Eyeing a solution – diagnosis and treatment of canine lens luxation

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Claudia Busse explains how the condition affects sight and how to diagnose it, as well as highlighting a range of treatment recommendations

Lens luxation is a condition commonly seen in ophthalmology referral centres.

The nature of the disease makes early recognition, and often surgical treatment, necessary to maintain a visual eye. After years of research, a genetic test is now available to assess the individual dog's genotype to try to predict whether a dog is going to develop lens luxation. Wisely used, this tool will help us to reduce primary lens luxation in the future. This article will give an overview on lens anatomy, physiology and function. It will discuss lens luxation, including monitoring for clinical signs, treatment options and prognosis, as well as how to deal with patients with different genotypes.

The lens

The lens is a highly-specialised tissue that is responsible for focusing sharp images on to the retina. To fulfil this function, it has to be transparent and requires a refractive surface. The lens is positioned in the anterior part of the eye just behind the iris. It is kept in place by inelastic microfibrils named zonules. The zonules insert near the lens equator at 360° and attach the lens to the surrounding ciliary body. The ciliary muscle can change the tension on the zonules, resulting in a change of the len's shape. This results in a change of its refractive power to allow focusing on objects that are seen at different distances (accommodation).

The accommodative power in dogs is one to three diopters. The lens lacks vascularisation and, therefore, depends on the aqueous humour and, to a lesser extent, the vitreous body for its metabolic needs.

Lens luxation

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

The primary lens luxation (PLL) has been found to be an inherited condition in many terrier breeds (such as Jack Russell terrier, Parson Russell terrier, wire-haired fox terrier, miniature bull terrier and Tibetan terrier) and also in some other dog breeds (such as Lancashire heeler, border collie and Shar Pei). In affected dogs, the zonules start to break down in early life.

Histological studies of affected eyes showed abnormal zonular fibre proteins as an underlying cause for their decreased stability. Loss of zonular attachment results in increasing movement of the lens. With further progression of the condition, the lens disinserts from the ciliary body and initially subluxates (partial disinsertion) and eventually luxates completely. As a consequence, the moving lens damages other intraocular tissues, which may lead to blindness.

A secondary lens luxation occurs when other eye conditions result in zonular defects. Glaucoma is probably the most common cause for a secondary lens luxation. A chronic increase in intraocular pressure (IOP) results in enlargement of the globe with stretching and, eventually, rupturing of the zonules. In cases of uveitis, the inflammatory process can also have a destructive effect on the zonules. Blunt, as well as sharp, traumas to the eye might also result in a luxation of the lens. However, the required force usually causes other, more severe, lesions, such as a rupture of the globe, intraocular haemorrhage and retinal detachment.

A luxation of the lens in the anterior chamber of the eye, anterior lens luxation, can result in acute blindness. The lens impairs the normal flow of aqueous humour through the pupil (pupil block), leading to a rapid increase of the IOP with subsequent damage to other ocular tissues. The retina and the optic nerve are sensitive to an increased IOP. Their damage can lead to transient or permanent blindness, depending on the altitude in IOP and the time span of the increase. Therefore, an anterior lens luxation should always be considered a surgical emergency. Removal of the lens is usually indicated.

The cornea is another structure that is easily damaged by a luxated lens traumatising its innermost layer, the corneal endothelium. This leads to sometimes transient, but often permanent, central corneal oedema.

How to diagnose a lens luxation

Clinical signs associated with lens luxation vary depending on the stage of the disease. In secondary lens luxation, other ocular changes will be dominant, such as a buphthalmic (engorged) globe in cases of glaucoma, intraocular haemorrhage following trauma or chronic inflammatory changes in cases of chronic uveitis.

In case of a PLL, however, clinical signs can be very subtle. With the loss of zonular fibres, the diaphragm between the anterior chamber and the posterior segment of the globe is broken, allowing vitreous to enter the anterior chamber, visible as a cloud-like structure extending from the pupillary aperture. Furthermore, the lens will become more movable, resulting in a shivering movement of lens and iris (phacodonesis and iridodonesis, respectively).

The dorsal zonular fibres are under the highest mechanical strain, due to gravity, and are often the first to break, resulting in a slight sinking of the lens. This can be visualised as an “aphakic crescent” easiest seen in patients with a dilated pupil. The lens also loses its ability to support the iris, resulting in a straight, curtain-like, rather than convex, contour of the iris surface.

A luxated lens can either be seen in the anterior chamber or is invisible in the posterior segment. The moving lens causes a varying degree of intraocular inflammation, represented by discomfort (blepharospasm, epiphora), conjunctival and deep perilimbal hyperaemia, a miotic pupil (if not obscured by the lens) and aqueous flare. Corneal oedema might be the consequence of a current or previous anterior lens luxation or can be caused by an increase in IOP.

If corneal oedema obscures visibility of the intraocular structures, an ocular ultrasound can help to identify the position of the lens. Tonometry should always be performed in patients with signs of intraocular inflammation and corneal oedema to rule out an increase in IOP.

Diagnosis: lens luxation, what should I do now?

It is important to explain to the owner that lens luxation is serious and a sight-threatening disease and that in cases of PLL both eyes are affected by the condition, even if the disease process might be asymmetric. Urgent referral to an ophthalmologist is recommended.

In case of an anterior lens luxation, this should be done immediately. The patient should be kept quiet and treated with systemic (non-steroidal) antiinflammatory medication. The use of topical anti-glaucoma medication can be considered in case of an increased IOP. However, topical prostaglandine analogues (travoprost, latanoprost) are contraindicated in patients with an anterior lens luxation as they might worsen the pupillary block and result in further increase of the IOP. The use of topical carboanhydrase inhibitors (brinzolamide, dorzolamide) is recommended. However, IOP control is often limited.

What are the treatment recommendations?

Different options exist for treatment of a lens luxation. However, as mentioned, an anterior lens luxation always presents as a surgical emergency. The anteriorly luxated lens is traditionally removed in total through an extensive excision of the peripheral cornea (— 160°), referred to as intracapsular lens extraction (ICLE).

However, over the past few years, the surgical equipment and techniques have dramatically improved so lens removal is more routinely done via phacoemulsification. This technique uses ultrasonic energy to fragment the lens into small pieces, allowing removal through a much smaller incision. The same technique is employed in cataract surgery. This has been shown to be less traumatic to the eye, resulting in a faster recovery and better long-term outcome for vision for our patients. A recent study found that 40 per cent and 75 per cent of eyes were visual after 2.7 years when using ICLE and phacoemulsification, respectively (Manning et al).

Lenses that are only partially dislocated (subluxation) or posteriorly luxated can also be surgically removed. However, retrieving them from the posterior segment of the eye may require more manipulation of the vitreous humour and may therefore carry a greater risk for retinal detachment. The option of surgical intervention in these patients depends on the position of the lens within the eye and the surgeon’s experience.

Successful placement of suture-in artificial lenses has been reported and is offered by some ophthalmologists. While it will improve the dog’s vision dramatically, it also increases the risk of intraocular haemorrhage, a potentially devastating complication in intraocular surgery. The most common complications after lens removal include glaucoma and retinal detachment. These can occur at any time after intraocular surgery, even years after, and are best addressed in the early stage.

Alternatively, the use of miosis-inducing eye drops can be considered in cases of subluxation or posterior luxation of the lens. Topical prostaglandine analogues, such as latanoprost or travoprost, are used for this purpose. By inducing a tight miosis, the lens is trapped in the posterior segment of the eye and less likely to luxate anteriorly. Additionally, the prostaglandine analogues are powerful anti-glaucoma drugs and might prevent or delay an increase in IOP, such as secondary glaucoma, a common sequela of lens luxation.

Selected patients are usually on twice-daily, long-term medication. However, the lens can still occasionally luxate anteriorly, in which case urgent surgical removal becomes necessary. The visual outcome of patients treated medically for a posterior lens luxation with prostaglandine analogues has not been reported; however, secondary glaucoma is a known complication that often leads to blindness.

Regardless of whether the chosen treatment is medical or surgical, owners should be encouraged to present their animals for regular re-examinations as advised by their ophthalmologist.

When the lens is removed, can the dog still see?

If a dog’s lens is removed and not replaced by an artificial lens, 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 “doggles” to compensate for this are commercially available, but are challenging to maintain or poorly tolerated by the dogs.

Unfortunately, some patients become blind regardless of treatment and many owners are concerned about the dog’s quality of life and even consider euthanasia. In the author’s experience, most dogs cope incredibly well with the loss of vision as they use their other senses to compensate. Management of a blind dog should be discussed in detail with the owner.

DNA test results

If a dog is homozygous for the mutation, it is highly likely to develop lens luxation in its lifetime. Regular ophthalmic examinations (every six to 12 months) from the age of 18 months are recommended so treatment can be instigated as soon as possible. A dog heterozygous for the mutation or carrier has got a much lower risk (two per cent to 20 per cent) to develop a primary lens luxation.

Nonetheless, regular ophthalmic examinations (every six to 12 months) are recommended from the age of two years. Dogs that are clear of the mutation are very unlikely to develop a primary lens luxation. No routine ophthalmic examinations are needed, but the dog should be examined if eye problems arise, as other ocular diseases can occur and a lens luxation can still be caused by other ocular problems or inborn defects that have yet to be discovered.

What about breeding?

Ocular health is important, but only one aspect of a dog’s general health. Removing all dogs that carry the mutation from the breeding population may result in a significant reduction in the genetic diversity of many breeds, which could lead to the emergence of new genetic diseases.

This is particularly likely to be the case for numerically small breeds and for breeds in which the mutation frequency is very high. Therefore, we strongly advise breeders to consider all their dogs for breeding, regardless of their PLL genotype. Homozygous and heterozygous dogs can be bred, but should only be bred with DNA-tested, clear dogs. All puppies from any litter that has at least one parent carrying the gene should be DNA tested, so the heterozygotes can be identified and followed clinically throughout their lives.

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