

Progressive Retinal Atrophy (PRA)

Ophthalmology Service



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Specialistssm

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What is PRA?

Progressive retinal atrophy is the name of a group of hereditary diseases that involve degeneration of the retina, and eventual blindness. At this time, there is no treatment for PRA. There are many different forms of PRA, and each of the forms are the result of a specific hereditary defect. The disease is inherited, and occurs in both eyes (bilateral). Pure-bred dogs are most commonly affected, and specific breeds are predisposed to specific types of PRA (see *Breed Specific Information section*).

Despite the form of PRA, most dogs present with similar clinical signs. The age of onset will vary between breeds. Diagnosis of PRA by your veterinarian can be made with an eye exam, and an electro-retinogram (ERG, detects activity of the retina). Diagnosis can be assisted with genetic testing (see *Genetic Testing section*).

Breed Specific Information

prcd-PRA (Progressive Rod-Cone Degeneration)

This form of PRA is the most common form. Rod cells degenerate first, causing night blindness. It is a recessive trait, which requires that both parents pass on a mutant, disease-causing gene. Parents can either be carriers (1 mutant gene, 1 normal gene), or affected animals (2 mutant genes). There is a genetic test available for this form of PRA in the **American & English Cocker Spaniels, American Eskimo, Australian Cattle Dogs, Australian Shepherds, Australian Stumpy Tail Cattle Dogs, Chesapeake Bay & Labrador Retrievers, Chinese Cresteds, Cockapoos, Entlebucher Mountain Dogs, Finnish & Swedish Lapphunds, Kuvasz, Labradoodles, Lapponian Herders, Miniature & Toy Poodles, Nova Scotia Duck Tolling Retrievers, Portuguese Water Dog, & the Spanish Water Dog**. The test requires a blood sample. *prcd*-PRA can be prevented through strategic breeding of tested dogs.

Dominant PRA of Mastiffs & Bullmastiffs

Mastiffs & Bullmastiffs are the only breeds known to be affected with a dominantly-inherited form of PRA. This means that only one copy of the mutant gene is required to cause the disease. Genetic testing can identify a dog as normal, heterozygous affected (1 mutant gene, 1 normal gene), or homozygous affected (2 mutant genes). About 50% of the puppies of a heterozygous animal will be affected, and 100% of the puppies of a homozygous animals will be affected.

Other Forms of PRA

Welsh Corgis are affected by a form of PRA called rcd3-PRA (rod & cone degeneration type 3). **Irish Setters** can be affected by rcd1-PRA. The mutation in **Miniature Schnauzers** is called Type A-PRA, because there are multiple causes in this breed. In **Samoyeds** and **Siberian Huskies** the disease is X-linked (on the X chromosome). This form is called XL-PRA.

Clinical Signs of PRA

- Impaired night vision, *nyctalopia*
- Dilated, non-responsive pupils
- Increased tapetal reflection, or “eye shine”
- Cataracts (late in the disease)
- Eventual blindness

Genetic Testing Available

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