

Lens luxation – when the lens gets wobbly

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Introduction

After many years of research a genetic test is now available to assess an individual dog's genotype (genetic status) to predict whether that dog is likely to develop lens luxation or pass the mutation on to its offspring. We hope this tool will help us to reduce and eventually eliminate primary lens luxation in the future. The test is currently available for the following breeds: Chinese crested dog, Jack Russell terrier, Lancashire Heeler, Miniature bull terrier, Parson Russell terrier, Patterdale terrier, Sealyham terrier and Tibetan terrier.

Although an extremely valuable tool for breeders the DNA test does mean that many owners and breeders will have to face the fact that their dog carries the mutation that has been associated with lens luxation. What now? The following article will give an overview of the lens anatomy and discuss the disease, including monitoring for clinical signs, treatment options and breeding recommendations.

The lens – what is it there for?

The lens is a highly specialised structure that is responsible for focusing images onto the retina. The retina is a thin membrane at the back of the eye where light is transformed into an electrical signal that is then transmitted to the brain. To fulfil its function the lens has to be transparent. The lens is also able to change its refractive power to allow for focusing on objects that are seen at different distances (accommodation).

The lens - anatomy

The lens is positioned just behind the iris (figure 1). It is kept in place by inelastic microfibrils named zonules, like miniature ligaments. The zonules insert near the lens equator at 360° and attach the lens to the surrounding tissue.

Lens luxation – What does that mean?

In cases of lens luxation the zonules that keep the lens in place lose their strength. Generally a lens luxation can be primary or secondary.

Primary lens luxation is caused by a genetic defect in the zonular anatomy. This has been found to be an inherited condition in many terrier breeds (Jack Russell terrier, Parson Russell terrier, Wire-haired Fox terrier, Miniature Bull terrier, Tibetan terrier etc.) but also in some other dog breeds (eg. Lancashire Heeler, Border collie). In an affected dog the zonules start to break down at some point during their early life (usually between 3 and 5 years of age) resulting in increased movement of the lens. Subsequently the lens disinserts from the surrounding tissue and initially subluxates (partial disinsertion) and eventually luxates (dislocates) completely.

A secondary lens luxation occurs when other conditions cause the zonular defects. An ocular trauma, inflammation or increased intraocular pressure (glaucoma) can result in damage and breakdown of the zonules.

When the lens moves inside the eye it can cause damage to other intraocular structures. Particularly the movement of the lens into the front of the eye (figure 3) where it can impair the normal flow of fluid inside the eye which leads to a rapid increase of the intraocular pressure. This can damage the retina and result in transient or permanent vision loss. Therefore an anterior lens luxation (the lens moving forward into the front chamber of the eye) presents an emergency and surgical removal of the lens is usually indicated.

Lens luxation - what to look out for?

Early signs of lens luxation can usually only be recognised by a veterinary ophthalmologist as sufficient illumination, magnification and experience are necessary to see the mild and early changes. Generally owners should seek advice from their veterinary surgeon if their dog's eye appears red, has a bluish tint, is uncomfortable (squinting, tearing, rubbing) or the dog shows visual impairment. All these signs can be caused by a variety of other diseases of the eye but may indicate a lens luxation.

Lens luxation – How can it be treated?

If the lens has dislocated into the front of the eye it is usually removed in-total through an extensive excision of the cornea (nearly 160 degree, figure 4).

Lenses that are only partially dislocated or luxated into the back of the eye can also be removed. However retrieving them from the back of the globe requires more manipulation inside the eye, which results in an increased risk of retinal damage and subsequent vision loss. Many veterinary ophthalmologists prefer the use of eye drops that result in a small pupil. This aims at entrapping the lens in the back to prevent it from moving to the front of the eye. Patients are usually on this medication long-term. However the lens can still luxate to the front in which case urgent surgical removal becomes necessary.

If the early signs of a loose lens are present surgical removal of the lens can be discussed with the ophthalmologist. At this stage removal might be possible though a smaller incision (few millimetres), comparable with cataract surgery, which is less invasive to the eye and carries a better long-term prognosis.

No lens – can my dog still see?

The dog will be able to see after the lens has been removed. However the dog will be long sighted. This means that close structures will appear blurry while objects in the distance can be seen without problems. Contact lenses or so called 'doggles' to compensate for this are commercially available however are challenging to maintain in the eye, or are poorly tolerated by the dogs, respectively.

Can my dog go blind? What if it does?

Sadly some dogs become blind despite treatment. Many owners are concerned about the dog's quality of life and even consider euthanasia. Most dogs cope incredibly well with the loss of vision as they use their other senses such as smell and hearing far more than humans. Particularly in a familiar environment the dog's behaviour might not change at all. Sometimes a fellow dog can be used as a "guide dog". The book "Living with blind dogs" by Caroline D. Levin is an

excellent help for owners dealing with their beloved pet's vision loss. It describes the ocular anatomy and explains in detail different reasons for blindness in dogs. Several chapters are dedicated to describe how dogs cope with blindness and ways to help them in this new situation.

DNA test – What do I do with the results?

The dog is homozygous for the mutation (two copies of the mutation) - genetically affected: The dog is highly likely to develop lens luxation in its lifetime. We would recommend that you contact an ophthalmologist close to you and discuss the situation with him/her. Your veterinary surgeon will be able to help you with this or you can contact the Small Animal Hospital of the Animal Health Trust for further help. Regular ophthalmic examinations (every 6-12 months) from the age of 18 months should be performed and treatment instigated as appropriate.

The dog is heterozygous for the mutation (one copy of the mutation, one normal gene) - carrier: Heterozygous dogs have been reported to suffer from lens luxation. Currently the risk of a heterozygote, or carrier, developing clinical signs of lens luxation is currently estimated to be between 2 and 20%; however the true risk is probably closer to 2% than 20% and further studies are underway to produce an accurate estimate for all breeds. Regular ophthalmic examinations (every 6-12 months) are recommended from the age of 2 years.

The dog is clear of the mutation (two normal genes): The dog is very unlikely to develop lens luxation. No ophthalmic examination is needed; however the dog should be examined by your veterinary surgeon if eye problems occur. Remember that other ocular diseases can occur and a lens luxation can still be caused by other ocular problems or trauma.

My dog carries the mutation – what about breeding?

Ocular health is important but only one aspect of a dog's general health. Removing all dogs that carry the mutation (homozygous and heterozygous dogs) from the breeding population may result in a significant reduction in the genetic

diversity of many breeds. This could mean that inbreeding would increase and allow other genetic diseases to emerge. This is particularly likely to be the case for numerically small breeds (eg. Miniature Bull terrier, Tibetan terrier) and for breeds for which the mutation frequency is very high. **Therefore, we strongly advise breeders to consider all their dogs for breeding, regardless of their PLL genotype. GENETICALLY AFFECTED and CARRIER dogs can be bred with, but should only be bred to DNA tested, CLEAR dogs. All puppies from any litter that has at least one CARRIER parent should be DNA tested, so that the CARRIERS can be identified and followed clinically throughout their lives.**

We **invite all dog owners** that make use of the new DNA-test, to update us with regards to their dog's ocular health throughout its life, regardless of whether the dog is genetically affected, a carrier or clear of the mutation, this will help us to find out more about the disease and give more detailed advice in the future.