

# WOODVALE PARK

## VETERINARY HOSPITAL



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# Retinal Degeneration

## Basics

### OVERVIEW

- “Retinal” refers to the retina; the retina is the innermost lining layer (located on the back surface) of the eyeball; it contains the light-sensitive rods and cones and other cells that convert images into signals and send messages to the brain, to allow for vision
- “Degeneration” is defined as a decline in function or structure
- “Retinal degeneration” is a decline in function or structure of the retina from any cause; the cause may be inherited or acquired (condition that develops sometime later in life/after birth)

### GENETICS

- Hereditary—inherited retinal degeneration is more frequent in dogs than in cats
- Inherited—a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time (known as “progressive retinal atrophy” or PRA); may be subdivided into abnormal development of the light-sensitive cells of the retina (known as “photoreceptor dysplasias”), which begin before the retina fully develops (at less than 12 weeks of age), and decline in function or structure of the light-sensitive cells of the retina (known as “photoreceptor degenerations”), which begin after the retina is fully developed and mature

#### Dogs

- Progressive retinal atrophy (a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—autosomal recessive in most breeds, such as collies, Irish setters, miniature poodles, cocker spaniels, briards, and Labrador retrievers; dominant in mastiffs, X-linked in Samoyeds and Siberian huskies
- Central progressive retinal atrophy (eye disease characterized by deterioration of the retina leading to loss of central vision, but retention of peripheral vision possibly for years)—autosomal dominant with incomplete penetrance in Labrador retrievers
- Inheritance in many breeds not determined
- Neuronal ceroid lipofuscinosis (a group of inherited, nervous system disorders with swelling and/or changes in the light-sensitive cells of the retina)—autosomal recessive (proven or presumed) in most breeds
- Inability to see clearly in bright light (known as “hemeralopia”)—autosomal recessive abnormal development of the light-sensitive cones in the retina (known as “cone dysplasia”) in Alaskan malamutes; undetermined inheritance in miniature poodles

#### Cats

- Abnormal development of the light-sensitive rods and cones in the retina (known as “rod–cone dysplasia”), Abyssinians have 2 forms—autosomal dominant: clinical signs at 4 months of age; autosomal recessive: may be blind by 2 years of age; also may have later onset of 2 years of age with vision problems by 4 years of age
- Isolated reports of both dominant and recessive inheritance in young Persians and domestic shorthairs

- Vision loss that becomes worse over time (known as “gyrate atrophy”)—autosomal recessive; caused by a buildup of a particular compound (ornithine) due to a lack of the enzyme that normally converts ornithine to glutamate (known as an “ornithine aminotransferase deficiency”)

## **SIGNALMENT/DESCRIPTION OF PET**

### **Species**

- Dogs
- Cats

## **SIGNALMENT/DESCRIPTION OF PET**

### **Species**

- Abnormal development of the skeleton (known as “skeletal dysplasia” or “dwarfism”) may be associated with Samoyeds and Labrador retrievers
- Retinal dysplasia also may be associated with multiple other eye abnormalities in Akitas and Doberman pinschers

### ***Hereditary—Dogs***

- Abnormal development of the retina (retinal dysplasia)—Bedlington terrier, Sealyham terrier, English springer spaniel, cocker spaniel
- Early onset progressive retinal atrophy (a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—Irish setter; collie; Norwegian elkhound; miniature schnauzer; Belgian shepherd, mastiff, Cardigan Welsh corgi, and briard
- Late-onset PRA (a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—miniature and toy poodle; American and English cocker spaniels; Labrador retriever; Tibetan terrier; miniature longhair dachshund; Akita; Samoyed; Siberian husky
- Central progressive retinal atrophy (eye disease characterized by deterioration of the retina leading to loss of central vision, but retention of peripheral vision possibly for years)—Labrador retriever; golden retriever; border collie; collie; Shetland sheepdog; briard
- Cone degeneration disease in which the function or structure of the cones has deteriorated (the retina contains the light-sensitive rods and cones and other cells that convert images into signals and send messages to the brain, to allow for vision)—German shorthaired pointers; Alaskan malamutes
- Neuronal ceroid lipofuscinosis (a group of inherited, nervous system disorders with swelling and/or changes in the light-sensitive cells of the retina)—English setter; border collie; American bulldog; Dalmatian; Tibetan terrier; collie
- Sudden blindness due to “sudden acquired retinal degeneration” or SARD—Brittany; miniature schnauzer; dachshund, any breed

### ***Hereditary—Cats***

- Abyssinian; Somali; Siamese; Persian

### **Mean Age and Range**

- Early progressive retinal atrophy (a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—3–4 months of age up to 2 years of age
- Late PRA (a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—clinical signs when the dog is greater than 4–6 years of age
- Cone degeneration disease in which the function or structure of the cones has deteriorated (the retina contains the light-sensitive rods and cones and other cells that convert images into signals and send messages to the brain, to allow for vision)—3–4 months
- Sudden blindness due to sudden acquired retinal degeneration—middle-aged to old dogs

### **Predominant Sex**

- Progressive retinal atrophy (a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—X-linked recessive condition in Siberian huskies and Samoyeds
- Sudden blindness due to sudden acquired retinal degeneration—70% are female

## **SIGNS/OBSERVED CHANGES IN THE PET**

- Progressive retinal atrophy (a group of eye diseases characterized by generalized deterioration of the retina,

- becoming increasingly worse over time) in dogs—gradually progressing night blindness (known as “nyctalopia”) that ultimately affects vision in bright light; may note dilated pupils or brighter tapetal reflex at night; may appear to be suddenly (acutely) blind (when pet becomes totally blind or is moved to unfamiliar surroundings)
- Inability to see clearly in bright light (hemeralopia) or cone degeneration disease—rare; light-sensitive cones degenerate; day vision lost
  - Central progressive retinal atrophy (eye disease characterized by deterioration of the retina leading to loss of central vision, but retention of peripheral vision possibly for years) in the dog—rare in the United States; central vision lost; may never become completely blind (especially hunting breeds)
  - Cone degeneration disease in which the function or structure of the cones has deteriorated (the retina contains the light-sensitive rods and cones and other cells that convert images into signals and send messages to the brain, to allow for vision)—puppies show avoidance of light (known as “photophobia”) and have trouble navigating their surroundings in bright light between 8 and 12 weeks of age; progresses to total day blindness; night vision remains normal
  - Sudden blindness due to sudden acquired retinal degeneration—vision lost in 1–4 weeks; increased urination (known as “polyuria”), increased thirst (known as “polydipsia”), and increased appetite (known as “polyphagia”) is common
  - If severe retinal degeneration—light reflexes of the pupil are impaired or nearly abolished; the “pupil” is the circular or elliptical opening in the center of the iris of the eye; light passes through the pupil to reach the back part of the eye (known as the “retina”); the iris is the colored or pigmented part of the eye; the pupil constricts or enlarges (dilates) based on the amount of light entering the eye; the pupil constricts with bright light and enlarges in dim light—these actions are the “light reflexes of the pupil”
  - Various changes in the appearance of the retina (light-sensitive lining of the back of the eye) may be noted when the veterinarian examines the back of the eye with an ophthalmoscope
  - Progressive retinal atrophy (a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time) in dogs—cataracts are common; a cataract is an opacity in the normally clear lens, if it is complete it prevents passage of light to the back part of the eye (retina)
  - Sudden blindness due to sudden acquired retinal degeneration in dogs—obesity; may note slow or absent light reflexes of the pupil; the pupil constricts or enlarges (dilates) based on the amount of light entering the eye; the pupil constricts with bright light and enlarges in dim light—these actions are the “light reflexes of the pupil”

## CAUSES

### Degenerative

- Progressive retinal atrophy (a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—affects both eyes symmetrically; most forms of PRA are inherited as recessive traits, except for PRA in English mastiffs and bullmastiffs, which is a dominant trait
- Long-term (chronic) or uncontrolled glaucoma (disease of the eye, in which the pressure within the eye is increased)—decrease in tissue of the retina (known as “retinal atrophy”; the retina is the light-sensitive lining of the back of the eye) and optic nerve (the nerve that runs from the back of the eye to the brain)
- Secondary to scarring from separation of the back part of the eye (retina) from the underlying, vascular part of the eyeball (known as the “choroid”; condition known as “retinal detachment”) or inflammation of the retina

### Anomalous (Abnormal Structure)

- Abnormal development of the light-sensitive rods and cones of the retina (known as “rod-cone photoreceptor dysplasias”)—inherited disease; affects both eyes
- Other types of abnormal development (dysplasias)—may be located in multiple areas of the retina (so-called “multifocal”) and non-blinding (for example, in English springer spaniels and Labrador retrievers)

### Metabolic

- Mucopolysaccharidosis (disease related to a lack of or insufficient amount of a particular enzyme)—mixed-breed dogs; Siamese and domestic shorthairs (cats)
- Ornithine aminotransferase deficiency—a mitochondrial enzyme; progressive and total gyrate atrophy of the choroid and retina due to a buildup of a particular compound (ornithine) due to a lack of the enzyme that normally converts ornithine to glutamate

### Cancer

- Cancer cells infiltrating the retina
- Scars from previous retinal detachment (separation of the back part of the eye (retina) from the underlying, vascular part of the eyeball), if treated

### **Nutritional**

- Severe deficiency of vitamin E or A (dogs and cats)—experimentally or dogs fed poor diets (high in polyunsaturated fats) may cause partial or complete degeneration of the retina
- Taurine deficiency (cats)—causes retinal degeneration and a heart-muscle disorder (known as “dilated cardiomyopathy”); taurine is an amino acid (protein) that is an important component of the diet of cats; cats cannot produce enough taurine in their bodies and so, must obtain taurine from their food to maintain the health of several organs, including the retina

### **Infectious/Immune**

- Infectious inflammation of the retina (known as “retinitis”) or inflammation of the choroid and retina (known as “chorioretinitis”); the “choroid” is located immediately under the retina and is part of the middle-layer of the eyeball that contains the blood vessels
- Infection may extend from or to the central nervous system (brain)

### **Chorioretinitis Idiopathic (Unknown Cause)**

- Sudden blindness due to sudden acquired retinal degeneration—dogs

### **Chorioretinitis Toxic**

- Individual pet is more likely to develop ill effects to a particular medication than other animals (known as “idiosyncratic reactions”)—griseofulvin or enrofloxacin (cats)
- Radiation treatment for cancer of the nose or central nervous system (dogs and cats)
- Concurrent administration of ketamine hydrochloride and methylnitrosourea induces widespread (diffuse) retinal degeneration (cats)

## **RISK FACTORS**

- Eye disease—cataracts; inflammation of the back part of the eye; inflammation of the choroid and retina (chorioretinitis); retinal detachment; glaucoma
- Taurine-deficient diet—dog food fed to cats (most cat foods now contain proper taurine levels)
- Genetics
- Cats—enrofloxacin (an antibiotic)

## **Treatment**

### **DIET**

- Cats—food should contain 500–750 ppm of taurine; taurine is an amino acid (protein) that is an important component of the diet of cats; cats cannot produce enough taurine in their bodies and so, must obtain taurine from their food to maintain the health of several organs, including the retina
- Dogs—balanced diet; avoid all meat diet, high in polyunsaturated fats

### **SURGERY**

- Not indicated in pets with blind, non-painful eyes

## **Medications**

Medications presented in this section are intended to provide general information about possible treatment. The treatment for a particular condition may evolve as medical advances are made; therefore, the medications should not be considered as all inclusive

- No medications currently are effective in treating retinal degeneration
- Pyridoxine supplementation (cats)—for ornithine aminotransferase deficiency; may increase activity of the enzyme; has not arrested or reversed degeneration of the retina
- Adequate dietary taurine—may halt the progression of the retinal deterioration due to inadequate levels of taurine (taurine-deficient retinopathy); taurine is an amino acid (protein) that is an important component of the diet of cats; cats cannot produce enough taurine in their bodies and so, must obtain taurine from their food to maintain the health of several organs, including the retina

# Follow-Up Care

## PATIENT MONITORING

- Repeated eye examinations, looking at the retina (light-sensitive lining of the back of the eye)— confirm progressive degeneration, if the diagnosis is in doubt; will note obvious signs of degeneration over weeks in the retinas of dogs with sudden blindness due to sudden acquired retinal degeneration
- Developing and progressing cataracts (opacities in the normally clear lens)—with progressive retinal atrophy; watch for painful complications (such as glaucoma [disease of the eye, in which the pressure within the eye is increased] and inflammation of the iris and other areas in the front part of the eye [known as “uveitis”])

## PREVENTIONS AND AVOIDANCE

- Do not breed pets suspected of having progressive retinal atrophy (a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)
- Do not breed known carriers (that is, offspring of an affected pet)

## POSSIBLE COMPLICATIONS

- Cataracts (opacities in the normally clear lens)
- Glaucoma (disease of the eye, in which the pressure within the eye is increased)
- Uveitis (inflammation of the iris and other areas in the front part of the eye)
- Eye trauma as a result of visual impairment
- Obesity—secondary to reduced activity

## EXPECTED COURSE AND PROGNOSIS

- Inherited progressive retinal atrophy (a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—progresses to complete blindness; progression often slow enough for the pet to adapt to visual loss; non-painful
- Degeneration from previous inflammation or trauma—usually does not progress, unless a generalized (systemic) disease causes persistent or recurrent eye inflammation
- Sudden blindness due to sudden acquired retinal degeneration—irreversible blindness
- Transient taurine deficiency (cats)—degeneration may halt at any stage

## Key Points

- Most blind pets function well
- Blind dogs can memorize their environment and function well unless the family moves or rearranges the furniture
- Blind dogs should be watched or kept on a leash, if they are outside, not in fenced yards, or in an area with a pool
- Suggest playing with toys that make sounds
- Some old blind pets with other problems (such as hearing loss or senility) may not adapt well to blindness
- Some blind pets experience behavioral changes (such as increased aggression or reduced activity)
- Pets with only one blind eye can function normally
- Blind cats should be kept indoors

# Notes

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