

What is Hypertrophic Cardiomyopathy?

Hypertrophic Cardiomyopathy (HCM) is a term used to describe a family of heart conditions that result in abnormal thickening of the heart wall muscle. It has also been called other names such as IHSS (idiopathic hypertrophic sub aortic stenosis), Obstructive cardiomyopathy, Familial muscular subaortic stenosis, and Brock's Disease. Many of these can be quite serious and can lead to death in relatively young individuals. This condition is in many cases inherited.

In the majority of patients with HCM the septum of the heart is thicker than it should be. The septum is a wall of muscle that separates the right ventricle from the left ventricle. In most cases the part of the septum that is thickened is the portion closest to the aortic valve. Many times the Mitral valve is involved as well. If you were to look at a piece of heart wall muscle under a microscope it would look abnormal. Usually in the heart the muscle cells are lined up in a fairly orderly arrangement that allows for the most efficient contraction of the heart muscle. In patients with HCM the muscle cells are aligned at different angles and sometimes can have unusual shapes. This pattern causes the individual muscle cells to work harder than normal and this causes them to abnormally multiply but thicken as well. For some reason this thickening process seems to be more concentrated in the area closer to the aortic valve. There can also be obstruction on the right side of the heart. There are three problems that this condition causes. 1. The abnormal thickening tends to create a pressure difference across the outflow tract of the left (or right) ventricle, in other words it becomes harder for the ventricle to push blood out the aorta (or pulmonary artery), This thickened muscle also decreases the amount of space inside the ventricle and can diminish the amount of blood that can be pumped with each beat. 2. The abnormal thickening of the septum causes the muscle to become stiff and it cannot relax normally. This causes blood to tend to back up into the lungs and body. This causes one to the major symptoms of shortness of breath with exercise and also angina (chest pain). 3. The thickened muscle is more prone to develop bad types of irregular heartbeats (ventricular tachycardia and ventricular fibrillation) and is the most common cause of sudden death in these patients.

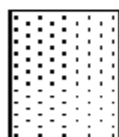
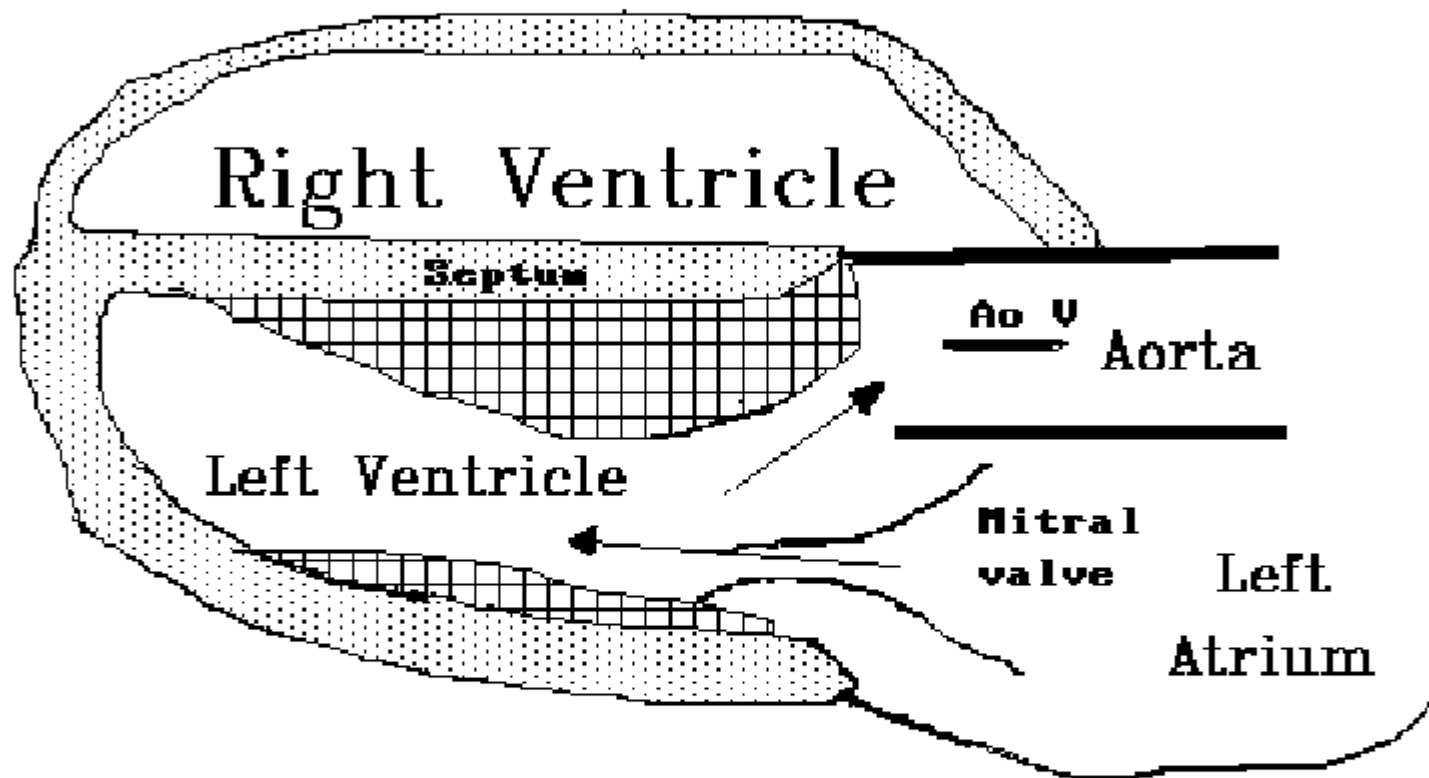
The diagnosis is based on several factors, the most important of which is the echocardiogram. This allows us to measure the wall thickness and heart size as well as the function of the heart. Most patients with HCM will have significant heart murmurs (extra heart sounds) that are caused by the turbulence of the blood going through the heart. More than 90% of these patients will have an abnormal EKG. There are some blood tests that can help determine if it is an inherited condition. Chromosome abnormalities have been found on chromosome numbers 1, 14, and 15. Chest X-Rays can be normal. The symptoms patients have vary considerably. Some patients have no symptoms; others may have shortness of breath or chest pain. Infants with this may appear more irritable than normal.

The treatment for this condition is limited. We use medicines called Inderal and verapamil to try to slow the heart rate and the speed of contraction of the heart. This will hopefully allow for better filling of the heart and better cardiac output. They can also prevent bad heart rhythms. We will also sometimes use Lasix to decrease the backpressure in the heart. On occasion the amount of blockage to the ventricle is so much that a portion of the muscle must be surgically removed. This procedure doesn't change the ultimate course of the disease, but does help some of the symptoms. Some centers are using pacemakers in selected cases to try and remodel the septum to make it less obstructive. Some centers are trying to purposely infarct portions of the septum to reduce the muscle mass. Transplantation is sometimes helpful. This is a very serious condition. When diagnosed in infants more than half die in the first year of life. The average risk for premature death in these patients is probably around 2-3 % per year. People with this diagnosis should not play competitive sports or engage in very vigorous activities. Because this is frequently an inherited condition it is important that family members be screened when the diagnosis is made. HCM can also be seen in pheochromocytoma, neurofibromatosis, tuberous sclerosis, Turner's syndrome, Noonans syndrome, Friedreich's ataxia and hyperthyroidism.

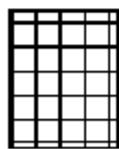
If you have any questions please ask one of the doctors.

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Long Axis Echocardiogram View



Normal Heart Muscle



Additional thickening
of heart muscle with
HCM