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DILATED CARDIOMYOPATHY: DEFINITION, DIFFERENTIALS AND DIAGNOSTIC WORK-UP

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Definition

Dilated cardiomyopathy (DCM) is the most common cardiomyopathy in dogs.

The diagnosis relies on the identification of a pre-defined phenotype (either on echocardiogram or post-mortem). The definition of DCM revolves around the identification of left- or biventricular dilatation associated with reduced systolic function in the absence of abnormal loading conditions or other disease processes that could cause systolic impairment.

Differentials

When we talk about DCM, we should be aware that a dilated left ventricle with decreased systolic function is not a single disease process and that differentials are broad to include a primary myocardial disease (which is what we refer to idiopathic/ primary DCM), but also other conditions. Because sometimes it is difficult to completely rule out secondary causes of DCM, it is preferable to talk about DCM phenotype, rather than DCM for this reason.

Here is a list of differentials that could present with a DCM phenotype:

1. **Primary**
    a. idiopathic DCM
2. **Secondary** DCM phenotype
    a. Inflammatory/ Myocarditis (infectious/ not infectious)
    b. Arrhythmia-induced
    c. Ischemic cardiomyopathy
    d. Athlete's heart (non-pathological condition)
    e. Diet-associated
    (taurine deficiency/ non taurine, vitamin E, Selenium, Toxin-related/ heavy metal, cobalt or environmental)
    f. Drug-induced (doxorubicin- other less commonly utilized drugs)
    g. Systemic diseases
    (hypothyroidism, pancreatic disease, hypoadrenocorticism, muscular dystrophies, glycogen storage disease/ amyloidosis)

Other cardiac conditions may also mimic a DCM phenotype and should be ruled out during the diagnostic investigations:

1. Chronic volume overload – congenital (PDA, VSD less likely)
2. Chronic volume overload- acquired (MR with systolic dysfunction)
3. End-stage subaortic stenosis
4. Aortic regurgitation

Primary dilated cardiomyopathy

The available data on DCM prevalence is probably not updated (from prevalence studies performed nearly 30 years ago), but was estimated to be between 0.5 to 1.1% in dogs referred to specialist centers.

Some breeds are known to be more prone to develop DCM, with higher prevalence rate in purebred dogs than mixed breeds (purebred dogs had a prevalence of 0.65 compared to 0.16 in mixed breeds). It mainly affects large breed dogs, with some exceptions. Because there is a wide variation in the clinical presentation and histopathology within breeds, there was a tendency to sometimes present DCM based on the breed (i.e., Dobermann Pinscher Cardiomyopathy, Boxer Cardiomyopathy, Cocker Spaniel Cardiomyopathy, Dalmatian Cardiomyopathy). This may be useful to present breed-specific characteristics, but diagnosis and treatment are most of the time very similar within breeds (with very few exceptions).

Here is a list of dog breeds more commonly affected by DCM (breed-related DCM): Dobermann Pinscher, Newfoundland, Cocker Spaniel, Dalmatian, Irish Wolfhound, Portuguese Water Dog, Estrela Mountain Dog, Toy Manchester Terrier, Great Dane, St Bernard, Scottish Deerhound and Giant Schnauzer. Please note that there are geographical variations in DCM prevalence, so this should not be a strict criterion for DCM diagnosis. Boxer dogs have been cited as affected by DCM, but because they are also prone to develop ARVC (which can affect all 4 chambers), it is difficult to ascertain if Boxer dogs with 4-chamber dilation and poor systolic function represent a different entity from ARVC, or if it is just an advanced, end-stage ARVC phenotype.

Generally, male dogs are more prone to develop DCM compared to females, but this predisposition may vary within the breed.

As this is considered an acquired disease condition, DCM tends to develop in adult dogs (6-8 years old), but it can be seen in younger (3 yo) or older (up to 12 yo) dogs. There are also exceptions depending on the breeds, ie, Portuguese Water dogs develop a juvenile form of DCM, affecting dogs as young as 4 months old and Great Danes tend to be affected earlier than Dobermann Pinschers on average (from 4 years of age).

Diagnostic work-up

For clinical purposes, the clinical presentation of dogs with DCM can be divided into two phases:

- *Preclinical ("occult") phase*:
  no clinical signs, but myocardial or electrical abnormalities are already present;
  this phase can last for months/ years
- *Clinical ("Overt") phase*:
  clinical signs are present (can be intermittent or persistent)

For this reason, a dog with DCM may be brought to visit for

1. non-cardiac/ unrelated reasons
   (i.e., annual visit, presence of a comorbidity or for a non-cardiac emergency),

2. non-specific clinical deterioration
   (i.e., weight loss, inappetence, lethargy)
3. cardiac-related reasons
   (i.e., exercise intolerance, syncope, coughing or tachypnea).

In some breeds, the owners may want to pursue cardiac screening as well.
In the overt phase, dogs may be brought to visit for different complaints. The complaints can be divided depending on the main manifestation of DCM :

- Left-sided congestive heart failure
  (tachypnoea, coughing, exercise intolerance, weakness)
- Biventricular congestive heart failure
  (tachypnoea, exercise intolerance, abdominal distension)
- Arrhythmias
  (exercise intolerance, syncope)
- Sudden cardiac death
  (unfortunately no prodromes to sudden cardiac death, but owners may report exercise intolerance and syncopal events prior to the event)

The diagnostic work-up of dogs with presumed or confirmed DCM depends highly on the stability of the patient.

A physical examination should be performed in all patients, with different degree of detail depending on the severity of clinical signs.

Diagnostic investigations range from point-of-care ultrasound, thoracic radiographs, to ECG or 24-hour Holter monitoring, echocardiogram and laboratory bloodwork, including cardiac biomarkers.

Below you can find a small summary of pros/cons of each technique.

Point-of-care US (POCUS)
PROs: quick, reduces patient restraint, diagnostic for overt cases of DCM.
CONs: may hinder concurrent respiratory diseases. Less helpful in equivocal cases of DCM.

Thoracic radiographs
PROs: provides diagnostic information in symptomatic patients and helps rule out non-cardiac causes of tachypnoea.
CONs: less sensitive for preclinical DCM or screening. Requires sedation or restraint which may be risky in unstable patients.

ECG
PROs: helps characterize arrhythmias and streamline arrhythmia management
CONs: not helpful in most cases of screening. Intermittent arrhythmias may be missed

24-hour Holter monitor
PROs: helpful to characterize cardiac rhythm over a longer period of time. May help with preclinical disease recognition

CONs: an additional cost to the client, may require renting of the device to other clinics. May slightly delay diagnosis (reading time)

Echocardiogram
PROs: gold standard for diagnosis and screening.
CONs: equivocal cases can still be challenging to diagnose (++ arrhythmic form)

The author will discuss these aspects in more detail in this session and the next one on management and treatment of DCM.

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