

Specialist Veterinary Cardiology Consultancy



Authored by: Dr Luca Ferasin, DVM PhD CertVC PGCert(HE) DipECVIM-CA (Cardiology) GPCert(B&PS) FRCVS EBVS and RCVS Specialist in Veterinary Cardiology

What is Arrhythmogenic Right Ventricular Cardiomyopathy?

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) or "Boxer Cardiomyopathy" or "Familial Arrhythmias" is the most common heart disease seen in this breed and is familial (inherited) in origin. The disease is always characterised by the presence of an abnormal heart rhythm (arrhythmia) and can be classified as:

- 1. **Type I:** no symptoms
- 2. **Type II:** episodic fainting (*syncope*), usually associated with exercise or excitement
- 3. **Type III:** a patient with echocardiographically and radiographically apparent myocardial abnormalities (cardiac dilation, poor function), with or without congestive heart failure (CHF)

An association has been found between ARVC in Boxers and a genetic mutation. The mutation is inherited, and while it seems to be associated with ARVC in Boxers, studies indicate that other unknown mutations also play a role in the disease.

The age of Boxers diagnosed with ARVC varies from 1 to 11 years, with a mean age of 5-7 years. Approximately 50% of these dogs present with syncope, while others may never develop clinical signs. Unfortunately, sudden death is also frequently reported. The disease is a condition affecting the muscle of the heart (*myocardium*). Fatty and fibrous tissue infiltrate into the muscle of the heart causing it to beat irregularly. The disease observed in Boxers appears

very similar to ARVC described in people, with sudden unexpected death reported in the majority of cases, especially in young fit adults, often during exercise.

The diagnosis of ARVC in Boxers can be challenging. An ultrasound scan of the heart (echocardiography) should be performed to rule out other heart diseases that can be responsible for fainting. The detection of an arrhythmia on electrocardiogram (ECG) is very suggestive of ARVC. However, due to the episodic nature of the arrhythmia, standard ECG recording (approximately 1 minute) may fail to demonstrate it. Therefore, 24h ambulatory ECG recording ("Holter", picture below) represents a much more sensitive test



to detect arrhythmias that occur on a daily basis. This is a small recorder connected to the dog's chest via self-adhesive electrodes and secured to a harness or vest in order to be carried by the patient for 24h. The presence of more than 500 abnormal beats (green arrow below) confirms ARVC.



Unfortunately, sometimes, the arrhythmia may occur less frequently than on a daily basis and even a 24h ECG may miss the ECG abnormalities. Therefore, more advanced ECG monitoring (implantable loop recorder, ILR) may be recommended in these cases. ILR have been used both in humans and pets to determine episodic arrhythmias for at least two decades. This small recorder is implanted under the skin and will provide up to 3 years of continuous leadless ECG monitoring. Arrhythmias can be captured automatically or by activating a

dedicated remote control during or immediately after a fainting episode.

g

What should be expected now?



ARVC is currently an untreatable disease. However, arrhythmias can often be controlled by omega-3 fatty acid supplementation (fish oil) with a significant reduction of the number of abnormal heart beats and fainting episodes. When fish oil

supplementation is not sufficient to control the abnormal rhythm, the addition of anti-arrhythmic drugs (e.g. atenolol, mexiletine, sotalol, amiodarone) can be considered. Unfortunately, there is no clear evidence that these drugs can increase life expectancy or reduce the risk of sudden death.

Implantable defibrillators are often used in human patients with ARVC to automatically convert life-threatening arrhythmias. These devices have also been tested in Boxers with ARVC but they are expensive and tend to discharge unnecessary electric shocks even when the dog is not experiencing arrhythmias. Prognosis is guarded and although many

affected dogs can survive for years, the risk of sudden death remains high.

As ARVC is suspected to be a genetic disease, it is important that affected animals are not

used for breeding to prevent passing the disease on to further generations.



This handout provides a general overview on this topic and may not apply to all patients.

Please do not hesitate to contact us if you require any additional information (www.cardiospecialist.co.uk)