

Hypertrophic Cardiomyopathy

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WHAT IS HYPERTROPHIC CARDIOMYOPATHY?

Hypertrophic cardiomyopathy (HCM) is the most common acquired heart diseases 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) The name “hypertrophic cardiomyopathy” means “thick” (hypertrophic), heart cardio =, myo = muscle, pathy = disease) (figure 1). This thickening has several consequences (see below). HCM is diagnosed once other secondary causes of left ventricular wall thickening (hypertrophy), such as hyperthyroidism, systemic hypertension, aortic stenosis and others, have been ruled out.

Like the similar disease in humans, HCM is often familial in cats, thought to be inherited in an autosomal dominant manner. A specific mutation has been identified in Maine Coon 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 most commonly diagnosed cat 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 several different genetic mutations have been identified as being able to cause the disease. To date, no viral or nutritional causes of HCM have been identified in humans or cats.

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 determine 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 stiff and noncompliant, 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 circulation. As the blood “backs up”, fluid is forced from the pulmonary capillaries into the lungs and chest cavity causing pulmonary edema and pleural effusion, respectively (commonly 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 this.

Other cats can suffer from 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.



Figure 1. On the left is a graphic representation of a normal left ventricle and on the right is an enlarged (concentric hypertrophy) left ventricle as noted in hypertrophic cardiomyopathy.

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 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 they listen 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 auscultated 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. Additional diagnostic tests are required to confirm both the presence of SAM and of HCM.

An echocardiogram (ultrasound of the heart) with color flow Doppler imaging 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 systemic hypertension 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.

In Maine Coon cats, a blood test is now available to determine if the pet has the genetic mutation associated with this condition. The test does not work for other cats with HCM. You can request this test from your veterinarian.

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 heart disease, such as hyperthyroidism, treatment of the primary disease may result in some or complete resolution of the heart condition.

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 recognize 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 as efficiently as possible are conjectural and may 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 actually 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 insure the patient is not placed at risk for

hemorrhage. Treatment does not guarantee that a blood clot will not form, nor is it designed to break down previously formed clots, since there are no studies showing that any of these medications actually do what they're claimed to do. Currently a study is underway examining the effect of one type of anti-clotting drug on survival of cats that have 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. Cats with congestive heart failure do not cough like dogs or people, 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, 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 very 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 a number of 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 is 12-18 months.

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 organs or limbs can be affected). Although treatment to break down or remove the clot is available, the high treatment costs of such procedure, death during administration of the drugs and high recurrence rate of thromboembolism persuades most from attempting this type of therapy. With supportive care, about 40-50% 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.

HOW CAN I PARTICIPATE IN CLINICAL TRIALS OF HCM?

Currently, there is a clinical trial examining the efficacy of anti-clotting medications in the prevention of recurrence of aortic thromboembolism (FATE). If your pet has suffered such an episode, your veterinarian can help you enroll your pet in this trial by visiting the study website: <http://www.vin.com/fatcat/>
