Lake Shore Pet Hospital

Retinal Degeneration

Basics

OVERVIEW

- "Retinal" refers to the retina; the retina is the 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 abnormal/incomplete development or 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 —autosomal recessive in most breeds, such as collie, Basenji, Irish setter, miniature poodle, cocker spaniel, Briard, and Labrador retriever, others
- Dominant in mastiff
- X-linked in Samoyed and Siberian husky
- 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 cone degeneration in the retina (known as "cone dysplasia")

Cats

Abnormal development of the light-sensitive rods and cones in the retina (known as "rod–cone dysplasia"), Abyssinians and mixed breed—autosomal dominant; Persians (autosomal recessive): clinical signs at 4 months of age: may be blind by 2 years of age; Abyssinians may have later onset, blind by 4 years of age

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, and others
- 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, American Staffordshire , pit bull terrier 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; Basenji, 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, and others
- 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; Australian shepherds
- 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 spaniel; miniature schnauzer; dachshund, any breed

Hereditary—Cats

- Abyssinian; Somali; Siamese; Persian as autosomal recessive

Mean Age and Range

- Early progressive retinal atrophy and dystrophies—3–4 months of age up to 2 years of age
- Late PRA —clinical signs when the dog is greater than 4–6 years of age
- Cone degeneration disease—3–4 months
- Sudden blindness due to sudden acquired retinal degeneration—middle-aged to old dogs

Predominant Sex

- Progressive retinal atrophy—X-linked recessive condition in Siberian huskies and Samoyeds, so more in males
- 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; may appear to be suddenly (acutely) blind; 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)
- Dysplasias will have early onset and dogs may be blind by 2 years of age
- Inability to see clearly in bright light (hemeralopia) or cone degeneration disease—rare; light-sensitive cones degenerate between 8 and 12 weeks of age puppies have trouble navigating in bright light though night vision remains normal; 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)
- 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
- Sudden blindness due to sudden acquired retinal degeneration (SARDS) 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
- Borzoi chorioretinopathy—multifocal retina lesions, hyperreflective and hyperpigmented
- Taurine-deficient retina disease in cats—begins in one spot in the middle of the retina then finally, diffuse degeneration and hyperreflectivity
- Retina dysplasia may be associated with multiple eye problems in Akita, Doberman pinscher or other breeds
- Storage diseases—may have cloudy eye surfaces (cornea), possible nervous system signs (example is neuronal ceroid lipofuscinosis)

CAUSES

Degenerative

- Progressive retinal atrophy — affects both eyes symmetrically; most forms of PRA are inherited as recessive traits, except for PRA in mastiffs, 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") and pressure on the optic nerve at the back of the eye
- Secondary to scarring from separation of the 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)
- Oculoskeletal abnormality in Labrador retriever, Samoyed

Metabolic

- Mucopolysaccharidosis (disease related to a lack of or insufficient amount of a particular enzyme) and other storage diseases, in English springer spaniel and others
- Ornithine aminotransferase deficiency—a mitochondrial enzyme; progressive and total 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, seen in older cats

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; multifocal, focal or generalized
- Infection may extend from or to the central nervous system (brain)

Chorioretinitis Idiopathic (Unknown Cause)
- Sudden blindness (SARDS) due to sudden acquired retinal degeneration post-inflammatory—dogs and cats

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)
- Phototoxicity (for example, exposure to welding light)

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; cats 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)

Follow-Up Care
PATIENT MONITORING
- Repeated eye examinations, looking at the retina (light-sensitive lining of the back of the eye)— confirm progressive retinal 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 (SARDS), with PRA changes evident over months
- 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"])—cataract formation with PRA or SARDS

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)