Clinical sheet - Cardiology

Hypertrophic cardiomyopathy in cats

Hypertrophic cardiomyopathy (HCM) is one of the most commonly encountered heart disease in cats. This disease is characterized by an abnormal thickening (hypertrophy) of one or several areas of the walls of the heart, usually of the left ventricle. Hypertrophic cardiomyopathy is also present in humans, and is caused by a variety of genetic anomalies of the cardiac muscle proteins. In cats, this disease is more prevalent in Ragdolls, Maine Coon, oriental breeds (Himalayan, Burmese, Sphynx, Persians), Devon Rex, but it is also commonly diagnosed in Domestic Short Hair cats. A specific genetic defect has been identified in Ragdolls and Maine Coon involving a contractile protein: the myosin binding protein C. This disease is usually diagnosed in middle-aged cats. However, there is also a juvenile form affecting young cats (usually Ragdolls).

Pathophysiology

The impact of the thickening of the ventricular wall on the heart function is quite variable, because they are many different forms of hypertrophy with this disease. If the hypertrophy is mild and focal, the cat may remain symptom-free for all his life. However, if the hypertrophy is severe, the ventricle will have a hard time distending, which leads to increased intra-cardiac pressure and congestive heart failure (CHF), with fluid build up in or around the lungs. In some cases, the ejection of blood from the left ventricle can be impeded because the mitral valve is aspirated against the inner wall of the ventricle, the septum, thus partially obstructing the passage. This phenomenon is called DOLVOT (Dynamic Obstruction of the Left Ventricular Outflow Tract) (Figure 1). In humans, this is associated with an increased risk of sudden death. Other complications of this disease include cardiac arrhythmias, leading to fainting or sudden death, and clot formation in the left atrium. This clot may fragment, travel in the aorta and obstruct a major artery (typically the iliac artery, which brings blood to the hind limb). This phenomenon is coined FATE (Feline Arterial ThromboEmbolism). This disease can be progressive: with time, the left ventricle may dilate, and its contractility may decline. Myocardial infarction may supervene, leading to destruction of areas of the cardiac muscle, a situation called remodeling.

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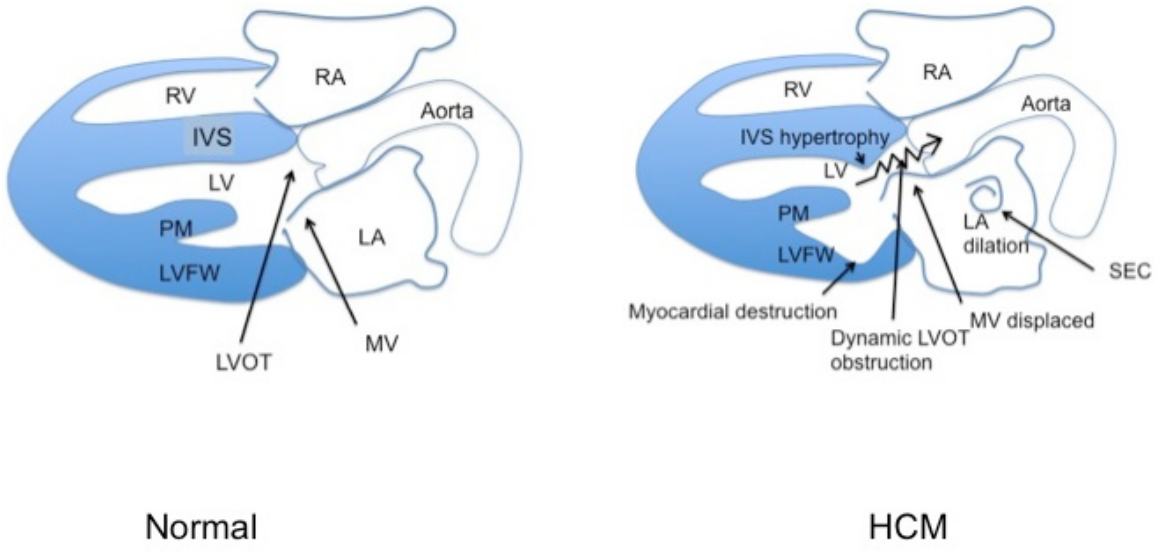

Figure 1: Salient features of hypertrophic cardiomyopathy (HCM) in cats.
On the left, a normal heart is represented. RV: right ventricle; LV: left ventricle, RA: right atrium, LA: left atrium; MV: mitral valve, LVOT: left ventricular outflow tract; IVS: interventricular septum; PM: papillary muscle; LVFW: left ventricular free wall. On the right, the hypertrophy represented here affects the base of the septum, but could affect any part of the left ventricular walls. The mitral valve is displaced against the septum, leading to dynamic obstruction of the LVOT (DOLVOT). Myocardial infarction may lead to focal destruction of the cardiac muscle. The left atrium is dilated, leading to blood stasis, and clot formation, often visible during echocardiography as a cloud of spontaneous echo contrast (SEC) whirling in the left atrium. (Art work: Eric de Madron)

Clinical signs

Many of these cats are asymptomatic. The disease may be discovered because of anomalies of the auscultation (heart murmur, arrhythmias, third heart sound), or in the context of HCM screening.

When CHF occurs, with fluid build-up in (pulmonary edema) or around (pleural effusion) the lungs, the cat will experience breathing difficulties, with increased breathing rate as well as labored breathing. Other non-specific symptoms such as vomiting and decreased appetite may also be present. Exercise intolerance, as seen in humans, is hard to assess in cats.

It is not uncommon to see the symptoms of CHF brought along by a triggering event such as anesthesia, IV fluids, or stress.

With FATE, there is sudden onset paralysis of one or several legs (hind or front legs) (Figure 2). The affected leg becomes flaccid and is dragged. The extremity of these legs becomes cold and pale.

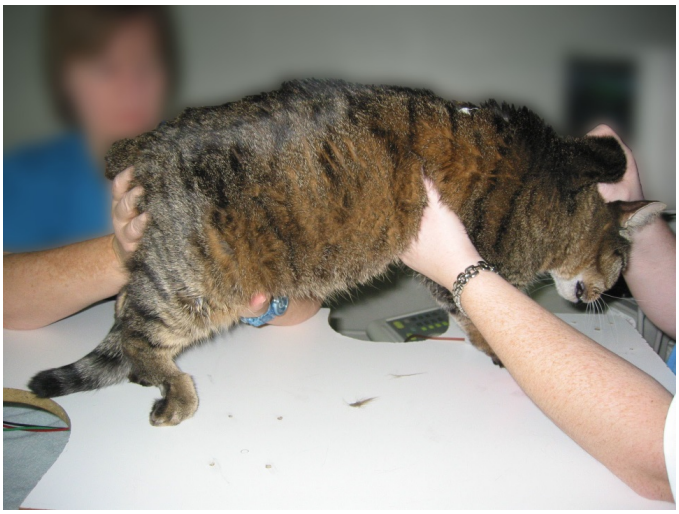

Figure 2: Arterial thromboembolism (FATE) in a cat with HCM.

A blood clot originating from the heart has obstructed the distal aorta, blocking blood flow to the hind limbs. His hind legs are paralyzed with inability to rectify the position of his paws (knuckling). (Photo: Eric de Madron)

Diagnosis

The diagnosis relies on the documentation of the ventricular wall anomalies by cardiac ultrasound (echocardiography), and the exclusion of other non-genetic causes of hypertrophy such as hyperthyroidism and hypertension. Ancillary testing may include thoracic radiographs, electrocardiogram (ECG), blood pressure measurement, and blood tests.

The goal of the cardiologist is not only to diagnose HCM, but also to establish risk factors to separate cats at low risk from cats at high risk of developing sudden death, CHF or FATE. Furthermore, periodic reassessments are required because of the progressive nature of some cases of HCM.

Treatment

In asymptomatic cats, no treatment has been proven to change the natural progression of the disease so far. However, there are some theoretical grounds behind the use of beta-blockers to attenuate the DOLVOT when present. In people, this has been shown to reduce the risk of sudden death. In cats, we do not know.

If the cat is thought to be at increased risk of FATE (typically because the atria are very dilated and SEC is present), then reduction of risk can be attempted by using blood thinners such as Clopidrogrel (Plavix) alone or in combination with aspirin.

If in CHF, then medications such as diuretics and ACEI (Enalapril, Benazepril) become required. Other medications stimulating contractility such as Pimobendan can be considered in very selected cases.

Prognosis

The prognosis is quite variable and depends on the pattern of anomalies present in the heart, the degree of disturbances in the circulation of the blood inside the heart, the degree of dilation of the atria, the heart rhythm, and blood pressure.

Cats with very stiff ventricles and profound disturbances of the blood circulation, very large atria, arrhythmias (such as atrial fibrillation), and low blood pressure have a poor prognosis. Juvenile forms of HCM, rapidly progressive, also carry a poor prognosis.

On the opposite end of the spectrum, cats with mild and focal hypertrophy without major blood circulation disturbances may live a completely normal life.

Screening

- **Genetic testing**

  The discovery of a specific genetic anomaly in Ragdolls and Maine Coon has allowed the development of a blood test. This test looks at the 2 copies of the gene affected (the one encoding myosin binding protein C). If the 2 copies are normal, risk of developing HCM is very low (but not nil, because other genes can be involved). If one copy is abnormal, then risk is moderate. If the 2 copies are abnormal, then the risk is high, and the disease is likely to be severe. Echocardiography will become necessary to see if the genetic anomalies have translated into real disease. This test works only in these 2 breeds. It can be useful for breeders.

- **Periodic echocardiography**

  This is the approach used in breeds at risk in which there is no genetic testing, such as Sphynx and Devon Rex.

- **NT- pro BNP testing**

  This blood test may help to identify asymptomatic cats with HCM, but is associated with a lot of false positives. Its best value is to predict the ABSENCE of heart disease when it is in the normal range.

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