Propriospinal myoclonus

Clinical reappraisal and review of literature

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ABSTRACT

Objective: Propriospinal myoclonus (PSM) is a rare disorder with repetitive, usually flexor arrhythmic brief jerks of the trunk, hips, and knees in a fixed pattern. It has a presumed generation in the spinal cord and diagnosis depends on characteristic features at polymyography. Recently, a historical paradigm shift took place as PSM has been reported to be a functional (or psychogenic) movement disorder (FMD) in most patients. This review aims to characterize the clinical features, etiology, electrophysiologic features, and treatment outcomes of PSM.

Methods: Re-evaluation of all published PSM cases and systematic scoring of clinical and electrophysiologic characteristics in all published cases since 1991.

Results: Of the 179 identified patients with PSM (55% male), the mean age at onset was 43 years (range 6–88 years). FMD was diagnosed in 104 (58%) cases. In 12 cases (26% of reported secondary cases, 7% of total cases), a structural spinal cord lesion was found. Clonazepam and botulinum toxin may be effective in reducing jerks.

Conclusions: FMD is more frequent than previously assumed. Structural lesions reported to underlie PSM are scarce. Based on our clinical experience and the reviewed literature, we recommend polymyography to assess recruitment variability combined with a Bereitschaftspotential recording in all cases. *Neurology*® 2014;83:1862–1870

GLOSSARY

BP = Bereitschaftspotential; **DTI** = diffusion tensor imaging; **FMD** = functional movement disorders; **PSM** = propriospinal myoclonus; **RLS** = restless legs syndrome.

Propriospinal myoclonus (PSM) is a rare hyperkinetic movement disorder involving the axial muscles.¹ Brown et al.¹ were the first to describe the clinical phenomenology based on 3 cases. PSM is characterized by painless, usually flexor arrhythmic jerks of the trunk, hips, and knees, which are often stimulus sensitive and typically increase when supine.^{1–3} Based on the initial publications, PSM had a presumed generation in the spinal cord and diagnosis depended on characteristic features at polymyography.^{1–4} The myoclonic activity in PSM was assumed to originate from a spinal generator and spread up and down the spinal cord via intrinsic propriospinal pathways, and these axial jerks were therefore termed propriospinal myoclonus.¹ Electrophysiologic features of PSM were published after the first clinical descriptions and included a fixed pattern of muscle activation, a slow conduction velocity (5–15 m/s) consistent with spread within the propriospinal tract, EMG burst duration of less than 1,000 ms, synchronous activation of agonist and antagonist muscles, and no facial involvement.⁴ Until recently, patients were subdivided into idiopathic PSM (i.e., primary, no underlying cause found) and secondary PSM. However, the diverse etiology of this disorder is currently highlighted by several movement disorders specialists, including our centres.^{5–7} A functional etiology was reported to underlie several PSM cases. Psychogenic or functional movement disorders (FMD) are common and characterized by abnormal movements inconsistent with a known organic etiology.⁸ Clinical clues indicative of

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FMD include acute onset, rapid progression of symptoms, patterns incongruent with organic movement disorders, distractibility, variability and simultaneous occurrence of abnormal movements and dysfunctions (multiple somatizations), plus emotional and psychiatric disturbances.^{8,9} Subsequent electrophysiologic studies may confirm the clinical diagnosis of FMD.^{10,11} The diagnosis of laboratory-supported FMD is made when clinically established diagnosis of FMD is supported by positive electrophysiologic tests indicating FMD.¹² Jerk-locked back-averaging of simultaneous EEG–EMG recording can demonstrate a Bereitschaftspotential (BP) preceding the axial jerks, indicating the type of motor preparatory activity usually associated with self-paced voluntary movement, and therefore FMD. However, in the first probable functional PSM case, a BP could not be demonstrated, questioning the electrophysiologic discriminating value of the BP in clinical practice.¹³ Other electrophysiologic features regarded to support FMD can consist of variable patterns of muscle recruitment and entrainment.^{8,14} Polymyography on its own may not be the optimal electrophysiologic test to discriminate between etiologies as it was demonstrated that healthy volunteers can mimic the EMG pattern that is considered typical for PSM.¹⁵

The current review of published PSM cases aims to evaluate clinical and electrophysiologic criteria that may identify FMD and PSM. Irrespective of the presumed etiology as reported by the authors, we systematically reviewed clinical features supporting organic disease and features indicative of FMD. Given the current debate on PSM etiology, our objective was to characterize idiopathic and secondary PSM and axial jerks due to FMD. We focus on the history, outcome, and etiology, and assess whether symptomatology, electrophysiologic features, therapeutic effect, and outcome are similar in the 3 etiologic groups. Finally, we determine red flags based on clinical cases with a proven structural lesion that should warn the clinician to suspect secondary PSM.

METHODS Search strategy and selection criteria. References for this review were identified by searches of PubMed from 1991 until May 2013 with combinations of the terms “proprioception,” “myoclonus,” “axial myoclonus,” and “spinal

myoclonus.” Conference proceedings were excluded. Only articles published in English were reviewed.

No new articles were found in the reference lists. Identical cases were included once.^{5,6,16} Thus, 47 articles describing 179 cases of PSM were systematically reviewed.^{1–7,13,17–55} Cases were divided based on etiology, with secondary cases further subdivided into cases with a structural lesion, defined by a spinal cord lesion on the MRI scan, i.e., myelopathy or syringomyelia, and secondary cases without structural cord abnormalities. All included articles were scored on presumed etiology, clinical data including tic characteristics,⁷ PSM and segmental myoclonus characteristics as proposed by Esposito et al.,⁶ FMD characteristics and radiologic findings, electrophysiologic data, and treatment. Regarding the previously listed clinical clues for FMD, we consider both functional neurologic symptoms and other somatization disorders as previous somatizations. At neurologic examination, special attention should be paid to facial movements, vocalizations, suppressibility, and improvement of symptoms during distraction. Electrophysiologic details of combined EEG–EMG and BP recordings were reviewed. Long-latency reflexes are too infrequently reported to be included in this review.¹⁷ As we found overlapping features with isolated tic disorders in our case series, we also scored the clinical tic characteristics of the ability to suppress jerks and premonitory sensations.⁷ Moreover, any items lacking in our own published case series were retrieved and scored systematically in order to increase the amount of available data to review.^{5,7}

RESULTS Epidemiology. The mean age at onset was 43 years (range 6¹⁸–88⁷ years). Mean duration of symptoms at presentation was 53.5 months (range 0–384, reported in 178 cases). Over half of the cases (n = 98) were male (55%; % in parentheses hereafter refers to % of total of 179 cases identified or % of the annotated subgroup).

Etiology: Historical shifts. The first historical shift was the observation of Brown et al.¹ in 1991 annotating axial jerks as PSM. To our knowledge, 3 cases were published prior to 1991 with axial flexion jerks resembling PSM.^{56,57} Two patients were classified as essential myoclonus.⁵⁶ A similar pattern of jerks was described in the third patient with complete cervical spinal cord transection who developed rhythmic myoclonic movements of the trunk and lower limbs, which was annotated as spinal myoclonus.⁵⁷

In 2008, the first case of possible FMD was published, causing a second historical paradigm shift.¹³ Thereafter, 129 cases were published, of which 104 (81%) were reported to be FMD, 10 (8%) idiopathic PSM, and 15 (12%) secondary PSM. Of the 179 cases, 104 (58%) were reported as FMD and 75 (42%) as organic. Of the latter, 29 (16%) were idiopathic and 46 (26%) had a secondary etiology. Reported etiologies of symptomatic PSM varied. A structural lesion in the spinal cord was found in 12 cases (7% of total cases and 26% of secondary cases) including ischemic myelopathy, cervical tumors, neuromyelitis optica, and 5 cases involving syringomyelia.^{1,2,23–25,31,35,39,40,46} In an additional case, although

"C5–C6 disc protrusion impinging on the cord was present on spinal MRI," the presence of myelopathy was not clearly stated and this was therefore considered as a nonstructural secondary case.⁴³ Other proposed secondary etiologies included cases with cervical disc herniation (no myelopathy)²⁰ and lumbar disc protrusion.¹⁷ An associated back trauma was reported in 10 cases.^{2,18,23,40} Single secondary cases were reported in herpes zoster,⁴ Lyme neuroborreliosis,²¹ hepatitis C,⁴⁸ gluten sensitivity,⁵⁵ a fragile X permutation,⁵² anti-MAG antibodies,⁴⁷ myasthenia gravis,² breast cancer,⁴¹ and inflammation.⁴² Furthermore, 2 PSM cases were related to drug abuse, 1 involving cannabis²⁹ and 1 with N₂O and vitamin B₁₂ deficiency.⁵³ Finally, secondary PSM was reported in 2 cases with a possible toxin from *Escherichia coli*²² and 4 secondary to administered medication, being interferon,¹⁹ ciprofloxacin,³⁸ bupivacaine,⁵⁴ and 1 after multiple drugs for a cesarean section.²⁷ Of note, temporal links between onset of PSM and reported causality were variable.

PSM triggered by relaxed wakefulness or the transition stage between wakefulness and sleep (predormitum stage) has been reported in 5 cases (classified as idiopathic).^{26,32,48} This could imply a clinical overlap of idiopathic PSM with restless legs syndrome (RLS) and axial jerks with a PSM pattern that represent another motor phenomenon associated with RLS.⁴⁹

Clinical phenomenology. PSM characteristics. Details of PSM characteristics can be found in table 1. In 77 (43%) of the total 179 cases, an increase in jerking when lying down was reported. PSM was position dependent in 89 (50%) cases. Only 9 (5%) were rhythmic and 90 (50%) arrhythmic. In 19 patients (idiopathic and secondary) jerks continued during sleep, proven with polysomnography in several cases. In none of the FMD cases did jerks continue during sleep. Twelve patients (7%) (7 secondary, 5 FMD) had extension instead of flexion jerks and 5 (3%) had both. Absence of pain during jerks was reported in 73 (41%) and 4 cases (2%) were painful. PSM jerks started axially in the majority of cases (150, 84%) and in 28 (16%, predominantly FMD cases) they did not.

Clinical clues suggestive of FMD. Prior to 2008, few clues suggestive of FMD were mentioned in the case reports, with missing information in up to 54% of published cases (for details, see table 1).¹³ In brief, PSM started acutely in 89 (50%) cases (34% idiopathic, 54% secondary, 52% FMD), and in 65 cases (36%) it did not start acutely.⁹ Acute onset and rapid progression do not seem to be helpful clinical clues to distinguish PSM from FMD, as jerks often start acutely. Some atypical details were reported as well, such as cessation of jerks during an arithmetic task.^{22,29,47} Jerks were mentioned as distractible in

25 (14%) cases, including 8 (28%) idiopathic and 3 (7%) secondary cases. Rapid progression of jerks was reported in 14 cases (8%) only, predominantly based on 11 FMD cases. However, rapid clinical progression is not necessarily indicative of FMD, as traumatic cord compression or myelitis could also have a rapid clinical course. Spontaneous remissions were mentioned in 11 cases (6%), including 4 (9%) secondary cases and no idiopathic cases. Previous somatizations were exclusively mentioned in 47 (45%) FMD cases. Psychiatric comorbidity was noted in 2 (7%) idiopathic, 1 (2%) secondary, and 33 (32%) FMD cases. Variability of jerks over time was noted in half of the FMD cases and in 3 (10%) idiopathic and 5 (11%) secondary cases. Inconsistency of the jerks at the time of examination was reported in 57 cases (all FMD). Involvement of facial musculature was reported in 12 (12%) of the FMD cases and in 1 idiopathic and 2 secondary cases. The neck muscles were involved in 62 (35%), and in 48 (27%) they were not involved.

Tic characteristics. Premonitory sensations were reported in 6 idiopathic (21%) and 7 secondary PSM cases (15%), and in 15 (14%) of the FMD cases. Seventeen cases (4 secondary [9%] and 13 FMD [13%]) could suppress the jerks (1 partially). In 40% of cases, tic characteristics were unreported.

Electrophysiology. Polymyography is regarded as the essential diagnostic procedure for PSM.⁴ The conduction velocity needs to be slow in order to involve the propriospinal pathway but was unreported in half of the published cases (49%). Conduction velocity was slow in 62 cases (35%), of which 20 (69%) were idiopathic, 22 (48%) were secondary, and 20 (19%) were FMD. Rapid conduction velocity was reported in a single secondary case (2%) and 21 functional cases (20%).

EMG burst duration was short (<300 ms) in 34% of idiopathic, 37% of secondary, and 52% of FMD cases and unreported in 14% of cases. Both long and variable burst durations were reported in presumed organic cases. In 6 (21%) idiopathic cases, the burst duration was longer than 300 ms, in 8 (17%) secondary cases and in 21 (20%) FMD cases. A variable EMG burst duration was found in 10 (34%) idiopathic cases, in 11 (24%) secondary cases, and in 17 (16%) FMD cases. Synchronous contraction of agonists and antagonists was present in 4 (14%) idiopathic cases, 10 (22%) secondary cases, and 40 (38%) FMD cases. Thus, in the majority of patients reported to be either secondary or idiopathic PSM, the polymyography findings were not compatible with PSM, as originally described by Brown et al.,¹ Chokroverty et al.,⁴ and Nogues et al.³⁵ A BP was found in 65 cases (63%) of FMD etiology. The BP was reported to be absent in 80 cases (45%), of which

Table 1 Confirmed presence of clinical characteristics indicative of propriospinal myoclonus, functional movement disorder, or tics per cases as divided per etiology

	Presence of characteristics per etiology, n (%)				All cases (n = 179), n (%)		
	Idiopathic (n = 29)	Secondary		FMD (n = 104)	Total present	Total absent	Missing data/ unreported
		All (n = 46)	Structural (n = 12)				
Classic PSM characteristics							
Aggravated supine	25 (86)	25 (54)	3 (25)	27 (26)	77 (43)	29 (16)	73 (41)
Position dependent	26 (90)	34 (74)	6 (50)	29 (28)	89 (50)	18 (10)	72 (40)
Stimulus sensitive	8 (28)	31 (67)	9 (75)	7 (7)	46 (26)	59 (33)	74 (41)
Rhythmic	2 (7)	11 (24)	3 (25)	0 (0)	9 (5)	90 (50) ^a	70 (39)
In sleep persistent	6 (21)	13 (28)	2 (17)	0 (0)	19 (11)	72 (40)	88 (49)
Extension	0 (0)	7 (15)	0 (0)	5 (5)	12 (7)	129 (72) ^b	33 (18)
Painful	0 (0)	4 (9)	2 (17)	0 (0)	4 (2)	73 (41)	102 (57)
Starts axial	29 (100)	43 (94)	9 (75)	78 (75)	150 (84)	28 (16)	1 (1)
FMD characteristics							
Acute onset	10 (34)	25 (54)	4 (33)	54 (52)	89 (50)	65 (36)	25 (14)
Distractible	8 (28)	3 (7)	1 (8)	14 (13)	25 (14)	63 (35)	91 (51)
Rapid progression	0 (0)	3 (7)	1 (8)	11 (11)	14 (8)	68 (38)	97 (54)
Spontaneous remissions	0 (0)	4 (9)	0 (0)	7 (7)	11 (6)	165 (92)	3 (2)
Previous somatizations	0 (0)	0 (0)	0 (0)	47 (45)	47 (26)	69 (39)	63 (35)
Psychiatric comorbidity	2 (7)	1 (2)	0 (0)	33 (32)	36 (20)	80 (45)	63 (35)
Variability over time	3 (10)	5 (11)	2 (17)	53 (51)	61 (34)	101 (56)	17 (10)
Inconsistency in jerks at time of visit	0 (0)	0 (0)	0 (0)	57 (55)	57 (32)	83 (46)	39 (22)
Facial involvement	1 (3)	2 (4)	1 (8)	12 (12)	15 (8)	164 (92)	0 (0)
Neck involvement	24 (83)	32 (70)	6 (50)	6 (6)	62 (35)	48 (27)	69 (39)
Tic characteristics							
Premonitory sensation	6 (21)	7 (15)	1 (8)	15 (14)	28 (16)	80 (45)	71 (40)
Suppressible	0 (0)	4 (9)	0 (0)	13 (13)	17 (9)	91 (51)	71 (40)

Abbreviations: FMD = functional movement disorder; PSM = propriospinal myoclonus.

PSM was originally described as flexor and painless and the reverse are stated in the table to illustrate the equal distribution of these relatively rare and atypical PSM characteristics. The total number of reported cases is 179, divided into 29 cases of idiopathic, 46 of secondary (of which 12 cases had structural lesions), and 104 of FMD etiology. Percentages are expressed as fraction of the group under consideration. In addition, the total number present and absent per characteristics is listed of all cases irrespective of etiology. The final column lists the missing data, being the unreported absence or presence of the clinical characteristics. Percentages are rounded to the nearest integer.

^a Absent indicates nonrhythmic jerks; however, in 10 patients (6%), jerks were both rhythmic and arrhythmic (not included in the table).

^b Absent means flexion jerks; both flexion and extension jerks were present in 5 patients (3%) (not included in the table).

24 (83%) were idiopathic, 36 (78%) were secondary, and 20 (19%) were still diagnosed as FMD. The BP was unreported in 34 cases (19%). An overview of reported electrophysiology findings is in table 2.

Red flags for PSM with underlying structural lesion. No clear red flags emerged that were suggestive of secondary cases with structural lesions.^{1,2,23–25,31,35,39,40,46} FMD cases were never reported to have rhythmic jerks or jerks persisting during sleep; however, the latter needs polysomnographic verification. Details of the characteristics of the structural cases can be found in tables 1 and 2. Of note, in the cases with a structural lesion, additional clinical signs were

present such as optic neuritis, urinary urgency, unsteady gait, abnormal reflexes, and thoracic sensory changes.^{1,2,23–25,31,35,39,40,46} These signs warrant investigation for a possible myelopathy.

Imaging. Imaging is required to exclude spinal lesions. MRI scanning was reported in 101 (78%) cases. In 29 (22%) cases, it was reported that no MRI scan was performed. Twelve cases (7%) had a structural lesion, as mentioned in the Etiology section. MRI with diffusion tensor imaging (DTI) was performed in a study of 10 idiopathic PSM cases and the spinal generator matched the microstructural abnormalities on MRI-DTI in 7 cases but in 3 it did not match the

Table 2 Confirmed reported electrophysiologic criterion per cases as divided per etiology

Electrophysiologic criterion	Idiopathic (n = 29), n (%)	Secondary, n (%)		FMD (n = 104), n (%)	Total present (n = 179), n (%)	Missing data/unreported, n (%)
		All secondary (n = 46)	Structural (n = 12)			
Conduction velocity						
Slow (1–15 m/s)	20 (69)	22 (48)	5 (42)	20 (19)	62 (35) ^a	88 (49)
Rapid (>15 m/s)	0 (0)	1 (2)	0 (0)	21 (20)	22 (12)	88 (49)
EMG burst duration						
Short (<300 ms)	10 (34)	17 (37)	2 (17)	54 (52)	81 (45)	25 (14)
Long (>300 ms)	6 (21)	8 (17)	4 (33)	21 (20)	35 (20)	25 (14)
Variable (short and long)	10 (34)	11 (24)	2 (17)	17 (16)	38 (21)	25 (14)
Synchronicity						
Present	4 (14)	10 (22)	2 (17)	40 (38)	54 (30)	46 (26)
BP						
Present	0 (0)	0 (0)	0 (0)	65 (63)	65 (36)	34 (19)
Absent	24 (83)	36 (78)	8 (67)	20 (19)	80 (45)	34 (19)

Abbreviations: BP = Bereitschaftspotential; FMD = functional movement disorder.

The total number of reported cases is 179, divided into 29 idiopathic, 46 secondary (of which 12 cases had structural lesions), and 104 FMD cases. Percentages are expressed as fraction of the group under consideration. In addition, the total number per characteristics is listed for all cases irrespective of etiology. The final column lists the missing data, being the unreported absence or presence of the specific electrophysiologic criterion. Percentages are rounded to the nearest integer.

^a Axial jerks with a variable conduction velocity (both slow and rapid) were described in 7 patients (4%) (not included in the table).

presumed level of spinal generation as determined by the onset of the jerks in the EMG.^{39,40} An alternative explanation for spinal tract integrity disruption with DTI could be movement artefacts. The possibility that the spinal cord microabnormalities detected on DTI imaging are consequential rather than causative of the jerks cannot be excluded and it would be of interest to repeat this study in patients with axial jerks with clear FMD.

Follow-up and therapy. Follow-up was reported in 126 (70%) cases (mean duration 30 months). PSM resolved in 41 (23%) cases (23 FMD [22%], 7 idiopathic [24%], 11 secondary [11%]). The distribution of clinical resolution is about equal between the idiopathic PSM and FMD. A relapse was observed in 24 (13%) cases. Treatment of PSM was predominantly with clonazepam, which was administered in 65 cases and was effective in 34 (52%). Botulinum toxin was applied and effective in 20 FMD cases. Selective serotonin reuptake inhibitors seem to be effective sporadically.⁵² Valproate was reported to be effective in 6 but ineffective in 10 cases. Additional administered drugs included carbamazepine, prednisone, and baclofen, and were mostly reported as ineffective. Other therapies included surgery of spinal lesions or discectomy, for instance Shprecher et al.⁴³ recently described the almost complete resolution of PSM after anterior cervical discectomy with fusion.^{1,20,24} In isolated cases, removal of surgical screws,¹³ plasmapheresis,⁴⁷ continuous positive airway pressure,³⁷ and a gluten-free diet⁵⁵ were reported

helpful. In FMD, the transient therapeutic response to electrical nerve stimulation⁵¹ and effect of cognitive behavioral therapy⁴⁴ prompted the diagnosis of FMD. In selected articles, patients with idiopathic PSM benefited from unconventional therapy, such as autogenic biofeedback training⁴⁵ and transcutaneous electrical nerve stimulation.³⁰

DISCUSSION AND RECOMMENDATIONS The current review demonstrates a lack of strict clinical and electrophysiologic criteria for PSM. The etiology, clinical characteristics, and EMG findings were heterogeneous.

The focus of the initial articles on PSM was on the presumed underlying etiologies and (structural) causation. The recent attention to FMD has doubled the number of reported cases and currently, the majority of cases (57%) are reported as FMD. The secondary etiologies of PSM include cervical lesions, e.g., spinal cord compression, tumors, and infections, but structural lesions were found in a small fraction (7%) of reported cases. This review tried to separate structural from nonstructural secondary cases. Causality of the structural lesions is likely. It is important to realize that in the cases with a structural lesion additional neurologic features point toward myelopathy, i.e., urinary urgency, unsteady gait, abnormal reflexes, and thoracic sensory changes.^{1,2,23–25,31,35,39,40,46} In a case with a medical history of bilateral optic neuritis, a demyelinating disorder was suspected.⁴⁶

Table 3 Proposed diagnostic criteria for idiopathic and secondary propriospinal myoclonus

Diagnostic category	Criteria
Idiopathic PSM	• Axial jerks without functional clinical clues in medical history or at neurologic examination
	• Normal imaging of the spinal axis (no evidence of myelopathy)
	• Consistent EMG pattern
	• Absent BP
Secondary PSM	• Axial jerks without functional clinical clues in medical history or at neurologic examination
	• Abnormal imaging of the spinal axis that is anatomically consistent with EMG pattern (recruitment order and onset)
	• Absent BP
	Clinical clues
Functional movement disorder	• Axial jerks with functional clinical clues in medical history (e.g., previous somatizations, spontaneous remissions) or at neurologic examination (importantly, distractibility and suppressibility)
	• Facial movements or vocalizations
	• Normal imaging of the spinal axis (no evidence of myelopathy)
	• Inconsistent EMG pattern or presence of BP

Abbreviations: BP = Bereitschaftspotential; FMD = functional movement disorder; PSM = propriospinal myoclonus. For idiopathic PSM, all criteria are required. For the diagnosis of FMD, clinical clues instead of criteria are listed indicating classification of axial jerks as FMD. Not all clues need to be present for the diagnosis of FMD.

In the nonstructural secondary cases, the causal relationship with the onset of PSM is less clear. Notably, physical precipitating factors (injury, illness) are common in patients with FMD.^{58,59} Some of the described secondary and idiopathic PSM could be considered as FMD, but based on the available information this remains speculative.

Several typical characteristics for PSM are noted in the literature, such as posture dependency, stimulus sensitivity, painless jerks, and axial initiation of the jerks, in this review not specific for any etiology. Regarding typical characteristics for PSM, rhythmicity of the jerks was never reported present in FMD cases. Axial jerks never persisted during sleep in FMD. In case FMD is considered, self-reports of the presence of jerks during sleep are not always reliable and we advise to objectify reported presence with polysomnography.

Regarding the evaluation of typical characteristics for FMD in published cases, this review demonstrates clinically overlapping features as well. However, we caution against taking the FMD clinical signs as a literal checklist as these clinical signs are merely indicative of FMD and it has been shown that the interrater agreement of the diagnosis of FMD is low based on these clinical signs alone.⁶⁰

Although patients with PSM are not typically considered to have a tic or functional disorder, idiopathic and secondary PSM cases were often reported to have atypical clinical symptoms such as grimacing, tonic contractions of the eye musculature, and “peculiar sensations of pressure and tingling migrating upward from the groin to the abdomen, trunk, and neck”

prior to the jerks.^{22,40} Along with the recent interest in functional characteristics in PSM, we noticed overlapping clinical features with tic disorders.⁷ We recommend clinicians to pay attention to facial movement or vocalizations in patients with axial jerks, as these do not concur with a spinal etiology and could indicate FMD.

As clinical presentations are heterogeneous and both PSM and FMD clinical clues can be present in individual cases, we advise additional electrophysiologic examination in all cases with axial jerks.

Identification of electrophysiologic characteristics specific for PSM is difficult. Overall, it is important to note that in most cases electrophysiologic details are missing, hampering comparison between cases. The duration of bursts is usually long but highly variable (between 16 and 5,000 ms). Synchronicity of contraction of agonist and antagonist muscles does not appear to be a prerequisite for PSM, as it is almost equally present and absent in reported cases. Of note, only in half of the cases are synchronicity and conduction velocities reported. A slow conduction velocity over the spinal cord has been suggested to indicate involvement of polysynaptic propriospinal pathways, but these velocities can also be found in voluntarily mimicked PSM and in 19% of published FMD cases.¹⁵ Still, slow conduction velocities were present in more than twofold of the remaining idiopathic and secondary (including structural) cases.

Jerk-locked back-averaging of simultaneous EEG–EMG was unreported in 19% of published cases. The BP was reported present in 63% of FMD cases, and reported absent in 19% of cases. This is in line with

our recent report on the diagnostic value of the BP in a separate clinical cohort indicating that the BP is essential in supporting a presumed FMD etiology, but an absent BP does not exclude FMD.¹¹

A limitation of the current review is report bias so that absence or presence of certain clinical or electrophysiologic features may be unreported or not performed at all, in particular BP testing and imaging. In the current review, lack of performance of imaging may lead to unambiguous assignment of cases to idiopathic PSM instead of secondary PSM. Future studies of prospectively included patients are needed to further elucidate the clinical and electrophysiologic features and their sensitivity and specificity for specific subtypes of PSM.

Suggested diagnostic criteria. Based on the current review, we suggest dividing axial jerks into 3 categories: idiopathic PSM, secondary PSM, and FMD. As clinical characteristics of PSM are heterogeneous and nonspecific, we recommend as a first step to clinically evaluate the presence of myelopathy or other neurologic symptoms such as optic neuropathy. We recommend conducting MRI of the spinal cord, especially to rule out myelopathy. If normal, we recommend conducting polymyography and coregistration of EEG–EMG for a BP. Polymyography is easily performed but might be of limited use, as only variability in muscle recruitment is indicative of FMD. Additional coregistration of EEG–EMG can evaluate the presence of a BP and is an important part of the diagnostic workup and superior to a polymyography, but not all institutions may have the facilities to conduct simultaneous EEG–EMG recordings. We would only consider a case idiopathic PSM if clinical evaluation is not indicative of a functional origin, combined with normal imaging of the spinal axis, consistent EMG pattern, plus an absent BP. Table 3 summarizes the suggested diagnostic criteria. We recommend clinical caution in patients in whom the BP is absent or not recordable, for instance due to a high jerk frequency or too much movement artefact. In such cases, imaging of the spinal axis is advised and follow-up should be performed in order to avoid misdiagnosis.

AUTHOR CONTRIBUTIONS

S.M.A.v.d.S. and M.A.J.T. conceived and designed the study. R.E. supplied additional patient data. S.M.A.v.d.S. and T.v.d.E. performed statistical analysis and interpreted the data. S.M.A.v.d.S. drafted the manuscript. R.E., C.C., M.J.E., J.H.T.M.K., K.P.B., A.-F.v.R., P.B., and M.A.J.T. revised the manuscript. All authors approved the final version to be submitted.

STUDY FUNDING

No targeted funding reported.

DISCLOSURE

S. van der Salm and R. Erro report no disclosures relevant to the manuscript. C. Cordivari received honoraria to support educational activities

from Ipsen. M. Edwards received honoraria for travel and speaking from the International Parkinson and Movement Disorders Society and UCB Pharma. He receives grant support from the Bachman-Strauss Foundation, the UK Dystonia Society, and NIHR. He is also supported by the UCL/UCLH Biomedical Research Centre. J. Koelman received educational grants from Allergan and Ipsen and a research grant from Ipsen. T. van den Ende reports no disclosures relevant to the manuscript. K. Bhatia received funding for travel from GlaxoSmithKline, Orion Corporation, Ipsen, and Merz Pharmaceuticals, LLC; serves on the editorial boards of *Movement Disorders* and *Therapeutic Advances in Neurological Disorders*; receives royalties from the publication of *Oxford Specialist Handbook of Parkinson's Disease and Other Movement Disorders* (Oxford University Press, 2008); received speaker honoraria from GlaxoSmithKline, Ipsen, Merz Pharmaceuticals, LLC, and Sun Pharmaceutical Industries Ltd.; received personal compensation for scientific advisory board for GSK and Boehringer Ingelheim; received research support from Ipsen and from the Halley Stewart Trust through Dystonia Society UK; and received the Wellcome Trust MRC strategic neurodegenerative disease initiative award (ref. WT089698), a grant from the Dystonia Coalition, and a grant from Parkinson's UK (ref. G-1009). A. Van Rootselaar reports no disclosures relevant to the manuscript. P. Brown received an honorarium from Medtronic. M. Tijssen is funded by STW Technology Society–NeuroSIPE, Netherlands Organization for Scientific Research–NWO Medium, Fonds Nuts-Ohra, Prinses Beatrix Fonds, Gossweiler Foundation, Stichting Wetenschapsfonds Dystonie Vereniging, and educational grants from Ipsen, Allergan, and Medtronic. Go to Neurology.org for full disclosures.

Received March 4, 2014. Accepted in final form August 11, 2014.

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