Proceedings of the 35th World Small Animal Veterinary Congress
WSAVA 2010

Geneva, Switzerland - 2010

Next WSAVA Congress:

2011 WSAVA

14 -17 October, 2011  Jeju, KOREA

Reprinted in IVIS with the permission of WSAVA
Definitions And Forms of primary, genetic HCM

Hypertrophic cardiomyopathy (HCM) is a clinical heterogeneous cardiac disease that is in most cases inherited as an autosomal dominant trait. According to the classification of the American Heart Association (AHA), HCM is a primary genetic heart disease. HCM is morphologically characterized as a hypertrophied, no dilated left ventricle (LV) without any underlying systemic or cardiac disease that could cause a hypertrophied LV, such as systemic hypertension or (sub-)aortic stenosis. HCM is a progressive disease that finally leads to congestive heart failure (CHF) or sudden death. Diagnosis is usually based upon two-dimensional (2D) echocardiography.

The echocardiographic picture is in humans, as in cats very heterogeneous concerning the severity of the hypertrophy and the myocardial hyperechogenicity, as well as concerning the localisation of the hypertrophy. The thickening may be global or regional, and symmetric and asymmetric forms can be distinguished. The symmetric, global form affects the LV homogenous, whereas the asymmetric form involves selectively only the papillary muscles, the LV posterior wall, the interventricular septal wall (IVS), or focal in the IVS, just below the aortic valve. In feline HCM, the LV papillary muscles are consistently enlarged.

Systolic anterior motion of the mitral valve (SAM) is common in cats with HCM. Cats with HCM and SAM are commonly said to have the obstructive form of HCM or hypertrophic obstructive cardiomyopathy (HOCM). Secondary obstruction of the LV outflow tract (LVOT) can worsen the LV hypertrophy through additional pressure overload.

HCM ➔ SAM; SAM = HCM ?

Systolic anterior motion of the mitral valve is the process of the septal (anterior) mitral valve leaflet or the chordal structures inserting on this leaflet being pulled into the LVOT during systole by the enlarged or displaced papillary muscles. Here it is caught in the blood flow and pushed toward and often ultimately against the interventricular septum. The initial pulling of the mitral valve leaflet toward the LVOT in systole can clearly be seen on many echocardiograms from cats with HCM to be due to the grossly enlarged papillary muscles encroaching on the LVOT (the region of the LV between the anterior leaflet of the mitral valve and the interventricular septum) and pulling the mitral apparatus structures into the basilar region of the outflow tract. SAM produces a dynamic subaortic stenosis that increases the velocity of blood flow through the subaortic region and usually produces turbulence. Simultaneously, when the septal leaflet is pulled toward the interventricular septum, a gap in the mitral valve is produced creating mitral regurgitation. These abnormalities are by far the most common causes of the heart murmur heard in cats with HCM.

Occasionally it has been reported that in some cats, the LV hypertrophy regressed with beta-blocker treatment for SAM and that SAM could be found in young, as well in older cats without LV hypertrophy. Therefore, it seems that occasional SAM may be a primary problem causing a secondary LV hypertrophy. A similar, but rare phenomenon has been observed rarely in dogs by D’Agnolo et al. Therefore the question arises concerning the development and the classification of this entity. SAM can be caused experimentally by displacement of the anterior papillary muscle. Potentially, the cause of "primary SAM" is different in young, and in older cats. In young cats a congenital abnormality of the anterior papillary muscle resulting in an abnormal attachment of the chordae tendinea, causing abnormal attachment of the mitral valve may be an explanation. However, this entity should be better classified as mitral valve dysplasia. In older cats with a new heart murmur and "primary SAM", myocardial and especially papillary muscle restructuration’s secondary to inflammatory, toxic, or ischemic insults may be suspected as the cause of SAM. These entities should be classified as secondary cardiomyopathies.
Myokarditis versus HCM

Myocarditis is known to cause abnormal myocardial function and cardiac dilation in humans for many years. Many cases of dilated cardiomyopathy (DCM) are actually caused by myocarditis. In humans, myocarditis is therefore classified as primary mixed cardiomyopathy. Myocarditis in cats is also a recognized entity. Interestingly, echocardiography in cats with myocarditis shows often not DCM, but a hypertrophied LV. In a recent pathological study, cats with cardiomyopathy were examined by histology and by PCR. 55% of the cats with HCM showed signs of active myocarditis on histology, and panleucopenia virus was found in some of these cats. The authors have examined several cats that developed pulmonary edema and had the typical picture of HCM on echocardiography, with LV hypertrophy and left atrial dilation, after previous anaesthesia or surgery. Supportive therapy including diuretics, oxygen and antibiotics led to resolution of the pulmonary edema and cats became clinically normal. Repeated echocardiography showed a successive regression of the LV hypertrophy and LA dilation. This presentation and disease course argues against primary HCM, but rather for a reversible defect, such as infectious or toxic myocarditis.

Steroid-induced „HCM“

A similar picture as in the suspected myocarditis cases, with acute pulmonary edema, has been reported in cats given steroids. Pulmonary edema and LV hypertrophy resolved in these cats, initially diagnosed with HCM due to LV hypertrophy and LA dilation seen on echocardiography. However, attempts to cause this form of cardiomyopathy were not successful. The question arises, if the cats need to have a genetic predisposition to react abnormal to steroids, or if the affected cases in fact had myocarditis associated with glucocorticoid application.

Hyperthyroidism-induced Cardiomyopathy

Thyroid hormones have important metabolic and cardiovascular effects. Hyperthyroidism causes an increased metabolic rate and this result in a demand for an increased cardiac output. In the peripheral blood vessels thyroid hormones cause a decreased resistance. This leads directly to an increased cardiac output. Additionally, thyroid hormones induce multiple genes within the myocardium causing LV hypertrophy. A volume overloaded LV with eccentric hypertrophy should be the resulting changes of these mechanisms. However, concentric hypertrophy secondary to hyperthyroidism has been reported as well.

Myocardial hypertrophy caused by infiltrations

Infiltrations of the myocardial wall causing LV hypertrophy such as amyloidosis have not been reported in veterinary medicine. Rarely, diffuse neoplastic infiltrations caused by lymphosarcoma have been reported, causing LV hypertrophy that can be found using echocardiography.

HCM AS CAUSE OF SYSTEMIC HYPERTENSION (?)

Occasionally, HCM has been reported as differential diagnosis for systemic hypertension. However, as blood pressure is a function of cardiac output and peripheral resistance, there is no rational explanation for such a relationship. LV hypertrophy with or without CHF and elevated blood pressure is therefore more likely a secondary change to the hypertension, and not the cause of hypertension. However, if LV hypertrophy is diagnosed, hypertension should be excluded as cause before the diagnosis of HCM can be made.

If hcm looks like dcm - the "burn out" cardiomyopathy

In some case of feline (and human) HCM a regression of the LV hypertrophy can be seen during the disease progression. The myocardial wall becomes thinner and the LV volume overloaded and a progressive systolic dysfunction develops. It is difficult to differentiate the echocardiographic picture from
a DCM, unless the cats has been examined and diagnosed at previous examinations with HCM. This disease entity is called "burn out" cardiomyopathy. Occasional LV hypertrophy is still present.

Conclusion
When LV hypertrophy is seen on echocardiography, several aspects should be considered, before HCM is diagnosed:

Is the hypertrophy only a pseudohypertrophy secondary to dehydration? Is a fixed or dynamic (sub-)aortic stenosis present? Is systemic hypertension present? Did the cat receive steroids? Is there a history of recent anaesthesia, or is there potentially systemic bacterial or viral disease present? Does the cat have hyperthyroidism?

If any of these questions is answered with "yes", treatment should be started according to the underlying entity. Follow-up examinations should be performed carefully and if echocardiographic changes are reversible, the cat has not primary HCM.

To summarize, several myocardial diseases can look like HCM on echocardiography. Clinical and echocardiographic follow-up examinations are necessary to differentiate primary HCM, from secondary LV hypertrophy. Various treatable diseases should be taken into consideration and treated accordingly, before the diagnosis of HCM is made.

Literatur