Eye disease in Dachshunds

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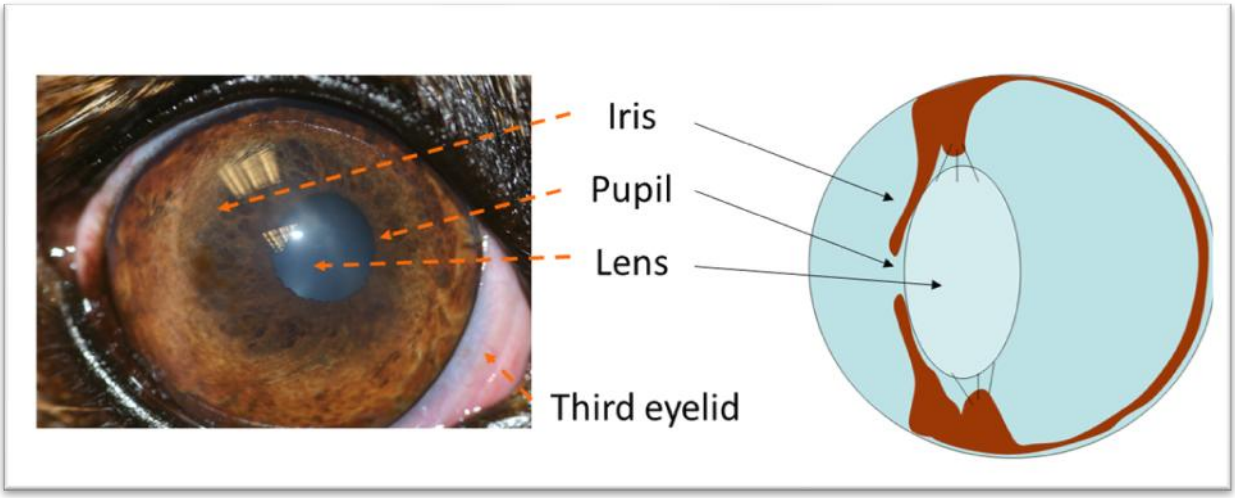
When we talk about diseases of the eye we distinguish conditions that affect the eyeball itself and conditions that affect the surrounding structures of the eye that are essential to keep the eyeball healthy and visual (the adnexa). To better understand the conditions that may affect the eye it is important to consider some of the anatomy of the eye.

Anatomy of the eye and its adnexa

The eye is protected by the eyelids and eyelashes. In comparison to humans dogs have three eyelids, the upper, the lower and the third eyelid (figure 1). The eyelids fulfill two main functions, one being protection of the eye from injuries by covering its surface in the event of a potential injury (objects moving towards the eye) and by wiping foreign particles (such as dust) off and the other is to spread the tears across the surface of the eye. The tears are mainly produced by two different glands (the lacrimal gland and the third eyelid gland) and they keep the eye moist, smooth, clean and healthy. The tears even contain substances that help fight infections.

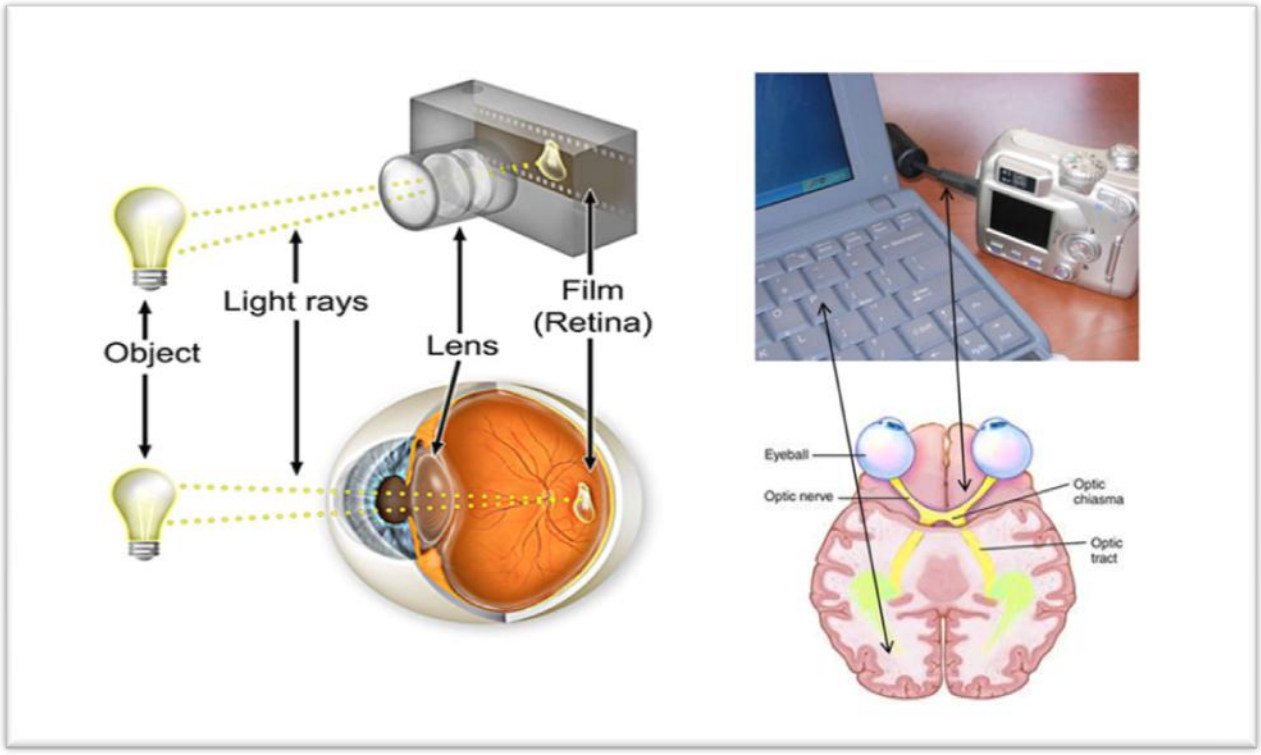
Figure 1: Anatomy of the eye



The eyeball itself is best compared with a camera (Figure 2). The wall of the eye consists of the white and non-transparent sclera and the central transparent cornea. Furthermore the structures of the eye can be divided in such that guide the light into the eye and the part where the light is finally converted into a nerve signal. The structures of the eye that guide the light in include the transparent cornea, the fluid filled front chamber of the eye, the transparent lens and the transparent jelly in the back of the eye. The pupil is formed by the iris and works as a shutter that allows the eye to regulate the amount of light that enters. The actual process of vision occurs in the retina, which is comparable to the film in a camera. The retina contains cells that are sensitive to light (the so called photoreceptors). When light reaches these photoreceptors a signal is created that is then sent on to the brain via a nerve (the optic nerve). Even here we can continue our comparison with the nerve being the cable of our camera

that is connected to the computer-like brain. All three, a functioning eye, a functioning connection (nerve) and a functioning brain are therefore necessary to be able to see. And for the eyeball the same applies, we need functional (transparent) structures to guide the light into the eye in the right manner and we need a functioning retina to get a signal that can be sent on.

Figure 2: The eye can be compared with a camera.



Vision is a fascinating and complicated sense. And maintaining best possible vision is one of the most important goals for us as veterinarians and particularly as ophthalmologists.

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Diseases of the eye

When we look at diseases that affect the eye we can divide them into two broad categories. There are diseases that are caused by genetic defects and others that are caused by other influences such as a trauma, an infection, a tumor, a toxin or defects of the metabolism such as in diabetes mellitus. While the latter cannot really be prevented the diseases caused by genetic defects can be dramatically reduced by employing a responsible breeding scheme. After all we all want to ensure that the dogs that we breed and that give us so much pleasure are healthy and comfortable.

Over the years different eye diseases became established in different breeds leading to the requirement of an eye examination prior to breeding. The aim is to choose animals for breeding that are likely to produce healthy progenies. Many of the eye disease found have been investigated in great detail so that the disease process is understood better and in some diseases the underlying genetic defect could be identified. This knowledge is now used by offering genetic testing of dogs, which is particularly helpful for conditions that develop later in life after an animal would have completed breeding such as several forms of progressive retinal atrophy (PRA, for further details see below) that can become apparent when the dog is already several years old.

Hereditary eye disease in the Dachshund

Different sources are available when establishing diseases that are hereditary in different breeds. Given that such disease can emerge at any time constant monitoring of the health of the eye is just as important as sharing findings with colleague ophthalmologists as well as dog breeders to monitor the frequency and distribution of a problem.

In the UK the information on hereditary eye diseases is collected when dogs are presented for an eye examination prior to breeding or as part of the screening process of litters for congenital eye diseases. These examinations are performed either by diploma holders of the European or American college of veterinary ophthalmologists (ECVO or ACVO) or registered BVA/KC/ISDS panelists.

The BVA scheme currently distinguishes diseases that are known to be inherited (Schedule A) and such that are under investigation and are considered suspicious of being inherited (Schedule B).

For the dachshund the following disease are named in the scheme:

Schedule A:

- GPRA in the Miniature Long Haired Dachshund (MLHD)

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Schedule B:

- Optic nerve hypoplasia in the MLHD
- GPRA in the Miniature Short Haired Dachshund (MSHD)
- Persistent pupillary membranes in the Miniature Wire Haired Dachshund (MWHD)

The American College of Veterinary Ophthalmologists publishes on a regular basis "Eye diseases that are presumed to be inherited in purebred dogs". Conditions listed in this book for the Dachshund include: microphthalmia and multiple ocular defects (too small eye with several other defects), distichiasis, dermoid (usually hair-bearing bits of skin tissue found on the surface of the eye), chronic superficial keratitis/pannus and punctated keratitis (immune-mediated inflammation of the cornea), corneal dystrophy (defects in the cornea that lead to opacities), iris coloboma (lack of iris tissue), persistent pupillary membranes (see below), uveodermatological syndrome (an immune disease that causes inflammation of the eye and the skin), cataracts (an opacity of the lens), persistent hyaloid artery (remnant of a blood vessel inside the eye), retinal folds, progressive retinal atrophy (PRA), optic nerve coloboma (part of the optic nerve is missing), optic nerve hypoplasia, retinal degeneration.

Hereditary eye diseases in the Dachshund explained

In the following section we will focus on the eye diseases that are most significant in Dachshunds in the UK.

GPRA

GPRA stands for generalised progressive retinal atrophy. It is a disease where a genetic defect results in ongoing damage and breakdown of the photoreceptors in the retina (the cells responsible for turning light into a nerve signal that is then sent on to the brain) with more and more of these photoreceptors dying over time. While the disease is not painful it invariably leads to reduced vision and eventually blindness in the affected dogs. The start of the disease can occur either early in life (early onset PRA), i.e. the affected dogs are only a few months old, or later in life (late onset PRA), when the dogs are usually several years old.

One genetic mutation associated with a form of GPRA in the MLHD has been identified and is currently used in a commercially available DNA-test. However when we examined outbred dogs that were homozygous for the mutation (carrying two defect genes) we found that many of them did not show any signs of the disease in their daily life. Just by using a more advanced diagnostic instrument (electroretinography) a reduced retinal function became visible. Researchers at the Animal Health Trust and Cambridge University have now found another area of DNA that

might influence whether a dog is getting PRA early or late in life. However further investigations are needed to classify this difference and to see whether another genetic test might be beneficial.

The same genetic defect that causes PRA in the MLHD has also been found in MSHD.

Persistent pupillary membranes

Persistent pupillary membranes are leftover blood vessels that used to fill the eye during the development. These vessels are needed for nutrition when the eye develops in the embryo and should disappear when they are not needed any more. These vessels lose their function when the eye becomes filled with fluid, which then takes over the nutrition of the structures inside the eye. If the process by which these vessels regress is disturbed some of them might persist in the eye. However this process can take some time so that the final result might not be reached before the dog is 2-3 months old or in some breeds even later than that. Persistent pupillary membranes look like little brown (pigmented) threads in the eye. A mild form of persistent pupillary membranes is very common and usually has no impact on the dogs' vision. However sometimes changes can be severe and even lead to a cloudy cornea or lens and therefore obstruct the light that goes into the eye. In these patients vision can be impaired. A mode of inheritance is not known for this condition, but given the complex nature of the regression process it is unlikely that a single gene defect causes the different degrees of persistent pupillary membranes. A diagnosis of persistent pupillary membranes can only be made when the eye has completed its development (in Dachshunds with 2-3 months).

Optic nerve hypoplasia

Optic nerve hypoplasia describes an underdevelopment of the nerve that connects the eye with the brain. The so-called hypoplastic nerves are either very thin and have very few nerve fibers or in severe cases might not contain functional nerve fibers at all. In patients that are affected by this condition the connection between eye and brain is impaired or missing, leading to severely reduced vision or even blindness. If affected, dogs are born with this condition and therefore the condition is visible during an eye examination at any age.

Distichiasis

Distichiasis has been described to be present in a large number of Dachshunds. It describes abnormal eye lashes that emerge from the eyelid margin and are directed to the surface of the eye. As the surface (cornea) of the eye is very

sensitive this might cause a considerable degree of discomfort resulting in signs such as tearing, squinting and rubbing of the eyes. While distichiasis does not have an impact on vision it can render dogs very uncomfortable, depending on how many extra lashes are present, how bristle they are and whether they actually point towards the cornea. Dogs that show signs of discomfort with the condition need to be treated to permanently remove the hair. A variety of techniques are available including cryotherapy (freezing of the hair) and electrolysis (burning of the hair's root with electricity). It is currently not known how distichiasis is inherited but it is likely that more than one gene is involved in the disease.

DNA testing and eye examination – why both?

Genetic tests are currently offered for a variety of diseases in different dog breeds (see www.optigen.com or www.aht.org.uk). These tests allow us to predict whether an animal will develop a specific disease caused by one particular gene defect. This is excellent for eradicating known hereditary diseases and helps to choose breeding partners for dogs to ensure healthy puppies. In case of a recessive mode of inheritance (when a dog only develops the disease if it carries two defective genes that originate from both parents) it even allows responsible breeding with affected dogs and carriers as long as they are mated with a clear partner (that carries two normal copies of the gene). As a result we might have dogs that carry the defective gene but also have a normal copy (so called carriers) and therefore will not suffer from the disease that we are looking at. This allows us to maintain a larger gene pool within a population of dogs, therefore a greater variation of genes and a smaller chance of new hereditary diseases evolving.

However there are drawbacks to the genetic testing that we try to overcome by continuing to examine the eyes of dogs that are going to be used for breeding. First of all very similar conditions can be caused by different genetic defects. If we use the example of a progressive retinal atrophy, we know that in people several gene defects have been shown to cause the same type of retinal atrophy. Therefore testing a person for one type of retinal atrophy will not rule out the development of the disease caused by one of the many other defects. In dogs for example we know that there are at least two different types of progressive retinal atrophy (PRA) in the Miniature Schnauzer, for one of which we can test the dogs, but not for the other.

Genetic testing for a certain disease also does not allow judgment on the health of the eye in general. Therefore it is important that we continue to examine dogs before breeding to identify emerging eye diseases that did not used to affect a certain breed. If a new eye disease is highlighted early because of the eye examinations are wide spread within the breed and therefore suffering or visual deficits of many dogs can subsequently be reduced or even prevented.