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What is progressive retinal atrophy?

The cells of the retina receive light stimuli from the external environment and transmit the information to the brain where it is interpreted to become vision. In progressive retinal atrophy (PRA), deterioration of the retinal cells causes blindness. The retina lines the back of the eye. The inner layer is the neural retina (called simply the retina) which has 9 layers, the outermost of which consists of the photoreceptor cells - the rods and cones. The outer layer of the retina is the retinal pigmented epithelium (RPE). In dogs the retina is not mature until 6 or 7 weeks of age.

The term progressive retinal atrophy covers several types of inherited degeneration (deterioration) of the retina. Sub-classifications of PRA are based on the age at which dogs show signs of the disease and the type of retinal cell which is affected.

Generalized PRA: These diseases affect primarily the photoreceptor cells. Both eyes are similarly affected and dogs eventually become totally blind.

i) **Early onset photoreceptor dysplasias:** In these conditions, the photoreceptor cells develop abnormally in the first few weeks after birth, and then degenerate along with the inner layers of the retina.

ii) **Later onset photoreceptor degeneration (progressive rod-cone degeneration):** Here the retina matures and functions apparently normally for varying periods of time before degenerating. Dogs are not usually clinically affected until 1 year of age or more, although abnormalities can be seen in the eye and on the electroretinogram (ERG) long before owners notice signs of visually impairment.

What does progressive retinal atrophy mean to your dog & you?

Generalized PRA - early onset: The first sign is generally failing night vision, as early as 6 weeks of age, and this progresses to complete loss of vision by about 1 - 2 years of age.

Generalized PRA (progressive rod-cone degeneration) - late onset: Generally night blindness is noticed between 2 and 5 years of age (depending on the breed) progressing to total blindness within a year or so. Peripheral vision is lost first.

How is progressive retinal atrophy diagnosed?

There are no obvious external changes to the eyes. You may notice that your dog has difficulty getting around when the lights are turned off, or when outside at night. If you suspect that your dog has impaired vision, your veterinarian will look for abnormalities with an ophthalmoscope. (editors note: currently in the lowchen, we are recommending that ALL lowchen be CERF tested by a certified canine ophthalmologist prior to breeding and no later than age two).

PRA may also be detected by electroretinogram (ERG) before your dog has any apparent visual difficulties. Electroretinography, which measures electrical patterns in the retina, is usually only available in specialty veterinary centres. (editors note: if the CERF exam shows ANY abnormalities or the pedigree indicates a known PRA dog or Carrier, an ERG exam should be done prior to the dog being introduced into a breeding program).

Genetic testing using individual blood samples is becoming available for different forms of PRA in different breeds. The advantage of such testing is that it can identify dogs whose sight is unaffected, but who are carriers of the disorder (heterozygotes).

How is progressive retinal atrophy treated?

There is no treatment for PRA. The degree of visual impairment varies with the breed and specific type of retinal degeneration as described above, but most affected dogs will ultimately be completely blind. With their acute senses of smell and hearing, dogs can compensate very well, particularly in familiar surroundings, to the point where owners

may be unaware of the extent of vision loss. You can help your dog by developing regular routes for exercise, maintaining consistent surroundings, introducing any necessary changes gradually, and being patient.

Breeding advice

Breeding is not advised for any dog with PRA, or for the parents (assumed to be carriers). Siblings should be carefully screened by electroretinogram if they are considered for breeding. Generalized PRA can often be detected by electroretinography at least a year before clinical signs are apparent.

Where to find more information?

American College of Veterinary Ophthalmologists. 1995. Ocular disorders presumed to be inherited in purebred dogs. This reference is helpful in differentiating disorders specific to different breeds.

Ackerman, L. 1999. The genetic connection. p. 162-167. AAHA Press. Lakewood, Colorado. This reference contains good information on inheritance, age of onset and form of PRA in different breeds.

CERF -Canine Eye Registration Foundation website: <http://www.vet.purdue.edu/%7Eyshen/cerf.html>

OptiGen - genetic testing: <http://www.optigen.com/>

VetGen - genetic testing: <http://www.vetgen.com/>

CERF Clinics and Vet Ophthalmologists in the United States

CERF stands for Canine Eye Registry Foundation. Although your regular veterinarian can check the eyes only a board certified ophthalmologist can conduct the necessary testing and issue the form which can be registered with CERF. Some breeders choose not to register with CERF however it is important to realize that dogs need an ANNUAL eye exam and the forms should be filed for reference should a problem develop.