access/expertise, comorbidities, HCM subtype, and response to prior therapies

Drug therapy
(generally attempted prior to invasive therapy)

Invasive septal reduction therapy
(generally reserved when medical therapy fails)

Disopyramide

Used for its long history of helping with heart rhythm issues.

- Non-invasive

Pros

Recurrent Shortage: This medication has been in short supply in recent years.

Side Effects: It can cause anticholinergic side effects like dry mouth, constipation, and blurred vision.

Cons

Dosing: Only short-acting versions are available in Canada, meaning it must be taken three times a day.

Potential Heart Risk: It can cause QT prolongation, which may affect the heart's electrical activity.

Effectiveness: The benefits are mostly based on observations from doctors rather than large studies

Cardiac myosin inhibitors (Mavacamten)

A newer medication that improves symptoms and exercise capacity

- Non-invasive

Shorter Experience: Mavacamten is a newer medication with less long-term data compared to other treatments.

Risk: It may cause systolic dysfunction, meaning the heart may not pump as well. Because of this, patients need heart scans (echocardiograms) every 1-3 months.

Cost: The medication is expensive.

Pediatric Trials: Studies for its use in children are still in progress

Alcohol septal ablation

A procedure to reduce the thickened part of the heart muscle. It's less invasive than surgery

Specialized Care: You need access to a medical center with experience in handling many cases.

Risk: There's a 10% chance of needing a pacemaker due to heart block (a disruption in the heart's electrical system).

Suitability: Whether the procedure works depends on the shape of the heart muscle and the surrounding arteries.

Not for Children: This treatment isn't used in pediatric patients.

Surgical myectomy (+/- mitral intervention)

Surgery to remove the thickened heart muscle, which is the most effective and durable treatment. It's more invasive but can greatly improve symptoms.

Specialized Care: This surgery needs to be done at a hospital that handles many similar cases (a high-volume center).

Invasive Procedure: This is a more involved surgery than some other treatments.

Risk: There's a 5% chance of needing a pacemaker due to heart block, which affects the heart's electrical system after surgery.

In summary, treatments for obstructive HCM range from medication to more invasive procedures, with choices depending on patient preferences and medical needs. Talk to your doctor about what's right for you.