PRA in Toy Poodles

Christine Scruggs, VMD

Progressive retinal atrophy, or PRA, is a genetic disease which is present in all three varieties of the poodle breed. PRA is actually a group of degenerative conditions involving the retina of the eye. Several of these conditions are inherited photoreceptor diseases which appear similar in clinical signs and are genetic in nature, i.e. they can be passed from generation to generation and can be traced throughout pedigrees. This group of diseases is present in several breeds of dogs, including the poodle, and is also seen in certain breeds of cats. The mode of inheritance for PRA in the toy and miniature poodle is described as a simple autosomal recessive gene. This means that an affected individual must carry two copies of the defective gene in order to express the disease. In any given population of poodles there will also be carrier individuals and clear individuals. Carriers are dogs which do not show any clinical signs of the disease, but can pass one copy of the defective gene on to their offspring; the other copy they carry is the normal gene, and will not express the disease. Clear individuals are dogs which carry two copies of the normal gene, and will never reproduce the disease. Below are described several proposed breedings between different types of individuals and the puppies they might produce:

Key:

 **= Clear**  **= Carrier**  **= Affected**

Breeding A: *Two carriers*

 Will statistically produce: 25 % Clear 

50 % Carrier 

25% Affected 

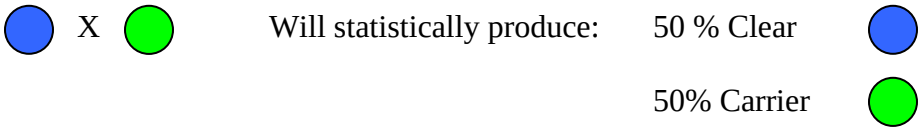
Breeding B: *Two clears*

 Will statistically produce: 100% Clear 

Breeding C: *Two affecteds*

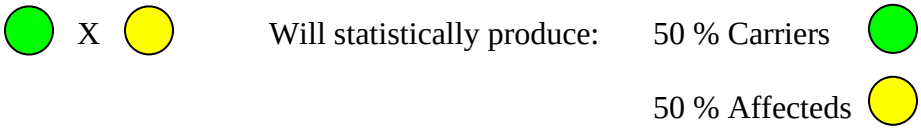
 Will statistically produce: 100% Affected 

Breeding D: *Clear to carrier*



Breeding E: *Clear to affected*

Breeding F: *Carrier to affected*



PRA has been researched in the toy poodle and the type of retinopathy for the toy and miniature breed has been described as progressive rod-cone dysplasia, or prcd. Typically, the owner of a PRA affected poodle will first notice night blindness, or loss of vision in dim or poor lighting. The night blindness is caused by the degeneration and loss of the rod cells within the retina, which are responsive to low or grey light levels. The night blindness will eventually progress to total blindness as the disease progresses and the cone cells (which detect higher light levels and color) of the retina also degenerate. The total loss of vision can occur over months to years and does not usually begin until the dog is 1 year of age or older, even as late as 3-5 years of age in the poodle. If an owner notes that night blindness is occurring in his/her dog, an ophthalmologic exam will determine if this is due to PRA or another ocular problem. Changes in the retina itself will show an increased reflectivity of the retina as well as decreased size of the blood vessels supplying the retinal layer, as the cells of the retina atrophy.

In the past, the only way to screen for PRA was to perform a CERF (canine eye registration foundation) exam on breeding animals every year. A board certified ophthalmologist examines the eye and notes any abnormalities present. If the dog has signs of PRA present in the retina, the ophthalmologist will note this on the examination form, and a CERF number is not assessed. If the eye is normal, or has abnormalities labeled "breeder's option" then this is also noted on the examination form, and a CERF number is assessed for that poodle, valid for a year from the date of the exam. This type of genetic screening is still the only one available to standard poodle breeders for PRA. However, toy and miniature poodles now have a DNA test offered by Optigen (http://www.optigen.com/opt9_test.html) to determine clear, carrier, and affected individuals for the prcd form of PRA. Because there are other ocular problems which are genetic in nature, toy and miniature poodles should still have yearly CERF exams.

There are many advantages to performing the PRA DNA test at an early age. Breeders can screen for clear and carrier poodles, and will know that an affected poodle

will eventually develop blindness. Breeders can also plan a breeding strategy for this disease within their kennel. This does not mean that all carrier and affected dogs should be spayed or neutered and placed in pet homes. What this does mean is that a breeder can use the genetic information on PRA for an individual poodle within his/her kennel and decide whether or not to breed a particular individual based upon the remainder of the genetic characteristics the breeder is trying to improve upon, avoid, and strengthen within his/her line.

For instance, an affected toy poodle may have other genetic advantages that preclude culling him or her from the genetic pool within a kennel. If the affected toy has strengths that the breeder does not want to lose within her line, she can still breed this dog. In order to avoid reproducing the disease, the breeder must find a match for the affected poodle who is clear for the disease. Therefore, all of the puppies will be carriers, but none of them will be affected by the disease. As time goes on, the breeder should replace her optimum breeding animal (in this case the affected dog) with a son or daughter that is one step better for the disease as well as having the other genes the breeder is trying to strengthen in the line. In other words, keep a carrier puppy from the first breeding that has the strengths the breeder is looking to improve upon in the line. In the next generation, keep a clear individual from the carrier puppy if possible. Over time, if breeders follow this strategy, the incidence of affecteds should drop dramatically, while clears should increase, and carriers will either increase or remain the same.

Many breeders would ask the question – why not just breed clear individuals only, and eradicate the disease altogether? The answer is if breeders focus on a single disease and attempt to eradicate that problem without considering the entire genetic package of the breed, they will encounter a "bottleneck" or loss of genetic diversity by eliminating a large number of potential breeding animals from the population. This can be extremely detrimental to a breed and can result in even more genetic problems, rather than solving them. It is much more reasonable to look at the whole picture of what a breeder is trying to accomplish, to focus on many generations of work rather than on individual poodles within a line. Therefore, it is entirely reasonable to breed a carrier or even an affected poodle, as long as he/she is bred to a clear individual, and as long as the focus is the entirety of a genetic picture for a kennel rather than on individual results.

The proposed breedings (A through F) mentioned above show what a breeder can expect to reproduce statistically when breeding clear, carrier, and affected toy poodles in all possible combinations. These are only valid results when breeding toy or miniature poodles who have been DNA tested with the Optigen PRA test, which is focused on the prcd form of PRA. As mentioned at the beginning of the article, PRA is a *group* of degenerative conditions affecting the retina of the eye. Therefore, even if a toy poodle is an Optigen clear dog, it does not mean this poodle is clear for other genetic causes of PRA. However, the prcd form of PRA is the most common cause of this disease (greater than 75% of cases) and most likely a toy or miniature poodle that is clear for this form of the disease will not reproduce PRA.

Overall, toy (and miniature) breeders have been very successful in reducing the incidence of PRA in their varieties, using the DNA test available. Breeders who utilize this test can eliminate reproducing this form of PRA from their lines, although they are not likely to be able to eliminate carriers for the disease without risking creating a bottleneck situation as mentioned earlier. It would be a dire mistake for breeders to only allow clear individuals to reproduce, focusing on this single disease rather than the genetic pool as a whole. Although perhaps considered a radical idea, in a rare case it may even be wise to breed an affected animal, if this individual has an overwhelming number of positive genes to contribute to the population. Of course, this individual would have to be bred to a clear poodle, to avoid reproducing affected puppies. Breeders must make informed decisions utilizing DNA tests such as that for PRA, as well as any other genetic tests available for their breed. The breeder must also focus on the larger picture – generations of accomplishment – rather than on producing a single spectacular dog. And, as always, open and honest exchange of information between all breeders would greatly facilitate improving the health of toy, miniature, and standard poodles.