Myocardial dysfunction, dilated cardiomyopathy (DCM). Prevalence and inheritance in Irish Wolfhounds

Impaired cardiac function may be a secondary condition caused by another disease. Most common causes of secondary cardiomyopathy are hypothyroid disease, myocarditis and heart base tumors. Diagnostic criteria vary between breeds and there is no single golden standard that applies to all breeds and cases.
Diagnosis is based on ultrasound examination and ECG examination, other examinations are supportive.
Treatment is aimed at boosting cardiac contractility, treating congestive heart failure and controlling arrhythmias.

DCM is a most common heart disease in large and giant breed dogs. In Irish Wolfhounds the incidence of DCM reaches approximately 20 percent, meaning that about 1 dog in 5 develops the disease at same point of its life. The mean age of onset has been estimated to be 4,5 years, most dogs developing the disease between 3-7 years of age. Males have a significantly higher risk than females and the onset in females also tends to happen at later age. Most affected dogs have a concurrent atrial fibrillation which may be a first sign of the disease.
As a breed Irish Wolfhounds may have a higher survivability than many other breeds. Despite the high incidende of DCM in Irish Wolfhounds, the left ventricular function does not often become severely compromised until the later stages of the disease, which improves the prognosis. DCM is still a major cause of death in this breed.
Two comprehensive studies trying to establish the mode of inheritance in Irish wolfhounds has been conducted. (O. Distl, A.C. Vollmar, C. Broschk, H. Hamann ja P.R. Fox 2007, sekä U. Philipp, A. Vollmar, J. Häggström, A. Thomas ja O. Distl 2012).
In both studies a simple monogenic mode of inheritance could be rejected. The model that best explained the results was a mixed monogenic-polygenic model with an autosomal dominant sex dependent major gene and further polygenic effects. The later study could identify 5 minor loci suggestively associated with DCM. The major gene is located on CFA37 and its gene action is significantly different between male and female dogs. The major risk allele is a common variant which might impair a big part of Irish wolfhound population.
The associated genes seem to affect lipid metabolism of the cells in the heart muscle.
The echocardiographic diagnostic criteria for DCM in Irish Wolfhounds have been agreed. Accepted cut values are end-diastolic dimension more than 61 mm, end-systolic dimension more than 41 mm, fractional shortening below 25 %, ejection fraction below 40-44 %.

In the last three years I have performed a cardiac examination for 45 Irish Wolfhounds. Several individuals have been followed for many years. So far 9 of these dogs have been diagnosed to have DCM. Only three dogs have developed classical form of the disease with loss of contractility and cardiac enlargement. Others have developed atrial fibrillation. 5 dogs are on medication either because of congestive heart failure or atrial fibrillation with rapid ventricular rate. Three dogs are assessed as equivocal because of premature ventricular complexes or tachycardic episodes. These dogs will probably develop DCM in the future. Findings are consistent with other similar studies. Results of heart examinations send to the Irish Wolfhound club do not reveal as many positive cases. This is because of three reasons. Many dogs are female which have smaller risk of developing the disease. Several dogs have been so young at the moment of examination that they do not show signs of the disease but may develop them later in life. Dogs with atrial fibrillation assessed equivocal should be changed to have DCM.

Measuring cardiac biomarker BNP might be beneficial in those cases that are otherwise uncertain.

In summary dilated cardiomyopathy is a highly prevalent and often lethal disease in Irish Wolfhounds. Most dogs develop the disease between 3-7 years of age but the onset may be at an older age.
Cardiac biomarkers alone are not sensitive enough to diagnose the disease. Genetic testing is not yet available for Irish Wolfhounds.
At the moment diagnosis is based on ultrasound examination supported by ECG analysis. 24 hour Holter monitoring is available to assess the risk for sudden death in dogs that have been diagnosed to have DCM. All the carriers will not be found before they are used for breeding with methods available for us at the moment. I can´t recommend routine testing before 3-4 years of age.

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