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# Paroxysmal Dyskinesia Fact Sheet

Paroxysmal dyskinesias (PDs) are episodic movement disorders in which abnormal movements are present only during attacks. Although increasingly being recognised they are often poorly characterised in veterinary literature and are commonly mistaken for an epileptic seizure, both by owners and by vets.

The term 'paroxysmal' indicates that the signs occur suddenly against a background of normality. The term 'dyskinesia' broadly refers to a movement of the body that is involuntary, which means that your dog has no control over the movement and remains fully aware of its surroundings. Between attacks, dogs are neurologically normal and there is no loss of consciousness during the attacks, though some dogs find the episodes disconcerting and do not respond normally. The attacks can last anything from a few minutes to a couple of hours and can sometime occur in clusters.

Most neurologists consider that PD results from dysfunction an area of the brain called the basal nuclei (often call the basal ganglia). Nerve cells in this area play an important role in initiating and controlling movement and any abnormal activity here can result in spontaneous and uncontrolled muscle activity. The underlying cause of many PDs is unknown, with the majority being described as idiopathic (meaning of unknown cause). In humans, some are recognised to be caused by genetic abnormalities and can be familial in origin. Some patients develop paroxysmal movement disorders as a result of a structural disease (e.g. tumour, inflammation, infection, stroke…) affecting the basal nuclei.

In veterinary medicine, PD have been described in a number of breeds (Cavalier King Charles spaniel, Border terrier, Cairn terrier, Scottish terrier, Dalmatian and Norwich terrier, Boxer, Bichon Frise, Pugs, Chinook, in which they have been 'labelled' as breed-specific entities (see below). Although not reported in the literature, similar paroxysmal movement disorders are increasingly seen in other breeds, particularly Jack Russell terriers (JRT) or Labrador retrievers in the UK. The most common appearance of affected dogs is 'cramping'/'spasm' involving the hind limbs, which is seen as an increase in the muscle tone of the limbs. While all four limbs may be affected, the hind limbs are often affected to a greater degree than the fore limbs. During an attack, animals can be severely incapacitated, since the spasm overcomes any attempts at voluntary movement; however, many dogs will still attempt to walk. In some cases, episodes can be triggered by excitement or exercise.

**How is Paroxysmal Dyskinesia (PD) diagnosed?**

Dogs and people with PD are often misdiagnosed as having unusual epileptic seizures (in some types of seizure, the patient remains conscious – as in an attack of PD). Correct identification of the exact nature of the paroxysmal event is therefore fundamental. Many involuntary muscle movement disorders are episodic in nature. Consequently, when examined between attacks, neurological evaluation is often completely normal. A thorough history is crucial in the evaluation

of affected animals, since many unrelated conditions can appear superficially similar. Paramount to establishing the existence of an involuntary muscle movement is ensuring that the affected animal maintains a normal mental state (normal consciousness) during the episode and that there is no loss of bladder/bowel function or excessive salivation. When possible, owners should be asked to provide a video recording of an episode to assist the clinician with their evaluation of the affected animal.

The most important differential diagnosis for paroxysmal movement disorders is simple, partial (focal) seizures. Such disorders are recognised in animals with stereotypic (i.e. repeatedly similar), episodic muscle movements. As such, they can be easily misconstrued as a paroxysmal movement disorder. Given the difficulty in the clinical differentiation from simple partial (focal) seizures, along with a lack of a defined diagnostic algorithm for PD, strong consideration should be placed on pursuing diagnostic testing aimed at eliminating structural disease of the CNS. Consequently, performing an MRI scan of the brain, with cerebrospinal fluid (CSF) analysis, is usually recommended. Complete blood count, serum biochemical evaluation and urinalysis to exclude underlying metabolic or endocrine disorders may also be considered prior to evaluation of possible structural brain disease.

**Selected breed-specific paroxysmal movement disorders**

Episodic falling syndrome (EFS) is a canine paroxysmal hypertonicity disorder found in Cavalier King Charles Spaniels. Episodes are triggered by exercise, stress or excitement and characterised by a gradually worsening muscle spasm in the fore and hind limbs during an attack, with the trunk also affected; this results in a characteristic 'deer-stalking' or 'praying' position. Episodes begin between fourteen weeks and four years of age and dogs are normal between episodes. The condition gets its name from the fact that all four limbs will often cramp during exercise, which can cause falling. Other conditions, including heart problems, can also cause collapse during exercise but EFS causes no loss of consciousness or colour change in the gums. Other clinical signs that sometimes occur include facial muscle stiffness, stumbling, a 'bunny-hopping' gait, arching of the back or vocalisation; again, other conditions can sometimes cause similar behaviour. A genetic test is available, so that suspected cases can have a blood sample taken and submitted via Laboklin (**http://www.laboklin.co.uk/**) or the Animal Health Trust (**http://www.aht.org.uk/**) for analysis. Treatment is possible, with most dogs responding to the use of a drug called acetazolamide. A ten-year breeder-led investigation into the inheritance of EFS has suggested an autosomal recessive mode of inheritance (**http://cavalierepisodicfalling.com**); this means that dogs can carry the disease and pass it on to their offspring, without necessarily being affected themselves. Clonazepam can be used as add-on treatment to acetazolamide in difficult to control cases, though its beneficial effects sometimes diminish with time; other drugs can also be considered.

Scottie cramp is a syndrome observed in young adult Scottish or Cairn terriers. In this condition, there is again sustained muscle contractions, primarily affecting the hind limbs. With excitement, the hind limbs typically assume a stiff, extended position, though affected dogs occasionally display exaggerated flexion of the limbs; the forelimbs can also cramp. During an attack, affected dogs develop a stiff, stilted gait over a few minutes. Severely affected dogs assume an arched posture over their back and may fall onto their side, with their head and tail flexed. The disease has a presumed autosomal recessive inheritance pattern with variable expression of the clinical signs. Diagnosis is based on a dog having typical episodes and no evidence of other conditions that might appear similar. Treatment is aimed at using drugs to improve muscle relaxation or to increase serotonin levels.

Another breed associated syndrome, which has been well documented in Border terriers, is often known as Canine Epileptoid Cramping Syndrome (CECS also known as 'Spike's disease'). Episodes in this condition are very variable, ranging from ataxia (a wobbly gait) to an inability to stand, contractions of abdominal, neck and back muscles; which results in abnormal posturing and contractions/cramping of the leg muscles (extensor rigidity or flexion of the limbs). The duration of the episode is also very variable, ranging from a few seconds to half an hour or longer; throughout this time, the dog remains aware of their surroundings, though they might be more subdued than normal. In many affected dogs, there is increased intestinal motility during an episode, which manifests as borborygmus (stomach rumbling). A genetic basis for the syndrome is suspected, but no specific genetic abnormality has been found. A recent study performed at Davie Veterinary Specialists has revealed a link between CECS and gluten sensitivity and we are in the process of validating a serological test for gluten sensitivity in this breed. Currently, the most effective way to manage CECS in Border terriers is to use a gluten-free diet, though it can take several weeks for an improvement to be noted and not all dogs will respond.

**Can we treat Paroxysmal Dyskinesia?**

Paroxysmal dyskinesia can be extremely frustrating to treat. Aside from Cavalier King Charles Spaniels, who respond to acetazolamide, and Border terriers who generally respond to exclusive gluten free diets, most cases of PD do not respond to medication. In particular, the large majority of PDs do not respond to anti-epileptic medication. We recently followed a group of affected Labradors and Jack Russell Terriers with PD receiving no treatment. Our findings suggest that these dogs with PD had a young onset, were often triggered by startle or sudden movements, and had a male bias (75%) with the majority being entire. A third of the dogs had at least one event comprising cluster episodes. Episode duration and frequency varied dramatically, even within an individual.

The natural history was self-limiting with a third entering remission and an improvement in three quarters of the dogs. Episodes significantly reduced in terms of frequency and duration in Labradors and Jack Russell Terriers respectively. Remission was significantly lower in dogs with cluster episodes than those without.

Based on these results and our experience of trying various medication for PD, we usually only advise treatment if the frequency of the episodes of PD is reaching one or more than one episode a week. Anecdotally, keppra (levetiracetam) has helped some affected Labradors.

***If you are concerned about the health of your pet you should contact your veterinary surgeon.***

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