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Approach to Canine Paroxysmal Dyskinesias

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11 movement.

Introduction

The term 'paroxysmal dyskinesia' (PD) describes a manifestation of abnormal involuntary muscle contraction which by definition is episodic in nature and self-limiting (Lowrie & Garosi 2017). There have been numerous published articles concerning PDs over recent years and the earliest report in veterinary literature dates back to 1942 concerning 'Scottie Cramp' in Scottish terriers (Klarenbeek 1942). Despite this, PDs remain a poorly understood and frequently under-recognised condition in veterinary patients (Richter et al. 2015; Strain 2016). Some useful terminology when considering this subject is included in table one. The purpose of this article is to review the basic classification and principles of recognition and diagnosis of PDs. This article will also introduce some of the breed-specific PDs, as well as the treatment/management options available and expected outcomes.

PDs encompass a number of clinical signs with specific terminology, as detailed in table two (Kent 2012). Dystonia tends to be the most common clinical sign which is characterised by sustained and often repetitive muscle contraction in one or several limbs (Lowrie & Garosi 2017). This results in abnormal postures or twisting and tremor-like movements (Richter et al. 2015), which can initially appear confusing and alarming to owners and veterinary surgeons alike. Affected animals may collapse and become recumbent as a result of their dystonic movements, but they will frequently remain standing and responsive to their external environment (Platt 2016). For example, affected animals may continue to attempt engagement in play or show interest in food (figure one). PDs can last from seconds to hours, often with an abrupt beginning and end (Lowrie & Garosi 2017). They occur in the conscious animal and neurological examination is typically normal between episodes. PDs may be triggered by stress, exercise or

excitement. There are also reports of drug-induced PDs (Kube et al. 2006; Mitek et al. 2013). The remainder of this article will largely concern primary PDs.

Current understanding suggests PDs are most likely the result of transient abnormal activity within deep collections of grey matter within the cerebral hemispheres (Lowrie & Garosi 2017); these areas are otherwise known as basal nuclei and are important in initiation and control of motor activity. Findings which help to support this theory include identification of lesions within the basal nuclei of patients with PDs (Bhatia & Marsden 1994; Gernert et al. 2000) and hyperactivity within basal nuclei during PD episodes, diagnosed using single photon emission computed tomography (Berti et al. 2011). Despite this evidence, the underlying cause of PDs remains controversial and ion channelopathies, as well as functional imbalances of neurotransmitters within the brain, are also implicated in their pathogenesis (Lee 1979). PDs have been linked to epilepsy on a pathophysiological basis (Crompton & Berkovic 2009) although PDs and epilepsy are now regarded as two very separate disorders.

Classification

Numerous clinical classification systems have been suggested in recent years for PDs. One of the more widely known classification systems adapted from human literature identifies three main groups of PDs: paroxysmal kinesigenic (action-induced) dyskinesia (PKD), paroxysmal non-kinesigenic dyskinesia (PNKD) and paroxysmal exertion-induced dyskinesia (PED) (Waln & Jankovic 2015). The details and differentiating features of each classification group are detailed in table three. Although it is useful to have an awareness of the characteristic features of each of these classification groups, the majority of cases in veterinary medicine are consistent with PNKD. Therefore, the direct

clinical relevance of this human classification is currently unclear. An additional sub-classification for veterinary patients was recently proposed based on suspected aetiology of PDs, which may have more relevant clinical application (Lowrie & Garosi 2017).

(1) Genetic causes – A genetic mutation of the brevican gene (*BCAN*) is implicated in episodic falling syndrome in Cavalier King Charles Spaniels (Gill et al. 2012). A mutation has also been identified in the *PIGN* gene which is linked to a PD in Soft-coated Wheaten terriers (Kolicheski et al. 2017).

(2) Dietary causes – Paroxysmal gluten sensitive dyskinesia (PGSD) is a type of PNKD well characterised in Border terriers (Black et al. 2014; Lowrie et al. 2018). The disorder shows a variable response to a gluten free diet with complete resolution of clinical signs in some cases (Lowrie et al. 2015).

(3) Secondary causes – Previous reports exist of PDs which occurred as a result of phenobarbital administration (Kube et al. 2006), or following the use of propofol (Mitek et al. 2013).

(4) Presumed genetic/ unidentified causes – An autosomal recessive mode of inheritance is presumed in several breed-related PDs, which have characteristic phenotypic features. These include 'Scottie Cramp' in Scottish terriers, as well as a familial occurrence of PDs reported in the Chinook breed of dog (Lowrie & Garosi 2016; Packer et al. 2010; Urkasemsin & Olby 2015). As of yet, no definitive genetic cause has been identified in these breeds, which can be directly linked to PDs.

Diagnosis

Diagnosis of PDs is often speculative and based on observation and assessment of key features of abnormal events. Due to the intrinsic nature of PDs, which are episodic and sometimes situation-specific, they are rarely observed on presentation to a veterinary

surgeon. Video recording and documentation of abnormal events has aided diagnosis in recent years with increased accessibility to smart-phone technology to record abnormal events at home. The marked heterogeneity and overlap with other transient disorders in terms of clinical features, can make accurate identification and recognition of PDs challenging (Lowrie & Garosi 2017). In addition, the co-occurrence of PDs with conditions such as epilepsy as seen in the Chinook dogs can further add to the diagnostic challenge associated with PDs (Packer et al. 2010).

Differential diagnosis for PDs can include seizure episodes, neuromuscular disease, idiopathic tremors, tetanic spasms, narcoleptic/cataplexic disorders, vestibular attacks, syncopal episodes, acute pain syndrome, and paroxysmal behavioural episodes (Richter et al. 2015). This list is by no means exhaustive and all of the above should be considered when making a diagnosis of PD. Table four identifies some of the main differentiating features between PDs and seizure episodes. One of the main challenges is the differentiation of PDs from simple focal seizures, which like PDs, are not associated with impaired consciousness. In contrast, simple focal seizures are often associated with obvious lateralisation of clinical signs due to unilateral cerebral involvement, while PDs often result in more generalised signs involving all limbs/both sides of the body. Although it is important to be aware of the limitations and potential inaccuracies of diagnosing by observation alone, it is a useful first step in identifying PDs.

The availability of advanced imaging along with time-consuming or invasive diagnostics (e.g. cerebrospinal fluid analysis and electrodiagnostics) is often limited in general practice, yet this does not preclude the possibility of making an accurate diagnosis of PDs (Lowrie & Garosi 2017). Advanced diagnostics are frequently of limited value

when making a diagnosis of PDs and often 'unremarkable'. Accurate clinical reasoning and judgement is vital when considering a case with possible PDs. Definitive diagnostic tests exist for very few PDs but serological testing in Border terriers for example, or genetic testing in Cavalier King Charles Spaniels and Soft-coated Wheaten terriers is available for breed-specific conditions and can easily be performed in a general practice setting. It is essential to obtain a thorough clinical history, full physical and neurological examination, in addition to obtaining a minimum database (routine blood work and urinalysis) as part of the diagnostic workup.

Breed-specific PDs in dogs (table five)

A. Paroxysmal dyskinesia of Scottish terriers (Scottie Cramp)

This episodic hyperkinetic syndrome described in this breed is now classified as a PNKD (Klarenbeek 1942; Lowrie & Garosi 2017). Clinical signs become evident from one month to seven years of age and females are overrepresented. Clinical signs consist of hypertonicity, arching of the lumbar spine, a stiff gait, flexion of the pelvic limbs, abduction of the thoracic limbs, dystonic postures (pillar-like stance, curling into a ball), skipping steps, and difficulty/inability to walk (Meyers 1970; Urkasemsin & Olby 2015). An autosomal-recessive inheritance pattern is presumed (Meyers 1970) and a defect in serotonin metabolism has been proposed as a possible cause of these episodes but the exact pathophysiological mechanism remains unknown (Meyers & Schab 1974; Peters & Meyers 1977). This is a non-progressive disease and severity can decrease with time. Avoidance of precipitating factors, such as excitement or stress, can help to reduce the frequency of episodes. Diazepam or acepromazine maleate can be used, but fluoxetine appears to be more effective in reducing the frequency and duration of the episodes (Geiger & Klopp 2009; Urkasemsin & Olby

2015).

B. Paroxysmal gluten-sensitive dyskinesia (PGSD) of Border terriers (BT)

PGSD is another term for a multisystem disorder previously known as canine epileptoid cramping syndrome (CECS) in BTs. An association between gluten sensitivity and this characteristic PD was demonstrated in BTs (Lowrie et al. 2018; Lowrie et al. 2015). Age of onset is six weeks to nine years (Black et al. 2014; Lowrie et al. 2018; Stassen et al. 2017). Dystonia of the limbs/ head/ neck, tremors, ataxia, difficulty walking, and inability to maintain a standing position are common associated findings and are often accompanied by borborygmi (Black et al. 2014; Lowrie et al. 2018; Lowrie & Garosi 2017; Lowrie et al. 2015; Urkasemsin & Olby 2015). Signs preceding the event include attention seeking, vomiting, and eating grass (Black et al. 2014). Concurrent dermatological and gastrointestinal disease is possible as seen in people with gluten sensitivity (Black et al. 2014; Hadjivassiliou et al. 2003; Lowrie et al. 2018). No genetic mutation could be identified in a cohort of 110 dogs indicating a complex mode of inheritance (Stassen et al. 2017). A link between dietary exclusion of gluten and resolution of clinical signs was shown (Lowrie et al. 2015). Serological testing for anti-transglutaminase-2 and anti-gliadin antibodies in addition to the clinical signs can aid diagnosis. However, these serological markers are not exclusive to PGSD (Lowrie et al. 2018). Institution of a strictly gluten-free diet can serve as a diagnostic and therapeutic tool (Lowrie et al. 2015; Lowrie et al. 2016). Possible PGSD has also been reported in a Yorkshire terrier (Park et al. 2014).

C. Episodic falling syndrome of Cavalier King Charles Spaniels (CKCS)

This familial PD is also known under the term ‘episodic hypertonicity of Cavalier King

Charles Spaniels' (Garosi et al 2002). There is currently disagreement in veterinary literature about whether this is a non-kinesigenic or exertion-induced PD (Forman et al. 2012; Lowrie & Garosi 2017). Age of onset is three months to four years. Ataxic pelvic limb gait, abduction of the limbs, progressive muscular hypertonicity, dystonic postures (arching of the spine, 'deer-stalking' or 'praying' posture), 'bunny hopping' and collapse are common associated clinical signs (Forman et al. 2012; Gill et al. 2012; Herrtage & Palmer 1983). An autosomal recessive mode of inheritance is suspected with around 13% of CKCS carrying the causative genetic mutation (Forman et al. 2012; Gill et al. 2012). A deletion involving the *BCAN* gene, which encodes an aggregating extracellular matrix proteoglycan has been demonstrated. DNA testing is available for diagnostic purposes but long-term may also be useful in the elimination of carrier dogs from breeding programs. Episodic hypertonicity can be a self-limiting disease in CKCS. Clonazepam can result in improvement of clinical signs although tolerance can occur with long-term therapy and acetazolamide represents an alternative therapeutic option (Forman et al. 2012; Garosi et al. 2002; Gill et al. 2012).

D. Paroxysmal dyskinesia of Soft-coated Wheaten terriers (SCWT)

Episodic dystonic movements and postures are reported in this breed as part of a familial PD. Age of onset is typically between eight months and three years (Kolicheski et al. 2017; O'Brien et. al 2015). Episodes are characterised by rapid flexion and extension of pelvic limbs with truncal dystonia and progressive involvement of thoracic limbs in severe cases (Kolicheski et al. 2017). An autosomal recessive trait of inheritance was proposed and a mutation in the gene *PIGN* was demonstrated. The diagnosis can be confirmed by DNA testing. Thus far, there is no proven benefit to treatment with

benzodiazepines, antiepileptic drugs and muscle relaxants. Clinical signs may be progressive over time (Kolicheski et al. 2017; O'Brien et. al 2015; Shelton 2004) but medical therapy with acetazolamide has been shown to be effective with some dogs achieving complete resolution of the dyskinesia (O'Brien et. al 2015).

E. Paroxysmal dyskinesia of Labrador retrievers

Episodes of dystonic involuntary movements and postures, resembling typical clinical features of PD, are occasionally reported in this breed. Age of onset is from nine months to ten years eight months and the majority of affected dogs are males. No genetic associations were investigated yet and the pathogenesis remains unknown. There is no specific treatment for PDs in this breed but a natural reduction in episode frequency was reported in the majority of dogs and spontaneous remission is possible (Lowrie & Garosi 2016).

G. Paroxysmal dyskinesia of Jack Russell terriers (JRT)

The natural history of PDs in JRTs was recently described. Age of onset was from one to eight years. Extremes of temperature preceded the episodes in 83% of dogs. Disease severity decreased over time in 57% of dogs and late spontaneous remission was achieved in 22% of dogs. The mode of inheritance and pathogenesis remain unknown. There is no specific treatment for this PD in JRTs (Lowrie & Garosi 2016).

H. Paroxysmal dyskinesia of Chinooks

A familial PD has been reported in this breed. Most dogs develop signs within their first three years of life. Affected dogs are unable to stand or walk during the episodes. Head tremors, flexion of one or more limbs, dystonia, repetitive limb contractions, and

collapse were reported. An autosomal recessive or polygenic pattern of inheritance is suspected based on pedigree analysis. Interestingly, the same breed lines, which suffered with PDs were found to suffer from epilepsy, but these two conditions appear to coexist in some dogs. There is no known treatment (Packer et al. 2010).

I. Sporadic reports of paroxysmal dyskinesia in other breeds

Typical paroxysmal dyskinetic events were reported in several other breeds including Wire-haired terrier, Norwich terrier, Dalmatians, West Highland White terriers, Cairn terriers, Norwich terriers and Bichon Frise (De Risio & Freeman 2015; Penderis & Franklin 2001; Urkasemsin & Olby 2015; Woods 1977). Episodic hypertonicity was also seen in Springer Spaniels and Boxer puppies (Ramsey et al. 1999; Shelton 2004). A 12-week-old female Golden Retriever with suspected PD was treated successfully with acetazolamide (Royaux et al. 2015). A phenobarbital-responsive PD was reported in a German shorthaired pointer (GSHP) (Harcourt-Brown 2008). In a recent publication, anecdotal evidence of another GSHP with similar signs that achieved full remission after phenobarbital therapy was presented (Lowrie & Garosi 2017).

Closing Comments

PDs comprise a heterogeneous group of disorders with a high degree of phenotypic variability. Video footage and documentation of abnormal episodes can be extremely useful as a first step in identification of PDs. Accurate clinical reasoning and judgement is vital when considering a case with possible PDs and differentiation from other paroxysmal disorders, for example seizure episodes, can be challenging. Although there are several breed-related PDs which are well characterised, PDs may occur in any breed and they remain a poorly understood and frequently under-

recognised condition in veterinary patients. A genetic and/or pathophysiological classification would not only facilitate the diagnosis of PDs, but it may also support the development of new therapeutic approaches.

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385 **Table one – Useful terms when considering the subject of PDs**

Paroxysmal	A sudden occurrence or intensification of clinical signs which is episodic in nature
Dyskinesia	Impairment or abnormal voluntary movement
Movement Disorder	A condition affecting the ability of an individual to initiate or control movement, often resulting in abnormal voluntary/involuntary movements
Paroxysmal kinesigenic dyskinesia (PKD)	A form of PDs precipitated by sudden movement
Paroxysmal non-kinesigenic dyskinesia (PNKD)	A form of PDs associated with stress or excitement, but not precipitated by movement
Paroxysmal exertion-induced dyskinesia (PED)	A form of PDs associated with heavy exercise

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387 **Table two – Clinical signs associated with PDs and their definitions**

Dystonia	Sustained involuntary muscle contraction causing abnormal postures
Athetosis	Prolonged, slow, involuntary contraction involving musculature of the trunk causing writhing and contortion of the body
Chorea	Unsustained involuntary muscle contraction causing abrupt movements
Choreoathetosis	Involuntary muscle contraction involving a combination of the athetosis and chorea
Ballism	Abrupt contraction of limb musculature causing flailing movements of the limbs, often unilateral

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390 **Table three – Summary of the key features associated with the three main categories of**
391 **PDs based on a human classification system**
392

Feature	PKD	PNKD	PED
Trigger	Sudden movement	Stress, caffeine, alcohol	Heavy exercise
Age of onset	Childhood/adolescent	Childhood/adolescent	Variable
Duration	< 5 minutes	2 – 4 minutes	5 minutes – 2 hours
Frequency	Variable – multiple attacks per day, may improve with age	Variable - several per week to several in a lifetime	Dependant on exercise
Treatment	Anticonvulsants including carbamazepine	Trigger avoidance, benzodiazepines	Trigger avoidance, ketogenic diet, gabapentin

PKD, paroxysmal kinesigenic dyskinesia; PNKD, paroxysmal non-kinesigenic dyskinesia; PED, paroxysmal exertion-induced dyskinesia.

393 **Table four – Differentiating clinical characteristics of PDs and seizure episodes**

Paroxysmal Dyskinesia	**Seizure Episode**
No impairment of consciousness	Reduced/ absent conscious responses
No autonomic signs	Autonomic signs may be present, e.g. hypersalivation, urination/defecation
Usually abrupt onset	Possible prodromal behavioural abnormalities
No abnormal post-ictal behaviours	Post-ictal behavioural changes may be present
Variable duration (seconds to hours)	Usually of short duration (<5 minutes)
Unremarkable neurological examination in between episodes	Possible persistent/ transient inter-ictal neurological abnormalities

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Table five – Breed specific PDs

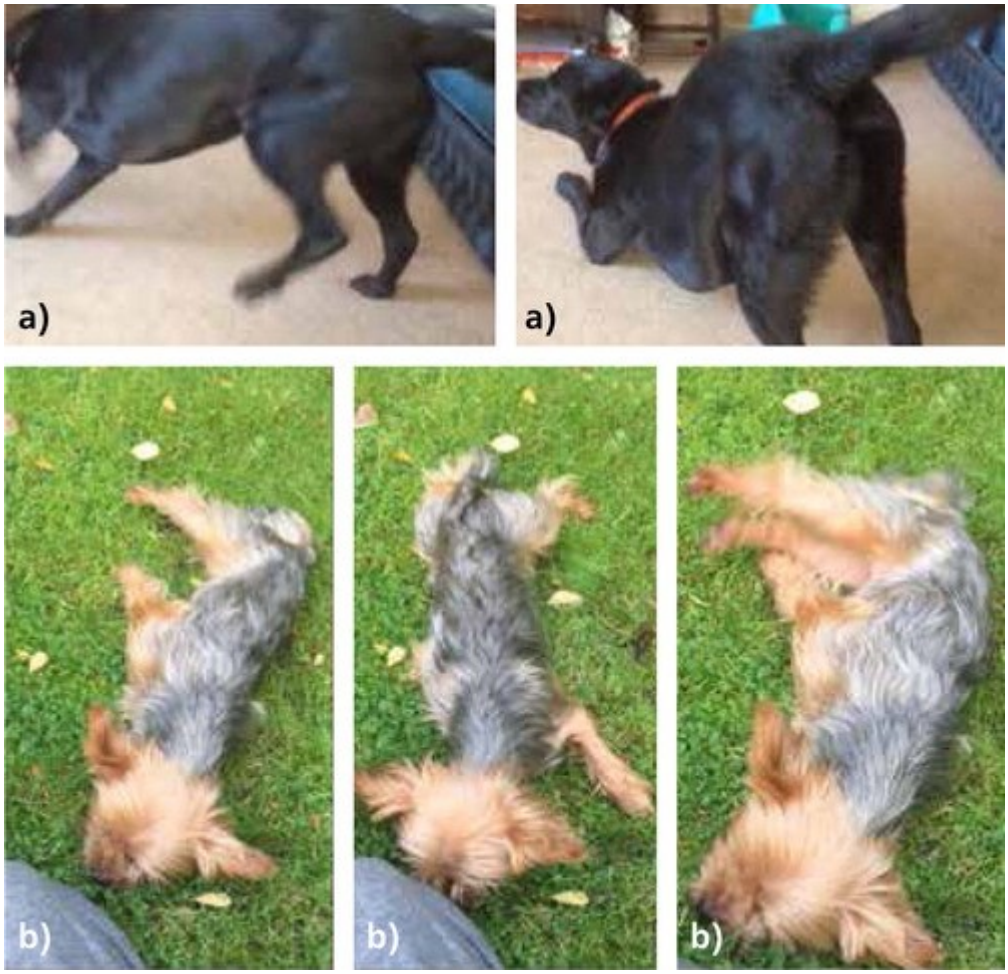
Breed specific PD	Reported age of onset	Suspected mode of inheritance	Genetic mutation	DNA test	Treatment	Prognosis
Scottie Cramp	One month to seven years	Autosomal recessive	Currently unknown	No	Fluoxetine Acepromazine Diazepam	Fair prognosis: non-progressive disease, severity can decrease with time
PGSD/CECS of Border terriers	Six weeks to nine years	Currently unknown	Currently unknown	No§	Gluten-free diet	Good prognosis: variable but generally good response to gluten-free diet
Episodic falling syndrome of CKCS	Three months to four years	Autosomal recessive	BCAN gene	Yes¥	Clonazepam, acetazolamide	Good prognosis: it can be a self-limiting disease
PD of Soft-coated Wheaten terrier	Median: two years	Autosomal recessive	PIGN gene	Yes	Acetazolamide	Guarded prognosis without treatment: generally progressive disease; Fair prognosis with treatment: improvement or resolution of signs is possible.
PD of Labrador retrievers	Nine months to ten years eight months	Currently unknown	Currently unknown	No	No known treatment*	Good prognosis: reduction in episode frequency can be seen in the majority of dogs and spontaneous remission is possible
PD of JRT	One to eight years	Currently unknown	Currently unknown	No	No known treatment*	Fair prognosis: Disease severity can decrease over time in some dogs and late spontaneous remission is possible
PD of Chinooks	Two months to five years	Autosomal recessive or polygenic trait	Currently unknown	No	No known treatment*	Unknown

* Medications such as clonazepam, acetazolamide or fluoxetine can be trialled for PD if episode frequency is not satisfactory.

§ Serological testing is available (anti-transglutaminase-2 IgA and anti-gliadin IgG antibodies).

¥ A number of homozygous dogs remain asymptomatic.

401 Figure one – a) dystonia, choreoathetosis and ballism, exhibited by an 18 month-old male
402 neutered Labrador retriever; b) dystonia and choreoathetosis resulting in collapse and
403 recumbency in a 4 year-old male entire Yorkshire terrier.



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