

THE CARDIOMYOPATHY CHRONICLES: AN UPDATE

By Rod Humphries

The pervasiveness of dilated cardiomyopathy (DCM) and its high transmission rates in Doberman Pinschers, as underlined by a recent genetic study, is beginning to resonate with breeders and owners.

The decision by Beverly Seielstad of Encore Dobermans — owner of one of America's top stud and show Dobermans, the four-year-old Ch. Foxfire's Magic Dragon — to publicly announce that he has DCM, has had a dramatic trickle down effect for the owners of 21 bitches bred to the dog and the owners of about 120 puppies, the last yet to be born. (See March 2008 issue and an advertisement/letter to breeders in this issue).

I have declared Magic Dragon, a two-time Top 20 contender, the "Poster Boy in the fight against Dilated Cardiomyopathy."

I have also had contact with a number of owners who are now doing cardiac testing as a result of my articles in the February 2008 issue chronicling the entire spectrum of DCM. Several have found their dogs to have early stages of DCM.

The articles also prompted potential puppy buyers to seek my advice on what to look for and what questions to ask breeders when purchasing a puppy. One puppy buyer quickly declined a potential sale because the breeder had no cardiac test for the sire or the dam... and brushed off testing as unnecessary.

Leading veterinary cardiologist and geneticist, Dr. Kathryn Meurs, of Washington State University, late last year released the results of an eight year study by a team of veterinary specialists (see Doberman Pinscher Magazine, February 2008 issue) which determined that DCM in Doberman Pinschers is transmitted by the autosomal dominant mode of inheritance. The study, which tracked four generations of an extended family of Dobermans, determined that 25 of the 35 dogs able to be fully diagnosed by the veterinary team had DCM, a staggering 71 per cent.

The disease, which can be clearly traced to America's foundation stock which had direct roots from Germany, is so prevalent it has invaded every country, every bloodline. It is everywhere and while no official figures are available because of a number of factors, it is obvious that many hundreds, maybe thousands are dying from either congestive heart failure or sudden death every year.

Many readers are still grappling with the genetics — especially the differences between the actual mode of inheritance, autosomal dominant, and the more familiar autosomal recessive which is not applicable to DCM in Dobermans. I have had phone calls and emails asking for clarification on various points, so I will reiterate the dramatic effects of the autosomal dominant mode of inheritance as it pertains to DCM in Dobermans:

(Note: Genes come in pairs, one inherited from the father and one from the mother. Two like copies of a gene are called "homozygous" and unmatched genes are "heterozygous").

- **Diseases and traits inherited in the autosomal dominant mode are expressed by a single dominant gene, not a recessive (hidden) gene.**
- **Autosomal dominant requires only one mutant gene from either the sire or dam of a mated pair to transmit the disease to a large percentage of offspring. Dog breeders are more familiar with the autosomal recessive mode which is not in play in DCM in Dobermans. For comparison only, any disease or trait inherited in the autosomal recessive mode requires two matching genes to produce affected offspring.**
- **Dobermans which are heterozygous for DCM (one mutant gene and one clear gene) are themselves afflicted with the disease. For comparison only, in autosomal recessive mode an animal with one mutant gene for any disease would be declared a "carrier" and would never contract the disease, but could transmit it when a like gene is matched from a breeding mate. Not so in autosomal dominant. There are no classic "carriers" in DCM in Dobermans.**
- **As an autosomal dominant disease or trait is passed openly from generation to generation, there is no skipping of generations. It is an unbroken stream of DCM from father or mother (or both) to daughters and sons, with no hidden mutant recessives.**
- **Autosomal dominant traits are normally easy to eliminate: to break the chain a breeder simply would not breed an affected animal. But without a DNA test -- and with the late onset of the disease in Dobermans — there are serious complications because many animals do not express the disease until after the peak breeding years.**
- **Because autosomal dominant needs only one mutant gene to pass the disease to offspring, the transmission rates are devastating and show why Dobermans have the highest incidence of DCM in the canine world.**

The Deadly Transmission Rates of DCM in Dobermans

The following autosomal dominant transmission facts were chronicled in the original articles, but cannot be hammered home often enough in the struggle to slow the disease:

- **An animal which is heterozygous for DCM (one mutant gene and one clear gene) is itself afflicted with the disease and will pass it to at least 50 per cent of its offspring. The 50 percent transmission rate applies even if the heterozygous animal is bred to a genetically clear animal.**
- **An animal which is homozygous dominant for DCM (two matching mutant genes) will transmit the disease to 100 percent of its offspring regardless of whether the breeding partner is genetically clear of DCM.**
- **Two breeding animals which are both heterozygous for DCM will transmit the disease to 75 percent of their offspring. This is the classic distinction of autosomal dominant: two affected breeding animals produce 25 per cent clear offspring.**

All the above transmission rates are true over a large sample size. (See the Genetic breakdown and Punnett squares in the February 2008 issue).

The frustration for Doberman breeders (and all breeds afflicted with DCM) is that there is currently no DNA test for DCM...a disease which can have a late onset and thereby creates havoc in breeding programs because it often surfaces after a generation or two down the breeding road.

Detection is often masked when a breeding animal is misdiagnosed at death or when it dies of some other disease or is accidentally killed before the DCM has been expressed. When no necropsy is performed by a cardiac specialist to determine if there was DCM, there is often surprise when the disease appears in offspring.

The other problem is the “code of silence” of the dog show culture in which some breeders and owners remain mute and even hide cases of DCM, thereby increasing the incidence of the disease. Some use avoidance maneuvers because the disease would affect sales and studs. “Well, they have to die of something,” is a familiar catch-cry. Unfortunately, the disease has been found in dogs as young as two-and-half years (and as old as 14.5 years) and in my own kennel I lost dogs at four, five and six years. The range of death in general studies is six to 10 years.

Annual cardiac testing by a heart specialist, while restricted in its impact because it is a mere snapshot at that time, nonetheless can often pick up early symptoms such as the case with Ch. Foxfire’s Magic Dragon who was diagnosed at four years of age.

An open registry to track the disease in pedigrees as an aid for breeders is the goal of this writer. Because the disease has to appear openly in every generation, every snippet of information on DCM in a bloodline can be a vital piece of the puzzle for a breeder planning a litter or a stud owner who wants to know the true status of his or her dog. That is exactly what happened with Ch. Foxfire’s Magic Dragon. Beverly Seielstad read the pedigrees of my nine dogs killed by DCM and decided that because there were some mutual animals, that she should have Magic Dragon checked.

Beverly’s decision to release all information on Magic Dragon has opened the door — and placed pressure on — other breeders and owners to test and tell. It also places enormous pressure on the owners of bitches bred to Magic Dragon. Beverly has asked that they contact all puppy owners with the information.

If Magic Dragon is heterozygous for DCM with one copy of the mutant gene, then at least 50 percent, or about 60 of his 120 puppies, will contract the disease. If he is homozygous with two copies of the mutant gene, then all 120 puppies will get the disease. (Percentages are true over a large sample size).

Owners who plan to breed Magic Dragon’s offspring will obviously need to be diligent and test before they make any move. The trickle down effect is quite devastating but Beverly was rightly adamant that she had to be honest... and had a moral and legal obligation to inform all those who used her dog that he had been diagnosed with the deadly disease. (I am looking into the legal ramifications of non-disclosure of the disease).

The public disclosure of my own personal saga of nine deaths at my home in Texas a number of years ago has had a powerful impact — but a death sentence announced on a current Top 20 Dog and sire of many champions who is only four years of age, has had more far reaching effects.

It jolted a lot of people in the Doberman breed and has put pressure on breeders and stud owners to do physical echocardiograms and electrocardiograms to determine if breeding stock has symptoms of the disease. Magic Dragon is an outwardly healthy animal who will probably continue to be shown and compete in the Top 20 at the DPCA National later this year. He is a dog with a death sentence and probably has one to three years to live.

Beverly wants to take her “poster boy” out to shows, and hopefully show him in the Top 20 at the National, because she believes that breeders and owners need to be reminded that such a healthy, outgoing dog has a ticking time bomb inside. “I am hoping it will be a stark reminder that healthy looking dogs can have this disease...and maybe it will push them to test and follow good breeding practices,” she said.