Progressive Retinal Atrophy

Progressive retinal atrophy or degeneration (PRA or PRD) is the name for several diseases that are progressive and lead to blindness. First recognized at the beginning of the 20th century in Gordon Setters, this inherited condition has been documented in over 100 breeds.

Anatomy of the eye

The eye is a very delicate, yet surprisingly durable organ. It consists of several layers. The cornea is a transparent layer that covers the front of the eye. The iris is the colored part of the eye and it is responsible for letting in more or less light. The lens gathers and 'bends' light in order to focus it on the retina. In between the cornea and lens is an area of fluid which bathes the lens and helps it focus. The retina lines the inside of the eye and converts light into signals which travel down the optic nerve to the brain. A large area between the lens and the retina contains a jelly-like fluid called 'vitreous'. The vitreous gives the eye its form and shape, provides nutrients, and removes waste products.

The retina

The retina is the structure affected in PRA. This important part of the eye receives the light gathered and focused by the other eye structures. It takes the light and essentially converts it into electrical nerve signals that the brain, via the optic nerve, interprets as vision. The retina contains photoreceptors, called rods and cones, which help the animal see in darkness (rods) and see certain colors (cones).

What is PRA?

There are multiple forms of PRA which differ in the age of onset and rate of progression of the disease. Some breeds experience an earlier onset than others do; other breeds do not develop PRA until later in life.

Normally, the photoreceptors in the retinas develop after birth to about 8 weeks of age. The retinas of dogs with PRA either have arrested development (termed retinal dysplasia) or early degeneration (deterioration) of the photoreceptors. Retinal dysplastic dogs are usually affected within two months of birth and may be completely blind by one year. Retinal degenerative dogs are affected from one year to eight years of age and the symptoms progress slowly.

PRA worsens over time. The affected animal experiences night blindness initially because the rods are affected first. The condition progresses to failed daytime vision.

What are the signs of PRA?

Signs may vary depending on the type of PRA and its rate of progression. PRA is non painful and outward appearance of the eye is often normal, i.e.; no redness, excess tearing, or squinting. Owners may notice a change in personality of their pet such as a reluctance to go down stairs or down a dark hallway. This is characteristic of night blindness, in which vision may appear to improve during the daytime. As the disease progresses, owners can observe a dilation of the pupils and the reflection of light from the back of the eye. If the blindness is progressing slowly, the owner may not notice any signs until the dog is in unfamiliar surroundings and the lack of vision is more apparent. In some animals, the lens of their eyes may become opaque or cloudy.

How is PRA diagnosed?

Depending on the form of PRA, characteristic changes in the retina and other parts of the eye may be observed through an ophthalmic examination by a veterinary opthalmologist. More sophisticated tests such as electroretinography may also be used. Both tests are painless and the animal does not have to be anesthetized. If no abnormalities are found during the exam by a board certified veterinary ophthalmologist, the dog can be certified free of heritable eye disease through the Canine Eye Registration Foundation (CERF).

How is PRA treated?

Unfortunately, there is no treatment for PRA, nor a way to slow the progression of the disease. Animals with PRA usually become blind. Dogs and cats are remarkably adaptable to progressive blindness, and can often seem to perform normally in their customary environments. Evidence of the blindness is more pronounced if the furniture is rearranged or the animals are in unfamiliar surroundings.

Can PRA be prevented?

PRA has been shown to have a genetic component. Puppies from parents who have no history of the disease and have been certified free of PRA will have less risk of developing the disease. Affected animals should not be bred and should be spayed or neutered. The littermates or parents of animals with PRA should also not be bred. If your dog develops PRA, notify the breeder, if possible.