Original article

Epileptic Disord 2005; 7 (2): 115-21

Language- and praxis-induced jerks in patients with juvenile myoclonic epilepsy

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Received November 8, 2004; Accepted February 14, 2005

ABSTRACT – Reflex traits have been described in patients with idiopathic generalized epilepsy. We report on four patients with juvenile myoclonic epilepsy in whom the coexistence of praxis- and language-induced jerks was documented in video-polygraphic EEG recordings.
[Published with video sequences]

Key words: reflex epilepsy, idiopathic generalized epilepsy, juvenile myoclonic epilepsy, neuropsychological EEG activation, language and praxis induction

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The 2001 Proposal for a Diagnostic Scheme for People with Epileptic Seizures and Epilepsy duly identified the different groups of various precipitating stimuli for both reflex seizures and reflex epilepsies (Engel 2001). The recognition of reflex traits is fundamental to treatment, since avoidance would be one of the determinant factors for seizure control.

Precipitating visual stimuli such as photosensitivity, pattern sensitivity, fixation-off and scotosensitivity, the first being by far the most common, represent the most well recognised group of reflex epileptic traits. More recently, some of those formerly considered as rare, such as praxis induction of myoclonic jerks in the arms (Inoue *et al.* 1994) and the talking/reading induction of perioral myoclonia (Wolf and Mayer 2000), have been broadly recognized in a number of epilepsy syndromes, but mainly in idiopathic generalized epilepsies (IGE), particularly in juvenile myoclonic epilepsy (JME).

Praxis-induction of myoclonic jerks was suggested by Inoue *et al.* (1994) who emphasized the sensitivity of some patients to situations in which they are required to consider complicated spatial tasks in a sequential fashion, specifically with the intention of making decisions and responding practically by using a part of their bodies, under stressful circumstances.

Perioral myoclonia, brief and repetitive jerks in the perioral region, the hallmark of reading epilepsy, is seen in both primary and secondary forms of this epilepsy syndrome. In the first, seizures would be exclusively precipitated by reading, while in the second, they could occur in other conditions (Bickford *et al.* 1956).

We have documented four cases of JME with concomitant signs and symptoms of these two reflex traits. All patients presented normal physical and neurological examinations, as well as routine blood tests and 1.5T MRI. A video-polygraphic EEG using the 10-20 International Electrode Sys-

tem, in addition to perioral and deltoid electrodes, was recorded with activation by hyperventilation and photic stimulation. A specific neuropsychological protocol including reading, speaking, writing, calculating, drawing and playing games, plus performing spatial tasks involving sequential processing and complicated movements and decision-making was also undertaken.

Case reports

Case 1

A 25-year-old journalist presented her first and only generalized tonic-clonic (GTC) seizure in the morning, preceded by myoclonic jerks in the right arm at the age of 14. Carbamazepine (CBZ) led to seizure control for four years. Then, at 18 she noticed repetitive, arrhythmic and irregular myoclonic jerks predominantly in the right arm when writing, typing, drawing and handling small objects. Clonazepam (CNZ) partially diminished the seizures. Moreover, at 22, in her locution-training laboratory she observed perioral myoclonic jerks while reading, silently or aloud, as well as while talking. The myoclonic jerks persisted at times associated with speech impairment. Valproate (VPA) improved the myoclonia induced by some actions but not those occurring when she talked or read quickly. VPA (900 mg/day) and CNZ (2 mg/day) partially reduced seizure frequency, nevertheless, she still presented very frequent episodes of isolated, abrupt, involuntary jaw jerks while talking or reading, although these were more frequent when reading out loud rather than silently, and was associated with difficulty in pronouncing words.

Occasional episodes of speech arrest as well as myoclonic jerks induced by praxis, were also noticed. Adding lamotrigine 50 mg/day to VPA and CNZ resulted in improvement in these symptoms. On the EEG, the background activity was normal, with some biphasic discharges of sharp waves mainly after closing the eyes. During somnolence and phase II sleep, the discharges were rare, appearing as isolated, occipital sharp waves and fast spikes bilaterally in the central areas. Silent reading and reading aloud, both in Portuguese and English, and spontaneous stressful speaking, brought about single complexes of a minute spike followed by a slow wave appearing asymmetrically in frontocentroparietal areas, some of them accompanied by perioral myoclonia. Activities such as writing, written calculation and games, as well as thinking and spatial decision-making, precipitated myoclonic jerks in both arms, but still prevailing in the arm involved in the action. During writing, the myoclonic jerks were accompanied by perioral myoclonia. The succession of the tasks exacerbated the symptoms, which were minimized by interruption of the activities and relaxation maneuvers. During the planning of spatial tasks without manual responses and also during the execution of spatial motor tasks, there were isolated fast spikes followed by slow waves, which were morphologically similar to those recorded while reading and involving the same areas *(figure 1)*.

Case 2

A 35-year-old, right-handed man, of Japanese origin, with no remarkable medical history himself, had however, a sister with epilepsy. At the age of 14, he presented his first GTC seizure during sleep. By the age of 16 he started presenting myoclonia during awakening mainly in the morning, sometimes provoking falls or leading to GTC seizures. At 17 there was an increase in seizure frequency, intensity and also precipitation by sleep deprivation, fasting, stress, sexual activity, flashing lights, speaking in public, hand activities, planning and decision-making, and calculating. Photosensitivity was confirmed by his EEG recordings obtained at this time, before the use of antiepileptic medication. Since then, he had been treated with multiple antiepileptic drugs in different combinations, including CBZ, VPA, phenobarbital (PB), phenytoin (PHT) and piracetam, with poor clinical control,. His present treatment regimen includes VPA (2,000 mg/day), clobazam (20 mg/day) and topiramate (200 mg/day). His neuropsychologically-activated video-polygraphic EEG showed rare paroxysms of generalized spike and polyspike waves exacerbated by reading out loud and silently, calculating, writing, drawing, and during praxis activities involving finger manipulation and decision-making *(figure 2)*. Yet, there was no photosensitivity.

Case 3

A 32-year-old, left-handed man, with unremarkable antecedents, including no family history of epilepsy, reported involuntary jerking of the arms while playing checkers and dominoes since the age of 12. The myoclonia appeared after playing for prolonged periods when he felt overwhelmed by the game, and obliging him to stop the action. At 14, during a checker game immediately after lunch, these jerks evolved to a GTC seizure. Since then he has avoided playing these games but has noticed spontaneous jerks in the arms after awakening, sometimes followed by a GTC seizure. PB was ineffective and, at 28, he noticed jaw jerking and abnormal sensations in the abdomen and throat, described as an ascendant shock-like feeling, when reading. These symptoms were so marked he had to stop studying. Sometimes, especially when anxious, there were hesitations and stuttering while talking and reading. PHT led to a worsening in seizure frequency. At 30 years of age, PB was switched to VPA (1,500 mg/day). As a result, there was a significant improvement since he only presented occasional GTC seizures not preceded by any myoclonic jerks when exposed to precipitant factors such as sleep deprivation. He stopped exposing himself to the already identified precipitants such as games and prolonged read-

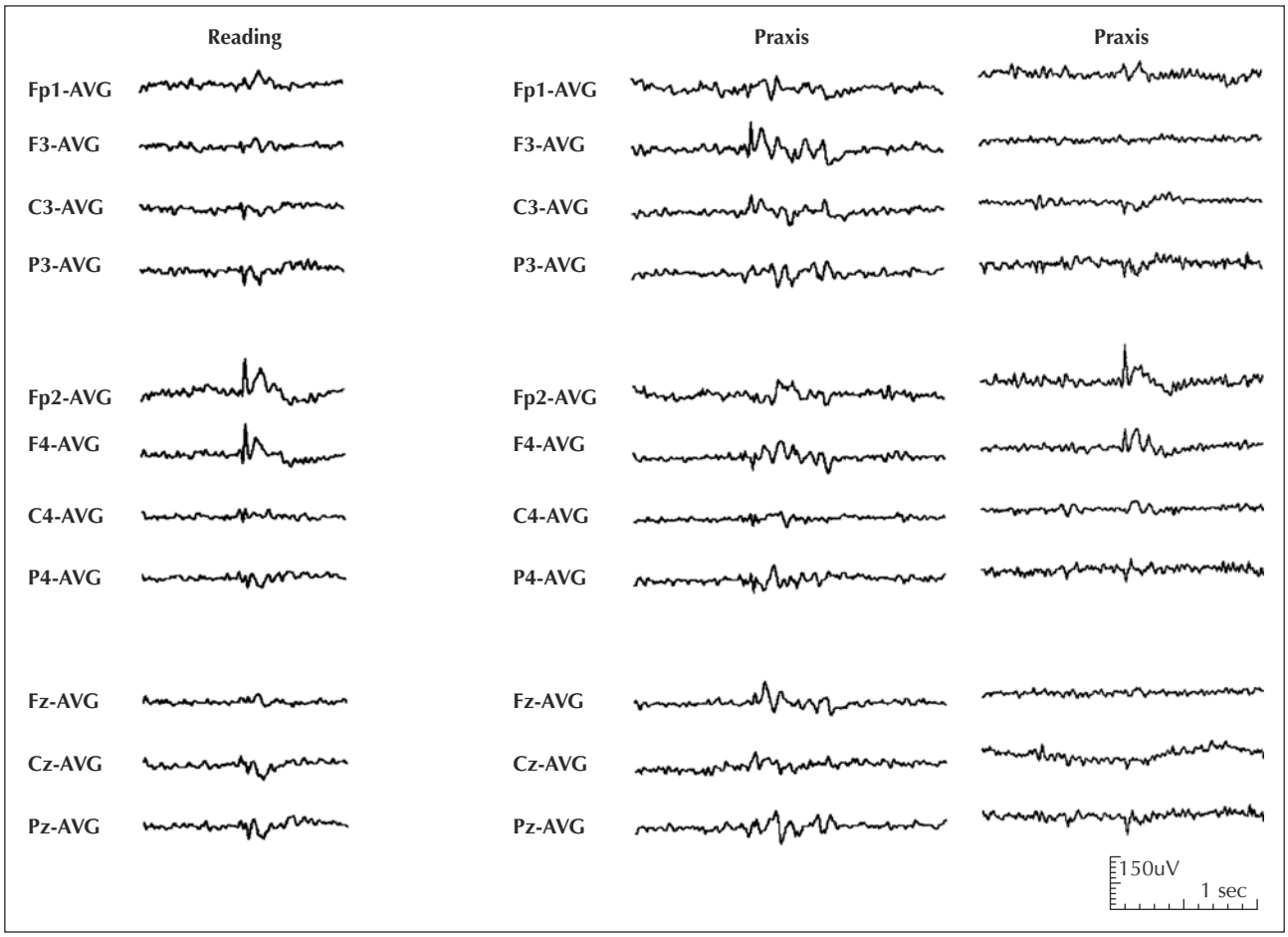

Figure 1. Fragments of EEG recorded during reading and praxis showing remarkable similarities between the fast spikes followed by slow waves in the frontocentroparietal areas. They were seen as isolated or short volleys of low amplitude, poorly formed, very fast and brief spike and wave complexes maximum sometimes on the left, sometimes on the right, mainly involving anterior areas.

ing. Normal background activity and spontaneous generalized spike and wave complexes, with anterior predominance, sometimes asymmetric, were seen in his neuropsychologically-activated video-polygraphic EEG. The only task that precipitated bilateral perioral myoclonia accompanied by the shock-like ascending sensation and interruption of speech was when reading Portuguese aloud. Generalized spike-wave complexes were recorded on the EEG at this time *(figure 3)*. The construction of a tower with plastic blocks led to a shock-like sensation in both arms.

Case 4

A 16-year-old, right-handed man with a strong family history of IGE in two aunts and three cousins, presented a cluster of myoclonic jerks on awakening following sleep deprivation due to prolonged exposure to a video-game, leading to a GTC seizure. Four months before he noticed jerks, mainly in his right arm, occurring at no particular time, but more often when tired. Occasionally, there was involvement of the lower limbs which resulted in falling. He noticed perioral myoclonia and an abnormal sensation in the throat that caused stuttering and interruption of the action especially when reading aloud or speaking in public during stressful situations. Treatment with CBZ was not successful, while VPA (750 mg) promoted control of the myoclonia and TCG seizures, although the perioral myoclonia persisted when exposed to precipitating factors. His neuropsychologically-activated videopolygraphic EEG showed normal background activity and generalized paroxysms of spike and polyspike waves exacerbated during sleep, praxis- and reading-induction. The clinical and video-polygraphic EEG findings are summarized in *table 1*.

Discussion

We describe four patients with reflex seizures induced by complex stimuli, language and praxis, triggered by com-

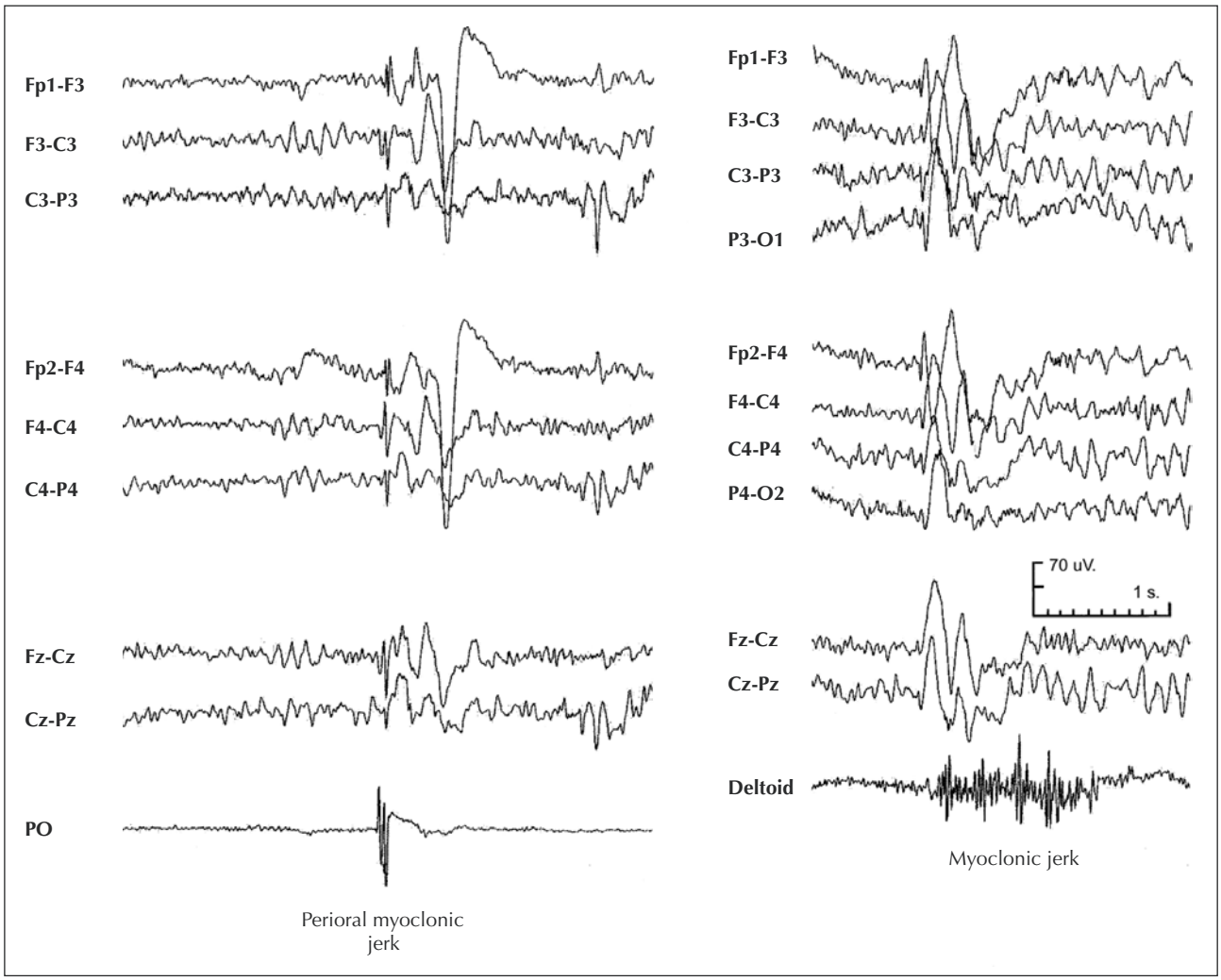

Figure 2. Brief paroxysms of very fast spikes followed by a slow wave, mainly in the central areas accompanied by jaw-jerking phenomenon while the patient was reading English aloud. Similar discharges during praxis were seen when the patient was doing puzzles. Once more notice the similarities between the epileptiform discharges during reading and praxis induction (70µV 1 sec).

bined specific activities such as visual-spatial tasks, praxis, decision-making and emotional components. These patients demonstrated common electroclinical characteristics such as onset at adolescence, family history of epilepsy, normal neurological examination, presence of two or three types of generalized seizures, normal background activity and generalized interictal discharges on EEG.

These findings are suggestive of IGE or more specifically, JME. All the patients presented myoclonic jerks during reading, speaking, planning and performing motor activities. When reading, characteristic perioral myoclonias, as classically described in reading epilepsy, were documented. These were accompanied by an abnormal sensation or movement in muscles which were involved in reading and talking, and sensations in parts of face such as twitching and interruption of the action, and stuttering.

On the other hand, there were also myoclonic jerks induced by motor and action-programming activities, which were reproduced in the video-polygraphic study. Interestingly, language- and praxis-induction were documented simultaneously. Usually these are considered independent symptoms of verbal and nonverbal cognitive cortical activation of generalized (or at least bilateral) ictogenesis, which are often a part of JME, and their coexistence is rare, although already described previously (Salas-Puig *et al.* 2001).

Considered uncommon, reflex seizures may occur in about 4-7% of the epilepsies in general (Panayiotopoulos 1996). Matsuoka *et al.* (2000) reported the results of neuropsychological EEG activation in 480 Japanese patients with different types of epilepsy, verifying induction of epileptic discharges in 38 (7.9%). This procedure was

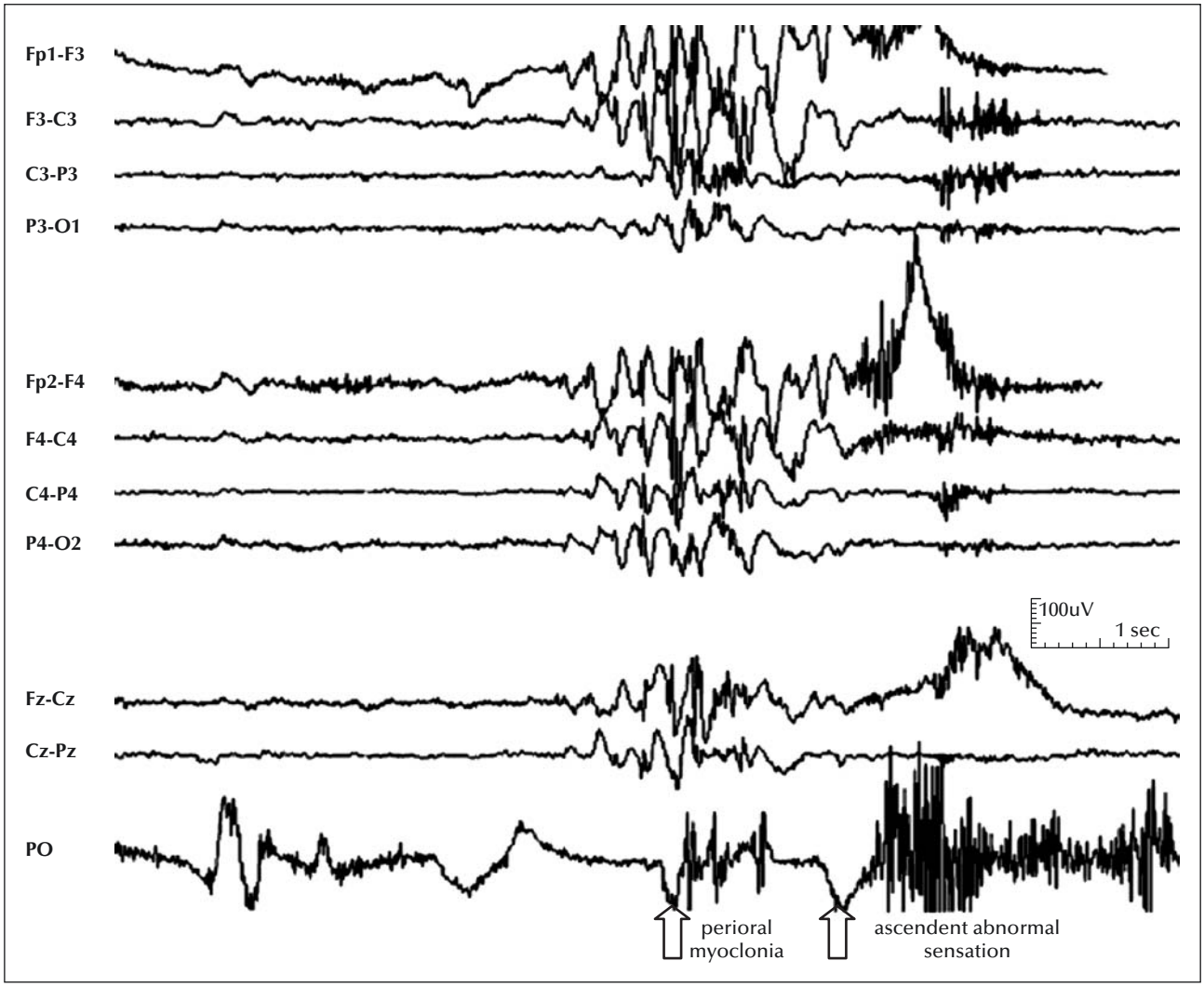

Figure 3. EEG tracing showing generalized spike wave complexes with consistent onset in the frontal regions while the patient was reading aloud, associated with perioral myoclonic jerks. The first arrow points to the perioral myoclonic jerks and the second to the moment at which the patient reported an abnormal, shock-like, ascendant sensation from the stomach to the throat.

effective in 36 (24.7%) of 146 IGE patients, especially in JME (22 of 45, 49%). Talking/reading induction was present in 14 (23%) of the 62 JME patients reported by Wolf and Mayer (2000), who described praxis-induction in 19 (31%) of them. Nine patients reported precipitation by both praxis and talking/reading. In Japan, Inoue and Kubota (2000) described 213 JME cases of which 27 (12%) were placed into in the praxis-sensitive group. Only in two patients did reading or speaking also induce a seizure.

An important fact however, is the observation that our patients continued having perioral myoclonia and praxis induction despite the use of VPA and other drugs considered reasonably effective in IGE and JME. This, therefore, suggests that the presence of reflex traits could represent an aggravating factor, especially when more than one is present in the same patient (Matsuoka *et al.* 2002). In agreement with this supposition, Inoue and Kubota (2000) reported that the rate of seizure-control over more than three years in patients with JME, had dropped from 69% when they presented only general precipitating factors, to 48% in praxis-sensitive patients.

Matsuoka *et al.* (2002) described the long-term course of seizure susceptibility in two patients with JME in whom the provocative effect of higher mental activities (praxis-induction) persisted, despite a decrease in spontaneous myoclonic seizures under the same drug regimen. These observations suggest that the pathophysiology of JME improves with time, but does persist for a long time. In these patients, a neural network involved in higher mental activities mainly associated with use of the hands might be predominantly involved rather than the circuits processing other physiological factors such as sleep deprivation, sud-

Table 1. Clinical and seizure characteristics documented in these cases showed by video-polygraphic EEG recordings.

Characteristics	**Case 1**	**Case 2**	**Case 3**	**Case 4**
Age (years)	25	32	35	16
Gender	female	male	male	male
Age of praxis-induction onset (years)	18	12	17	16
Type of seizures	Myoclonia in the acting upper limb, sometimes bilateral	Myoclonia in the acting upper limb, sometimes bilateral	Myoclonia in the acting upper limb, sometimes bilateral	Myoclonia in the acting upper limb, sometimes bilateral
Activities	Writing, typing, drawing, handling small objects, planning and performing slight movements	Planning and playing games, and slight movements	Writing, drawing, calculating, planning and performing slight movements	Planning and playing games, and slight movements
Age at language-induction onset (years)	22	28	17	16
Type of seizures	Perioral myoclonia	Perioral myoclonia accompanied by abnormal sensation in the abdomen and throat	Perioral myoclonia	Perioral myoclonia accompanied by abnormal sensation in the throat, stuttering and interruption of the action
Activities	Silent and reading aloud, speaking in public	Reading aloud and speaking in public	Silent and reading aloud, speaking in public	Reading aloud and speaking in public

den awakening, fatigue and flickering lights emphasized in prior reports. However, these circuits could also activate hyperexcitable rolandic areas.

In support of this hypothesis, there were quite localized, frontocentroparietal epileptiform discharges on the EEG in three patients, despite bilateral jerks. Interestingly, in these patients, both reading- and praxis-induction were accompanied by brief paroxysms of very fast spikes followed by a slow wave, mainly in the frontocentroparietal areas.
These findings had already been observed by Matsuoka (1990) who reported that mental activity involving the hands, provoked central-dominant discharges with unilateral myoclonic seizure as well as generalized discharges with bilateral myoclonic seizures. Conversely, Chiafari *et al.* (2004) reported reflex writing seizures in two siblings with JME who obtained good control with VPA. We could postulate that the presence of only one of these reflex traits might represent a more benign epileptic vulnerability.

The results of this study further support the notion that "language- and praxis-induced" reflex seizures precipitated by specific stimuli occur in the context of JME. All the patients described here presented both language- and praxis-induced jerks, a rare coexistence in epileptology. □

Acknowledgements. We would like to thank Pr. Peter Wolf and Dr. Thomas Mayer, for having kindly shared their valuable expertise with us including their protocol. CAPES, DAAD and FAPESP from Brazil supported this study.

Legends for video sequences

Language- and praxis-induction

Case 1
– Reading English aloud (perioral myoclonia)
– Making plastic models (praxis-induction)
– Matchstick pattern reproduction (praxis-induction)

Case 2
– Reading Portuguese aloud (perioral myoclonia)
– Writing, calculation (praxis-induction)
– Making plastic models (praxis-induction)
– Doing puzzles (praxis-induction)

Case 3
– Reading Portuguese aloud (perioral myoclonia and abnormal sensation)

Case 4
– Reading Portuguese aloud (perioral myoclonia)

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