What is Juvenile Dilated Cardiomyopathy?

Juvenile Dilated Cardiomyopathy in Toy Manchester Terriers is characterized by the death of young animals (typically less than 12 months of age) as a result of a sudden and fatal arrhythmia. Genetic research has determined that the condition is caused by a mutation in a gene associated with potassium channel formation, which is an integral part of the process regulating electrical activity in the heart.

What causes JDCM?

Each beat of an animal's heart results from rhythmic contractions of the heart muscle. Those contractions are regulated by electrical signals resulting from the controlled flow of ions into and out of heart muscle cells through channels in their membranes. There are a number of different ions involved in this process (including sodium, calcium and potassium) and each one has a role to play in depolarizing or repolarizing the heart muscle cells so they contract regularly. Juvenile Dilated Cardiomyopathy in Toy Manchesters is caused by a mutation in a gene governing one of the potassium channels involved in this process. Researchers theorize that the mutation interrupts the function of normal potassium channels, causing damage to heart tissue at the microscopic level. Over time, normal heart tissue is replaced by scar tissue, interfering with the heart's ability to conduct the electrical current needed to make it beat. The eventual result is a fatal arrhythmia (abnormal heart rhythm) leading to sudden death.

What are the symptoms of JDCM?

Affected dogs usually appear healthy with no signs of heart disease present before the sudden passing. The only external abnormality is that affected male puppies may have unilateral or bilateral cryptorchidism (undescended testicles on one or both sides). For a number of the puppies, their sudden passing occurred within a day of general anesthesia/surgery or exercise.

How do I know if my dog has JDCM?

Until the fatal arrhythmia happens, affected dogs appear healthy (even during vet exams) and behave like normal puppies. As a result, the only way to know conclusively if a dog or puppy may be affected by JDCM is to perform a test to determine their genetic status.

Is Juvenile Cardiomyopathy ever seen in English Toy Terriers?

This is a complicated question. Cases of juvenile cardiomyopathy have been observed in English Toy Terriers, however all cases reported **to this study** have had pedigrees that include Toy Manchester Terriers. It is recommended that English Toy Terrier breeders with Toy Manchester Terrier behind their dog(s) test their breeding stock. It is also recommended that English Toy Terrier Clubs or individual breeders interested in determining whether the gene exists in the pure ETT population consider further investigation through testing.

Juvenile Dilated Cardiomyopathy (JDCM) Test Result Interpretation

The JDCM test screens for a specific DNA mutation that has been discovered in a cardiac potassium channel gene. The mutation is inherited in an autosomal recessive manner. We have designated the letter D to indicate the deleterious (JDCM) form of the gene and N to indicate the normal form of the gene. A dog's particular combination of N or D forms of the gene is known as its genotype.

Clear (N/N) Clear dogs have no copies of the mutation, and cannot pass the mutation on to offspring.

Carrier (D/N): A carrier dog has only one copy of the mutation. Since the mutation is recessive, one copy will not cause JDCM. Carriers will, on average, pass the mutation on to half of their offspring. This does not mean that they need to be taken out of the breeding pool, but they should only be bred to clear dogs to avoid producing affected puppies.

Affected (D/D): An affected dog has two copies of the mutation that causes JDCM (this is also referred to as being homozygous affected). We do not know if these dogs are fertile, as few have survived to sexual maturity. However, we do not recommend breeding an affected dog**.** Based on the data available so far, we believe that this is a fully penetrant mutation. This means that all affected dogs will develop JDCM, and pass away at a young age (most likely in their first year of life). Males often have cryptor chidism (undescended testicles). However, affected dogs otherwise can appear healthy with no signs of heart disease until they suddenly pass.

Breeding Outcomes

* Clear (N/N) x Clear (N/N) = 100% Clear (N/N)
* Clear (N/N) x Carrier (D/N) = 50%
Clear (N/N), 50% Carrier (D/N)
(This is an average, individual litters may see anywhere from 100% Clear to 100% Carrier)
* Carrier (D/N) x Carrier (D/N) = 25
% Clear (N/N), 50% Carrier (D/N),
25% Affected (D/D)
(This is an average, individual litters may see more or less)

Further details can be found at: http://www.canadamt.com/juvenile-cardiomyopathy-study.html

Test Submission forms can be downloaded at
https://drive.google.com/file/d/0B2_OEHvXGzcMTmk4a3pOVnJzbHc/view