

# FACTS ABOUT PROGRESSIVE RETINAL ATROPHY

#### By Leona Domino

PRA is a general term for a number of eye diseases.

These diseases have included certain features. One is that the diseases begin at a very specific time in each breed. The diseases progress at a very predictable rate. The dogs that are affected with the disease become blind at an age very specific to the breed. Some become blind early, some late. The end result is blindness.

#### **PAPILLON PRA:**

There are a number of different kinds of PRA. PRA can be divided into either dysplastic disease, where the cells develop abnormally, or degenerative disease, where the cells develop normally but then undergo a damaging change. Usually, dogs affected with PRA first experience difficulty seeing at night or in dim light. As the disease progresses, the dog are finally blind both day and night. Papillons are affected with a slowly progressive degeneration that causes blindness at seven to eight years old. At present there is no cure and no treatment to arrest the course of degeneration in the affected dogs.

# HOW IS PRA TRANSMITTED ?

PRA IS NOT CONTAGIOUS. It is an inherited disorder, passed along through the generations by a simple autosomal recessive gene. Simple autosomal recessive inheritance means that if a dog is PRA affected ( is either blind or going blind ) BOTH of the parents must possess the PRA gene. The dog has inherited the PRA gene from BOTH sire and dam, giving him a double-dose of the gene. Only when a dog has inherited TWO PRA genes, one from each parents, can he be actively affected with PRA blindness.

The parents of a PRA affected animal may have normal vision themselves, even though they both "CARRY" the PRA gene. They are referred to as CARRIERS. A carrier shows NO physical signs of possessing the PRA gene. A carrier has no outward signs that it carries the PRA gene. The way a PRA carrier reveals itself as a carrier is by producing a PRA AFFECTED (blind) offspring.

TERMS :

Dogs have 78 chromosomes and the sex chromosomes are 39 or  $\frac{1}{2}$  of the dog's genetic picture. When breeding each parent donates  $\frac{1}{2}$  of its genetic material to the new puppy. Therefore you will note that a carrier will not have signs of PRA but can continue the disease 50% of the time. If bred to a bitch with the same gene then an affected puppy can be produced. It takes 2 donated genes to produce affected.

# WHY ARE CARRIERS OF SUCH CONCERN ?

Since carriers appear normal outwardly, the problem becomes complicated. The situation can be compared to an iceberg. Blind animals make up the tip of the iceberg that shows, but underneath the surface are the carriers. www.pcagenetics.com

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They cannot be easily detected as of this date. It is possible for carriers to remain "hidden" in breeding programs before they finally, if ever, produce an affected puppy. Each time the carrier is used for breeding, a percentage of more "little-carriers" are added to the breeding population. As time goes on the more carriers there are within a breed, the greater chances are that affected will ultimately be produced by breeding two "hidden" carriers together.

## ARE PRA CARRIERS A BIG PROBLEM IN OUR BREED ?

That is impossible to answer. Since there is no reliable way to identify the carrier status until it reveals itself by producing a blind puppy, many carriers will never be identified.

#### WHAT IS BEING DONE TO FIND CARRIERS ?

Research is being conducted at Michigan State University (Michigan, USA) to find the marker on the gene that would result in breeders having a test that would tell them the genetic state of the breeding dog. Dr. Simon Petersen-Jones is the scientist working on this project. The Papillon Club of America (PCA) is assisting with getting a colony of affected dogs available to the doctor to use in his research.

## WITH NO TEST AVAILABLE YET,

HOW DO I BREED TO AVOID PRODUCING A CARRIER OR AFFECTED DOG ?

Until a test is developed, you can have your dogs examined by a veterinary ophthalmologist , record the finding with CERF and stay vigilant regarding your dog's vision. Keep in mind that night blindness or dim night vision is the first sign.( ARTICLE : Night Blindness Test )

Do not breed to a dog that has produced an affected puppy or that has not been cleared by an eye exam. Eye exams by a V.O. should start by 2 years and be yearly until 8-9 years of age.

You can give your puppies a night eye test yourself.

Keep in mind : very young puppies will show signs of poor night vision. This may not be visible to the V.O. doctor until 2 to 3 years of age, -do the night test on all your puppies.

3-4 month old puppies may show night vision hesitancy. This would be a serious indication of vision problems to come.