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Progressive Retinal Degeneration in Dogs

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Progressive retinal degeneration (PRD) is a group of heritable blinding diseases affecting many species of animals. It is also known as progressive retinal atrophy (PRA), and is a major cause of blindness in dogs. This condition has been recognized in numerous canine breeds. It is characterized by the progressive, untreatable degeneration or destruction of the retina.

The retina is the thin, light-sensitive membrane lining the inside back of the eye. It is easiest to think of it as the film in a camera. Similarly to the film, the retina converts light into an image. If the retina is defective, a poor image will result. As the retina becomes more damaged, the animal sees less and less. Because of the arrangement of cells in the retina, night vision is usually lost first. As degeneration proceeds, day vision becomes compromised, and eventually the dog becomes blind.

Progressive retinal degeneration manifests in two basic forms: early onset and late onset. It has been shown that the early onset variety is due to defective development of the cells of the retina. Because of this maldevelopment, this type of PRD can be diagnosed early in life. Breeds such as the collie (including the Border Collie) and the Irish Setter are prone to this type of PRD.

The late onset PRD occurs sometime after the retinal tissue matures which is about six weeks of age. Although this may begin within a few weeks of this time, clinically detectable signs of retinal disease may not be manifest until the dog is two to five years old, hence the descriptive term, late onset PRD. Although this type of PRD has been seen in numerous breeds, the miniature and toy poodle, and American and English Cocker Spaniel, perhaps by virtue of their popularity, are among the most common breeds affected. It should be noted that breeds in which early onset PRD is prevalent may also have an unrelated late onset PRD.

Although the heritable nature of PRD has been proven, the exact mode of inheritance is unknown for the majority of breeds in which PRD is seen. It is, however, known for some breeds and in these has been shown to be a simple autosomal recessive gene. This means that the disease is not related to sex. This also means that both the dam and sire must carry the gene in order for puppies to get the disease. The dam and sire may either be affected or unaffected. Not all puppies will have the disease but, unaffected puppies may also be carriers of the disease. The collie, Irish Setter, Norwegian Elkhound, and poodle are in this category. There is some evidence in the poodle, however, that there may be some influence based on sex of the parents when an affected dog is mated with an unaffected carrier (that is carrying just one defective gene instead of the two required for the disease be observed). In the simplest situation, one would expect about 50% of the offspring to be affected with the disease. In one research institution, however, it was found that when the male was affected and the female was a carrier (not affected but carrying the defective gene), the number of affected pups was far fewer than would be expected.

Although there continues to be no effective treatment of PRD, there have been some advances in our understanding of these group of diseases in the last decade. Two of these may have some bearing on the ability to detect carriers or to diagnose the disease earlier than presently clinically possible. It has been demonstrated that there is a specific biochemical defect in the retina of breeds such as the collie and the Irish Setter. It is believed that this defect is responsible for the lack of proper retinal development in certain forms of early onset PRD. There also has been demonstration of improper formation of some of the retinal proteins which may be responsible for causing or modifying the disease. These discoveries may be quite useful in the near future.

For early onset PRD, the diagnosis of the disease can be made within the first year of life using standard clinical techniques such as retinal examination with the ophthalmoscope. Electrical testing of the retina, termed electroretinography or ERG, can detect this subset of PRD within the first few weeks of life in many instances. In late onset PRD, clinical diagnosis usually is not definitive until the disease is moderately advanced, as mentioned earlier. The ERG can be a valuable diagnostic tool in these cases, in that sophisticated equipment may detect affected individuals within the first year of life