

Most Americans with HCM live normal life spans

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Most of the 600,000 Americans with hypertrophic cardiomyopathy (HCM) -- a genetic heart muscle disease -- can live normal life spans, according to the first science-based guideline for diagnosing and treating this disorder.

The joint recommendations of the American College of Cardiology Foundation and the American [Heart](#) Association are published online in *Circulation: Journal of the [American Heart Association](#)* and *Journal of the American College of Cardiology*.

"HCM is widely misperceived as a fatal condition, but a diagnosis of HCM is not a diagnosis of [sudden cardiac death](#). We now have effective treatments to help most patients manage the condition, although it remains the most common cause of sudden death in the young and in competitive athletes," said Bernard J. Gersh, M.D., Ph.D., co-chairman of the guideline writing committee and professor of medicine at the Mayo Clinic College of Medicine, Rochester, Minn.

HCM is a genetic disorder which changes the way the heart functions, including erratic heart beats and obstructed blood flow from the left side of the heart. It can also cause sudden cardiac death, which occurs when lethal [heart rhythms](#) intervene. However, for many people, HCM is symptomless and benign.

Heart enlargement thickening in HCM most commonly involves the ventricular septum, the muscular wall that separates the left and right

sides of the heart. Abnormal motion of the mitral valve may obstruct the flow of blood from the [left ventricle](#) to the aorta artery causing the heart to work harder to keep an adequate amount of blood flowing throughout the body.

"A variety of mutations (more than 1,400) in one or more genes of the sarcomere (contractile unit of the heart) cause HCM, but account for only about 50 percent of clinically identified patients. Many other genes and mutations will be discovered to explain HCM in other patients," said Barry Maron, M.D., co-chairman of the guideline writing committee and director of the Hypertrophic Cardiomyopathy Center at the Minneapolis Heart Institute Foundation, Minneapolis, Minn.

HCM is the most common inherited heart disease, occurring in one per 500 people and therefore at least 600,000 Americans.

"The typical cardiologist might see one new HCM case a year. Many patients with HCM will benefit by seeking treatment from clinical centers of excellence focused on this disease, which have the most contemporary medical and surgical treatments available," Maron said.

Managing HCM-related heart failure can be complicated, particularly in patients who do not respond to standard drug therapy, such as beta- and calcium-channel blockers.

"There are highly successful surgical and catheter-based approaches, best carried out in centers of excellence," Maron said.

Among the other recommendations in the newly released guideline:

- Drug therapy with beta-blockers, verapamil or disopyramide, can control symptoms such as shortness of breath.
- The most appropriate treatment for advanced heart failure, due to

obstruction in patients who don't respond to medication, remains transaortic surgical septal myectomy – a procedure to remove excessive muscle from the septum and improve the symptoms.

-- For patients who are not candidates for surgery, catheter-based alcohol septal ablation is an alternative treatment option. The procedure introduces alcohol in the heart and causes a myocardial infarction (heart attack) to reduce the muscle mass of the septum.

-- All patients diagnosed with HCM should undergo a comprehensive evaluation of their risk of sudden cardiac arrest, including a personal and family history and noninvasive assessment with an echocardiogram.

-- HCM patients should be provided genetic counseling and genetic testing if appropriate.

-- HCM with or without obstruction should disqualify individuals from participating in most competition sports. Patients can participate in low-intensity competitive sports, such as golf and a range of recreational sporting activities.

-- Prevention of sudden death is possible in HCM using the implantable defibrillator in selected patients judged to be at high risk based on risk markers such as fainting or a history of a family member with sudden death.

Provided by American Heart Association

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