

## Paroxysmal movement disorders in dogs

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**Laurent Garosi, Robert J Harvey** discuss diagnosis and treatment of this disorder in breeds such as cavalier King Charles spaniels

**MOVEMENT disorders in animals are increasingly being recognised, although they are often poorly characterised in the veterinary literature.**

Strictly speaking, the term “involuntary movement disorder” can be used to describe any movement that is not normal. However, it is primarily used for movement disorders that do not impair consciousness or cannot be classified as ataxias (cerebellar, vestibular, proprioceptive), lameness or weakness. The term dyskinesia specifically means diminished voluntary movements. This is often due to other involuntary movements of the body that prohibit voluntary movements. Dystonia is defined as an involuntary sustained contraction of a group of muscles, producing twisting/repetitive movements or abnormal postures.

Various terms have been used for the different forms. Chorea is an abrupt, unsustained contraction of different muscle groups. Athetosis is a prolonged contraction of the trunk muscles, resulting in bending and writhing of the body. Involuntary movements that have characteristics of both are described as choreoathetosis. Ballism is an abrupt contraction of the limb muscles, resulting in flailing movement of the limbs.

Paroxysmal dystonia or dyskinesia is the designation most commonly used when dystonic movements or postures are not continuous, but occur paroxysmally, followed by a return to normality with no neurological deficit in between attacks. The term paroxysmal indicates signs that occur suddenly against a background of normal motor functioning. Depending upon precipitating

factors, frequency, duration of the attacks and response to treatment, four major types are recognised in humans.

- Paroxysmal kinesigenic choreoathetosis/ dyskinesias (PKC/PKD): typically the paroxysms consist of dystonic posturing and choreoathetotic or ballistic movements. Consciousness is never lost. All attacks are brief (usually less than two minutes) and are precipitated by sudden movements. The frequency may be as high as 100 attacks each day. This disorder usually manifests in childhood or early adolescence and affects males four times more often than females. The family history is positive in half of the patients, usually suggesting autosomal dominant inheritance. PKC patients often respond to antiepileptic drugs, particularly carbamazepine, phenytoin and valproate.
- Paroxysmal dystonic choreoathetosis/ paroxysmal nonkinesigenic dyskinesias (PDC/ PNKD): the attacks are mainly choreoathetotic, can last from five minutes to four hours, and are often triggered by alcohol, fatigue, coffee, tea or excitement, but not by sudden movement. Frequency ranges from a few attacks a day to none for months. Between attacks, neurological examination is usually normal. PNKD often manifests during childhood and early adolescence. The disease is usually inherited in an autosomal dominant fashion, and sporadic cases are rare. Antiepileptic drugs are generally not effective and management consists of avoiding precipitating factors (such as coffee or fatigue). Limited success is achieved with anticholinergics, levodopa, acetazolamide, carbamazepine, gabapentin and benzodiazepine (particularly clonazepam).
- Paroxysmal exertion-induced dyskinesia (PED): dystonic attacks are precipitated by prolonged muscular exertion and last for five to 30 minutes. Antiepileptic drugs are also generally unhelpful.
- Paroxysmal hypnogenic dyskinesia (PHD): attacks occur during sleep and consist of ballism, dystonia, or choreoathetoid movements during non-REM sleep.

Although the pathogenesis of many paroxysmal movement disorders is unknown in humans, the overwhelming majority of cases are of idiopathic or familial aetiology. Few patients develop paroxysmal movement disorders following central nervous system lesions, such as multiple sclerosis, head trauma, cerebral palsy, stroke or encephalitis.

## **Canine paroxysmal movement disorders**

In veterinary medicine, paroxysmal movement disorders 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, chinook) where they have been “labelled” as breed-specific entities (see below). Although not reported in the literature, similar paroxysmal movement disorders can be seen in other breeds, especially young Labrador retrievers in the UK. In an affected animal, abnormal movements may not be stereotypic, varying not only in duration and frequency, but also in affected muscle groups. The most common clinical presentation is dystonia involving the pelvic

limbs, which clinically appears as increased extensor tone of the limbs. While all four limbs may be affected, the pelvic limbs are often affected to a greater degree than the thoracic limbs.

Affected animals can also be severely incapacitated by an inability to move limbs with sustained contraction of extensor or flexor muscle groups. Episodes are often triggered by excitement or exercise. Loss of consciousness and awareness is not a feature of these conditions, which may help to differentiate from some epileptic seizure activities. The neuroanatomical basis for this collection of involuntary muscle movements is unknown. In most instances, these conditions are rarely lifethreatening or incapacitating to the affected animal, therefore large-scale studies with thorough histological evaluation of the nervous system of affected animals have not occurred. Where investigations have been performed, a functional disorder probably involving abnormal neurotransmitters or their receptors has been speculated.

## **Diagnostic approach**

Paroxysmal movement disorders are often misdiagnosed as epileptic seizures (especially simple partial seizures in which the patient is still conscious), as there is significant variability and overlap in the clinical presentation for both conditions. Correct identification of the exact nature of the paroxysmal event is, therefore, fundamental. Many involuntary muscle movement disorders are episodic in nature. Consequently, neurological evaluation is often completely normal at the time of presentation. Therefore, a thorough anamnesis is crucial in evaluating affected animals. Paramount to establishing the existence of an involuntary muscle movement is ensuring the affected animal maintains a normal mental state (normal consciousness) during the episode. Descriptions of the involuntary muscle movements should include whether the movement was limited to an isolated anatomic area, such as a single limb or the head (focal), or involved the body and limbs (generalised). Knowing whether the movement persists during relaxation or sleep is also helpful. Questioning owners about a possible trigger for the movement is also important information to consider. When possible, owners should be asked to provide a video recording of an episode to assist the clinician with the 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, 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 paroxysmal movement disorder, strong consideration should be placed on pursuing diagnostic testing aimed at eliminating structural disease of the central nervous system from consideration. Consequently, performing an MRI scan of the brain, along with cerebrospinal fluid (CSF) analysis, is recommended. Complete blood count, biochemical evaluation, and urinalysis to exclude underlying metabolic (including organic acidurias) or endocrine disorders should also be considered prior to evaluating possible structural brain disease. Drugs (such as phenobarbital) known to potentially cause paroxysmal movement disorders should be discontinued. Another possibility that is rarely considered is startle disease (hyperekplexia),

which manifests as neonatal hypertonia, a hyperextended posture and breathing difficulties. Handling, or unexpected sights and sounds, may trigger episodes. Startle disease results from genetic deficits in glycinergic transmission and cases have been reported in Irish wolfhounds and Labrador retrievers. Startle disease in animals is often associated with early mortality, although early intervention with clonazepam, used to treat human startle disease, may prove useful.

## **Breed-specific 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 progressive hypertonicity throughout thoracic and pelvic limbs, resulting in a characteristic “deer-stalking” or “praying” position. Episodes begin between 14 weeks and four years of age. Stiffening of all four limbs during exercise can cause falling, although there is no loss of consciousness or cyanosis. Other clinical signs may include facial muscle stiffness, stumbling, a “bunny-hopping” gait, arching of the back or vocalisation. Affected dogs are typically normal neurologically between episodes. Most dogs respond to the use of the carbonic anhydrase inhibitor acetazolamide. A 10-year breeder-led investigation into the inheritance of EFS has suggested an autosomal recessive mode of inheritance (<http://cavalierepisodicfalling.com>). Clonazepam can be used as add-on to acetazolamide in refractory cases, but its effects may not be life-long, as functional tolerance tends to develop.

Scottie cramp is a syndrome observed in young adult Scottish or cairn terriers, consisting of involuntary sustained muscle contractions primarily affecting the hindlimbs. With excitement, the hindlimbs assume a hypertonic, extended position or they may occasionally display exaggerated flexion of the limbs. The forelimbs become abducted and develop increased extensor tone. Affected dogs progressively develop a stiff, stilted gait over a few minutes. Severely affected dogs assume an arched posture over their back and may fall into lateral recumbency with their head and tail flexed. The disease has a presumed autosomal recessive inheritance pattern with variable expression of the clinical signs. A functional deficiency in serotonin modulation of motor neuron function has been postulated. Diagnosis is based on signalment and characteristic clinical signs. Signs can be induced with exercise two hours after administering methylsergide (0.3mg/kg orally), a serotonin antagonist. Treatment is aimed at muscle relaxation or increasing serotonin levels. Although not currently documented, the use of serotonin re-uptake inhibitors may be useful for treatment of this condition.

A syndrome known as canine epileptoid cramping syndrome (also known as Spike’s disease) has been observed in border terriers. Episodes consist of gait abnormalities ranging from ataxia to an inability to stand, contractions of abdominal, neck and back muscles resulting in abnormal posturing and contractions/cramping of the appendicular muscle (extensor rigidity or flexion of the limbs). Duration of the episodes can vary from seconds to half an hour or longer during which the dog remains aware of its surroundings. Increased intestinal motility is suspected, based on hearing borborygmus. Affected dogs may experience pain during the episodes. A genetic basis for the

syndrome is also suspected. Hypoallergenic dietary therapy has been proposed with variable success ([www.borderterrier-cecs.com](http://www.borderterrier-cecs.com)).

## Future research

Two distinct studies have recently used genome-wide association strategies to identify a microdeletion affecting the gene BCAN in cavalier King Charles spaniels with EFS. BCAN encodes the brain-specific extracellular matrix proteoglycan brevican. These findings have allowed the development of rapid genotyping tests for this condition, which are now available via Laboklin ([www.laboklin.co.uk](http://www.laboklin.co.uk)) or the Animal Health Trust ([www.aht.org.uk](http://www.aht.org.uk)). We are seeking from colleagues cases of suspected canine epileptoid cramping syndrome in border terriers, suspected Scottie cramp in Scottish or cairn terriers or suspected startle disease in Irish wolfhounds or Labrador retrievers for the purpose of genetic research. Identification of a DNA marker for these conditions may help us to identify the cause of these disorders, accurately diagnose them and advise breeders in their breeding programmes.

If you have a case of suspected canine epileptoid cramping syndrome, Scottie cramp or startle disease, please contact Dr Laurent Garosi at Davies Veterinary Specialists at [lsg@vetspecialists.co.uk](mailto:lsg@vetspecialists.co.uk) or telephone 01582 883950.