

# Role of gluten in canine epileptoid cramping syndrome

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**Canine epileptoid cramping syndrome (CECS) has been known to border terrier breeders and owners since the turn of the century.**

Some owners know the condition by the name “Spike’s disease”, as Spike was one of the first dogs to be recognised with the disorder. The term CECS is a misnomer, as epileptoid implies an epileptic disorder, but work has shown this condition is separate to epilepsy.

## Definition and clinical signs of CECS



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CECS is a type of paroxysmal dyskinesia (PD) specific to border terriers. Dyskinesia is a Greek word meaning “bad movement” with paroxysmal depicting the intermittent nature of the problem. Therefore, it is a type of disorder where dogs have episodes of abnormal, uncontrolled movement.

These signs can be seen in any breed of any dog, but CECS is unique in that it appears associated with signs suggestive of gastrointestinal disease, such as intermittent vomiting, diarrhoea, borborygmi (loud gut noises) and abdominal cramping. Some affected dogs will also show frequent signs suggestive of atopy, such as scratching, chewing and licking at the skin.

PDs are episodic movement disorders in which abnormal movements are present only during

attacks. Between attacks, most individuals are generally neurologically normal.

Some border terriers can have just a few episodes and then appear to go into complete remission – having no further episodes or very occasional ones – but others experience these episodes for the remainder of their lives with a frequency of anything from one a year to several per week.

Border terriers mostly begin having episodes of CECS while young – often experiencing their first by the age of two. In some border terriers, the episodes can be triggered by excitement, a sudden burst of energy or startle, but others appear to have episodes without any apparent trigger.

## **Recognising CECS**

During an episode of dyskinesia a dog will remain fully alert. This is a key factor to consider because any loss of consciousness or awareness would rule out this condition.

During an episode a dog will exhibit involuntary movements of one or more limbs. These abnormal movements can sometimes be brief and fairly mild, with the dog showing a little bit of unsteadiness or incoordination of a single limb. However, other border terriers have very severe episodes despite being fully aware, resulting in collapse and involving the entire body, which can be very distressing to both dog and owner.

During these extreme episodes, very severe muscle contractions may occur. While some mild episodes may be fairly short, the severe episodes can last more than an hour. Once an episode has ceased, recovery is immediate – a dog will get to its feet and almost instantly revert back to normal. No “aura” appears before the episode or postictal signs following the episode.

Dogs are completely normal between episodes and show no problems until the next episode occurs. Frequency, severity and episode length can vary dramatically between dogs, but also in individuals. Importantly, PD is not thought to be life threatening or influence life expectancy, with affected dogs often living long, full lives.

In addition to the signs of PD, up to 50% of border terriers with CECS may also show gastrointestinal signs between or during episodes.

Signs that may be observed include vomiting, diarrhoea or non-specific episodes where the dog may stare vacantly, licking its lips, and appearing to be full of pain with an arched back and tense abdominal muscles. It is thought the latter sign may represent a manifestation of oesophageal reflux, or “heartburn”, which can cause significant discomfort.

Other signs occasionally reported in border terriers with CECS are frequent itching of the skin and ears or frequent licking or chewing at the paws. It is these features that make CECS unique to other types of PD.

## **Characterising paroxysmal episodes**

When managing dogs presenting with paroxysmal episodes, it is important to characterise what the episodes most likely represent. CECS is most commonly mistaken for epileptic seizures by vets and owners; however, certain features can be assessed that allow a correct diagnosis to be achieved.

### **Awareness**

As already mentioned, through any episode of dyskinesia a dog should maintain normal awareness, such as looking in the direction of a name call. Any loss of awareness would exclude a diagnosis of PD.

### **Autonomic signs**

One of the most useful signs in distinguishing PD from epileptic seizures is the absence of autonomic signs. The majority of epileptic seizures will be accompanied by autonomic signs, most commonly salivation/frothing at the mouth and/or urination.

### **Duration and recovery**

PD can continue for hours with rapid recovery, which differentiates them from prolonged epileptic seizures. Seizures usually have a short ictal duration, usually less than one minute, with the presence of abnormal behaviour after each event (the postictal phase).

However, when seizures are prolonged, such as in status epilepticus or cluster seizures, a relatively long period (hours) of abnormal behaviour would be anticipated on recovery, including blindness, pacing, disorientation and ataxia. Therefore, a slow recovery following cessation of an episode would make a PD less likely.

### **Muscle tone**

Other disorders that can mimic PD include syncope and cataplexy/narcolepsy (sudden loss of muscle tone with normal consciousness). Both disorders may be distinguished from PD by manifesting with a sudden acute loss of muscle tone. Dogs with PD should have a normal or increased muscle tone during an episode.

### **Movement**

A PD, as the name implies, should involve some form of abnormal movement of one or more limbs. Dogs with paroxysmal episodes that involve focal or whole body tremors, or twitches may be

mistaken as having PD. However, if a dog shows no obvious movement during an episode this is enough to rule out CECS.

Examples of episodes involving no movement include idiopathic benign head bobbing, in which dogs present with prolonged episodes of head tremors in either a horizontal or vertical plane. Myokymia refers to the presence of episodic focal or generalised continuous muscle twitching that can appear like worms crawling under the skin. Myoclonus is a sudden, brief, muscular jerk occurring as a single or irregularly recurrent event.

## **Stereotypy**

Epileptic seizures tend to be uniform in terms of appearance and duration. This is in contrast to PD, where episodes can vary dramatically in terms of the movements observed, frequency and duration.

## **Triggers**

PD is often triggered by excitement or exercise that is distinct to epileptic seizures, in which they most commonly occur from a relaxed state or during sleep.

Therefore, epileptic seizures tend to occur during periods of rest or at night, whereas PD may occur in response to a doorbell ringing (as an example of a sudden “startle” response), or form after a sudden rise following lying down for a period.

## **Antiepileptic medication response**

Finally, a complete lack of response to antiepileptic medication is rare in dogs with epileptic seizures, but relatively common in dogs with PD.

Therefore, dogs thought to have epileptic seizures that do not respond to appropriate treatment should be considered to have some other paroxysmal condition requiring an alternative management strategy.

## **Causes**

As is the case with most PD, CECS is believed to result from a dysfunction in the area of the brain called the basal ganglia. However, much is to be learned about how and why PDs occur.

The majority of PDs are thought to be genetic in nature and have been confirmed by studies in cavalier King Charles spaniels (so-called hypertonicity syndrome) and soft-coated wheaten terriers, in which a genetic mutation has been found that can be used as a diagnostic test for conditions in these breeds (Gill et al, 2012; O'Brien et al, 2015). Other known affected breeds include Jack

Russell terriers, Labrador retrievers, chinooks, Scottish terriers and Norwich terriers. This implicates a familial link.

Since early reports of CECS, owners and breeders have become aware of their border terriers responding to specific diets.

An initial study was performed asking owners with affected dogs what types of diet had helped. It revealed almost half had responded to a hypoallergenic or gluten-free diet. Research in people has demonstrated gluten sensitivity is a common cause for many conditions (Czaja-Bulsa, 2015). One of the best-known gluten disorders in people is coeliac disease.

Coeliac disease is caused by the immune system mistakenly producing antibodies against gluten that damage the hair-like villi that line the gut, leading to malnutrition. Blood tests can be performed to look for these antibodies to help condition diagnosis and has helped to diagnose mild symptoms in those often unaware of their condition. Although specificity is excellent, sensitivity is low, so combinations of tests are usually performed to maximise sensitivity.

Gluten is blamed for causing bloating, gut pain, headaches and lethargy. This syndrome in people has been dubbed non-coeliac gluten sensitivity and there have been claims up to a fifth have it. A neurological condition in people called gluten ataxia has shown how gluten can affect the brain without causing signs of gut disease, although, in a small proportion, diarrhoea and stomach cramps may still be seen. This has led to research in border terriers with CECS, as these dogs have neurological signs, but may also have gastrointestinal signs. Is it possible CECS is a canine equivalent of gluten sensitivity?

Six border terriers with CECS were investigated and antibodies to gluten (anti-gliadin IgG antibodies and transglutaminase-2 IgA antibodies) were measured by ELISA before they were started on a gluten-free diet. They were tested again at three, six and nine months after starting the diet.

It was found border terriers with CECS had much higher levels of gluten antibodies before starting a gluten-free diet compared to healthy border terriers. The antibody levels decreased after the start of a gluten-free diet, and were back to a normal level nine months later. Furthermore, dogs stopped having PD episodes, although one dog did scavenge horse manure, which is rich in gluten, and continued to have episodes until the owners became aware of this and prevented its consumption.

Two other dogs responded well, but were inadvertently given treats containing gluten after the study finished and this caused a relapse of episodes. However, by returning back to a gluten-free diet these dogs went into remission (Lowrie et al, 2015).

## Diagnoses

Videos remain an integral way to make a diagnosis. The consulting room is rarely the correct

environment to diagnose dogs with such obvious intermittent episodes as they are inevitably normal by the time they present to the veterinary practice. Therefore, it is always advisable to capture an episode on film to show a vet so he or she can pass this on to a suitably qualified neurologist.

Many of the dogs diagnosed with PD have undergone thorough neurological testing and, in each case, all results were found to be normal. However, testing is important to ensure other conditions are not present that may cause life-threatening problems.

Once an episode typical of this condition has been observed, the opportunity exists to perform a blood test looking for antibodies to gluten (anti-gliadin antibodies and transglutaminase-2 antibodies). However, this test is only reliable if a dog is not receiving a gluten-free diet. If dogs are on a gluten-free diet, the levels of gluten antibodies will be artificially decreased, so a negative result may be obtained despite the dog suffering from CECS.

## **Treatment**

It is important to emphasise CECS or any form of PD is not life threatening. Although episodes may be very disturbing and concerning to observe, no dog has ever died from an episode of dyskinesia. However, these episodes clearly impinge on a dog's quality of life, so any treatment that can be offered is worth pursuing.

The author's studies have shown a gluten-free diet can help. Gluten is a protein source that is a composite of two amino acid chains (gliadin and glutenin) present in the endosperm of grass-related grains, including wheat, barley and rye. Proteins of maize and rice are often mislabelled as gluten when, in fact, they lack gliadin. Going truly gluten-free means excluding the vast number of foods containing these grains and often things such as oats, which are processed with the same machinery as wheat.

Royal Canin's hypoallergenic diet has been found to be the most effective (Lowrie et al, 2015). Although other diets may well be effective, this diet has consistently improved dogs suffering from CECS. This diet is recommended for trial if you suspect a dog of having CECS, although it is recommended serum antibody tests are performed first.

## **Monitoring diet**

Once a positive test for gluten antibodies has been obtained and a gluten-free diet is started, antibody levels should be checked every three months to ensure they are returning to normal. It is also important to keep a diary of episodes to ensure they reduce in frequency once the diet starts.

If the antibody levels are not decreasing and the border terrier continues to have episodes, it is very important to ensure the dog is compliant to the diet and has not been scavenging or

inadvertently receiving other foods.

## Prognosis

As mentioned, a dog with PD or CECS is an otherwise happy dog other than when episodes are seen. Life expectancy is not reduced and, therefore, the institution of a gluten-free diet after a diagnosis has been made through serological testing should result in a good long-term remission from this condition.

## References

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