Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC)

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 in the UK. It is familial in origin and it appears to be increasing in prevalence. The disease is always characterised by 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:* congestive heart failure (with or without syncope)

Males and females seem to be equally affected without a specific gender predisposition. The disease is inheritable and could be caused by a variety of genetic mutations, although a specific cause has not been clearly identified yet. The age of Boxers diagnosed with ARVC varies from 1 to 11 years, with a mean age of 6-9 years. Approximately 50% of these dogs present 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, especially during exercise. In people, ARVC is probably the most common cause of sudden death in

young adults, after trauma and suicide.

The diagnosis of ARVC in Boxers can be challenging. An ultrasound scanning of the heart (*echocardiography*) should be performed to rule out other heart diseases that can be responsible of syncope. The detection of 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 (or "Holter") 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 (see picture above). Unfortunately, the arrhythmia may occur less frequently than on a daily basis and even a 24h ECG may miss the abnormality. Recently, a new device (implantable loop recorder or 'Reveal') has been used both in humans and pets to determine episodic arrhythmias. This small recorder (picture below) is implanted under the skin using light anaesthesia 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 after a fainting episode.

What should be expected now?

ARVC is currently an untreatable disease. However, arrhythmias can often be controlled by anti-arrhythmic drugs (eg atenolol, mexiletine, sotalol, amiodarone) with a significant reduction of the number of fainting episodes and improvement of quality of life . 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 ARVC patients to convert automatically life-threatening arrhythmias and they have also been used in Boxers with ARVC. However they are expensive and require a very careful programming to avoid un-

necessary electroshocks.

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.