Idiopathic dilated cardiomyopathy in the dog: diagnostic approach

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This is the second in a series of three articles on canine cardiomyopathy, reviewing the diagnostic approach to idiopathic dilated cardiomyopathy. The third article will discuss treatment modalities.

DEFINITION

Idiopathic dilated cardiomyopathy (IDCM) is a heart muscle disease of unknown aetiology characterised by progressive systolic dysfunction (loss of contractility) of the ventricular myocardium. This leads to dilation of first the ventricle(s) and later the atria. Some degree of diastolic dysfunction will also be present. In some breeds (see Table 3 in article about breed characteristics UK Vet Vol 8 No 7) ventricular arrhythmias rather than systolic dysfunction indicate the presence of IDCM.

DIAGNOSIS

The diagnosis of IDCM is a diagnosis of exclusion. It is based on the combination of the history, clinical findings and complementary exams like electrocardiography, radiography and colour Doppler echocardiography.

One has to be aware that different phases in the natural evolution of the disease are recognised:

1. **Occult phase**: the disease is present but cannot be confirmed by the means of electrocardiography nor echocardiography.
2. **Subclinical phase:** compensatory mechanisms are responsible for the lack of clinical signs in these animals but the presence of the disease can be shown by further investigations.
3. **Clinical phase:** there are clinical signs of congestive heart failure, arrhythmias or sudden death.

History

At presentation most, but not all, dogs with DCM show clinical signs. Cough and exercise intolerance are the most common complaints. It is important to remember that large breed dogs mainly cough because of advanced pulmonary oedema and rarely because of left mainstem bronchus compression by an enlarged left atrium, in contrast to small breed dogs with chronic mitral valve disease. An exception is the Dobermann, where the upright position of the heart and the relatively enlarged left atrium compresses the bronchus. Dyspnoea, anorexia and lethargy are commonly reported. In certain breeds (Boxer, Dobermann, Great Dane) syncope and sudden death are unfortunately often the only clinical signs indicating the presence of the cardiomyopathy.

Due to the familial aspect of IDCM always obtain a family history. Unfortunately in many breeds (see Table 3 article about breed characteristics: UK Vet Vol 8 No 7) IDCM develops at a more advanced age and progeny will already be present at the time of clinical manifestation of the disease.

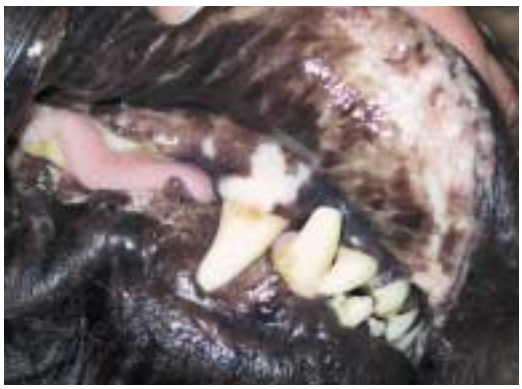

Fig. 1: *Pale mucous membranes in a dog with DCM.*

Clinical examination

Pallor of the mucosae (Fig. 1) with sluggish capillary refill time is a common finding in advanced DCM. Affected dogs often have a stronger apex beat than a dog with a normal heart, an observation that is commonly mistaken for a strongly contracting heart. However, the precordial impulse is not an indicator of contractility but is generated by the wall stress of myocardial contraction. Since wall stress is a factor of left ventricular pressure and radius the wall tension will be greater in a dilated heart. Thoracic palpation often reveals the presence of arrhythmias. Sinus tachycardia (indicating cardiac decompensation), atrial fibrillation and/or ventricular arrhythmias are often noticed. Thoracic auscultation will reveal the presence of a soft systolic murmur (the grading rarely exceeds III/VI and might be variable) at the level of the left heart apex and/or

a gallop rhythm. The mitral (and tricuspid) insufficiency is secondary to the annular dilation of the valvular apparatus. A thrill is extremely rare and if present it probably indicates a congenital problem or chronic valvular disease. The femoral pulse quality will be poor and pulse deficits are commonly noticed. The presence of pulsus alternans (alternating pulse quality in the absence of an arrhythmia) indicates severe myocardial failure and it is occasionally reported in dogs with IDCM, mainly in English Cocker Spaniels. Cold extremities indicate poor peripheral perfusion due to poor cardiac output and secondary compensatory peripheral vasoconstriction. Muscle wasting is noticed in the advanced cases of DCM and has been attributed to the release of cytokines (TNF-alpha and interleukins) and poor muscle perfusion.

Animals with DCM present in left and/or right-sided (biventricular) heart failure. The presence of biventricular failure is a poor prognostic indicator. Left-sided congestive heart failure is characterised by expiratory dyspnoea and perihilar or more generalised crepitations on auscultation due to the presence of pulmonary oedema. Dogs in biventricular failure will additionally show signs of ascites (distended abdomen with fluid thrill), pleural effusion (discordant dyspnoea, muffled heart and lung sounds ventrally, dull percussion ventrally) and distended jugular veins due to increased central venous pressures. Hepatomegaly and less commonly splenomegaly are other indicators of right-sided heart failure. Animals with right-sided heart failure can also develop diarrhoea as a result of protein losing enteropathy due to acquired intestinal lymphangiectasia.

Thoracocentesis and abdominocentesis

Effusions should be tapped to determine their quality. Pleural effusions due to right-sided heart failure are mainly modified transudates but they can be chylous (Fig. 2).

Fig. 2: *Chylous pleural effusion from a dog in biventricular failure.*

Ascitic fluid secondary to congestive heart failure is often a modified transudate. Real transudates are rare in case of right-sided congestive heart failure and more commonly indicate hypoproteinaemia or portal hypertension.

Thoracocentesis is required to alleviate respiratory distress due to severe pleural effusion. It is not advisable to drain ascites unless the effusion is compromising respiration.

Blood pressure

Blood pressure monitoring (Fig. 3) should become an essential part of every dog's routine clinical examination. A low systemic blood pressure in a dog with IDCM can indicate severe myocardial failure and cardiogenic shock. The blood pressure is also important for treatment choice (see article treatment of DCM).

Fig. 3: *Equipment for indirect blood pressure monitoring via Doppler technique.*

Serum biochemistry

Pre-renal azotaemia due to the low cardiac output is common. Use of diuretics can worsen this prerenal azotaemia. Alanine aminotransferase and alkaline phosphatases are often mildly elevated due to chronic hepatic congestion.

If DCM is confirmed serum carnitine and plasma taurine determination (Fig 4) is optional (available in the UK from Idexx). Plasma taurine levels are recommended in at least American Cocker Spaniels, Golden Retrievers, Dalmatians and Newfoundland dogs.

Plasma Carnitine

Total: 12-38 µM/l
Free: 8-36 µM/l
Esterified: 0-7 mM/l
Plasma in heparin tube, centrifugate immediately

Plasma Taurine
Normal >50 nmol/ml
DCM >20 nmol/ml
Plasma in heparin tube, centrifugate immediately

Fig. 4: *Normal plasma taurine and carnitine levels.*

Atrial natriuretic peptide (ANP) is released from the atrial myocardium under stretch and increased transmural pressures and appears to be increased significantly in dogs in heart failure. The precursor Pro-ANP is also released from the atria, has a longer half-life and is therefore easier to measure than ANP. Recently a PRO-ANP Elisa assay (Idexx) has been commercialised as a marker for heart failure. It is the author's opinion that this blood test will never replace proper clinical judgment and radiography for the diagnosis of heart failure. Although some promising data have been published for the use of PRO-ANP as a diagnostic and a prognostic marker for heart failure, too few numbers of dogs with dilated cardiomyopathy (and in general) were present to make definite conclusions. One needs to be aware that collection and transport of the samples needs to be highly controlled for accuracy.

ECG

Common P-QRS-T complex abnormalities in DCM include tall R-waves (left ventricular enlargement), deep Q-waves (in case of biventricular enlargement) and ST-coving.

However, these days electrocardiography is mainly used to characterise arrhythmias, rather than to indicate chamber enlargement. Atrial fibrillation develops as a consequence of atrial stretch and dilation and is often a sign of the presence of DCM in large breed dogs. However, in giant breeds slow atrial fibrillation (Fig. 5) can be present without atrial dilation or other overt signs of DCM. Clinical experience has shown that most of these animals develop DCM within 3-5 years. Ventricular premature complexes (couplets and triplets) and ventricular tachycardias (Fig. 6) are also a hallmark of DCM and are considered risk factors for sudden death.

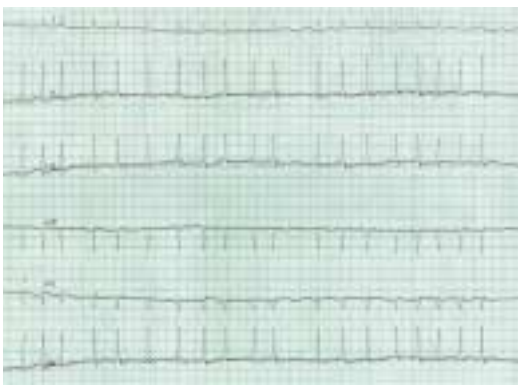

Fig. 5: *Slow atrial fibrillation with a ventricular response rate at 110 BPM (Irish Wolfhound).*

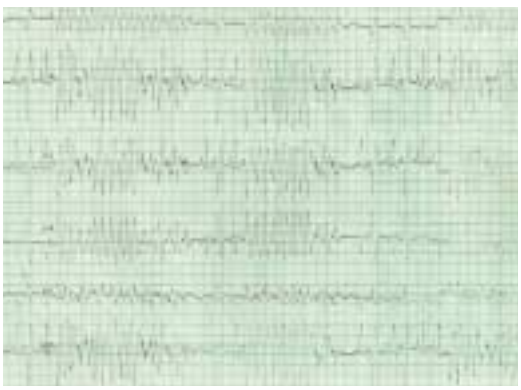

Fig. 6: *Malignant ventricular arrhythmias in a Great Dane with DCM that died suddenly.*

Holter

Since the ability to detect the presence of arrhythmias by the use of a 2 minute ECG-strip is very limited in asymptomatic dogs a 24-hour ECG monitoring (Holter, Fig. 7) is often advocated.

Fig. 7: *Holter monitoring in a syncopal German Shepherd Dog.*

In animals with no overt clinical signs Holter monitoring might be indicated to detect paroxysmal arrhythmias. Breed standards have been established for the Boxer and Dobermann.

Holter monitoring or event recording has become an essential part of the diagnostic approach in syncopal animals.

Radiography

Radiography remains essential for the confirmation of congestive heart failure. In case of left-sided failure left atrial enlargement, venous congestion, interstitial and alveolar infiltrates can be observed (Fig. 8). Cardiogenic

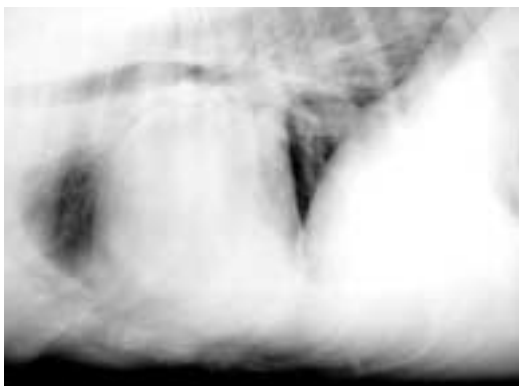

Fig. 8a: *Thoracic radiograph of a Dobermann with DCM in failure (lateral).*

pulmonary oedema has a perihilar distribution on lateral radiographs but on the dorsoventral projection the infiltrates often start in the right caudal lung lobe. Right-sided heart failure is characterised by a wide caudal vena cava, the presence of pleural effusion, hepatomegaly and/or ascites.

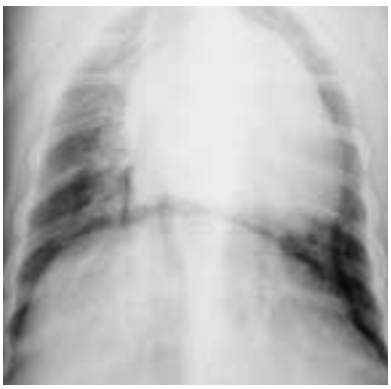

Fig. 8b: *Thoracic radiograph of a Dobermann with DCM in failure (dorsoventral).*

Despite radiography being extremely useful for the detection of cardiomegaly (Vertebral Heart Size as a guideline), it should not be used as a prognostic indicator. One has to remember that most giant breeds and Spaniels with DCM will show massive cardiomegaly on radiographs but that Dobermanns often show only very mild ventricular enlargement (but with obvious left atrial dilation) and that Boxers almost never have an enlarged heart. Care should be taken not to confuse the globular cardiac shape (without any chamber indentation at all) seen with pericardial effusion with the generalised dilated heart of cardiomyopathy. In the former there is never any sign of left-sided heart failure (venous congestion, pulmonary interstitial and or alveolar infiltrates) but signs of right-sided failure (wide caudal vena cava, ascites and pleural effusion) are often present in both diseases.

Radiography is also a very useful tool to evaluate response to therapy.

Doppler echocardiography (Fig. 9)

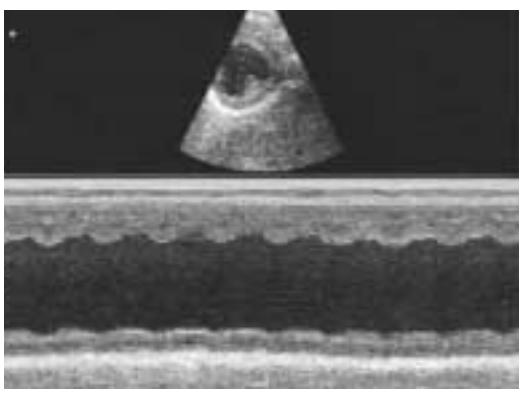

Fig. 9a: *M-Mode echocardiography of the left ventricle showing poor contractility.*

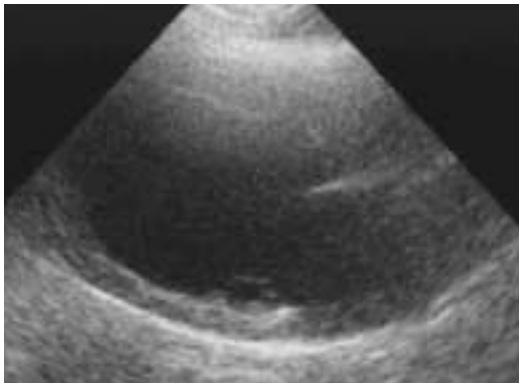

Fig. 9b: *Two-dimensional echocardiography showing a dilated spherical left ventricle with thinned walls (fractional shortening was 11%).*

Due to the remodelling that occurs in DCM the left ventricle becomes more spherical in shape and the myocardial walls become thinner. The left atrium is often only moderately dilated compared to animals with mitral valve disease (the exception is the Dobermann, see above). Myocardial failure is defined as a decrease in myocardial contractility. However, myocardial contractility depends on preload, afterload and heart rate and is therefore difficult to measure accurately by echocardiography. Therefore multiple indicators of contractility (left ventricular diameter in end-systole, fractional shortening, ejection fraction, systolic time intervals) are used to estimate systolic myocardial function. The primary echo-abnormality in myocardial failure will be an increased left ventricular end-systolic diameter. Fractional shortening (left ventricular diameter in diastole - left ventricular diameter in systole/ left ventricular diameter in diastole) is the parameter most often used in the clinical setting (FS<25% indicates myocardial failure). One has to remember that this index is very breed specific. Other indices like ejection fraction (left ventricular volume in diastole-left ventricular volume in systole/left ventricular volume in diastole), E-point septal separation (EPSS) and the systolic time intervals (pre-ejection period PEP and ejection time ET) should always be measured in case of doubt.

Mitral and tricuspid regurgitation are often present and are secondary to annular dilation of the valve apparatus due to the ventricular enlargement (Fig. 10). If atrial fibrillation is present the velocity of the regurgitation fraction can be quite variable explaining the variation in murmur intensity often heard with atrial fibrillation (Fig. 11).

DNA testing

Considering the familial history in most breeds pedigree information is being used to establish heritability patterns (see Table 3 in article about breed characteristics UK Vet Vol 8 No 7). At the time of writing there is no genetic test available to screen for DCM gene carriers or affected

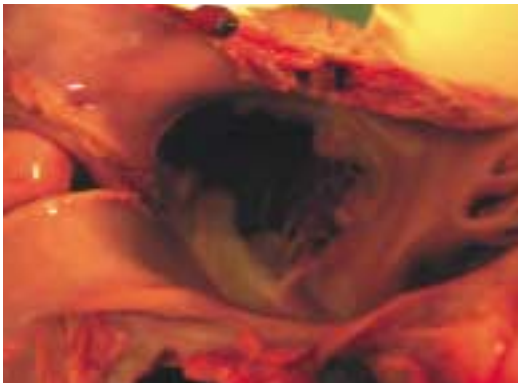

Fig. 10: *Post-mortem specimen showing severe mitral annular dilation.*

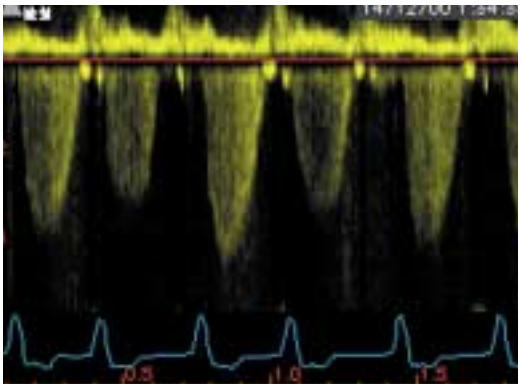

Fig. 11: *Doppler echocardiography showing the variation in mitral regurgitation (and consequently murmur grading) with atrial fibrillation.*

animals. However with the worldwide research into genetic linkage in DCM a genetic marker of DCM might become available in the next years.

CONCLUSION

The diagnosis of idiopathic dilated cardiomyopathy is not always straightforward and may demand multiple ancillary tests. In those animals with equivocal results or in breeding stock repetitive evaluation by experienced individuals is strongly recommended.

FURTHER READING AND REFERENCES

KITTLESON and KIENLE (1998) Small Animal Cardiovascular Medicine (Mosby).

FOX, SISSON, MOISE (2000) Textbook of canine and feline cardiomyopathy (W. B. Saunders).

DUKES-MCEWAN (2000) Canine dilated cardiomyopathy. In practice p 520-530; 620-626.

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These multiple choice questions are based on the above text. Readers are invited to answer the questions as part of the RCVS CPD remote learning program. Answers appear on the inside back cover. In the editorial panel's view, the percentage scored, should reflect the appropriate proportion of the total time spent reading the article, which can then be recorded on the RCVS CPD recording form.

1. Which statement is true regarding the occult phase of Idiopathic DCM in the dog:

a. Compensatory mechanisms are responsible for the lack of clinical signs in these animals but the presence of the disease can be shown by further investigations.
b. The disease is present but cannot be confirmed by the means of electrocardiography nor echocardiography.
c. A DNA test is available to screen for these animals.
d. There are clinical signs of congestive heart failure, arrhythmias or sudden death.
e. The disease is present and can only be confirmed by echocardiography.

2. Which statement is false regarding the clinical examination in a dog with Idiopathic DCM:

a. Pallor of the mucosae with sluggish capillary refill time is a common finding in advanced DCM.
b. A dog with DCM often has a stronger apex beat than a dog with a normal heart.
c. Arrhythmias are often noticed on auscultation.
d. Thoracic auscultation will show the presence of a systolic murmur with a grading exceeding IV/VI.
e. The presence of pulsus alternans indicates severe myocardial failure.

3. Which statement is false regarding radiography in Idiopathic DCM:

a. Radiographic cardiomegaly is extremely useful as a prognostic indicator.
b. Cardiogenic pulmonary oedema often has a perihilar distribution on lateral radiographs but on the dorsoventral projection the infiltrates often start in the right caudal lung lobe.
c. Boxers almost never have an enlarged heart.
d. Spaniels with DCM will show massive cardiomegaly on radiographs
e. Dobermanns often show only very mild ventricular enlargement but with obvious left atrial dilation.

4. Which statement is false regarding ECG and Holter monitoring in Idiopathic DCM:

a. Common QRS complex abnormalities include tall R-waves and ST-coving.
b. Atrial fibrillation is always an indicator of congestive heart failure.
c. In animals with no overt clinical signs Holter monitoring might be indicated to detect paroxysmal arrhythmias
d. Ventricular couplets and triplets and ventricular tachycardias are considered risk factors for sudden death.
e. Holter monitoring or event recording has become an essential part of the diagnostic approach in syncopal animals.

5. Which statement is false regarding echocardiography and Idiopathic DCM:

a. Myocardial contractility is difficult to measure accurately by echocardiography.
b. The primary echo-abnormality in myocardial failure will be an increased end-systolic diameter.
c. Fractional shortening is a very accurate parameter to determine myocardial contractility.
d. Mitral and tricuspid regurgitation are often visualised
e. If atrial fibrillation is present the velocity of the regurgitation fraction can be quite variable explaining the variation in murmur intensity often heard with atrial fibrillation.