

BORDER TERRIER BREED HEALTH GROUP

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CANINE EPILEPTOID CRAMPING SYNDROME (CECS)

A commentary for use by Veterinary Surgeons and Owners

CECS, sometimes also known as paroxysmal gluten-sensitive dyskinesia (PGSD) and as Spike's Disease, is a seizure like disorder recorded in a number of breeds but is of particular concern in the Border Terrier. It is one of the group of conditions classed as paroxysmal dyskinesias. Symptoms are highly variable but a common factor is that the animal remains fully conscious during the episode. Also, paroxysmal implies that the abnormal activity comes in episodes with the dog being normal at other times.

The condition has almost certainly been present in the breed for a very long time but the true prevalence isn't known. It came to prominence during the eighties and nineties when a substantial number of cases were reported from other countries in Europe. This was perhaps due to breeders working with a limited gene pool and unwittingly practising close breeding on dogs from affected lines. It does seem to be more common in Europe than in the UK but it has been reported from all countries where the breed is kept.

DIAGNOSIS

Diagnosis of CECS isn't straightforward and is largely by eliminating other neurological and digestive problems such as epilepsy, portosystemic (liver) shunt, liver disease and brain lesions e.g. tumours, all of which are known to occur occasionally in Border Terriers. Because of the possibility of confusing CECS with other potentially life-threatening conditions, it is important to get a professional diagnosis. Episodes can start as young as 6 weeks through to 7 years of age with most commencing in young adulthood, the same age at which epilepsy will usually surface.

As it is unlikely that any of these episodes will be witnessed by a vet, it is extremely helpful if the owner is able to video one. In fact, definitive diagnosis would, at present, require such evidence and it would be preferable for the video to be viewed by a neurologist with an interest in the condition such as Mark Lowrie of Dovecote Veterinary Hospital, Castle Donnington, Derby or one of those working at the Animal Health Trust (AHT).

Episodes vary greatly in severity but during them dogs have difficulty walking, this varying from mild ataxia to a complete inability to stand. There may be tremors of the limbs, head and neck, sometimes with dystonia (uncontrolled muscle spasms). Some dogs show abdominal discomfort with borborygmi (gut noises) and flatulence and there may be loose stools passed before or after the event, sometimes mucoid or blood tinged. Many dogs exhibit lip smacking but this can be caused by any condition where there is abdominal discomfort. The dog remains conscious and responsive throughout. CECS cases do NOT show signs of autonomic disturbance such as salivation or loss of bowel and bladder control during the attack. Episodes can last from a few minutes to several hours and may occur frequently or only once every few months.

Some owners report that episodes may be triggered by excitement or stress and others note that episodes in their dogs may be preceded by gastrointestinal signs such as flatulence or passing loose stools.

In the majority of CECS cases drugs such as phenobarbitone or potassium bromide which are used in the treatment of epilepsy will have very little, if any, effect either in reducing the frequency or severity of episodes. Some dogs appear to benefit from diazepam or buscopan given during the attack but there don't appear to be any drugs which consistently help.

BREEDING

CECS is believed to have an inherited predisposition. Even after many years of research, not just in the UK but worldwide, we still do not know the means of inheritance but it is likely to involve multiple genes and at present there are no DNA tests available to identify animals likely to be affected or carriers of the condition.

Common sense dictates that CECS sufferers should not be bred from nor should matings be repeated which have produced affected animals. It would be prudent to breed away from lines which seem to produce a lot of cases but in order to establish if some lines do, in fact, carry a higher frequency of the genes responsible we need to have pedigrees of confirmed cases to study. These, along with veterinary confirmation of diagnosis and a sample video, should be sent to the Breed Health Co-ordinator Steve Dean (Email: stevedean@tyrianborder.com)

It is also worth sending a blood sample or a cheek swab to the AHT with full details of the case including the video, pedigree and microchip number. Sample will be useful at a later date when progress on genetic research has been achieved

DIET

Although the propensity towards CECS may be inherited it would appear that an environmental trigger may be needed to induce clinical episodes in genetically susceptible individuals. It has long been suspected that dietary proteins may be the trigger for CECS episodes and recent research suggests that, for some cases at least, the protein responsible is gluten. A study by Lowrie et al* showed remission of CECS signs in a study group of Border Terriers fed on a gluten-free diet. These animals at the start of the study were also shown to have raised levels of the antibodies Serum anti-transglutaminase 2 (TG2,IgA) and anti-gliadin (AGA,IgG). The antibody levels fell during the study but TG2 and AGA levels are NOT a conclusive test for diagnosing CECS.

From the raised TG2 and AGA levels in affected dogs at the start of the study, coupled with their reduction and the improvement in clinical signs when the animals were fed a gluten-free diet, the researchers have suggested that CECS in dogs bears many similarities to coeliac disease in humans. In coeliac disease, some sufferers exhibit non enteric signs including neurological ones i.e. gluten present in the diet can provoke symptoms in a variety of organs, not just the gastrointestinal tract. In Border Terriers with CECS, up to half of them may show other signs of immunological disturbances such as food intolerances or allergic skin disease.

ADVICE TO OWNERS

At present the best advice to owners who suspect that their dog may suffer from CECS would be to consult their vet to try and get the diagnosis confirmed. Taking a video of an episode along to the consultation would be most helpful as would taking along a completed questionnaire, downloadable from the Border Terrier Breed Health Group Website.

If CECS is confirmed it would be worth trying a gluten free diet. This must be strictly adhered to as even tiny traces of gluten may be sufficient to trigger an attack in a susceptible animal. It is also worth investigating any other dietary intolerances which the dog may suffer from. We would be interested in hearing about how many dogs respond positively to this change in feeding. Information on this should be sent to the Breed Health Co-ordinator Steve Dean. (Email: stevedean@tyrianborder.com)

During the actual episodes, there is little that the owner can do other than be there to comfort and reassure their dog. It might be worth trying buscopan or diazepam to see if this helps to relieve the cramping but in many cases it will not.

NEED FOR RELIABLE INFORMATION

The Animal Health Trust are interested in DNA samples from affected dogs, preferably confirmed by a neurologist or accompanied by a video clip of an episode. At present the samples are being stored but they will be useful if progress is made in developing a test for the disease.

The Breed Health Group is very interested in the pedigrees of confirmed cases (veterinary neurologist diagnosis) preferably forwarded with a completed questionnaire to the Breed Health Coordinator.

REFERENCES/FURTHER READING.

Phenotypic characterisation of canine epileptoid cramping syndrome in the Border Terrier--
V.Black,L.Garosi,M.Lowrie,R.J Harvey and J.Gale. Journal of Small Animal Practice 2013

A presumptive case of gluten sensitivity in a Border Terrier: a multisystem disorder?
M.Lowrie,M.Hadjivassiliou,D.S Sanzders,O.A Gardner.Veterinary Record,December 3rd 2016

The clinical and serological effect of a gluten free diet in Border Terriers with Epileptoid Cramping Syndrome - M.Lowrie,O.A Gordon,M.Hadjivassiliou,R.j Harvey,D.S Sanders,R.Powell and L.Garosi
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