



Movement disorders phenomenology in focal motor seizures

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ABSTRACT

Introduction: Although focal motor seizures may resemble one or more movement disorders their phenomenology and prevalence remain uncertain.

Methods: To examine the extent to which focal motor seizures can present with a phenomenology fulfilling diagnostic criteria for movement disorders, 100 consecutive patients with focal motor seizures were rated by movement disorders experts, epileptologists, and general neurologists.

Results: A focal motor seizure phenomenologically manifested as a defined movement disorder in 29% of the patients from a consecutive video-EEG documented cohort as per consensus among experts: myoclonus and dystonia (10 and 9 cases, respectively) were the most common movement disorders, followed by chorea (4), stereotypies (3) myoclonus-dystonia (2), and tremor (1).

Conclusions: Movement disorders and focal motor epilepsy share overlapping movement phenomenology.

1. Introduction

An epileptic seizure is defined as “a transient occurrence of signs and/or symptoms due to abnormal excessive or synchronous neuronal activity in the brain” [1]. Since the diagnosis and classification of a movement disorder relies on phenomenology, a motor seizure can clinically resemble one or more movement disorders. This clinical overlap is reflected by an inconsistent use of nomenclature such as myoclonic and dystonic crises and, more recently, ‘faciobrachial dystonic seizure’, a manifestation of LGI-1 antibody encephalitis [2].

We sought to examine the extent to which focal motor seizures can present with a phenomenology fulfilling diagnostic criteria for one or more movement disorders using a series of video electroencephalography (vEEG)-confirmed focal motor seizures.

2. Methods

Epilepsy cohort and video-EEG. A series of 100 consecutive patients with vEEG-confirmed focal motor seizures of different etiologies (Table 1) [3] were referred to the Epilepsy Unit in the Department of

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Table 1
Demographic and clinical features of the patients.

Sex, n, M/F	50/50
Age at baseline, mean [range], years	48.0 [17–85]
Epilepsy etiology	
Focal structural/immune/infectious	34
- Malformation	9
- Immune-mediated (autoimmune)	6
- Neoplasm	4
- Vascular	4
- Post-traumatic	3
- Infective	2
- Neurocutaneous	2
- Perinatal hypoxia	1
- Hippocampal sclerosis	1
- Multiple sclerosis	1
- Hydrocephalus	1
Focal of unknown etiology	18
Focal genetic ('idiopathic')	8
Undetermined whether focal or generalized epilepsy	4
Epileptic encephalopathy	5
Reflex	3
Status epilepticus/seizure cluster	
In epileptic patients	2
Acute symptomatic	24
- Vascular	18
- Neoplasm	4
- Infectious	1
- Immuno-mediated	1
Unknown etiology	2

Human Neurosciences of Sapienza University, Rome. All patients included in the study underwent vEEG monitoring (Telefactor, Micromed System Plus and Xltek devices, 21 Channels, International 10–20 System), during the pre-ictal, ictal and post-ictal periods. Seizures and epileptic syndromes were categorized according to the new classification of the International League Against Epilepsy [4,5]. EEG tracings, clinical records and neuroimaging studies were reviewed by three epileptologists not included among the EPI cohort.

Raters. Videos were rated by three groups of physicians: five movement disorders experts (MDE), four epileptologists (EPI), and four general neurologists (GEN) (Supplemental Table 1). A standardized datasheet was used for rating body sites and movement disorders. Tremor, dystonia and stereotypies were defined according to recently published criteria [6–8]; other movement disorders were diagnosed according to expert-derived definitions [9]. Raters received no clinical information with the videos. In cases containing multiple episodes, raters were asked to rate each episode separately.

2.1. Statistical analysis

Values were expressed as mean \pm SD or number (%), as appropriate. Inter-rater agreement was calculated with Cohen's kappa. The level of agreement was evaluated according to the six levels of Landis and Koch [10]. Comparisons between the continuous variables were performed using ANOVA followed by Student *t*-test for unpaired sample as post-hoc. Comparisons between the distributions of categorical variables were performed by Fisher's exact test. A *p*-value $<$ 0.05 was deemed statistically significant. All analyses were performed using the Statistical Package for the Social Sciences (SPSS) version 20.0 for Mac.

3. Results

The three groups of raters identified a similar number of focal motor episodes: 1.4 ± 0.6 , 1.2 ± 0.6 and 1.3 ± 0.5 for MDE, EPI and GEN, respectively (*p* $>$ 0.15). Within each group, the inter-rater agreement of the number and type of involved body sites and the movement disorders classification was poor with the exception of upper limb

Table 2

Inter-rater agreement among neurologist groups; bold-typed values indicate 'moderate' agreement (remaining ones were 'poor').

	MDE		EPI		GEN	
	κ	Z	κ	Z	κ	Z
N. of body sites	0.2871	9.65	0.1846	7.07	0.2400	7.92
Face	0.3452	15.38	0.2960	13.08	0.1668	7.47
Neck	0.3644	12.17	0.1372	4.82	0.1966	7.97
Trunk	0.2457	10.17	0.1693	5.41	0.1402	5.73
UL	0.4073	20.51	0.3202	15.75	0.3513	17.72
LL	0.3672	15.22	0.3186	12.33	0.5282	20.47

Combined kappa, all assessment with *p* $<$ 0.00001. Abbreviations: EPI: epileptologists; GEN: recently board certificated neurologists; MDE: movement disorders experts.

diagnosis among MDE and lower limb diagnosis among GEN, where it was moderate (0.41 and 0.53 respectively; Table 2).

Recognition of movement disorders by MDE. Because of insufficient inter-rater agreement, we conservatively restricted further analyses on episodes with full agreement among MDE ($\kappa = 1$). These 29 cases (29%, Table 3) were characterized as: myoclonus (10, 34.5% of this sample – Videos 1 and 2), dystonia (9, 31.0% – Videos 3–6), dyskinesias (4, 13.8% – Videos 7 and 8), stereotypies (3, 10.3%), myoclonus-dystonia (2, 6.9%), and tremor (1, 3.4%).

Supplementary video related to this article can be found at <https://doi.org/10.1016/j.parkreldis.2018.10.021>.

Upper limb was most commonly involved (17 episodes) followed by face (9), neck and lower limb (4 each), shoulder (2), and trunk (1). Symptoms were unilateral in all episodes except eight (three cases of 'blepharospasm', 2 lower-limb dyskinesias, oromandibular dystonia, antecollis and upper-limb stereotypy, 1 case each; Table 3).

The remaining 71 patients were diagnosed by MDE as affected by a movement disorder by all raters in 36 cases, by 4 raters in 11, by 3 in 14, by 2 in 9 and by 1 in 1. Myoclonus was the most common nosology (101 times) followed by dystonia (86), stereotypies (36), dyskinesias (35), tic (16) and tremor (11).

Recognition of movement disorders by EPI and GEN. In the 29 patients with full agreement among MDE, consensus was obtained in 4/29 and 5/29 patients among EPI and GEN, respectively. Five categories were concordant with the MDE for all cases for EPI: hyperekplexia, upper limb dystonia, upper limb myoclonus, and hemi-facial spasm; three cases for GEN: lower limb myoclonus and two cases of upper limb myoclonus. Despite an over-recognition of stereotypies (8/29 episodes, Table 3), GEN reached full consensus in only two of such patients.

EEG. In five of the patients with bilateral manifestations there was bi-hemispheric EEG activity (8, 15, 49, 52 and 79), also found in three with unilateral phenomenology (22, 28 and 34). Nine episodes originated in the left hemisphere, 13 in the right hemisphere (Table 3). Eight patients had status epilepticus with predominant motor signs. There was no correlation between the two most common movement disorders (myoclonus and dystonia) and either location or pattern of EEG abnormalities (Fig. 1).

4. Discussion

In this series, focal motor seizures phenomenologically manifested as recognizable movement disorders in 29% of the patients. However, the low inter-rater agreement suggested substantial difficulties in the application of existing diagnostic criteria to patients with involuntary movements caused by an epileptic seizure. The poor agreement among observers might also suggest that current definitions of movement disorders cannot be considered as diagnostic criteria too, and validated diagnostic guidelines are needed. Nevertheless, these difficulties may also be related to the expertise of the raters as EPI and GEN reached an

Table 3
Epileptic patients diagnosed as movement disorders by all MDE (definitions according to [6–9]). Bold-types text indicates at least 75% concordance among EPI and GEN with MDE diagnoses.

Case	Etiology	Diagnosis						
		Category	In MDE	In EPI (n of raters)**	In GEN (n of raters)**	Side	Onset	Spreading
1	Symptomatic (UK)	Myoclonus	Hyperkplexia	Hyperkplexia (3)	Hyperkplexia (2)	L	Parietal	Temporal
8	Symptomatic (UK)	Dystonia	Blepharospasm	Myoclonus (2)	HFS (1)	Bil	Frontal	Parietal
11	Symptomatic* (AVM)	Myoclonus	HFS	HFS (1)	HFS (2)	R	Fronto-Parietal	Frontal
15	Symptomatic (UK)	Dyskinesias	Copulatory dyskinesias and lower limbs tremor	None	None	Bil	Fronto (Insular)-Temporal	Fronto (Insular)-Temporal
18	Cryptogenic	Dystonia	Blepharospasm	Myoclonus (2)	Tics (1), myoclonus (1)	R	Frontal	Frontal
20	Cryptogenic	Dystonia	Oromandibular dystonia	None	Stereotypies (1), HFS (1)	L	Frontal	Frontal
22	Cryptogenic	Dystonia	Upper limb dystonia	Dystonia (3)	Dystonia (2)	Bil	UK	Temporal (mesial)
28	Idiopathic	Myoclonus	Limbs oscillatory myoclonus	Myoclonus (1)	Myoclonus (2)	Bil	Fronto-Temporal	Fronto-Temporal
34	Symptomatic (UK)	Myoclonus	Lower limb myoclonus	Myoclonus (1)	Myoclonus (3)	Bil	Frontal	Frontal
35	Symptomatic* (UK)	Stereotypies	Upper limb stereotypies	None	Stereotypies (1)	R	Frontal	Frontal
42	Symptomatic (meningioma)	Dystonia	Torticollis with upper limb dystonia	None	Stereotypies (1), myoclonus (1), dystonia (1)	R	Frontal	Temporal (anterior)
44	Symptomatic (cortical dysplasia)	Dystonia	Blepharospasm	Dystonia (1)	Stereotypies (1)	R	Parietal	Parietal
49	Symptomatic (cortical dysplasia)	Dystonia	Antecollis	None	Hyperkplexia (1), ballism (1)	Bil	Parietal	Frontal
52	Symptomatic* (UK)	Stereotypies	Bilateral upper limb stereotypies	None	Stereotypies (3)	Bil	Occipital	Parieto-Temporal (posterior)
63	Cryptogenic	Myoclonus	Upper limb myoclonus	Myoclonus (1)	Myoclonus (2)	R	Frontal (Insular)	Frontal (Insular)
66	Cryptogenic	Myoclonus	Upper limb myoclonus	Myoclonus (3)	Myoclonus (3)	L	Frontal (Rolandic)	Frontal (Rolandic)
79	Symptomatic (CP)	Dyskinesias	Copulatory dyskinesias	None	None	Bil	Parietal	Frontal (L)
82	Symptomatic* (hemorrhagic)	Dyskinesias	Upper limb chorea-athetosis	Dyskinesias (1)	Myoclonus (2)	L	Frontal	Frontal
83	Symptomatic (UK)	Myoclonus	HFS	HFS (2)	HFS (1)	R	Temporal (posterior)	Parietal
86	Symptomatic* (UK)	Myoclonus	HFS	HFS (3)	Myoclonus (1)	L	Fronto-Parietal-Temporal-Occipital	Parietal
87	Symptomatic (cortical dysplasia)	Stereotypies	Upper limb stereotypies	None	Stereotypies (2)	L	Frontal	Frontal
88	Symptomatic* (UK)	Tremor	Upper limb tremor	Tremor (1)	Tremor (1), myoclonus (1), dystonia (1)	R	Parietal	Frontal (Rolandic)
90	Symptomatic (UK)	Dyskinesias	Upper limb chorea-athetosis	None	Stereotypies (2)	R	Parietal	Frontal (Rolandic)
91	Cryptogenic*	Myoclonus dystonia	HFS and hand dystonic tremor	HFS (1)	HFS and hand dystonic tremor (1), facio-brachial dystonia (1)	L	Frontal (Insular)	Frontal (Insular)
92	Symptomatic (cortical dysplasia)	Myoclonus dystonia	HFS and hand dystonic tremor	HFS (1)	HFS and hand dystonic tremor (1), brachial dystonia (1)	L	Frontal (Insular)	Temporal
93	Cryptogenic	Myoclonus	Shoulder myoclonus	Myoclonus (2)	Myoclonus (2)	R	Frontal	Frontal
95	Symptomatic (NF)	Myoclonus	Upper limb myoclonus	Myoclonus (2)	Myoclonus (4)	R	Frontal	Temporal (mesial)
99	Idiopathic*	Dystonia	Torticollis with upper limb dystonia	Torticollis with upper limb dystonia (1)	Torticollis with upper limb dystonia (2)	L	Frontal (Insular)	Frontal (Insular)
100	Symptomatic (Goldenhhar syndrome)	Dystonia	Torticollis with upper limb dystonia	Torticollis with upper limb dystonia (1)	Dystonia (2)	R	Fronto-Temporal	Fronto-Temporal

Abbreviations*: status epilepticus; **: the most common diagnosis is listed; AVM: arteriovenous malformation; Bil: bilateral; EEG: electroencephalogram; EPI: epileptologist; GEN: recently board certified neurologists; HFS: hemifacial spasm; CP: cerebral palsy; L: left; MDE: movement disorders experts; NF: neurofibromatosis; R: right; UK: unknown.

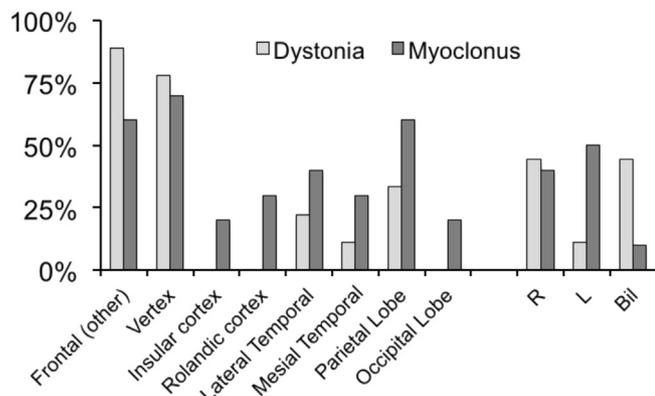


Fig. 1. Electrographic localization in the two most common movement disorders. Three epileptologists not part of the blind EPI cohort reviewed EEGs for morphology, frequency, duration and topography, which were defined for electro-clinical correlations.

acceptable consensus very rarely, only characterizing hemifacial spasm and myoclonus better than other involuntary movements.

4.1. Myoclonus

Predictably given the cortical localization, the majority of seizures by MDE fulfilled the diagnostic criteria for myoclonus [11]. This was the case particularly for unilateral and appendicular movements involving one limb and/or one hemi-face (6/8 of these seizures had a focus in the frontal lobe). Indeed, a focal seizure can be mistaken for hemifacial spasm, which is particularly challenging in cases of *epilepsia partialis continua* [12,13]. In some, the epileptic spread to neighboring brain regions and may have contributed to changes of semiology from myoclonus to dystonic postures (e.g., 91 and 92). Most cases of unilateral myoclonus (10/11) were associated with a contralateral discharge (Table 3) [14]. In a few cases, seizures resembled a myoclonus-dystonia phenotype due to the coexistence of these two disorders. However, unlike myoclonus-dystonia, the face was often affected. This relationship is complicated by the rarely reported association between genetically confirmed myoclonus-dystonia and generalized seizures [15–17].

One of the myoclonic patients fit clinical criteria for hyperekplexia, which is usually caused by a brainstem generator but can rarely be the manifestation of a reflex seizure caused by bilateral discharges in the mesial frontal lobe, as seen in our patient (so-called ‘startle epilepsy’) [18].

4.2. Dystonia

The second largest group of patients included patients with tonic contractions resembling dystonia. ‘Ictal dystonia’ has been described as being characterized by a rotatory component of the hand (into extreme pronation or supination) in the context of temporal lobe epilepsy [19,20]. Associated head deviation and the so-called ‘M2e’ are other classical epileptic phenotypes [14]. These cases (42, 99 and 100) have frontal or parietal focus, exclusively unilateral, in keeping with the good lateralization value reported for this type of seizure [14]. These episodes were sustained, lasting much longer than faciobrachial dystonic seizures [2]. One case (22, Video 4) had isolated upper limb dystonia, which was associated with bilateral discharges arising from the parietal lobe. The role of the parietal lobe is difficult to ascertain in light of the widespread projections of parietal neurons, known to be responsible for inaccurate localization readings on scalp EEG [21].

We identified patients with bilateral dystonia: antecollis, oromandibular dystonia and blepharospasm, which were caused by bilateral discharges (in absence of consciousness impairment) involving

the frontal lobe, as previously recognized [22]. Although antecollis has been reported as a non-epileptic feature in patients with Dravet syndrome [23], herein we observed an epileptic tonic head flexion (49, Video 5), also different from the much faster and well-known epileptic head drop [24]. Ictal semiology comparable to blepharospasm was observed in patients with eyelid myoclonia. Although a proper electrophysiological classification of these movements is lacking [25,26], more sustained eyelid contractions can challenge its recognition from blepharospasm (Video 3).

4.3. Other movement disorders

Four patients (13.8%) had choreiform movements, involving both lower limbs (15 and 79, Video 7) or one upper limb (82 and 90, Video 8), as previously reported [27,28]. Three cases (35, 52, and 87) presented with stereotypies associated with frontal discharges, in keeping with the notion that frontal seizures can manifest with complex gestures, which may appear integrated (quasi-naturalistic) [22]. Finally, case 88 was labeled as having tremor in light of the rhythmic muscle activation of one upper limb, similar to episodes of paroxysmal ‘oscillatory myoclonus’ mimicking tremor [29].

This study has some limitations, namely among them the low inter-rater agreement, which prompted us to adopt a more conservative approach. Other limitations include the lack of electrophysiological assessments other than EEG (e.g., EMG) and the limitations inherent to EEG resolution, which might not always identify the epileptogenic zone itself or the involvement of subcortical structures for discharge propagation or “liberation” phenomena [21].

In conclusion, movement disorders and epilepsy are overlapping nosological entities manifesting along a phenomenological and a pathophysiological (and instrumental-based) continuum. As a consequence they may share both phenomenology and pathophysiological bases, in line with the modern conception of neurological diseases as network dysregulations [30]. Our study systematically explored these overlaps and assisted in the differential diagnosis between focal seizures and paroxysmal movement disorders. Although the episodic nature and additional signs (such as mouth automatisms or impaired awareness) can help in the diagnostic process, clinicians should recognize that some movement disorders are paroxysmal and some focal seizures are continuous, such as in focal status epilepticus or *epilepsia partialis continua* [13].

As a consequence, patients with common movement disorders need to be approached in a judicious manner because of the differing therapeutic implications (e.g., the use of antiepileptic drugs instead of botulinum neurotoxin in a patient with blepharospasm or hemifacial spasm of epileptic nature).

Altogether, our observations emphasize the crucial role of video-EEG and the importance of a multidimensional diagnostic approach, highlighting the need for improved phenomenological classification of these motor manifestations. Further studies on the abnormalities of movement in epileptic disorders will contribute to a better understand of the overlap between epilepsy and movement disorders and may contribute to a revision of criteria for movement classification and nosology.

Conflicts of interest

None.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.parkreldis.2018.10.021>.

References

- [1] R.S. Fisher, W. van Emde Boas, W. Blume, C. Elger, P. Genton, P. Lee, J. Engel Jr., Epileptic seizures and epilepsy: definitions proposed by the international League against epilepsy (ILAE) and the international bureau for epilepsy (IBE), *Epilepsia* 46 (2005) 470–472.
- [2] S.R. Irani, A.W. Michell, B. Lang, P. Pettingill, P. Waters, M.R. Johnson, J.M. Schott, R.J. Armstrong, S.Z. A, A. Bleasel, E.R. Somerville, S.M. Smith, A. Vincent, Faciobrachial dystonic seizures precede Lgi1 antibody limbic encephalitis, *Ann. Neurol.* 69 (2011) 892–900.
- [3] J. Engel Jr., The etiologic classification of epilepsy, *Epilepsia* 52 (2011) 1195–1197 discussion 1205–1199.
- [4] R.S. Fisher, J.H. Cross, J.A. French, N. Higurashi, E. Hirsch, F.E. Jansen, L. Lagae, S.L. Moshe, J. Peltola, E. Roulet Perez, I.E. Scheffer, S.M. Zuberi, Operational classification of seizure types by the International League against Epilepsy: position paper of the ILAE commission for classification and terminology, *Epilepsia* (2017).
- [5] R.S. Fisher, An overview of the 2017 ILAE operational classification of seizure types, *Epilepsy Behav.* 70 (2017) 271–273.
- [6] K.P. Bhatia, P. Bain, N. Bajaj, R.J. Elble, M. Hallett, E.D. Louis, J. Raethjen, M. Stamelou, C.M. Testa, G. Deuschl, P. Tremor, Task Force of the International, S. Movement Disorder, Consensus Statement on the Classification of Tremors, from the Task Force on Tremor of the International Parkinson and Movement Disorder Society, *Mov Disord* (2017).
- [7] A. Albanese, K. Bhatia, S.B. Bressman, M.R. Delong, S. Fahn, V.S. Fung, M. Hallett, J. Jankovic, H.A. Jinnah, C. Klein, A.E. Lang, J.W. Mink, J.K. Teller, Phenomenology and classification of dystonia: a consensus update, *Mov. Disord.* 28 (2013) 863–873.
- [8] M.J. Edwards, A.E. Lang, K.P. Bhatia, Stereotypies: a critical appraisal and suggestion of a clinically useful definition, *Mov. Disord.* 27 (2012) 179–185.
- [9] S. Fahn, J. Jankovic, M. Hallett, Clinical overview and phenomenology of movement disorders, in: S. Fahn, J. Jankovic, M. Hallett (Eds.), *Principles and Practice of Movement Disorders*, Elsevier Inc, Edinburgh London New York Oxford Philadelphia St Louis Sydney Toronto, 2011.
- [10] J.R. Landis, G.G. Koch, The measurement of observer agreement for categorical data, *Biometrics* 33 (1977) 159–174.
- [11] P. Brown, M.C. Ridding, K.J. Werhahn, J.C. Rothwell, C.D. Marsden, Abnormalities of the balance between inhibition and excitation in the motor cortex of patients with cortical myoclonus, *Brain* 119 (Pt 1) (1996) 309–317.
- [12] C. Deluca, G. Tommasi, G. Moretto, A. Fiaschi, M. Tinazzi, Focal motor seizures mimicking hemifacial spasm, *Park. Relat. Disord.* 14 (2008) 649–651.
- [13] A.J. Espay, V.J. Schmthorst, J.P. Szaflarski, Chronic isolated hemifacial spasm as a manifestation of epilepsy partialis continua, *Epilepsy Behav.* 12 (2008) 332–336.
- [14] A. Marashly, A. Ewida, R. Agarwal, K. Younes, H.O. Luders, Ictal motor sequences: lateralization and localization values, *Epilepsia* 57 (2016) 369–375.
- [15] E.M. Foncke, C. Klein, J.H. Koelman, P.L. Kramer, K. Schilling, B. Muller, J. Garrels, P. de Carvalho Aguiar, L. Liu, A. de Froe, J.D. Speelman, L.J. Ozelius, M.A. Tijssen, Hereditary myoclonus-dystonia associated with epilepsy, *Neurology* 60 (2003) 1988–1990.
- [16] S. O’Riordan, L.J. Ozelius, P. de Carvalho Aguiar, M. Hutchinson, M. King, T. Lynch, Inherited myoclonus-dystonia and epilepsy: further evidence of an association? *Mov. Disord.* 19 (2004) 1456–1459.
- [17] K. Haugarvoll, C. Tzoulis, G.T. Tran, B. Karlsen, B.A. Engelsen, P.M. Knappskog, L.A. Bindoff, Myoclonus-dystonia and epilepsy in a family with a novel epsilon-sarcoglycan mutation, *J. Neurol.* 261 (2014) 358–362.
- [18] S. Fernandez, A. Donaire, I. Maestro, E. Seres, X. Setoain, N. Bargallo, J. Rumia, T. Boget, C. Falcon, M. Carreno, Functional neuroimaging in startle epilepsy: involvement of a mesial frontoparietal network, *Epilepsia* 52 (2011) 1725–1732.
- [19] P. Kotagal, H. Luders, H.H. Morris, D.S. Dinner, E. Wyllie, J. Godoy, A.D. Rother, Dystonic posturing in complex partial seizures of temporal lobe onset: a new lateralizing sign, *Neurology* 39 (1989) 196–201.
- [20] H.Y. Yu, C.H. Yiu, D.J. Yen, C. Chen, Y.C. Guo, S.Y. Kwan, Y.Y. Lin, Y.H. Shih, Lateralizing value of early head turning and ictal dystonia in temporal lobe seizures: a video-EEG study, *Seizure* 10 (2001) 428–432.
- [21] A.J. Ristic, A.V. Alexopoulos, N. So, C. Wong, I.M. Najm, Parietal lobe epilepsy: the great imitator among focal epilepsies, *Epileptic Disord.* 14 (2012) 22–31.
- [22] P. Chauvel, A. McGonigal, Emergence of semiology in epileptic seizures, *Epilepsy Behav.* 38 (2014) 94–103.
- [23] A. Fasano, F. Borlot, A.E. Lang, D.M. Andrade, Antecollis and levodopa-responsive parkinsonism are late features of Dravet syndrome, *Neurology* 82 (2014) 2250–2251.
- [24] E. Antelmi, G. Plazzi, R. Erro, P. Tinuper, B. Balint, R. Liguori, K.P. Bhatia, Intermittent head drops: the differential spectrum, *J. Neurol. Neurosurg. Psychiatry* 87 (2016) 414–419.
- [25] R.H. Caraballo, E. Fontana, F. Darra, S. Chacon, N. Ross, E. Fiorini, N. Fejerman, B. Dalla Bernardina, A study of 63 cases with eyelid myoclonia with or without absences: type of seizure or an epileptic syndrome? *Seizure* 18 (2009) 440–445.
- [26] P.O. da Conceicao, M.S. Guaranha, C.G. Uchida, K. Carvalho, L.M. Guilhoto, G.M. De Araujo-Filho, H.C. Junior, P. Wolf, E.M. Yacubian, Blinking and eyelid myoclonia: characteristics and correlations of eyelid movements, *Seizure* 24 (2015) 12–16.
- [27] J. Chorobski, T. Bacia, On the coexistence of epileptic seizures and abnormal involuntary movements, *J. Neurol. Neurosurg. Psychiatry* 24 (1961) 151–157.
- [28] T.W. van Strien, A.F. van Rootselaar, A.A. Hilgevoord, W.H. Linszen, A.J. Groffen, M.A. Tijssen, Paroxysmal kinesigenic dyskinesia: cortical or non-cortical origin, *Park. Relat. Disord.* 18 (2012) 645–648.
- [29] J.A. Obeso, A.E. Lang, J.C. Rothwell, C.D. Marsden, Postanoxic symptomatic oscillatory myoclonus, *Neurology* 33 (1983) 240–243.
- [30] E. Vytyarova, R. Marecek, J. Fousek, O. Strycek, I. Rektor, Large-scale cortico-subcortical functional networks in focal epilepsies: the role of the basal ganglia, *Neuroimage Clin* 14 (2017) 28–36.