

## **CONGENITAL STATIONARY NIGHT BLINDNESS :**

being a report to the ISAE following the initial warning that this disease may be present in the Irish Setter.

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The light sensitive cells in the retina (the photoreceptors) are of two types and are called rods and cones. In summary the rods deal with awareness, motion and vision in low levels of illumination whilst the cones are involved in acuity of vision and colour interpretation. Thus a congenital rod abnormality and primary or acquired rod degenerations can be heralded by poor vision or blindness in low levels of illumination ("night blindness"), but providing there is no cone involvement day vision is retained. A high incidence of a gene mutation for a congenital type of "stationary" night blindness (CSNB) has been discovered in recent years in the Briard breed, but fortunately the mutation has been identified, a DNA test developed and disease control established.

At a routine BVA eye examination clinic in late 2008 I examined an Irish Setter with a described history suggestive of a stationary night blindness. The dog's eyes were clinically normal but a subsequent electrophysiological test called an electroretinogram (ERG) carried out under a general anaesthetic demonstrated a low response to light stimulation. In isolation this result suggested that a rod defect was present, but fortunately one year later there has been no further degeneration in sight and the ERG result is the same. It was obvious that some related and unrelated dogs should be examined both clinically and using the ERG to check the significance this initial finding. As such I have subsequently tested a further 13 Irish Setters of variable ages, some of which are related to this first dog. There was a suspicion of possible night blindness in two of the series. In none of the dogs did I find ophthalmoscopic evidence of retinal disease. However, using the same standard protocol for the electroretinography, the ERG responses were lower than values recorded routinely for other breeds. There was the expected age related reduction in values. In only one dog in this small series has the clinical status changed, its night blindness reportedly progressing to a loss of some day vision too over the course of a year. I have not had the opportunity to re-examine the dog, nor repeat its ERG evaluation, but as it is now some 9 yrs. of age it may well be that it is developing the late onset form of progressive retinal atrophy (PRA).

So where does this small series take us :

1. Though small the series demonstrates that the ERG response of the normal Irish Setter can be lower than that recorded for other breeds. My comparators are dogs of all ages that are screened for cataract surgery at the College. It is important to stress that all the electroretinography was completed using the same protocol under general anaesthesia. Recording the small potential difference generated by the retina is subject to the vagaries of the type of equipment used and for comparative reasons it is essential that there is no variation in procedure.
2. Work in the Briard breed has shown that not all affected dogs have a stationary disease and in some, ophthalmoscopic changes related to abnormal pigmentation can occur in the longer standing cases. The term "retinal dystrophy" is now preferred to CSNB and three of the four Irish Setters have tested negative for this disease.
3. It will be remembered that night blindness also occurs in PRA. Your breed appears to be clear for the early onset PRA called rcd-1, but there have been reports of a late onset PRA in both this country and on the continent. It becomes essential then that any dog demonstrating poor vision at night time or in darkened areas should be checked as routine for the ophthalmoscopic signs of this disease.
4. On the basis of this small series I cannot conclude that a CSNB/retinal dystrophy type of disease is present in the Irish Setter breed, but you should be aware of the suspicion. The fact that new inherited ocular diseases can appear in any breed from time to time should maintain a constant level of awareness that is best addressed currently by the routine testing of all dogs in any breeding programme.

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The ISAE Committee would like to emphasise that we are aware of the position regarding late onset PRA both here and overseas (see our 2007 Annual Review). Professor Bedford instigated the placing of a "watch" on the problem in Irish Setters at the BVA. There have been too few cases as yet to take this further. The main message to the breed is to say, once again, that we should all remain vigilant, that owners should have their dogs' eyes checked regularly at a BVA/KC/ISDS clinic with an ophthalmoscope; check their dogs at home with the night blindness test (see the ISAE 2007 Annual Review and this web-site). It is important to ensure that DNA in the form of blood or cheek swabs is stored at the Animal Health Trust for every one of their dogs so that it is there in readiness for developing a DNA test should the need arise (see the link to the AHT on this web-site or contact me).

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