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RIGHT VENTRICULAR ARRHYTHMIC CARDIOMYOPATHY: AN UPDATE ON BOXER CARDIOMYOPATHY

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Myocardial disease in the Boxer dog has typically been classified under a variety of different names including boxer cardiomyopathy, dilated cardiomyopathy, and familial ventricular arrhythmias, among others. However, more recent studies have demonstrated that the disease has many similarities to a human disease called Arrhythmogenic Right Ventricular Cardiomyopathy. The similarities between the diseases include clinical presentation, etiology and histopathology among others. Additionally, since the disease is most commonly characterized by ventricular arrhythmias, syncope and sudden death as opposed to systolic dysfunction and ventricular dilation, it seems appropriate to stress the arrhythmic form of the disease. Therefore, more recently, the disease in the Boxer has become referred to as Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC).

ETIOLOGY

Arrhythmogenic right ventricular cardiomyopathy is a familial disease in the boxer and appears to be inherited as an autosomal dominant trait. Therefore, theoretically, there should not be any silent carriers. Unfortunately, the disease also appears to be a disease of variable genetic penetrance and affected dogs can have many different presentations ranging from totally asymptomatic to severe forms where dogs die of sudden cardiac death at a young age.

DIAGNOSIS

The presentation in the adult Boxer may take one of three forms as originally proposed by Dr. Harpster. Although previously classified as forms I-III, they may also be considered as: concealed (asymptomatic with VPCs), overt (syncope) or myocardial dysfunction (left ventricular systolic dysfunction). The ventricular premature complex (VPCs) in boxers with ARVC typically have a wide, upright QRS in leads I, II, III, and AVF (left bundle branch block morphology), consistent with the right ventricular origin of this arrhythmia. The arrhythmia may be quite intermittent and in many cases, may require a 24 hour Holter monitor for documentation. Interpretation of the Holter results can sometimes be challenging because strict criteria for this diagnosis does not exist. However since it is unusual for a normal dog to have any VPCs in a 24-hour period, the observation of > 100 VPCs, or periods of couplets, triplets or runs of ventricular tachycardia are abnormal. A larger number of VPCs or a greater complexity of the arrhythmia (ventricular tachycardia, bigeminy, etc) has been associated with the development of clinical signs. Supraventricular premature complexes may be seen but not frequently, and are more commonly associated with the myocardial dysfunction form of the disease.

SCREENING

The familial etiology has led to a wide spread interest in screening dogs before selecting them for breeding. Since this disease appears to present as an electrical abnormality more often than one of myocardial dysfunction, screening efforts should be based on annual Holter monitoring and possibly,

annual echocardiography. Unfortunately, clear criteria for affected status are still being determined and day to day variability of arrhythmias exist. However, dogs that are symptomatic (syncope, heart failure) or have evidence of ventricular tachycardia on a Holter should not be used for breeding. Additionally, dogs that have over 100 left bundle branch block morphology VPCs/24 hours are probably highly suspicious. To help decrease the risk of making an error when adding or removing a dog from a breeding program, owners should be encouraged to screen annually rather than put to 100% emphasis on a single Holter reading. Since the disease is adult onset and an increase in VPCs has been observed with age in affected animals, an animal that is clear at the age of two is not guaranteed to stay clear. Additionally, an animal with a few hundred VPCs at the age of two years may have more, less or the same number the next year!

TREATMENT

At this time, there is no evidence that treatment will significantly alter the outcome for affected dogs. However, there is no evidence that treatment will not alter the outcome. Therefore, our current recommendations are the following:

ASYMPTOMATIC DOGS WITH VENTRICULAR TACHYARRHYTHMIAS

If an arrhythmia is detected on routine examination, a Holter monitor should be performed to evaluate for the frequency and complexity of the arrhythmia. Although a strict relationship between the development of symptoms and the number of VPCs does not exist, treatment is generally started if > 1000 VPCs/24 hours, runs of ventricular tachycardia or evidence of the R on T phenomenon exist. Owners should be advised that ventricular antiarrhythmics have the potential for proarrhythmic effects and that treatment is not known to decrease risk of sudden death.

DOGS WITH SYNCOPE

Dogs with syncope and ventricular arrhythmias are generally started on treatment. There are two choices for treatment that are well tolerated and have been shown to decrease VPC number and complexity, sotalol (1.5-3.5 mg/kg, q 12hr, orally) and the combination of mexiletine (5-8 mg/kg, q 8hr, orally) and atenolol (12.5 mg/DOG, q12h, orally). It is likely that there is individual variation for drug response and if a poor response is observed with one drug, a different one may prove to be more effective. Ideally, a Holter monitor would be placed before starting therapy and repeated two to three weeks after starting therapy to demonstrate the effect of therapy. Significant day-to-day variation in VPC number exists and a therapeutic effect is likely to exist if at least an 85% reduction in VPC number while on medication is observed.

DOGS WITH SYSTOLIC DYSFUNCTION AND HEART FAILURE

If echocardiography demonstrates significant systolic dysfunction and ventricular dilation, treatment as for DCM is indicated.

Symptoms of heart failure should be alleviated with furosemide (1-3 mg/kg, q8-12h) and ACE inhibitors (enalapril, 0.25-0.5 mg/kg, q12h, orally). As heart failure becomes more refractory, the addition of spironolactone (1-2 mg/kg, q12h, orally) should be considered for aldosterone blocking effects. Digoxin may be added when the heart failure becomes refractory or atrial fibrillation is observed. Pimobendan can be

added on a case by case basis with approval from the FDA at a dose of 0.3 mg/kg orally twice a day. Pimobendan may have significant difference for dogs with DCM and heart failure in terms of both quality of life (increased appetite, activity, etc) and survival. Dogs with DCM and chronic heart failure often develop weight loss and cardiac cachexia. Fish oil supplementation has been shown to decrease cardiac cachexia in some cases. It may be dosed at 40 mg/kg EPA and 25 mg/kg DHA . For easy dosing, most 1.0 gm capsules contain 180 mg EPA and 120 mg DHA.

Additionally, supplementation with L-carnitine might be considered (50 mg/kg, q8-12h, orally), since a small number of affected dogs have demonstrated improvement in systolic function and prognosis after supplementation.

PROGNOSIS

Sudden death is always possible. However, many dogs may live for years on antiarrhythmics without symptoms, some of these may eventually develop ventricular dilation and systolic dysfunction.

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