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, especially 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 studied
- 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 build-up 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 ANIMAL

Species

- Dogs and cats

Breed Predilections

- Abnormal development of the retina (retinal dysplasia) 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 (many breeds)

- Abnormal development of the retina (retinal dysplasia)—Bedlington terrier, Sealyham terrier, English springer spaniel, cocker spaniel
- Early-onset progressive retinal atrophy (PRA; 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 (congenital [present at birth] stationary night blindness)
- Late-onset progressive retinal atrophy (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
- 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; Dalmatian; Tibetan terrier; collie

• Sudden blindness due to "sudden acquired retinal degeneration" or "SARD"—Brittany; miniature schnauzer; dachshund, any breed

Hereditary—Cats

• Abyssinian, Siamese, Persian, domestic shorthair

Mean Age and Range

• Early progressive retinal atrophy (PRA; a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—3 to 4 months of age up to 2 years of age

• Late progressive retinal atrophy (PRA; a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—clinical signs when the animal is greater than 4 to 6 years of age

• Sudden blindness due to sudden acquired retinal degeneration or SARD—middle-aged to old dogs

Predominant Sex

• Progressive retinal atrophy (PRA; a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—none, except possibly X-linked recessive condition in Siberian huskies

• Sudden blindness due to sudden acquired retinal degeneration or SARD—70% are female

SIGNS/OBSERVED CHANGES in the ANIMAL

• Progressive retinal atrophy (PRA; 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 patient finally becomes totally blind or is moved to unfamiliar surroundings)

• Inability to see clearly in bright light (hemeralopia)—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; may have difficulty locating stationary objects in bright light (especially hunting breeds)

• Sudden blindness due to sudden acquired retinal degeneration or SARD—vision lost in 1 to 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; **t**he 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 (PRA; 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 or SARD in dogs—obesity; enlargement of the liver (known as "hepatomegaly"); may note slow or absent light reflexes of the pupil; **t**he 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 (PRA; a group of eye diseases characterized by generalized deterioration of the retina, becoming increasingly worse over time)—genetic; affects both eyes symmetrically

• 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 build-up 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)

Idiopathic (Unknown Cause)

• Sudden blindness due to sudden acquired retinal degeneration or SARD—dogs

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)
• 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 to 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 patients with blind, nonpainful 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)—at 3 to 6-month intervals; 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 or SARD
• Developing and progressing cataracts (opacities in the normally clear lens)—with progressive retinal atrophy (PRA); 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 animals suspected of having inherited progressive retinal atrophy (PRA; 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 animal)

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 (PRA; 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 patient to adapt to visual loss; nonpainful
- Degeneration from previous inflammation or trauma—usually does not progress, unless a generalized (systemic) disease (such as toxoplasmosis) causes persistent or recurrent eye inflammation
- Sudden blindness due to sudden acquired retinal degeneration or SARD—irreversible blindness
- Transient taurine deficiency (cats)—degeneration may halt at any stage

KEY POINTS

- Patient visually impaired—condition is irreversible, but nonpainful
- 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
- Dogs can memorize their environment and unless the family moves or rearranges the furniture, most blind animals function well
- Apply perfume to legs of furniture to help the patient memorize the environment and identify the location of objects
- Some old blind animals with other problems (such as hearing loss or senility) may not adapt well to blindness
- Some blind animals experience behavioral changes (such as increased aggression or reduced activity)
- Animals with only one blind eye can function normally
- Blind cats may adapt better than dogs, but probably should be kept indoors