Lens Luxation in the Rat Terrier?

Recently a Rat Terrier was diagnosed with an anterior luxated lens. A few breeders closely attached to this Rat Terrier and her lineage pulled together to begin researching what we now know is a genetic eye disease. We have elected to share our findings with other breeders in hopes that we can educate other Rat Terrier breeders. The details and our findings are outlined below:

The female Rat Terrier was born on 01/27/2004 and is one month shy of three years old. She was recently bred and had a litter of puppies only 5 weeks prior to her diagnoses.

On the morning (6:00 AM) on Wed 11/06/2006 the Rat Terrier was found holding her left eye closed with no additional discomfort and ate her breakfast as normal. A vet appointment was made that same day for 6:00 PM. With Dr. Paulding of Harleysville, Veterinary clinic. Within 12 hours of the onset of the eye discomfort above said veterinarian along with an additional vet at the same hospital, Dr. Stutz diagnosed the Rat Terrier with an Anterior Luxated Lens.

The following day 11/07/06 Rat Terrier was taken to a specialist Dr. Glickstein (optometrist) where the diagnosis was confirmed. The owners of the Rat Terrier after a consultation elected to remove the left eye. Upon surgery, the dog was placed under anesthesia, and as soon as she was under and surgery was to begin, the lens slid back into place. As we now know is a miracle. This specialist was surprised that the lens went back into place and the dog was taken off anesthesia.

The dog is now on one drop in the left eye of Travamax twice daily every 12 hours to maintain the pressure in the eye. We elected to save the eye, so we can learn from this Rat Terrier how this disease may effect her and possibly others if diagnosed in the future.

We would be happy to discuss this Rat Terrier findings and details in further detail, should you have an interest. As well as post updates on her health.
Lens Luxation

This presentation is being compiled to gather information on this eye disease. This project is being researched and studied by Kari Jepson (Bellridge), Barbie Trammell (Burway), Donna Sullivan (Four Paws), Shelly Heist (Rat Pack), and Stacy McWilliams (River Ridge) to help us better understand and document facts not theories.

The lens is the transparent structure within the eye that assists in focusing. It is a flattened sphere held in place by tiny ligaments around its circumference.

Cataracts and lens luxation are the two major diseases that affect the lens. In some breeds, especially in Terriers, the ligaments can break down allowing the lens to dislocate from its normal position. When the lens falls backwards into the eye it is called posterior luxation. If it falls forward ( anterior luxation) it blocks the drainage of fluid from the eye allowing pressure to build up which is extremely painful, causing permanent blindness or glaucoma to develop. Surgical removal of the lens is the only solution.

Very little is actually known about this eye disease. It is presumed to be a hereditary and genetically recessive trait. Opinions vary on how dominate or prevalent this eye disease is.

Currently a DNA study is under way by Gary Johnson, DMV of the University of Missouri. This study is funded by AKC and the Jack Russell Terrier Association.
**What is a lens luxation?**

- With this disorder there is abnormal positioning of the lens within the eye. Normally the lens is suspended between the iris and the retina, held in position by the lens zonules and the adjacent vitreous (see diagram). There can be partial (sub-luxation) or complete displacement (luxation) of the lens from its normal site, either forward into the anterior chamber of the eye (in front of the pupil) or backward into the vitreous.
- Forward (anterior) lens luxation in particular may cause an increase in the pressure within the eye (**glaucoma**), which if untreated leads to blindness.
- Lens luxation may be primary (inherited), with certain breeds predisposed as listed below. Secondary luxation may occur in any breed as a result of trauma, inflammation, glaucoma or an intraocular tumour.

**How is lens luxation inherited?**

- The mode of inheritance is not defined.

**What breeds are affected by lens luxation?**

- Primary lens luxation is most commonly seen in the border collie and in the smaller terrier breeds including the Cardigan Welsh corgi, wire- and smooth-haired fox terrier, Jack Russell terrier, Manchester terrier (toy and standard), miniature bull terrier, Scottish terrier, Sealyham terrier, Skye terrier, Tibetan terrier, and Welsh terrier. It is also seen in the Brittany spaniel.

For many breeds and many disorders, the studies to determine the mode of inheritance or the frequency in the breed have not been carried out, or are inconclusive. We have listed breeds for which there is a consensus among those investigating in this field and among veterinary practitioners, that the condition is significant in this breed.

**How is a lens luxation diagnosed?**

- With an anterior luxation, your dog may show intense pain (rubbing, pawing at the eye), tearing and visual impairment associated with glaucoma. Alternately, your dog may show no clinical signs associated with the lens luxation (usually a sub- or posterior luxation) and your veterinarian may observe an ocular abnormality during a routine physical examination. He/she will examine your dog’s eye with an ophthalmoscope and measure the intraocular pressure. This can usually be done with local anaesthetic drops placed in your dog's eye.
The canine eye

Figure 2

Components of the Canine Eye
- Conjunctiva
- Upper Eyelid
- Lens
- Cornea
- Lower Eyelid
- Third Eyelid
- Drainage Pore
- Retina
- Optic Nerve
- Vitreous Body
- Ciliary Body
What the Veterinarian may see:

- PHYSICAL EXAM: may see blepharospasm, epiphora, central corneal edema
- OPHTHALMOSCOPIC EXAM: may see increased or decreased anterior chamber depth, iridodonesis, aphakic crescent, central corneal edema; with anterior displacement the IOP is generally elevated (IOP of 50 mm Hg or more will lead to permanent optic nerve and retinal damage within hours if not relieved); IOP may be decreased due to uveitis caused by lens irritation.

How is a lens luxation treated?

- Treatment depends on the location of the lens (anterior or posterior), the presence or absence of acute glaucoma, and the potential for vision.
- With sudden anterior lens luxation, your veterinarian will immediately start medical therapy for glaucoma. The lens should be surgically removed as soon as possible. If the intraocular pressure is elevated, then surgery is urgent to prevent permanent damage to the retina and optic nerve. Pressures over 50 mm Hg will cause such damage within hours.
- For dogs with anterior lens luxation that have become blind, glaucoma can be treated by removing the globe of the eye (enucleation). This will eliminate the pain for your dog. There are also procedures that can be done that preserve the globe such as placing a prosthesis.
- Posterior luxated lenses are difficult to remove surgically. As long as the lens can be maintained in that position, problems with vision are less likely. Long-term eyedrops can be used to keep the pupil small and the lens behind it.

Breeding advice

- Dogs that have experienced lens luxation should not be used for breeding. However this condition often does not occur before 4 to 7 years of age, making it difficult to identify affected dogs before they are used for breeding.

Website reference for above material on page 3 & 4: http://www.upei.ca/~cidd/intro.htm
Breeds reported to be affected by Lens Luxation

• Primary lens luxation is reportedly most commonly seen in the Border Collie and in the smaller terrier breeds including the Cardigan Welsh Corgi, Wire- and Smooth-haired Fox Terrier, Jack Russell Terrier, Manchester Terrier (toy and standard), Miniature Bull Terrier, Scottish Terrier, Sealyham Terrier, Skye Terrier, Tibetan Terrier, and Welsh Terrier. It is also seen in the Brittany Spaniel.

**I can not verify this disease exists in the above listed breeds as referenced on the internet, except for the Jack Russell Terrier and Miniature Bull Terrier. The breeders I spoke to who raise these breeds, have never encountered it. Even Jack Russell breeders who have encountered this problem, have not had it reoccur even when the exact same breeding was repeated.**

The Miniature Bull Terrier is a breed listed as affected. This is from a web site on the Miniature Bull Terrier:

Luxation may also be associated with the following:

1. Trauma to the eye
2. Glaucoma and enlargement of the eye with breakage of the zonules
3. Disorders that affect the strength of collagen such as Ehlers-Danlos Syndrome which result in weakening the zonules
4. Congenital (present at birth) deformities of the lens
5. Idiopathic luxations, which means there is no known cause detected

Web site reference on the Miniature Bull terrier from:
http://www.ch-w.demon.co.uk/billy/pll.htm
Anterior Luxated lens
In this Wirehair Fox Terrier, detection of the anterior luxation is difficult but it obscures visualization of the central iris and pupil. Courtesy of Dr. Kirk N. Gelatt

Cataractous Lens
In this mixed breed dog, the cataractous lens is clearly displaced in the anterior chamber. Therapy consists of early lens removal and control of intraocular pressure. Courtesy of Dr. Kirk N. Gelatt
Treatments

- Treatment consists of lowering IOP to normal levels (usually with mannitol, 1-2 g/kg IV); transpupillary aqueous humor flow may be reestablished with moderate dilation with 10% phenylephrine.

- Lens removal by intracapsular extraction or phacoemulsification is performed as soon as possible.

- Postoperative treatment consists of topical and systemic antibiotics and corticosteroids, and maintenance of a moderate but moving pupil. IOP is closely monitored and any increases treated with topical β-blockers, topical and systemic carbonic anhydrase inhibitors, and prostaglandin analogs.

- Longterm postoperative complications include anterior uveitis, secondary glaucoma, and retinal detachment.
People Interviewed

• Cindy Cooke – Scottish Terrier breeder (Anstam Kennel) and former UKC Vice President. I called to ask if she or any other Scottie breeders she knows of ever had this issue. She has not. We discussed that if this is such a hereditary problem and is suppose to be displayed in Scotties, why she and other fellow breeders have not had it. Her breed has issues with PPM (persistent pupillary membranes), not Anterior Lens Luxation. We also discussed whether this was actually a hereditary disease or if it is congenital.

• Linda Reese – UKC Judge and Smooth Fox Terrier breeder – has not seen this displayed in her breed either even though her breed is listed on the internet as predisposed to this supposed hereditary eye disease.

• Genie Franklin of Farmcliffe Farms – JRT breeder – has seen this in her breed on occasion. Genie shared with me a JRT experienced this problem after being involved in a performance event. At the end of the event the dog seemed to have a “dry irritated” eye. When brought to the vet, it was discovered the dog had a luxated lens. The doctor questioned whether the physical exercise could have produced the onset of it. Genie said this disease is being researched and studied now in conjunction with the University of Missouri and their associates in England.

• Gary Johnson, DMV – started a study on anterior lens luxation. Study was under funded and is currently limited in activity. He told me they were analyzing DNA to identify markers common to this disease. He told me even though a disease is hereditary, it can also be a rare genetic occurrence. He is willing to receive DNA from any affected dog to add this information to their data base. He referred my email inquiry to his associate, who will contact me directly. Once I have heard more, I will share that new information.
People Interviewed Continued

• Laura Harakaly of Hillside Kennel – JRT breeder - had this issue one time in a dog – both eyes were affected not just one. To better track the dominance or hereditary likelihood, she repeated the exact breeding that produced this dog with luxating lenses. Not one of the dogs from the prior litter (littermates) or the new litter produced this disease. She has monitored the offspring to track any problems. Eight generations later, she still has not had a recurrence.

• David Mordasky, DMV, is a licensed veterinarian who has seen this disease about 3 times in 26 years. He told me that one was a JRT (Laura’s dog) and the other two were Labradors, breeds not even listed or believed to be predisposed to this problem. Both Labs were older dogs and he felt their age (geriatric/senior dogs) were a issue as well. Doctor Mordasky also questioned that if this was a truly dominant inheritable genetic problem, why wouldn’t we see this problem more regularly like we do in luxating patella's, hip dysplacia, and other known hereditary problems. He also discussed that various factors need to be considered – not only the hereditary genetic makeup – but environmental (dog’s activities, aging, whelping) issues may also factor into the onset of some diseases.

** Footnotes: definitions

Hereditary - genetically transmitted or transmittable from parent to offspring

Congenital - acquired during development in the uterus and not heredity
Web site Reference links

- http://www.caninegeneticdiseases.net/GLX/Lx-basics.htm
- http://www.geocities.com/welshealth/Articles/Luxation-Glaucoma.htm (current AKC study on lens luxation)
- http://www.ch-w.demon.co.uk/billy/pll.htm
Unanswered Questions

• It is suggested that dogs 3 – 7 years of age are affected. If this is such a hereditary disease, why hasn’t it been exhibited in numerous Rat Terriers prior?
• It is suggested that this is both a recessive and dominant gene. No study has been completed so therefore all current information is based on theories not actual data.
• CERF testing can not detect this disease. Other than onset, to date there is no test for prevention or detection.
• Breeders contacted with breeds listed that are suppose to be predisposed to this eye disease, have not experienced it and do not know anyone who has, except for the Jack Russell Terriers.
• Various vets contacted have not treated this disease but their clientele consists of owners of these listed breeds.
• It has been suggested this is a connective tissue issue. How do we prove this?
• It has been suggested males should be more susceptible to this disease.
• The American Hairless Terrier would also be an affected breed if this theory is correct, as it is a breed developed from the Rat Terrier. I do not know of any AHT breeder who has experienced this disease either.
• Is it possible that this disease is present and the onset can be brought on by trauma, extreme physical exertion, or activity?
• Several Rat Terrier breeders with 10+ years of breeding have not seen this problem in their dogs or offspring.
• It was suggested that all dilutes or extreme white dogs from any litter affected by this lens disease, be neutered to reduce the risk of passing it on genetically.
• It was suggested that we repeat a breeding of the same and similar lines to better track any possibility of a reoccurrence.
• It was suggested we breed at least one of the offspring of the affected dog to track any possibility of a future lens problem.