Health Matters - GPRA

In 2014 the ESSC committee learnt of a case of GPRA (generalised progressive retinal atrophy) in a Shetland Sheepdog in the UK. Information about the disease and some of the actions that are being taken in response to this news has been posted on the Club's website, and also sent out to members in a recent mailing. The purpose of this article is to explain in more detail what GPRA is and what Sheltie owners can do about it.

What is GPRA?

Generalised progressive retinal atrophy is a disease of dogs which causes progressive blindness over a period of months or years. The retina is a light-sensitive layer of tissue, lining the inner surface of the eye, which when damaged causes loss of vision. GPRA can be divided into either dysplastic disease, where the cells of the retina develop abnormally in puppies and young dogs, or degenerative, where the cells develop normally but then undergo a destructive change. GPRA can affect the rod cells in the retina which detect shape and motion, and function in dim light, or it can affect cone cells which detect color and definition, and function in bright light.

How likely is it that my Sheltie will develop GPRA?

On the basis of current evidence not very likely. The ESSC is only aware of one confirmed case in the UK, but is taking steps to gather as much information as possible to determine whether the incidence is higher than originally thought.

At what age will a dog develop symptoms?

The UK Sheltie which has been diagnosed with GPRA was 10 years old when the first signs became apparent, suggesting that this type of GPRA is a late onset, or degenerative, variety.

In Scandinavia in recent years cases of both GPRA and retinopathy, a different disease but with similar symptoms, have been identified. Some of the Scandinavian dogs have presented with symptoms at an early age of between 1 to 2 years, but there have also been cases in older Shelties.

How do I know if my dog could be affected?

The first sign you may notice is poor vision at night or in low light conditions. The dog's pupils may appear dilated and you may witness increased 'eye shine'. In cases of advanced disease secondary cataracts may occur, in which case you could observe cloudiness in the eye. As soon as any sight problems become apparent you would be advised to make an appointment with your veterinary surgeon who could give you a referral to an eye specialist, or to arrange an eye examination direct with a BVA panellist.

Diagnosis is made by an opthalmoscopic examination of the back of the eye to see if there are any signs of degeneration. This may be supplemented by an ERG (electroretinogram) test which measures the electrical responses of the light sensitive rod and cone cells in the eye.

Is there any treatment for GPRA?

There is no cure for GPRA. Because the disease is progressive many affected dogs learn to adapt to their reduction in vision, but there are no proven effective treatments at present.

What can I do?

It is recommended that your dog should have eye examinations at intervals throughout its life, and not just as a puppy to look for congenital defects. Regular checks are essential both for general eye health, and for other conditions which might first become apparent through changes in the eye. This is exactly the same as with humans, where regular eye check ups are vital in the early diagnosis

of many health problems. Please take advantage of the facilities (described below) that the Club is offering to help us find out more about the incidence of GPRA, and if necessary to develop strategies for reducing it.

Is GPRA inherited?

GPRA is an inherited disease, and the normal pattern of inheritance is through an autosomal recessive gene – i.e. to become clinically affected a dog must inherit one defective gene from each of its parents. This is similar to the mode of inheritance of Collie Eye Anomaly (CEA/CH).

It is now recognised that GPRA is actually a family of diseases and that there is no single genetic cause across all dog breeds. Research on Shelties in Scandinavia has confirmed that at least one variety is inherited through an autosomal recessive gene, and a genetic test will soon be available for it. However there are other types for which the defective gene has still to be discovered, and therefore no tests have been developed to date. Tests on the DNA of the one affected UK Sheltie have shown that it is one of these other types of GPRA for which no test is yet available.

What is the ESSC doing about GPRA?

To improve the quality of the data available on GPRA, the ESSC will arrange a BVA/KC eye testing session each year at its Championship show. To encourage Sheltie owners to participate the Club will subsidise BVA/KC eye tests on the following basis:

- 100% refund for dogs of 5 years of age and over
- 50% refund for dogs of 3 and 4 years of age
- Not For Competition entry fees for dogs entered solely for eye testing will be refunded

Refunds will be made on production of a copy of the eye certificate, payment receipt and agreement to provide a DNA sample (which would be by cheek swab) whether or not the diagnosis is positive for GPRA. This is because any research which might be undertaken in the future would require DNA samples of both affected and unaffected Shelties. Also, as with the case of the known affected UK Sheltie, a negative result at one test does not mean that a positive result will not be found at a subsequent eye examination. If the result does turn out to be positive, then the Club will expect closely related Shelties to be presented for eye examination and to provide DNA samples.

The above subsidy, up to the value of the BVA group testing rate, will also be available for individually arranged examinations and group testing sessions with BVA panellists, organised by branches of the ESSC or other recognised bodies such as breed clubs or the Pastoral Breeds Health Foundation.

Details supplied to the ESSC of any positive tests will be strictly confidential. Owners of affected dogs will be consulted if and when the incidence of GPRA is found to require further action.

Where can I find out more about GPRA?

More information on hereditary eye disease in dogs, including GPRA, can be found in the publication on the ESSC website at http://www.essc.org.uk/pdf/eyes.pdf

For details about GPRA and retinopathy in Scandinavian Shetland Sheepdogs and the research project in Norway see http://www.nssk.no/PRA/PRA.html

General information about GPRA in other dog breeds can be found in the following Wikipedia article http://en.wikipedia.org/wiki/Progressive retinal atrophy