

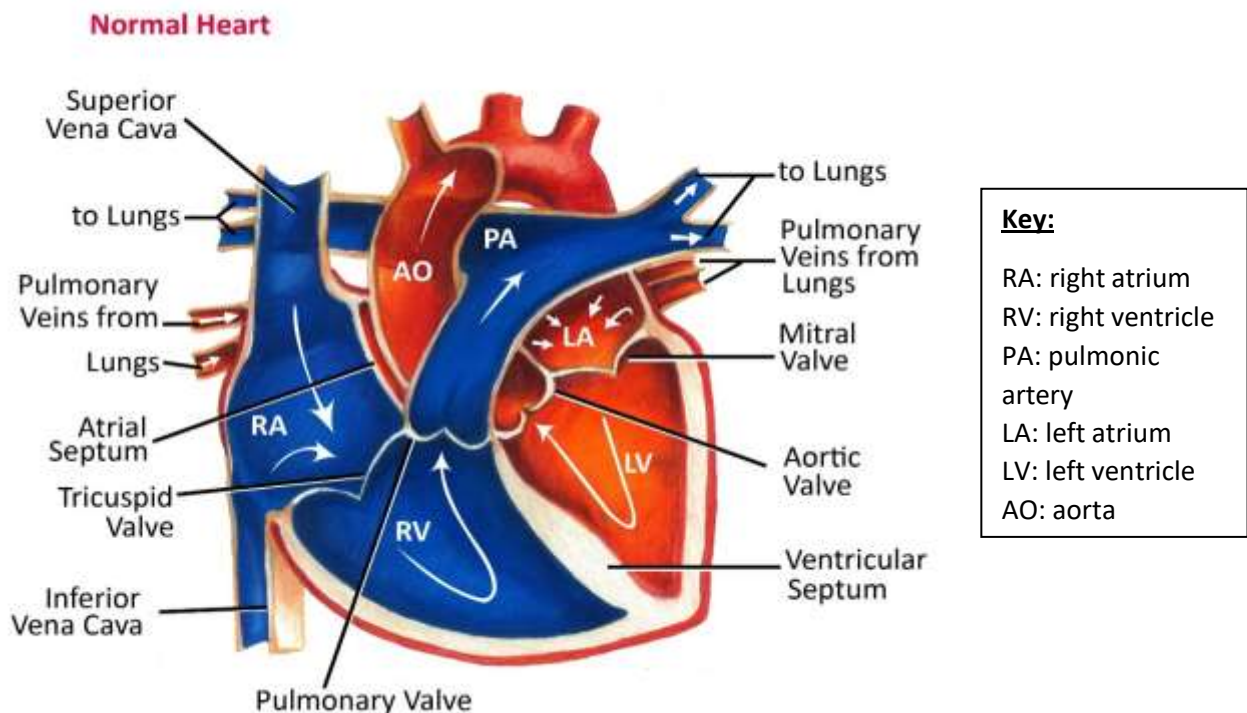


Hypertrophic Cardiomyopathy (HCM)

How does the heart work?

The heart is the organ responsible for pumping blood to and from all tissues of the body. The heart is divided into right and left sides. The job of the right side is to pump oxygen-deficient blood returning from the body into the lungs where fresh oxygen is collected and carbon dioxide is removed. The oxygen-rich blood returning from the lungs enters the left side of the heart where it is pumped into the aorta then to the rest of the body via the arterial system.

Each side of the heart has two chambers, an upper atrium and a lower ventricle. Between the atrium and ventricle on each side lies a valve – the tricuspid on the right and the mitral on the left – that regulates blood flow into the chambers. As the heart pumps, these valves act as one-way gates allowing blood to flow from the atrium above to the ventricle below and preventing blood from flowing back into the atrium. From the ventricles, blood is then forced to flow out into the pulmonary artery (on the right) or the aorta (on the left) through a second series of one-way valves called the pulmonic valve and the aortic valve, respectively.

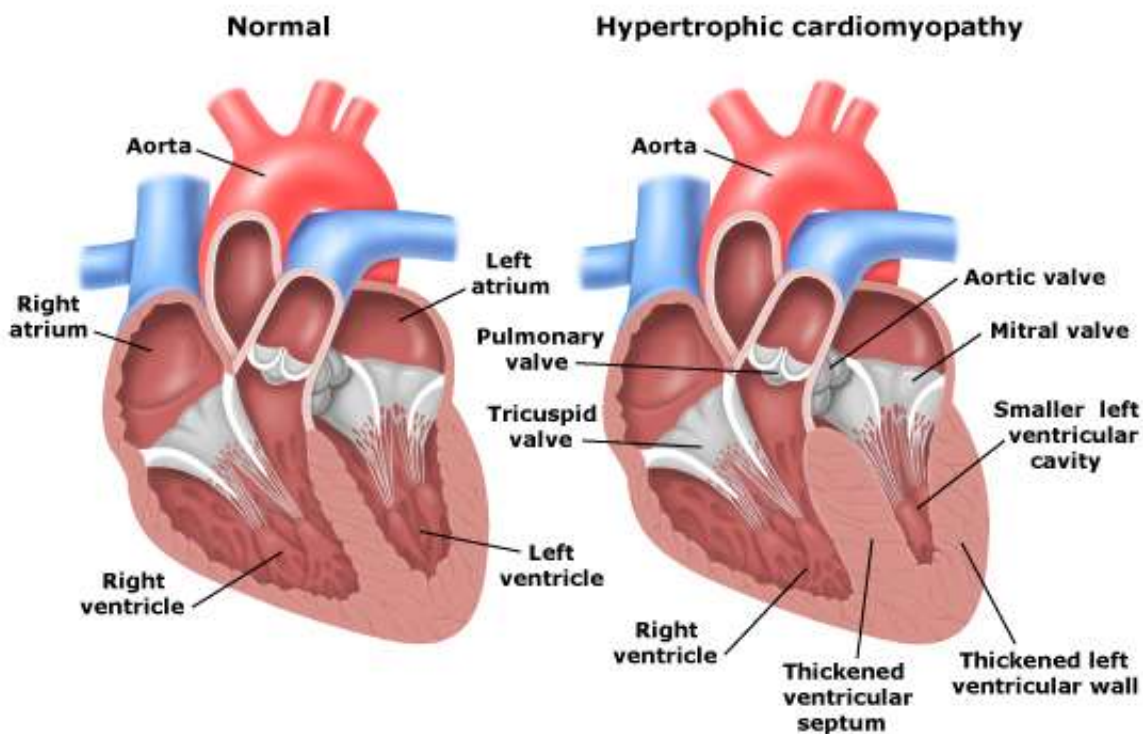




What is HCM?

Cardiomyopathy literally translates into heart (*cardio*) muscle (*myo*) disease (*pathy*). Hypertrophic means thickening so fully translated HCM means 'thick heart muscle disease.' Hypertrophic cardiomyopathy is the most common acquired heart disease in cats but very rare in dogs. HCM is a primary heart muscle disease where the muscular walls of the ventricles become abnormally thickened (hypertrophied). This thickening has several consequences (see below). HCM is diagnosed once other secondary causes of left ventricular wall thickening (hypertrophy), such as hyperthyroidism, high blood pressure, aortic stenosis and others have been ruled out.

Like the similar disease in humans, HCM is often familial in cats. A specific genetic mutation has been identified in Maine Coon and Ragdoll cats and a genetic test exists to identify affected cats. While many purebred cats (such as American shorthairs, oriental breeds and Persians) are predisposed to the disease, the domestic short hair (regular house cat) is the type most commonly diagnosed with HCM. Cats are usually middle-aged to older; however, the disease can be diagnosed at any age, often less than 5 years in purebred cats. In humans, HCM is also familial and over 200 different genetic mutations in more than a dozen genes have been identified as being able to cause the disease.





What are the consequences of the thickened left ventricle?

Unlike thickening of the walls in response to a physical stress (e.g. weight lifting or running marathons) where the thickening occurs to deal with the extra workload placed on the heart, the thickening with HCM is not normal. The degree to which the muscle walls thicken ultimately determines the clinical severity of the disease – some cats (and people) have mild disease, others have severe disease.

As HCM develops and progresses, the structure and function of the heart is altered in several ways. The hallmark problem with HCM lies in the inability of the left ventricle to relax appropriately. The thickened left ventricular walls become less flexible which prevents the left ventricle from relaxing or stretching sufficiently to fill with blood from the left atrium. This abnormal relaxation and inability to stretch may ultimately result in a build-up of blood “upstream” of the left ventricle – namely the left atrium and the pulmonary (lung) circulation. As the blood backs up, fluid is forced from the pulmonary capillaries into the lungs and chest cavity causing pulmonary oedema and pleural effusion, respectively (called congestive heart failure, or CHF).

In some cats, the thickening leads to arrhythmias and can result in sudden death (akin to what is seen in young basketball players that suddenly die on the court). It is difficult to predict which cats are likely to do, but sudden death is relatively uncommon in cats (compared to humans with HCM).

A small number of cats can suffer from feline aortic thromboembolism (FATE) – a blood clot in the aorta that causes blockage of blood flow to the back legs (most commonly), leading to sudden paralysis, severe pain, and often, death.

How is HCM diagnosed?

The clinical signs of HCM are variable. To some degree, the clinical signs depend on the severity – mild disease doesn't cause obvious problems, but severe disease often does. Additionally, cats are masters at masking problems until they become severe, so cats with severe HCM may appear completely normal or have only subtle signs that go unnoticed (i.e. mildly increased respiratory rate) or they may be very nonspecific to heart disease (i.e. decreased appetite). On the other hand, an owner may notice signs such as respiratory distress secondary to congestive heart failure or leg paralysis secondary to a thromboembolic (blood clot) event. In addition, your veterinarian can clue in on signs when he or she listens to your cat's chest during their physical exam. An increased heart rate, heart murmur, and/or gallop rhythm (extra heart sound) may be appreciated as the disease advances.

A common feature of HCM is termed systolic anterior motion (SAM) of the mitral valve. This abnormal motion of the mitral valve partially obstructs the outflow of blood from



the left ventricle into the aorta, resulting in a heart murmur which can be heard by your veterinarian with a stethoscope during your pet's physical examination. However, it is important to note that not all murmurs in cats are due to SAM. Echocardiography is required to confirm both the presence and severity of SAM.

An echocardiogram (ultrasound of the heart) offers the best means to diagnose HCM. Echocardiography allows a veterinarian to observe the physical structure and dynamic function of the heart. Fortunately, the test is non-invasive and poses essentially no risk to the cat.

Electrocardiograms and radiographs provide additional useful information and are often used to assist a veterinarian in diagnosing HCM but cannot be used alone to diagnose the disease. Since very subtle structural and functional changes can occur within the heart in the early stages of HCM, it is strongly recommended that a veterinary cardiologist be consulted for diagnosis as well as subsequent management of the disease.

As previously mentioned, additional tests might be needed to rule out underlying diseases such as high blood pressure or hyperthyroidism which may cause similar hypertrophy of the left ventricle as HCM. If no other causes are found, the diagnosis of HCM is made.

Two breeds (Maine Coon and Ragdoll) have been identified with mutations in the same gene (Myosin-binding protein C) as a cause of HCM. A genetic test has been developed to test these breeds for the mutation. The test does not work in other breeds.

How is HCM treated?

Currently, there is no cure for HCM. The changes occurring to the heart muscle are irreversible. However, if your pet's left ventricular hypertrophy is secondary to some other underlying disease, such as hyperthyroidism, treatment of the primary disease may result in partial or complete resolution of the heart changes.

As previously mentioned, hypertrophy of the heart muscle affects the ability of the left ventricle to relax properly, and therefore, function appropriately. While veterinarians may prescribe one or more medications to try to improve the heart's relaxing ability, it is important to recognise that no drugs have been shown to be effective in achieving this. Thus, attempts to reduce the risk of heart failure and to help the heart function efficiently are largely theoretical and may ultimately be of no value.



Some treatment options that may be prescribed include:

- Drugs that are thought to alter relaxation of heart muscle or slow down the heart rate to allow a longer time for the heart to fill, or both. No proof exists that either of these approaches benefits cats with HCM. It is quite acceptable to not treat cats with HCM prior to the onset of CHF; when information becomes available showing a benefit of any treatment, guidelines may change.
- Drugs to treat congestive heart failure (diuretics and ACE inhibitors). These medications are not specific for HCM but are used in cats with heart failure secondary to any heart condition. With severe fluid build-up in the chest cavity, the veterinarian may physically remove the fluid with a catheter to help the cat breathe.
- Drugs that are thought to reduce the risk of clot formation, or clot recurrence. A medication that reduces the ability of the blood to clot may be prescribed if the patient is felt to be at risk for blood clot formation or currently has a blood clot in one of its arteries or heart chambers. The use of certain drugs for this purpose must be closely monitored to ensure the patient is not placed at risk for haemorrhage (bleeding). Treatment does not guarantee that a blood clot will not form, nor is it designed to break down previously formed clots. The anti-platelet agent clopidogrel has been shown to significantly reduce the likelihood of recurrent thromboembolism (i.e. in cats that have previously had a thromboembolic episode).

As an owner of a cat with HCM, you should be very sensitive to changes in your pet's condition and should not hesitate to seek veterinary advice. Your veterinarian may show you how to monitor your cat's respiratory rate as an increased rate may be a sign that congestive heart failure is developing or worsening (please see the separate sleeping respiratory rate form). Cats with congestive heart failure do not cough like people with CHF, but often exhibit open mouth breathing and panting. A cat that is having difficulty breathing from heart failure or has loss of function of hindlimbs or front limbs, requires veterinary care as quickly as possible. In the acute setting, these problems may need specific treatments (oxygen therapy, injectable medications, anticoagulation medications, or pain medications) that can only be offered by a veterinarian.

What is the prognosis?

The prognosis of a cat with HCM is highly variable. Some cats may develop only mild hypertrophy and suffer little compromise of heart function, while others progress to more severe disease. HCM may worsen quickly over a period of months, or it may progress slowly over several years. Its severity may not change for many years and then suddenly worsen. Some cats with HCM die suddenly even though they had no clinical signs of heart disease.



A cat with mild to moderate disease may enjoy an essentially normal life for several years. However, the prognosis is much more guarded once the cat has more severe disease. The risk of developing congestive heart failure is proportional to disease severity, which is often classified by measuring wall thicknesses and left atrial size. Although congestive heart failure can be treated medically, severe heart failure may become difficult to manage over time as the disease progresses. The prognosis for a cat with heart failure, unfortunately, is guarded to poor. On average, survival for cats with HCM and heart failure is 12 to 18 months after diagnosis.

Thromboembolism is a severe uncommon complicating condition in HCM. It can cause acute pain and various clinical signs such as loss of function of the hindlimbs (most commonly, although other limbs or organs can be affected). Although treatment to break down or remove the clot is available, the high treatment costs of the procedure, death during administration of the drugs and high recurrence rate of thromboembolism deters most from attempting this type of therapy. With supportive care, about 40 to 50% of patients with thromboembolic disease will break down clots on their own and regain limb function over time. However, despite the best medical efforts to prevent their reoccurrence, a cat that has survived a thromboembolic event has a significant risk of developing another over the following weeks to months.

To breed or not to breed?

In HCM, an autosomal dominant pattern of inheritance has been documented. Not only does that mean it can be passed on genetically, but also that males and females are equally affected; every affected cat will have at least one affected parent; and all carriers of even a single copy of the gene can show the disease and transmit a mutant gene to half of their offspring.

In two breeds (Maine Coon and Ragdoll), a specific genetic mutation has been identified and a test has been developed to identify cats with the mutation. So, what does this mean if you want to breed Maine Coon or Ragdoll cats?

If the DNA test is positive for that mutant gene, the cat may develop HCM. As in people, not all individuals with the mutation will develop the disease. Breeding recommendations are currently all over the place. Some people recommend that if the cat has two copies of the gene, then it should not be used for breeding. Rather, the cat should be screened periodically to see if it develops the disease. If the cat has one copy of the gene, it should also be screened periodically for the disease, although his or her status for breeding is much better.

On the flip side, some veterinarians feel that any cat with the mutation should be spayed or neutered. However, the gene pool for purebred cats is quite small. If all the cats with the mutation in one generation are not bred, you might end up with far too



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much inbreeding, which would ruin the breed anyway. Therefore, some people recommend that cats who have one copy of the mutation and who have no clinical evidence of the disease may be used to breed to a mutation-negative cat. Offspring of that cat should be carefully evaluated and, if possible, a mutation-negative kitten should be used for a breeding replacement.

Before breeding your cat, be sure you understand all of the possibilities of what could happen. Genetics is an area that not all veterinarians are comfortable discussing. Discuss with your veterinarian whether referral to a geneticist or cardiologist would be best for you and your pet.

Additional Resource:

<http://vetmed.tufts.edu/heartsmart/>

This is a very useful and well-written resource, providing pet owners with a clear and credible source of information about veterinary cardiology.