

## Topic 9: Juvenile Cataracts in West Highland White Terriers

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### Introduction to this Topic

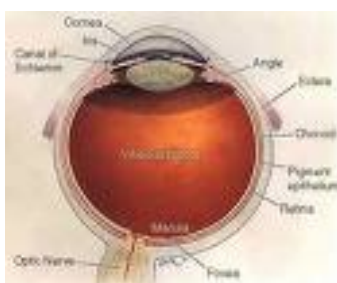
To be able to prosper in their environment, dogs need healthy eyes. Evolution has provided dogs with a very keen sense of vision and the ability to see in extreme conditions of light and darkness and to be highly perceptive of movement. Although we generally don't think of them as primal predators, every Westie is born with this in their blood, and predators need sharp vision.

At times, injuries to the eye and the surrounding lids and gland, or disease, will disrupt vision. Unfortunately, highly complex structures such as the eye, may also suffer from defects in ocular development, such as the formation of juvenile cataracts. Simply defined, a **cataract** is an irregularity and opacity in the lens.

It may be helpful to readers to give a few short definitions that will help when reading material included in this Topic. Most cataracts are seen in older dogs. If the cataract forms as a result of disease (like infections or diabetes mellitus), exposure to some chemicals and intense irradiation (cataracts are an unwanted side effect of radiation therapy of the head and neck), or injury, the cataract is considered to be "acquired" as a result of the injurious process. A **juvenile cataract** is one that has formed before birth (congenital juvenile cataract) or which develops shortly after birth, as the eyes of the dog mature (a developmental juvenile cataract). Juvenile cataracts may be caused by the expression of defective genes and/or problems affecting the dam during gestation. In some breeds of dogs, the incidence of cataracts increases with age and is considered hereditary (inherited as a genetic disease from one or both parents). The outcome of all is the same – a decrease in visual acuity for the dog.

### The anatomy of the eye and the phenomenon of vision

The parts that make up the eye are shown in the figure appearing below (Source: The School of Biomedical Engineering at Georgia Tech). The eye is essentially made up of two connected parts. The smaller part, called the anterior chamber, is bounded by the transparent cornea at the front of the eye and the lens. The larger part of the eye, the posterior chamber is formed externally by the tough white part called the sclera, and contains the lens, the gelatinous vitreous humor, and the neural receptor membrane of vision, the retina. The eye is composed of several layers of cells, blood vessels (in the uveal tract), and connective tissue.



The lens is a living tissue. It is a tight clustering of specialized cells, enclosed in a capsule, located behind the iris and in front of the vitreous body, and is held in place by fibers, the anterior vitreous face, and the iris

(Magrane, 1972). The lens is normally quite flexible. Tension and relaxation on the edges of the lens, controlled by the small muscles and fibers, alters lens shape. By changing its shape, the lens can alter its refractive power. As a living tissue, the lens grows with advancing age, needs nutrition (acquired by diffusion through the aqueous and vitreous humors, can be damaged by injury and disease, and can show limited healing. Lens shape varies throughout life.

Let's discuss a few important points about the lens, since cataracts form within the lens. The capsule, surface layer or epithelium, and fibers are the three main parts components of the lens. The capsule is a thickened smooth membrane made of collagen and produced by the lens epithelium and fibers. It completely surrounds the lens and possesses elastic properties, so when not under tension, the lens assumes a rounded shape. The epithelium is comprised of cells that elongate over time and are eventually transformed into lens fibers, which contain high concentrations of the protein crystallin (which help the lens to refract and transmit light). These fibers are tightly packed and extend the full length of the lens. Continual growth of the lens adds more elongated cells and fibers and produces an arrangement similar to the layers of an onion (Magrane, 1972). Damage to any of the components of the lens can result in formation of a cataract.

One final point – the formation of the lens helps orchestrate the overall development of the eye. The lens forms relatively early during the development of the eye and helps induce the formation of both chambers and many of the components of the eye. This pivotal role of the lens in controlling development of the eye is important for several reasons. First, if the lens does not properly form, this can affect the development of other components of the eye. Second, disease or defective gene expression that occurs during pregnancy can significantly affect lens formation and, by extension, the development of healthy eyes. Third, the presence of cataracts at the time of birth clearly indicates problems with lens development, but also may signal the potential for problems elsewhere in the eye. Finally, it is important to remember that puppies are born with incompletely matured eyes and some of the process of development takes place after birth. A good rule of thumb is that formation of the eye is complete by about 12 weeks of age, in most breeds of dog. Dogs should have good visual acuity by this age.

### **Normal vision – summarized!**

Light energy from the surroundings produces electrochemical changes in specialized nerve cells (the rods and cones) in the retina. These changes result in the generation of signals (called 'nerve action potentials') that are relayed to the brain, where they are processed and consciously appreciated as a vision (Magrane, 1972). The lens is a key part of the system that focuses and transmits light to the retina so that signals that eventually produce vision are received on the retina.

## Examination of eyes – you and your veterinarian

Owners and breeders are often the first to detect a problem with the eyes and vision. Some common signs that something is not right include:

- The eyes, lids, and membranes of the eye just don't look right; there may be discoloration, irregularities in shape and size, or perhaps the eyes are inappropriately proportioned to the size of the head. Shown at the right is the eye of a dog in which there is a cataract, causing the lens to appear milky white/opaque, and in which the pupil is dilated.
- Puppies may bump into things in their path (this has to be differentiated from just clumsiness or poor coordination),
- Puppies appear reluctant to move about or are overly shy (most Westie puppies are pretty affable and playful),
- Puppies are reluctant to go in to darkened areas during exploration,
- Puppies appear to cue interactions based on hearing rather than on both hearing and seeing (that is, they may search for sounds instead of searching visually).



If a problem with vision is suspected, you should take your Westie to your veterinarian.

All thorough physical examinations of dogs include an evaluation of the eyes. Most evaluations by veterinarians have common elements, and some of the routine evaluation is done with simple tests:

- Evaluation of the gross appearance of both eyes, comparison of one eye with the other and with the head in terms of size, shape, coloration, tonicity (see below), and integration with facial shape,
- Detection of visual signal processing: the veterinarian will shine a light in one eye and then the other to see if there is constriction of the pupils in response to light. This is actually a simple test of a complex process, since it tests whether light is focused on the retina, which then creates a visual 'signal' that is then transmitted on nerve fibers to the brain. There the signal is interpreted as vision, at the same time that automatic mechanisms in the central nervous system regulate the size of the pupils, (Figure, right, [not a Westie!])
- The ability to track movement in a lighted area is assessed by the response of the dog to hand movements near the eyes. In many examinations, the veterinarian will see if the dog will blink in response to movement near the eyes – assessing both visual perception and the automatic blink response,
- Tonicity (firmness) of the eyes can be first evaluated by gently applied pressure to the eyes through the lids. Most dogs do not mind this part of



- the examination and it helps determine if the eyes are firm, but not too firm, and if there are irregularities or pain,
- Ophthalmic evaluation of the anterior chamber, posterior chamber, and intraocular structures (lens, iris, pupil, retina).

Many eye problems can be detected with these simple tests. In some cases, owners and veterinarians may wish to seek out a specialist (a veterinary ophthalmologist) for further testing. Specialists commonly see patients by referral and have specialized equipment and facilities for examination and treatment of eye diseases. Many specialists are certified by examination boards after years of advanced training on treating diseases of the eye. Shown at the right, Dr.



Phillip Pickett, a board-certified veterinary ophthalmologist, and his veterinary ophthalmology technician, Ms. Betsy Midkiff, perform an examination of a Golden Retriever service dog, using an external lens and light source, which allows the retina to be more closely examined. In the next figure, shown below, left, a hand-held ophthalmoscopic evaluation is conducted. The device being used is a more sophisticated version of the ophthalmoscope used by most veterinarians and has increased magnification to allow detection of smaller defects.

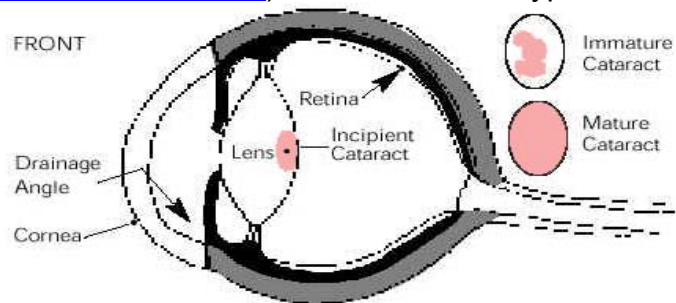


The value to Westie owners in seeking regular evaluations of their dogs should be evident. Health problems can be detected, diagnosed and many (most) are treated effectively. When more complex treatments, such as surgery, are needed to treat problems and to correct defects, it makes sense to seek the services of a specialist. These specialists may have facilities and tools for treatment and, since they concentrate exclusively on treating diseases of the eye, they are generally more practiced (have seen more cases and more difficult cases), and may have access to the latest research findings and tools.

## **Cataracts - An Overview**

A cataract is an opacity occurring in the lens, usually appearing as a cloudy white discoloration, and is a site of injury and cell death within the lens. Unfortunately, cataracts are one of a number of diseases that Westie owners have to consider when breeding and when raising Westies. Cataracts can affect only one or both eyes. Cataracts are among the most common intraocular lesions and are a

leading cause of vision loss in dogs. They can be classified according to several criteria: age at presentation, localization, etiology (cause), appearance, and stage (Adkins, et al., 2005). In the diagram at the lower right (Source: <http://www.animaleyecare.net/diseases/cataract.htm>) several different types of cataracts are diagrammed. In the eye shown, a small, focal, incipient (developing) cataract is shown. Unfortunately, the position of this cataract, in the center of the lens, will interfere with the direct path of light energy to the retina at the back of the eye, and will interfere with visual acuity.



Also in this figure are simplified diagrams of immature and mature cataracts. These terms, 'immature' and 'mature', refer to the developmental stage **of the cataract**. Immature cataracts are newly formed and may occupy only a portion of the lens; mature cataracts have been present longer and may involve the entire lens. In some mature cataracts, the cells in the lens have degenerated and liquefied. This debris is held within the lens capsule, a membrane that surrounds the lens.

Cataracts are more common in adult dogs than young dogs. Common causes of cataracts in adult dogs include direct penetrating trauma to the eye that damages the lens capsule and lens cells, ocular diseases that interfere with nutrition of the lens (such as glaucoma) or inflame the eye (including panophthalmitis and uveitis, see following), and some systemic diseases such as diabetes mellitus.



Cataracts diagnosed in younger dogs (usually less than 6 months old), called juvenile or congenital cataracts, can be hereditary or occur as a result of problems during gestation. Cataracts may also be acquired in association with viral infections during gestation or in newborns

In many species (including people and dogs), cataracts can be associated with other ocular congenital abnormalities. These include coloboma, which present as holes or craters in the posterior lining of the eye; retinal dysplasia, or failure of the retina to mature; and aniridia, when the iris fails to develop normally (Magrane, 1972).

Exposure to chemical substances (both local and systemic exposures), radiation, and electricity have been known to cause cataracts (Magrane, 1972).

Uveitis, an inflammation of the vascular ('uveal') layer of the eye, can be caused directly by degeneration of the lens, in some cases. With the formation and



that in some cases the development of cataracts has a complex mode of inheritance (that is, it is a polygenic trait).

Gelatt and coworkers (2003), and others (Gelatt, et. al., 1985; Barnett, et.al., 1985, Leppanen, et. al., 2001; Mellersh, et. al., 2006; Starr, et.al. 2007; Muller, et.al, 2008) noted that in the 300 breeds of dogs, 20 are known to have hereditary cataracts and the presence of defective genes leading to cataract formation is suspected to affect another 125 breeds, making hereditary/congenital cataracts one of the most common genetic diseases in dogs. Commonly affected breeds are shown in **Table 1, found near the end of this chapter.**

Hejtmancik (2008) recently reviewed the molecular genetics of congenital cataracts in people. In people, congenital or infantile cataracts are those that occur within the first year of life, and juvenile cataracts are defined as those occurring up to age 10. Approximately 8-25% of congenital cataracts were considered to be inherited – the result of transmission and expression of faulty genes. Mutations in 26 specific genes have been identified as causes of human congenital cataracts, with about half of the mutations affecting the development of lens crystallin proteins, and the remainder affecting expression of connexins, heat shock proteins and other structural proteins. Hunter and coworkers (2006) took a first step to identify linkages between breed and cataract formation in dogs, using a highly complex genomic analysis, known as radiation hybrid mapping. Their preliminary work allowed them to locate 21 canine homolog regions to human cataract gene defects and to begin analysis of these homolog regions for specific defects. This work is ongoing.

Several groups of researchers have conducted genetic and genomic analyses on dogs to see if similar mutations cause juvenile cataracts. In an early study, Zhang and coworkers (1991) were unable to find mutations in key genes encoding lens crystallins in Miniature Schnauzers. More recently, Sidjanin and coworkers (2005) were unable to identify mutations in a human gene linked to congenital cataracts (galactokinase-1) in a pedigree of Labrador Retrievers affected with juvenile cataracts.

Mellersh and coworkers (2006) were able to identify mutations in genes regulating heat shock proteins (HSF4) in Staffordshire, Bull, and Boston Terriers affected with hereditary cataracts. However, Muller and coworkers (2008) were not able to identify mutations in HSF4 in Dachshunds or Entlebucher Mountain Dogs with hereditary cataracts.

Clearly, the search for the complex mix of genes responsible for causing hereditary cataracts in dogs must continue. This is not going to be a trivial (or inexpensive) task. Havanese dogs are a toy breed variant of the Bichon breed that have multiple congenital abnormalities, including cataracts, liver shunts, heart murmurs, and a high incidence of osteochondrodysplasia. Gelatt and

coworkers (2005) noted an incidence of hereditary cataracts in over 11% of Havanese dogs, a figure substantiated by the breed club. Starr and coworkers (2007), using a canine specific microarray, found 113 differentially regulated genes in this breed and now are attempting to determine the single, multiple and linked expressions that may be responsible for the multiple congenital anomalies. Given a cost of about \$1000 per analysis using the Affymetrix Canine GeneChip 2.0, it may take time to sort this out.

## **Prevention of Cataracts**

Since we know that some types of cataracts have a hereditary basis, it's essential for dog breeders to keep thorough records of litters and diseases affecting each pup. Breeders should keep in regular contact with the owners of pups from their litters throughout the life of the dogs. The presence of cataracts in young dogs (less than 6 months old) and in multiple dogs from the same breeding is very suggestive of an underlying genetic problem. One caveat – if, during gestation, there is evidence of ill-health in the dam, cataracts may be the result of damage to the developing puppies. Most experienced breeders are very aware of the need to keep pregnant dams well-nourished and free from exposure to potentially damaging viruses and chemicals in the environment.

In the event that a litter is delivered and one or more pups develop cataracts, the breeder has a responsibility to 1) seek veterinary diagnosis and discuss treatment options for affected pups, 2) examine the breeding and pedigree of both dam and sire for similar problems (or the presence of other congenital defects from this paired breeding), and 3) refrain from breeding either sire or dam until the relationship of breeding to cataract development can be clearly determined. As noted above, hereditary cataracts were identified as a significant problem in the Miniature Schnauzer breed in the 1970s and 1980s. Following the leadership provided by breed associations, veterinarians and research scientists, this autosomal recessive trait was identified and bred against, resulting in a substantial decrease in the incidence of the disease in Miniature Schnauzers today. By identifying those dogs that are carriers of the disease and not breeding them, juvenile cataracts can be controlled and eventually eliminated. The importance of keeping accurate breeding records and long-term follow-up on litters cannot be understated.

Potential owners need to do an extensive “background check” before purchasing a Westie from a breeder. These potential Westie owners need to be sure there are accurate records for each dam and sire, a solid bloodline, and no overt problems or diseases noted in each litter from the time of whelping. If Westie owners and breeders work together, this disease will eventually be eliminated from breeding stock.



## Treatment of Cataracts

Although there is no cure for cataracts, treatment can be highly successful and is recommended for two reasons. First, the presence of cataractous lenses can lead to the development of serious eye diseases such as glaucoma. The breakdown of cataracts can cause inflammation in the eye (panophthalmitis), which leads to squinting, tearing or watering of the eye, and increased redness of the conjunctiva, the “white part” of the eye. Both glaucoma and panophthalmitis can be serious and progressive problems, potentially leading to blindness. Second, the presence of even small cataracts can lead to losses of visual acuity for affected dogs. In many cases, dogs will learn to compensate for small visual defects by head turning and memorizing their environment. Well-compensated dogs may not need to have treatment for their small cataracts. However, if there is evidence that dogs with cataracts cannot find their way in the environment (bumping into objects, reluctance to walk or run, difficulty in finding sources of food and water), consultation should be sought with your veterinarian.

Surgery to remove lenses with cataracts and replacement with artificial lenses is a common and effective method of treatment for dogs with cataracts. Surgery can be performed to remove the cataract, as long as the rest of the eye is healthy and the dog is healthy enough to undergo general anesthesia (although cataract removal in humans does not require general anesthesia, canines must be completely anesthetized for lens removal/replacement). A good outcome of surgery is dependent upon the selection of the patient, surgical technique, and perioperative (referring to the time period surrounding surgery, before, during, and after) screening and therapy. Patients are selected based on a number of different criteria including age, health, and progression of the disease. As with any procedure, the risk of failure for the operation increases with age. Increased age makes surgery slightly more risky because it will be harder for the animal to tolerate anesthesia and for the body to recover and heal itself post operatively.

In order to prepare for surgery, your veterinarian may want you to administer a few medications to your pet. These could include a topical non-steroidal anti-inflammatory agent or corticosteroids, placed on the eye, and you could be asked to give aspirin orally to reduce the pain (Na-Young, et al., 2006; Kecova, et al., 2004). Your veterinarian will also be likely to perform a complete ophthalmic examination, collect blood for a complete blood count and serum chemistry profile (to help evaluate anesthetic risk), and urinalysis, in order to determine that your animal is healthy enough to undergo surgery (Adkins, et al., 2005).

An electroretinogram (ERG) may be obtained by the veterinary ophthalmologist before cataract surgery is performed. This measurement involves placing one or more sensors on the head of the dog and then stimulating the visual pathway (generally with variably pulsed light). The dog is generally sedated while this procedure is done, to minimize movement, allow placement of sensors, and to maximize transmission of light to the retina. The reason that the ophthalmologist

may recommend an ERG is to be sure that the dog has an intact vision system and that the only significant ocular problem is the presence of the cataract. Dogs that have other eye problems (such as glaucoma with retinal degeneration leading to “retinal blindness”) will not have significant improvement in vision after cataract surgery. However, if the lens with cataract is an element in causing glaucoma, lens extraction may be helpful in preserving the eye, even with limited or no vision.

Several different surgical treatments are used for cataracts. Typically, dogs are anesthetized and prepared for surgery. The pupil is dilated with drugs and topical anesthetics and anti-infective are placed on the surface of the eye. Then, the surgeon will make a small incision that will allow access to the inside of the eye.

For some dogs, the surgeon will then extract the lens (and cataract) by cutting any ligament connections to the lens, holding the lens capsule, and gently removing the lens, taking care not to disturb fluids in the posterior portion of the eye.

Depending on the dog and surgeon, a prosthetic lens (see figure on the right) may then be implanted and the surgical site closed with very fine suture. (Source of this figure: <http://www.animaleyecare.net/diseases/cataract.htm>)



Phacoemulsification refers to a type of cataract surgery in which the lens is emulsified (essentially liquefied) with an ultrasonic instrument, and then the lenticular debris is removed from the eye. After the removal of all the residual debris from the emulsified lens, an intraocular prosthetic lens is implanted, in order to reduce the possibility of the patient suffering from extreme farsightedness. The surgical site is sutured after lens replacement.

In some cases, owners and surgeons may elect not to have a prosthetic lens implanted. Dogs without a functional lens are unable to focus light on the retina, and will only be able to perceive vague patterns due to variations in light and darkness. While this may seem to be very undesirable to us (people are very visual), many dogs can get along well with limited vision. In fact, one of the co-authors of this Topic (JR) has examined dogs in his clinical practice and found them to be blind – without owners knowing this! These dogs, if blindness has had gradual onset, compensate with memory and other senses.

With every surgery there is a risk, and cataract surgery is no exception. The prognosis for a return of some degree of visual acuity after surgery is normally very good (approximately 90%) depending on the stage of the cataract and other concurrent findings. Complications of surgery may include (Williams, et al., 1996):

- Post-operative pain and infection (diligent aftercare of the affected eye is needed for several weeks) (see below),
- Glaucoma, uveitis or panophthalmitis, or keratitis,
- Retinal detachment

Post-operative care is very important in the healing process. The eye will be bandaged in order to immobilize the eye, provide added warmth (conducive to healing) and prevent infection. In some cases, the veterinarian may temporarily suture the eyelid closed to provide additional protection to the eye during healing. Your veterinarian may ask that you apply eyedrops, ocular lubricant, or other topical treatment to the eye following surgery. Oral corticosteroids and antibiotics may also be given for a short time after surgery. A week following surgery, the eyelid suture (if used) will be removed. Owners must be cautious in handling their dogs and may wish to use a protective collar to prevent the dog from scratching at the eye. They should also restrict movement and lessen excitement during the first week prior to surgery (Na-Young et al., 2006).

Cataract surgery has significantly improved over the past few decades. At first, surgeons simply removed the intracapsular lens and used irrigation-aspiration techniques to extract any remaining lens cortical material. Recently, phacoemulsification and aspiration have become common methods for lens extraction. New surgical instruments have also been developed that allow surgeons to choose from a one-handed or two-handed system for fragmentation and aspiration. An operating microscope allows the surgeon to perform delicate work on the eye and lens fragments during surgery (see below).



(Source: [http://www.spokesmanreview.com/stories/2005/dec/15/cit\\_cataracts.IMG\\_12-15-2005\\_8F69DK0.jpg](http://www.spokesmanreview.com/stories/2005/dec/15/cit_cataracts.IMG_12-15-2005_8F69DK0.jpg))

Research is currently underway to develop ocular surgery lasers which optimize cutting, welding, and coagulation during surgery. Not only would this make cataract surgery more precise, but it would decrease the chances of certain

post-operative complications.

Research is also being done to compare the different types of intraocular lenses available. These include acryl foldable, silicone, and polymethylmethacrylate (PMMA) (Na-Young, et al., 2006). Developing the best surgical technique, as well as material for the intraocular lenses, will be significant steps in ensuring cataract surgery will be even more successful and cause fewer problems in the future.

New techniques for cataract surgery are currently being tested to minimize the size of the incision and create a more accurate suture placement. Wilkie and colleagues (2008) have modified the current approach for suture fixation of an intraocular lens (IOL) implant in canines. This new technique uses smaller suture needles in order to suture the IOL into the eye and position it appropriately. Surgeons, using an intraoperative microscope, could visualize the suture and needle during the entire procedure, and, using a foldable IOL, implant the lens through the small incision, creating less trauma and complications. Though there is a concern that this new technique, which uses two sutures, could have complications, Wilkie et al. (2008) report no instances of suture slippage or instability to date in cases followed for many months.

In an attempt to find an alternative to ultrasonographic phacoemulsification, several researchers have been investigating the use of visible, ultraviolet, and infrared radiation to remove the cataractous lens. With each of these methods, certain complications and risks arise. For example, UV radiation is potentially carcinogenic and mutagenic, and infrared lasers can result in a larger area of damage. Duran and Zato (2001) found that by using an Erbium YAG infrared laser they could obtain optimal cutting, welding, and coagulating qualities because the wavelength of the Erbium laser is strongly absorbed by the water contained in biological tissue. When clinically tested against phacoemulsification there was no significant advantage found; both methods showed improved visual acuity. Duran et al., (2001) stated that there are certain disadvantages to using the Erbium laser. For example, the laser does not penetrate very deeply and therefore creates narrow furrows. Because the laser has a low aspiration rate, removal of the flaky nuclear material can be hazardous and time-consuming. More research is needed to improve the laser technique in order for it to surpass the current phacoemulsification procedure for cataract surgery.

## Prevention

There is no real way to prevent cataracts unless they are known to be hereditary (inherited as recessive genes). If that is the case, breeding is not recommended to ensure the disease will not be passed on within the breed. The Canine Eye Registration Foundation (CERF) (<http://www.vmdb.org/feb01.html#dxspot>) warns breeders against using those dogs affected with cataracts and also those that are carriers of the disease.

**Table 1.** Inherited cataracts in the dog may occur independently or in association with other ocular diseases. The breeds that appear to develop inherited cataracts along with their age of onset are listed below. If a dog is diagnosed with

inherited cataracts, the dog should obviously not be used for breeding because of the likelihood of perpetuating the disease in the offspring. (Source: <http://www.peteducation.com/article.cfm?cls=2&cat=1606&articleid=407>)

Breed	Age of Onset
Afghan Hound	6-12 months
American Cocker Spaniel	6 + months
Boston Terrier	Congenital
Chesapeake Bay Retriever	1 + years
German Shepherd	8 + weeks
Golden Retriever	6 + months
Labrador Retriever	6 + months
Miniature Schnauzer	Congenital or 6 + months
Old English Sheepdog	Congenital
Siberian Husky	6 + months
Staffordshire Bull Terrier	6 + months
Standard Poodle	1 + years
Welsh Springer Spaniel	Congenital
West Highland White Terrier	Congenital

If cataracts are seen in young dogs (less than 6 months old and more commonly at an early age) the prudent approach is to assume cataracts to be hereditary. These dogs should not be bred, except in unusual cases specifically known to be associated with other causes. As long as owners and breeders work together juvenile cataracts could be controlled and even eliminated in some breeds.

Currently genetic research is being done in order to identify those dogs that carry the recessive gene for juvenile cataracts. If all goes well there will be a simple DNA test that can be administered to determine which dogs are affected by the disease and therefore should not be bred. This research is being conducted by the Veterinary Genetic Services (VetGen) collaboration (<http://www.vetgen.com>). They are currently obtaining samples of DNA (via swab) from the families of dogs where two or more siblings are affected with the disease. By creating a large database with affected dogs in comparison to members of their family that are not affected with the disease it will be possible for VetGen to identify the gene that causes juvenile cataracts.

### References for this Topic

Adkins E, Hendrix D, "Outcomes of Dogs Presented for Cataract Evaluation: A Retrospective Study," *Journal of American Animal Hospital Association* 41: 235-240, 2005

Andley UP, "Crystallins and hereditary cataracts: molecular mechanisms and potential for therapy," *Expert Rev Mole Med* 8: 1-19, 2006

Barnett, K, Startup, F, "Hereditary cataract in the Standard Poodle," *Vet Rec* 117:15-16, 1985

Duncan, M, Kozmik, Z, C, Cveklova, K, Piatigorsky, J, Cveki, A, "Overexpression of PAX6(5a) in lens fiber cells results in cataract and upregulation of  $\alpha 5\beta 1$  integrin expression," *Jour Cell Science* 113: 3173-3185, 2000

Duran, s, Zato, M, "Erbium:YAG laser emulsification of the cataractous lens," *J Cataract Refract Surg.* 27:1025-32, 2001

Gelatt, K, Samuelson, D, Bauer, J, Das, N, Wolf, E, Barrie, K, Andresen, T, "Inheritance of congenital cataracts and microphthalmia in the Miniature Schnauzer," *Amer J Vet Res* 44: 1130-1132, 1983

Gelatt K, Wallace M, Andrew S, MacKay E, Samuelson D, "Cataracts in the Bichon Frise," *Vet Ophth* 6:3-9, 2003

Gelatt, K, MacKay, E, "Prevalence of primary breed-related cataracts in the dog in North America," *Vet Ophth* 8: 101-111, 2005

Hejtmancik, J, "Congenital cataracts and their molecular genetics," *Sem Cell Dev Biol* 19:134-149, 2008-09-21

Hunter, L, Sidjanin, D, Johnson, J, Zangerl, B, Galibert, F, Andre, C, Kirkness, E, Talamas, E, Aclund, G, Aguirre, G, "Radiation hybrid mapping of cataract genes in the dog," *Mol Vision* 12:588-596, 2006

Kecova H, Necas A, "Phacoemulsification and Intraocular Lens Implantation: Recent Trends in Cataract Surgery," *Acta Veterinarian Brno* 73: 85-92, 2004

Leppanen, M, Martenson, J, Maki, K, "Results of ophthalmic screening examinations of German Pinschers in Finland – a retrospective analysis," *Vet Ophth* 4:165-169, 2001

Magrane, W, *Canine Ophthalmology*, 2<sup>nd</sup> ed. Lea and Febiger, Philadelphia, 1972

Mellersh, CS, Pettitt L, Forman OP, Vaudin M, Barnett KC, "Identification of mutations in HSF4 in dogs of three different breeds with hereditary cataracts.," *Vet Ophth* 5: 369-378, 2006

Müller C, Wöhlke A, Distl O, " Evaluation of canine heat shock transcription factor 4 (HSF4) as a candidate gene for primary cataracts in the Dachshund and the Entlebucher Mountain dog," Vet Ophth11: 34-37, 2008

Na-Young Yi, Shin-Ae Park, Man-Bok Jeong, Won-Tae Kim, Se-Eun Kim, Je-Min Chae, Kang-Moon Seo, "Phacoemulsification and acryl foldable intraocular lens implantation in dogs: 32 cases," Journ Vet Science 7: 281-285, 2006

Sidjanin, D, McElwee, J, Miller, B, Aguirre, G, "Cloning of canine galactokinase (GALK1) and evaluation as a candidate gene for hereditary cataracts in Labrador Retrievers," Animal Genetics 36:265-266, 2005

Starr, A, Famula, T, Markward, M, Baldwin, J, Fowler, K, Klumb, D, Simpson, N, Murphy, K, "Hereditary evaluation of multiple developmental abnormalities in the Havanese dog breed," Jour Heredity 98:510-517, 2007

Williams DL, Boydell IP, Long RD, "Current concepts in the management of canine cataract: a survey of techniques used by surgeons in Britain, Europe and the USA and a review of recent literature," Vet Rec 138: 347-353, 1996

Wilkie, D, Gemensky-Metzler, A, Stone, S, Basham, C, Norris, K, "A modified *ab externo* approach for suture fixation of an intraocular lens implant in the dog," Vet Ophth 11:43-48, 2008

Zhang, R, Samuelson, D, Zhang, Z, Reddy, V, Shastry, B, "Analysis of eye lens-specific genes in congenital hereditary cataracts and microphthalmia of the Miniature Schnauzer dog," Invest Ophth Vis Science 32:2662-2665, 1991