Arrhythmogenic Right Ventricular Cardiomyopathy (Boxer Cardiomyopathy)

ABOUT THE DIAGNOSIS

Arrhythmogenic right ventricular cardiomyopathy is also called ARVC for short, or boxer cardiomyopathy, because it occurs predominantly in boxer dogs. ARVC is a heart disease that affects the heart muscle tissue, is almost always of genetic origin (inherited from the father, the mother, or both), and is usually first detected in adult dogs, not puppies. That is, the genetic defect that causes ARVC is inherited from one or both parents, but the first detectable sign (typically an electrocardiogram [ECG] or Holter monitor) is usually only detected after the age of 2 or 3 years old.

The heart tissue is made up mainly of muscle cells. In the normal, healthy heart, the heartbeat is triggered by a normal, organized wave of electricity that travels through these muscle cells to cause the heart to pump. When the heart pumps, the blood within the heart chambers moves to the rest of the body, providing oxygen and various nutrients to the body’s tissues. In the heart affected by ARVC, the origin of some heartbeats may be abnormal, causing an erratic—and often very fast—beat of the heart (abnormal rhythm, “arrhythmia”). Boxers with this disease may have episodes of collapse that may or may not be triggered by excitement or exercise. Some boxers faint (syncope) when the heart begins to beat irregularly because it may beat so irregularly and so fast that the pumping action of the heart is compromised and not enough circulation reaches the brain to maintain consciousness. Between these episodes, a dog with ARVC usually appears healthy, comfortable, and externally normal. Much less commonly, and as a more serious form of ARVC, the heart muscle tissue may hamper the circulation such that fluid accumulates in the dog’s lungs and causes breathing problems, or in the abdomen/belly causing abdominal enlargement.

The problem is recognized in the boxer breed internationally, but not all boxers have ARVC. Many boxers have perfectly healthy hearts. Some boxers with ARVC have an abnormal heart rhythm but no symptoms; these may be carriers of the disease who go on to develop symptoms later in life. The diagnosis is serious, because any boxer with this disease has an increased risk of sudden cardiac death.

Diagnosis: Boxer cardiomyopathy may be suspected by your veterinarian based on your dog’s history and symptoms, if any. A cardiac arrhythmia may be present at the time of examination. An electrocardiogram (ECG, EKG) is the definitive test. For this test, small metal clips are gently attached to the dog’s skin in several places as the dog rests quietly. These clips are attached to a machine that can convert the heart’s electrical impulses to a graphic representation on paper; it is the same type of ECG/EKG as in human medicine. The abnormal heart rhythm that is characteristic of ARVC is sporadic, however; it comes and goes with no predictable pattern. In some dogs with ARVC, the abnormal heart rhythm (an arrhythmia called premature ventricular complexes) is much worse with excitement, whereas excitement makes no difference to some other dogs with ARVC. Therefore an electrocardiogram performed in the clinic may or may not reveal the abnormality. If there is uncertainty, portable heart monitors (Holter monitor, cardiac event monitor) may be used for assessing the heartbeat over 24 hours or intermittently over several days, respectively. Finally, since many other disorders unrelated to ARVC can cause irregularities in the heart rhythm, an accurate diagnosis usually requires further assessment (e.g., x-rays of the chest and abdomen, ultrasound of the abdomen and heart, blood tests), which can be tailored based on the veterinarian’s initial findings.

Cause: The specific cause of ARVC is unknown. It is genetic in origin. Striatin is a protein in normal cardiac muscle cells and a mutation in this protein is present in many boxer dogs with ARVC. There is a blood test available for this mutation, but it is not perfect because some dogs with ARVC do not have the mutation. The genetic test can be used to help screen for the disease, reducing or eliminating ARVC depends on screening adult boxers prior to breeding using a Holter monitor (24-hour portable ECG/EKG monitor that the dog wears at home for a day), and excluding dogs that have a high number of irregular heartbeats from any breeding program. Importantly, healthy boxers may have a heartbeat that is irregular but healthy (“respiratory sinus arrhythmia”), a common finding in healthy human athletes. Therefore, be sure not to jump to conclusions if you notice that your boxer’s heart rhythm seems irregular: only an ECG/EKG can tell whether this is healthy or abnormal.

LIVING WITH THE DIAGNOSIS

A diagnosis of ARVC carries a guarded prognosis (life span may be shortened) because a sudden deterioration or even a fatal event is possible if the heart rhythm suddenly becomes very erratic. Depending on the results of the diagnostic tests (see above), different medications can be given, including omega-3 fatty acids (all cases) and cardiac antarrrhythmic medications (some cases, based on the veterinarian’s assessment). However, because ARVC is genetic in origin, these drugs only treat symptoms and are not a cure. Despite taking drugs, the disease may continue to progress and the goal of treatment is to reduce the symptomatic episodes. Since no two dogs are the same, it may be necessary to give several medications until one is found that helps your dog best.

Boxers diagnosed with this disease should not be bred. Breeding restriction like this is in order to avoid perpetuating the problem in offspring. If your boxer is involved in a breeding program, your veterinarian may recommend regular (perhaps annual) testing with a Holter monitor to make sure there are no signs of ARVC. If possible, dogs diagnosed with ARVC should avoid stressful situations or events that cause excitement. This can be a difficult task for owners, but common-sense attention to minimizing environmental triggers of excitement (without severely compromising quality of life) may reduce the risk of severe or even life-threatening complications. Examples include disconnecting a doorbell and posting a “please knock” sign if the doorbell makes the dog very agitated and limiting the intensity of play with other dogs. This can be difficult in a happy-go-lucky dog like the boxer, so the point is to limit, not abolish, physical activity; to make the dog active and playful in a slow, consistent way (slow, longer leash-walks, for example); and to avoid sudden bursts of intense physical exertion (cut back on sprinting, ball-chasing, etc.).

TREATMENT

Medications are available to help control the abnormal cardiac rhythm caused by ARVC. Your veterinarian can prescribe medications based on the results of x-rays, ultrasound exam, and/or electrocardiogram. These medications help some dogs but not others, and unfortunately some dogs continue to exhibit symptoms
related to this diagnosis even after taking medications. In such cases, other types of antiarrhythmic medications may be used, or further testing may be recommended to ensure that another disease process is not to blame.

Omega-3 fatty acid supplementation (fish oil; 780 mg eicosapentaenoic acid [EPA] and 497 mg docosahexaenoic acid [DHA] per dog, given by mouth or with food once a day) has been shown to reduce the number of erratic heartbeats in boxers with ARVC and should be given to every dog with ARVC.

Each dog diagnosed with this disease requires a tailored treatment regimen that may need to be changed periodically, depending on the dog’s response. Your veterinarian will determine the best plan for your pet.

DOs
• If your boxer has periods of weakness and/or fainting, take him or her to your veterinarian for an evaluation. Be sure to keep the dog as calm and comfortable as possible.
• Remember that even though boxers with ARVC look normal for more than 99% of the time, having ARVC means a risk of sudden collapse and even death and excessive excitement may increase the risk.
• Give medications exactly as directed. If you are concerned that the medications are not working, or are working differently than expected, contact your veterinarian before making any changes yourself.
• Talk to your veterinarian about having your boxer tested regularly for this disease, especially if involved in a breeding program.
• Remember that ARVC can be a complex and difficult disease, and you may wish to seek a second opinion from a veterinary cardiologist: see www.acvim.org or www.vetspecialists.com (North America) and www.ecvim-ca.org (Europe) for a list of veterinarians that are board-certified specialists in veterinary cardiology.

DON’Ts
• Do not breed a boxer that is diagnosed with this disease.
• Do not assume that your boxer does not have this disease if no symptoms are evident, because it is common for irregular heartbeats to go unnoticed if an ECG/EKG or Holter monitor has not been performed.
• Do not stop giving a drug without talking to your veterinarian. Some drugs must be gradually decreased or serious side effects can result.
• Do not allow a dog with ARVC to swim in deep water or walk/hike near ledges or cliffs. When ARVC produces symptoms, fainting is the most common one. Fainting when swimming (risk of drowning) or when near a ledge or cliff (risk of disastrous fall from a height) can only be prevented by avoiding those locations.

WHEN TO CALL YOUR VETERINARIAN
• If your dog shows negative reactions to the drugs including weakness, anxiety or nervousness, vomiting, diarrhea, hives (bumps under the skin), constipation, seizures, etc. These are uncommon with the medications used for treating ARVC.
• If you cannot return for a scheduled visit.

Also available in Spanish.