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MORE THAN MEETS THE EYE

Inherited ocular diseases

THE EYES OF A DOG are located within a protective bony socket called the orbit. In addition to the orbital bones, the eye is protected by upper and lower eyelids.

The exposed portion of the eye is covered with tears, which are made up of three main layers: a fatty layer, a water layer and a mucous layer. The eyelids protect the eye and distribute the tears across its surface to keep it moist.

The eye itself is made up of three main layers: the outer layer, called the fibrous coat; the blood-vessel-rich layer, called the uvea; and the inner nervous-tissue layer at the back of the eye, called the retina. The retina has special receptors called rods and cones, which provide dim light, and bright light and colour vision, respectively. Contrary to popular belief, dogs have the ability to see not only shades of grey, but also colours, namely blues and yellows.

In addition to these three layers, the inside of the eye contains transparent fluid (aqueous humor) in the front, and a transparent gel (vitreous) at the back, both of which help provide nourishment to the eye's various structures. A lens sitting behind the iris and centred in the pupil helps focus images on the retina at the back of the eye. The eye's function is to transmit images from the retina through the optic nerve to the brain for vision.

Eyelid disorders

Entropion is an inward turning of the eyelid margin. This condition is presumed to be inherited when it occurs at a young age (usually within the first two years) and as a dog grows.

Depending on the breed, entropion may affect various portions of the eyelid(s). For example, large- and giant-breed dogs are predisposed to entropion affecting the outer aspect of the lower eyelid, so-called lateral or lateral ventral entropion. Small breeds have a predisposition for entropion of the inner corner of the eyelids, so-called medial or medial ventral entropion.

Depending on its severity, entropion may cause signs ranging from mild tearing to severe eye discomfort with squinting, copious tearing and damage to the surface of the eye including an ulcer, brown pigment and/or blood-vessel growth in the normally transparent cornea. Some affected dogs outgrow their entropion as their facial bones and eyelids develop. However, permanent entropion associated with signs of eye irritation will require some form of surgical correction to prevent continued discomfort and damage.

Ectropion is an outward turning of the eyelid margin that results in varying degrees of exposure of the conjunctiva (pink tissue lining the eyelids), and an inability of the eyelids to completely close during blinking. Ectropion is characteristic of breeds such as the Bloodhound, Bullmastiff, Newfoundland and many spaniel breeds. Most cases of ectropion don't cause significant eye irritation and, hence, do not require surgery. Some dogs with ectropion experience constant or intermittent eye discharge, conjunctivitis or disease of the cornea and therefore benefit from surgical correction of the ectropion.

Distichiasis. Distichia are hairs on the eyelid margin that come from an abnormality of the meibomian (tarsal) glands. These hairs are, in a sense, misplaced eyelashes and most do not cause any problems. Distichiasis is the condition caused by distichia that generates signs of eye irritation, including tearing, squinting, conjunctivitis, and inflammation/ulcers of the cornea. It is treated by microsurgical removal of the abnormal gland and hair follicle, electrocautery, electrolysis or freezing/cryotherapy.

Ectopic cilia are hairs arising from the meibomian gland and growing through the pink tissue lining the inside

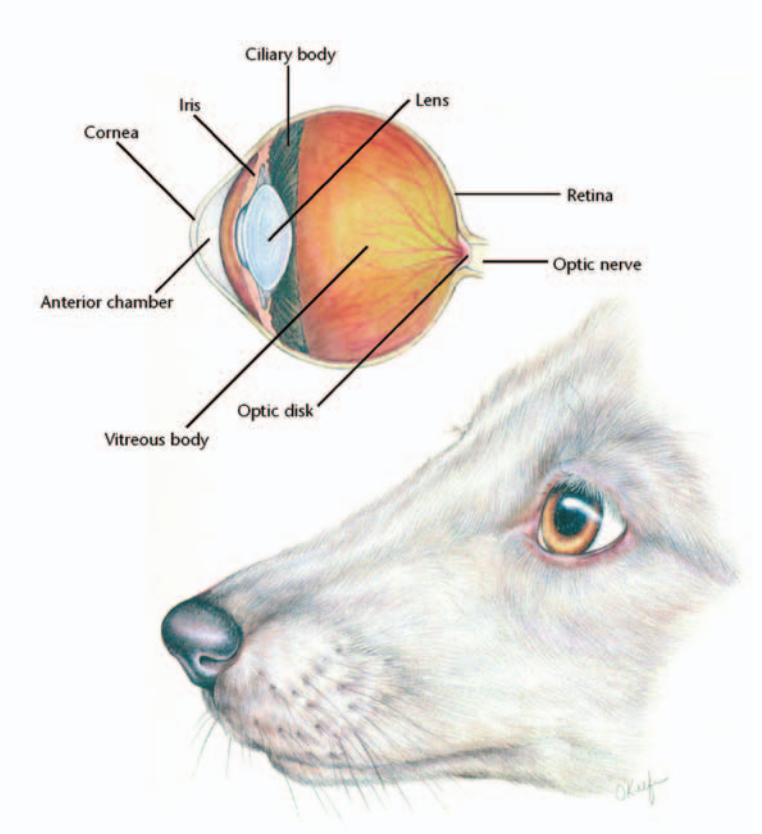

ILLUSTRATION: COURTESY *HILL'S ATLAS OF VETERINARY CLINICAL ANATOMY*

of the eyelid, causing eye irritation. Small breeds appear predisposed to this condition. These abnormal small hairs rub on the surface of the eye, causing squinting, excessive tearing, conjunctivitis, and ulcers of the cornea. Because ectopic cilia are small, diagnosis is difficult without the aid of magnification. The treatment of choice is surgery to remove the ectopic cilia. They are often multiple. Younger affected dogs have an increased likelihood of developing additional ectopic cilia.

Trichiasis is a condition in which hairs located in normal sites around the eye (e.g., skin folds around the nose, eyelids) are directed at and rubbing against the surface of the eye, resulting in tearing and inflammation of the cornea. This is a common condition in small-breed dogs. Mild degrees of trichiasis not causing disease of the cornea do not require therapy. In certain cases, simply keeping the offending hairs clipped short, so they are no longer in contact with the eye, is adequate. If the trichiasis is severe, surgical correction, although variably successful, is required.

Trichomegaly is an abnormally long eyelash. This condition is typically an incidental finding in a variety of breeds, the genetics of which have not been determined. Trichomegaly is most commonly seen in the American Cocker Spaniel, and Toy and Miniature Poodles.

Corneal disorders

Corneal dystrophy is an inherited or breed-related whitish deposit of cholesterol or fat that occurs at varying depths within the cornea. Corneal dystrophy affects both eyes and appears symmetrical between the eyes, typically in a central to off-central location.

When the epithelium (outer layer of the cornea) is involved, recurrent eye irritation from erosions/ulcers of the cornea is common. *Corneal epithelial dystrophy* is reported most commonly in the Shetland Sheepdog, Basset Hound, Dachshund and Border Collie.

Corneal stromal dystrophy, affecting the mid region of the cornea, may be seen in any breed of dog, and usually develops between two and four years of age. There is typically no treatment required for this form of dystrophy as the deposits tend to be small and progress minimally. However, when the dystrophy involves the endothelium (the cornea's innermost layer), severe progressive cloudiness or bluish haziness to the surface of the eye is common. *Corneal endothelial dystrophy* shows up later in life (eight to 12 years) and is diagnosed in several breeds. Treatment includes eye drops or ointment. Corneal transplant remains the treatment of choice for maintaining vision.

Pigmentary keratitis is a disease of small-breed dogs in which brown pigment starts to cover the cornea. Blood vessels and scarring often accompany the pigment, and all three opacities interfere with vision. Several factors contribute to this syndrome and are related to the eyelid structure of dogs with "pushed in" noses (i.e., brachycephalic breeds), such as Lhasa Apsos, Pekingese and Pugs. These breeds have shallow bony sockets, resulting in varying degrees of eye prominence and inability to completely close the eyelids with blinking; hairs at the inner corners of the eyes; and medial ventral entropion. Exposure of the eyes, and hairs rubbing on the eyes, result in irritation to the surface of the eyes, inducing pigmentation, blood-vessel growth and scarring on the cornea. Surgery to reconstruct the eyelids is recommended, especially if the pigmentary keratitis is progressing.

Chronic superficial keratitis (pannus) is a progressive inflammatory and potentially blinding disease of the cornea that always affects both eyes. This condition develops most commonly in German Shepherd Dogs and German Shepherd crosses, but also occurs in the Belgian Shepherd Dog, Border Collie, Bouvier des Flandres, Greyhound and large crossbred dogs, among others. Affected dogs develop a progressive cloudiness or "growth" of brown pigment and/or blood vessels starting at the outer edge of the cornea and advancing toward the centre. Without treatment, the entire cornea will become affected.

The exact cause of this inflammatory condition is unknown, although it is assumed to be a result of a defective immune reaction. Other factors have been associated with the development of pannus, including a genetic component for predisposed breeds, and dogs living at high altitudes (above 4,000 feet – related to ultraviolet light exposure). Treatment involves the use of medications – eye drops or ointment – to help suppress the immune reaction. Complete remission of pannus signs can result following prolonged therapy; however, lifelong treatment is required.

Disorders of the blood-vessel-rich layer (uvea)

Persistent pupillary membranes (PPMs). Pupillary membranes are tiny remnants of blood vessels that are normally present in puppies before birth and typically disappear between three and five weeks of age. These remnant blood vessels appear as brown-pigmented strands of uveal tissue arising from the iris. If they fail to disappear, these tissues are called "persistent."

There are four main forms of PPMs based on where

CERF examinations

The Canine Eye Registration Foundation (CERF) was founded by breeders in conjunction with board-certified veterinary ophthalmologists to assist with the elimination of inherited eye disease in purebred dogs by forming a centralized North American registry.

The CERF registry keeps data on dogs examined by veterinary ophthalmologists who are members of the American College of Veterinary Ophthalmologists (i.e., ACVO Diplomates). Both inherited and non-inherited eye diseases are recorded and reported by the registry.

A CERF examination involves dilation of the dog's pupils using eye drops to permit complete examination of the eyes from front to back. These drops usually wear off within four to six hours. After the pupils are fully dilated (about 20 minutes), the ACVO Diplomate will complete a CERF form and indicate any specific eye disease(s) found. Breeding advice will be offered based on guidelines established for that particular breed.

Generally, the recommendation for dogs with eye diseases that have a tendency toward significant consequences to the eye (e.g., large cataracts and/or those located in a typical inherited position within the lens, and progressive retinal atrophy/degeneration) is "Do not breed." Breeding advice for other, less-threatening conditions deemed to be heritable (e.g., distichia, and iris-to-iris persistent pupillary membranes) is "breeder option."

If the dog is certified to be free of inherited eye disease, the breeder or owner can submit a completed owner's copy of the CERF form with the appropriate fee to CERF. The certification is good for one year from the date of the examination. Because many forms of inherited eye disease develop over time, a dog must be re-examined and re-certified to maintain registration with CERF.

Regardless of the outcome of the dog's examination, the research copy of the form will be sent to the CERF office where its information will be entered into the database for that breed. This information will be used in generating research reports, but the individual dog's identification becomes confidential and will never be released. In particular, this data is used to form the CERF database, which helps determine trends in eye disease for veterinarians, interested breed clubs, and individual breeders and owners of purebred dogs.

they extend: 1) iris-to-iris (the most common form); 2) iris-to-lens; 3) iris-to-cornea; and 4) iris sheets. PPMs are non-progressive and do not require treatment. Inheritance of PPMs has been documented or suspected in several breeds.

Iris coloboma is a notch-like defect or "hole" in the iris that can occur in any region but usually develops at the 6 o'clock position. Colobomas may occur alone without other eye abnormalities, but more commonly they are associated with additional eye defects, as noted in colour-dilute breeds such as the merle Australian Shepherd. There is no treatment for this condition; however, selective breeding can prevent it.

Uveal cysts are variably brown/pigmented, round, fluid-filled structures that arise from the blood-vessel-rich layer of the eye, namely the iris and ciliary body. Uveal cysts may occur in one eye as a single cyst or multiple cysts, or they may be present in both eyes. The Boston Terrier, Great Dane, Labrador and Golden Retrievers, and Norwegian Elkhound are predisposed to this disease.

Uveal cysts may also develop due to trauma or long-standing inflammation inside the eye but because they can mimic melanomas or other tumours inside the eye, a complete eye examination is recommended. Treatment for uveal cysts is typically not required. In rare instances, the cysts (mainly those arising from the ciliary body) may be associated with glaucoma. In addition, dogs with uveal cysts that interfere with vision may require referral to a veterinary ophthalmologist for laser ablation or deflation using a needle.

Lens disorders

Cataracts, a whitish opacity or cloudiness of the lens, may range from non-progressive, tiny and non-vision-threatening to rapidly progressive, complete and blinding. Genetics is the most common cause of cataracts in dogs. Inherited cataracts have been reported in several breeds. Dogs affected with inherited cataracts and carriers of the genetic defect should not be bred. Treatment is indicated if the cataracts are large and interfere with vision and/or cause cataract-related inflammation inside the eye(s). Medical therapy can help control the inflammation. Surgery to remove the cataracts is required to restore vision.

Lens luxation is displacement of the lens caused by disruption of the fibres (zonules) that hold it in place. An inherited disorder of the zonules affecting both eyes (one eye may be affected first), so-called primary lens luxation, has been reported in dogs. This condition occurs due to an inherited deterioration/degeneration of the zonules.

Breeds predisposed to this condition include the terrier breeds, American Cocker Spaniel, Beagle, Border Collie, Chihuahua, Chinese Shar Pei, Miniature and Toy Poodles, Miniature Schnauzer and Pembroke Welsh Corgi. Clinical signs include red eye, cloudy eye and vision impairment, often as a result of the development of glaucoma with progressive lens displacement.

Lens luxations are typically considered an emergency, requiring early diagnosis and medical treatment initially, and prompt referral to a veterinary ophthalmologist if possible.

It is important to determine the cause of the lens luxation. In addition to inherited primary lens luxation, there are secondary causes of lens displacement, such as trauma, inflammation or tumour inside the eye, and glaucoma, all of which should be distinguished from primary lens luxation to help determine the prognosis and whether or not the affected dog should be used for breeding. If a diagnosis of lens luxation is made early in the course of the disease and the eye maintains vision, referral to a veterinary ophthalmologist for surgery to remove the lens is recommended.

Retinal disorder

Progressive retinal atrophy/degeneration (PRA/PRD) is an inherited slow deterioration/degeneration of the retina. PRA/PRD has been described in numerous breeds. Depending on the affected breed, PRA/PRD may be early-onset (six to 10 weeks of age) or late-onset (as late as seven years of age).

As the name of the condition implies, PRA/PRD is progressive and leads to blindness over months to years, depending on the affected breed. PRA/PRD affects both eyes equally, and results in loss of night vision followed by loss of day vision, hence blindness.

Most of these retinal degenerations are inherited as an autosomal recessive condition, meaning that affected dogs carry both mutated genes and the clinical signs of the disease, while other dogs in the affected litter may be carriers of one mutated gene with no clinical signs of disease. The diagnosis of PRA/PRD can be confirmed by an eye examination (see CERF sidebar) and/or electroretinography (electrodiagnostic test used to assess retinal function). As well, DNA testing using blood from dogs in question has now become available in some breeds.

For further details on DNA tests available for this and other eye conditions, refer to the Optigen web site: *www.optigen.com/opt9_test.html*. ●

American College of Veterinary Ophthalmologists (ACVO) Diplomates capable of offering Canine Eye Registration Foundation (CERF) examinations in Canada

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