

# Motor impairment on awakening in a patient with an EEG pattern of “unilateral, continuous spikes and waves during slow sleep”

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**ABSTRACT** – Movement disorders are rarely described in association with the “continuous spikes and waves during slow sleep (CSWS)” EEG pattern. We report the case of a young girl affected by an epileptic encephalopathy who, from the age of seven years and four months, has twice presented a movement disorder affecting the right arm, manifesting on awakening and disappearing by early afternoon. Sleep EEG during these periods showed continuous, high-amplitude, diphasic spikes and slow waves over the left hemisphere. Association of clobazam, valproic acid and, on the second occasion, ethosuccimide led to disappearance of the above-described EEG picture and associated motor symptoms. Neurophysiological investigations excluded other possible aetiologies. In view of this, and of the close relationship between the EEG picture and clinical course, we interpret the patient’s impairment as “motor neglect” secondary to the continuous electrical activity recorded during sleep over the left hemisphere and involving the associative areas. This electrical activity in sleep, may be regarded as a “functional lesion” whose clinical consequences can be correlated with the site of the abnormalities.

*[Published with video sequences]*

**Key words:** CSWS, functional motor impairment, childhood epilepsy, motor neglect, continuous spike-wave



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Continuous spikes and waves during slow sleep (CSWS) is a well-known EEG pattern that can be associated with cognitive and behavioural decline (Veggiotti *et al.* 1999). Movement disorders, on the other hand, are rarely described in association with this EEG pattern. Dystonia, dyspraxia,

ataxia and apraxia (Neville and Boyd 1995, Maquet *et al.* 1995) and/or unilateral deficit and negative myoclonus are considered the most disabling movement disorders occurring during CSWS, but reports, even of these conditions, are sporadic (Tassinari *et al.* 2000).

In a previous study (Veggiotti *et al.* 1999), we described three patients with unilateral CSWS who, for a few hours after awakening, exhibited a worsening of spontaneous movements in the hemibody contralateral to CSWS activity. All the patients had symptomatic epilepsy due to perinatal occlusion of the middle cerebral artery.

We report the long-term clinical and EEG findings in a patient with cryptogenic epilepsy and unilateral CSWS, who presented on awakening a transitory movement disorder affecting the contralateral arm.

## Case report

We describe the case of a girl, born on 26/07/91, following a normal pregnancy and delivery. From the age of 10 days, the patient presented, during sleep, right partial motor seizures, and from 40 days she also began to present infantile spasms, which were treated with ACTH and valproic acid (VPA).

Although the patient showed severe psychomotor retardation from the first months of life, hemispheric lateralization of handedness was achieved (she is right-handed).

At the age of 6 years, following a period of relative well-being during which sporadic partial seizures triggered by fever were the only episodes of note, the patient began to present partial motor seizures lasting around two minutes and involving the right arm and face (flexion-extension of the right hand and rightward twisting of the lips). An EEG focus in the left, central-temporal region was evident from the very first examination. VPA dosage was increased and the seizures disappeared.

When the patient was 7 years and 4 months old, her parents and physiotherapist noticed that she was making less use of her right arm upon awakening: this deficit, albeit fluctuating, persisted throughout the morning, before disappearing in the early afternoon. The patient tended to let her arm lie passively against her body, and her spontaneous right arm movements were reduced. She used her right arm only in order to point, and even this movement was almost entirely restricted to the hand, as she appeared to have difficulty flexing her arm and forearm. When invited to reach for objects presented in mid-line position, she responded by moving her left arm, and would attempt to move the right one only when the object was within easy reach of the right hand; bimanual prehension of large objects was absent. Her difficulties became more pronounced when she attempted to take hold of the object, especially when this required the use of fine motor skills. Once she had managed, with difficulty, to grasp the object, the problem then became one of letting it go; this was achieved only after various attempts and always through a throwing action. She was able to accomplish this same task more smoothly and efficiently using her left arm. The impairment of the right arm was made worse by reduced coordination. The patient's parents also drew

attention to differences between her performance of certain tasks (brushing teeth and taking biscuits) in the morning, just after awakening, and in the evening. In the morning, motor activity involving the right arm was practically absent: the patient used this arm only when specifically asked to do so. By contrast, in the evening she used her right arm spontaneously.

The EEG at this time revealed high-amplitude, diphasic spikes and slow waves over the left centroparietal region, which were very clearly activated when falling asleep and during sleep, and then became continuous and diffuse over the left hemisphere (*figure 1*). This EEG pattern disappeared after awakening (*figure 2*). The addition of clobazam to the therapeutic regimen was followed by disappearance of the EEG pattern described above, and of the motor impairment.

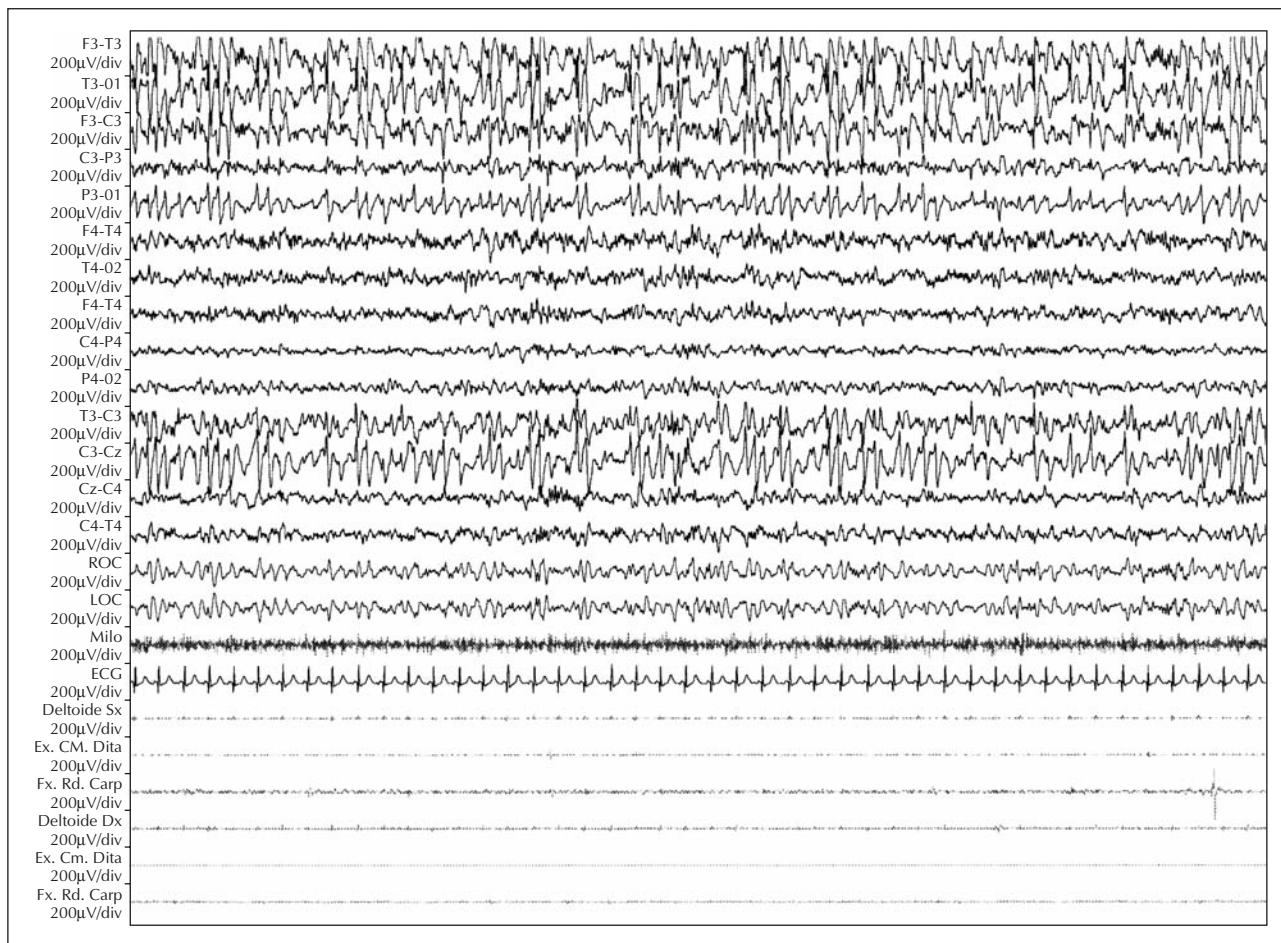
At age eight years and nine months, our patient once more began making less use of her right arm, and a clinical picture developed that was similar to the one she had presented at the age of seven. Once again, the sleep EEG showed continuous spikes and waves over the left hemisphere, mainly in the centroparietal region. Polysomnographic recordings failed to document seizures during nocturnal sleep. Polygraphic recording was performed and this allowed us to exclude epileptic negative myoclonus, as shown in *figure 3*.

The clinical picture resolved with a combination of CLB, ethosuccimide (ETS) and VPA. Both the EEG pattern and the clinical picture responded well to antiepileptic drug (AED) treatment. Repeated metabolic investigations (including plasma, urine and CSF amino acid concentrations and urinary organic acid analysis) were negative. Brain MRI revealed mild cerebellar vermis atrophy. Neurologically, the patient currently presents an ataxic gait with intentional tremor of the upper limbs. Our patient is cognitively impaired; she understands only simple, contextualized sentences, and her play is stereotyped and nonsymbolic.

The patient's seizures have not recurred since 2000; EEG traces show rare spikes and waves in the left centroparietal region, not activated by sleep: she is still on ETS, clobazam and VPA therapy.

## Discussion

The case reported here is that of a patient presenting a sudden-onset movement impairment affecting the right arm. Despite severely delayed psychomotor development, the patient had achieved hemispheric lateralization of handedness, and displayed a right arm preference. Because of this, the patient's parents and therapists were quick to spot the motor dysfunction and were able to describe it clearly and accurately. A peculiar characteristic of this "motor dysfunction" was its temporal pattern: indeed, the symptoms were constantly found to be more



**Figure 1.** Nocturnal polysomnographic recording made at the age of seven years and four months during stage 2 NREM sleep: it is possible to observe a picture of continuous spikes and waves over the left derivations that can be defined as a pattern of “unilateral continuous spikes and waves during slow sleep”. In fact the right derivations are completely spared.

marked upon awakening from nocturnal sleep, and had always disappeared by early afternoon.

The EEG pattern coinciding with the motor impairment was one of continuous spikes and waves during slow sleep over the left hemisphere, giving rise to a picture of “unilateral CSWS”. However, to avoid confusion over terminology and also in view of the fact that the “CSWS” pattern is, by definition, bilateral and symmetric, we preferred to define the EEG picture found in our patient as “unilateral continuous spikes and waves during slow sleep”.

The correlation between the clinical signs and the EEG pattern was confirmed by the fact that disappearance of the motor impairment, following the introduction of appropriate AED therapy, coincided with disappearance of the “unilateral continuous spikes and waves during slow sleep”. In fact, on both occasions, AED therapy successfully controlled the EEG pattern and the associated clinical

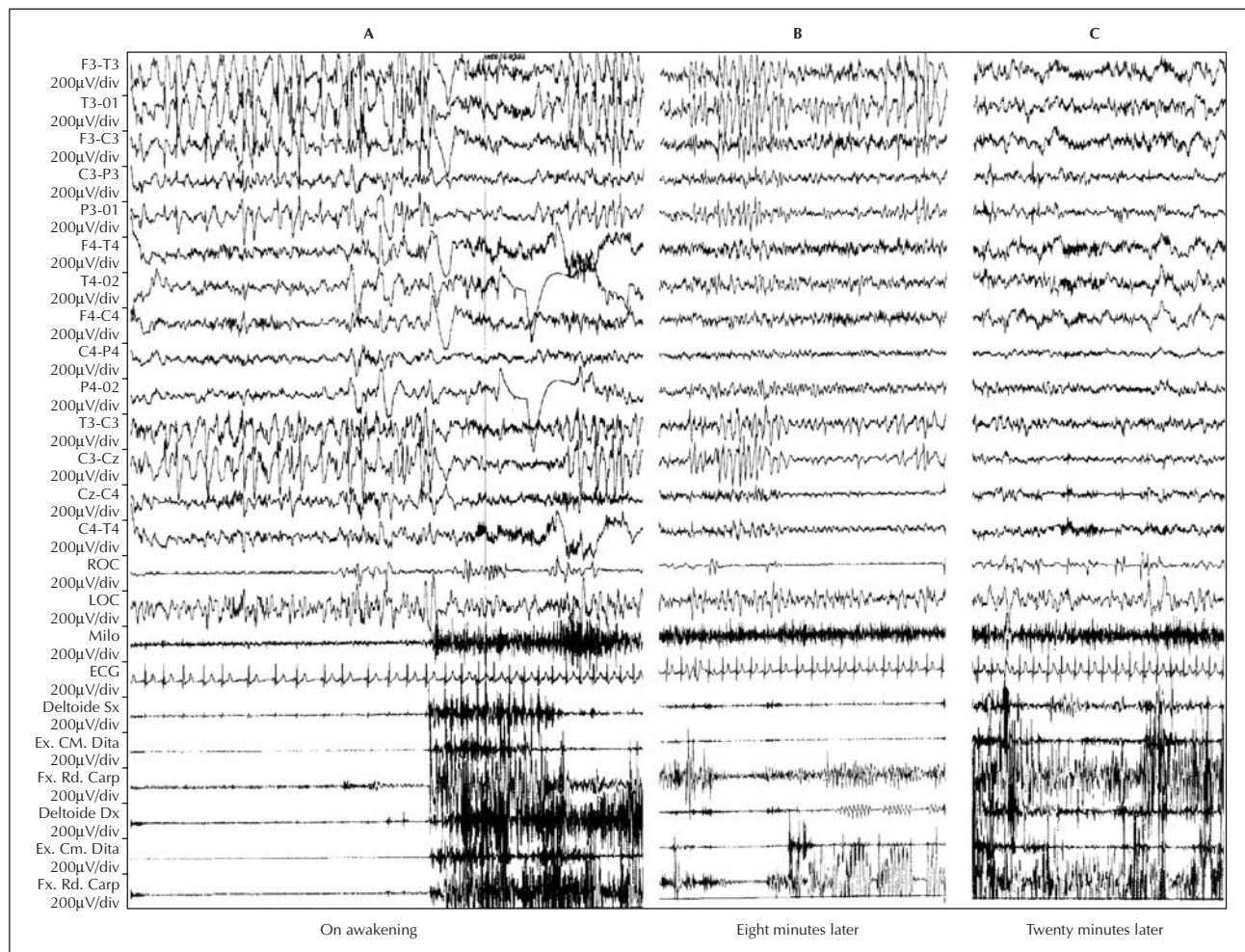
picture. The motor impairment manifested by this patient is difficult to interpret and may be caused by different nosological entities.

The movement disorder did not present the characteristics of post-ictal Todd paralysis: indeed, the patient presented no frank strength deficit upon awakening, and the disturbance persisted for many hours after awakening, a feature not associated with post-ictal paralysis. Furthermore, repeated nocturnal polysomnographic recordings failed to document the occurrence of seizures.

Combined EEG-polygraphic recording also ruled out the presence of “epileptic negative myoclonus”, responsible for the movement disorder as described by Guerrini *et al.* (1993) in a five-patient series.

The relationship between CSWS and movement disorders has been the focus of few studies: Veggiotti *et al.* (1999) described three patients with congenital hemiplegia and





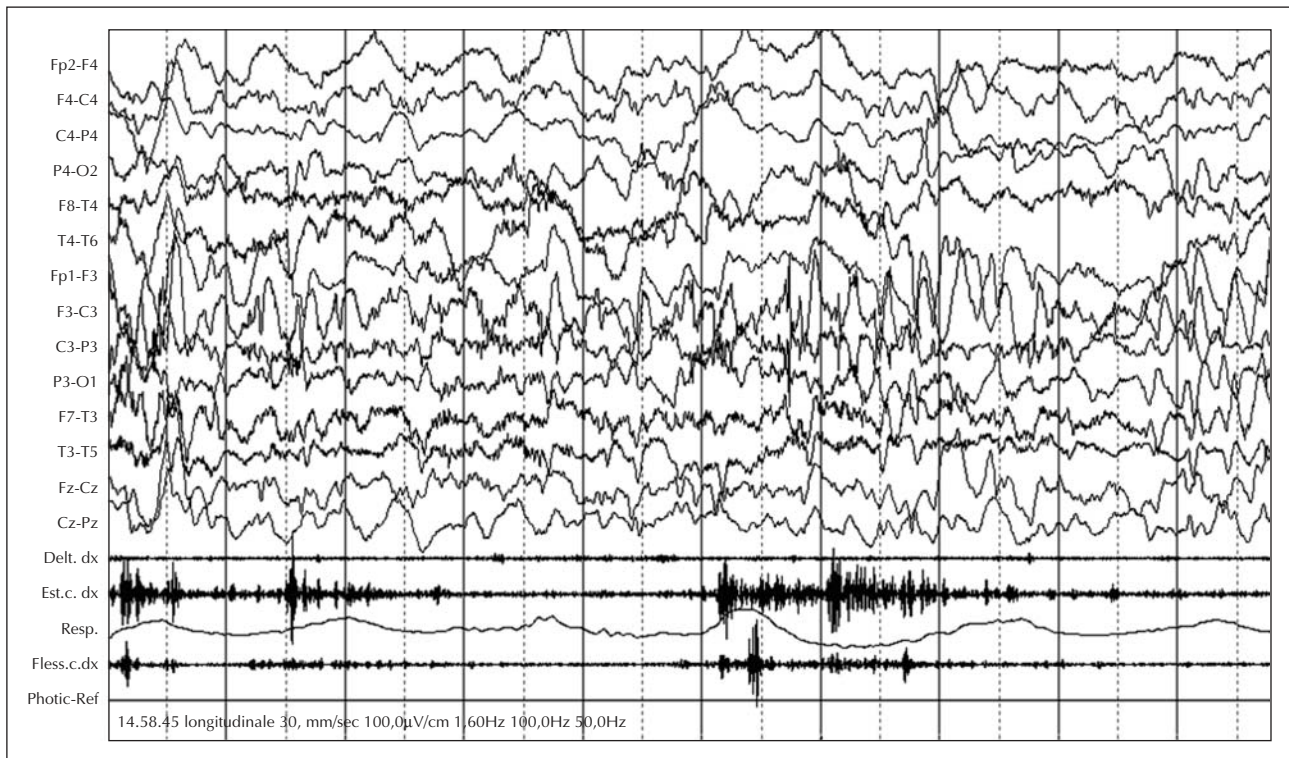
**Figure 2.** The same nocturnal polysomnographic recording. Here we show the clear changes in EEG activity from sleep through awakening. **A)** the continuous spike-and-wave activity during sleep is interrupted by induced awakening. **B)** eight minutes after awakening, this activity starts to appear fragmented. **C)** twenty minutes after awakening it has disappeared; isolated diphasic spikes can be observed in the left centroparietal region.

an EEG picture of unilateral CSWS who, in the absence of nocturnal seizures, showed reduced use of the hemibody contralateral to the CSWS activity, which lasted a few hours after awakening. Like our patient, these subjects presented a movement disorder that was clearly apparent upon awakening and that showed a correlation with epileptiform activity during sleep; in the latter patients, however, the disorder appeared in the presence of a pathological anatomical substrate.

More recent studies (Veggiotti *et al.* 2002, Praline *et al.* 2003) have stressed the significance of the location of the electrical focus in CSWS.

It has been reported that the presence of continuous epileptiform activity in a delicate phase of cortical development can result in severe functional impairment of the

areas involved; it thus becomes fundamentally important to identify the areas most involved in this pathological activity, as these determine the clinical outcome. Many authors agree that in cases defined as Landau Kleffner Syndrome, the focus is located in the temporal region and the main clinical symptom is acquired, auditory agnosia or aphasia; conversely, in cases of CSWS, in which the main focus is frontal, the neuropsychological impact is global (although greater on the executive functions and/or in terms of behavioural symptoms). Recent publications have considered extending the clinical spectrum associated with the CSWS syndrome. The finding of visual agnosia in a child with non-lesional, occipito-temporal CSWS (Eriksson *et al.* 2003) is consistent with the presence of a severe visuospatial deficit in adults with a history of



**Figure 3.** Polygraphic recording with a time scale of 30/mm/sec, which allowed us to exclude the presence of epileptic negative myoclonus.

CSWS (in these same patients the location of the electrical focus had been occipital and parietal-occipital) (Praline *et al.* 2003).

These data prompt us to hypothesize that the movement disorder presented by our patient might be regarded as “motor neglect” secondary to the continuous slow spike and spike-and-wave activity recorded over the left centro-parietal regions and involving the associative areas. These areas are indeed considered fundamental in motor planning and their impairment could explain the presence, in our patient, of “motor neglect” secondary to a transitory motor planning disorder. By “motor neglect” we mean the condition defined by Castaigne (Castaigne *et al.* 1970), *i.e.*, a movement disorder restricted to one side of the body, characterized by motor hemi-neglect, in the absence of paralysis, hypertonia, sensory impairments, asomatognosia and anosognosia. In the case of our patient however, this “motor neglect” has no organic substrate: we failed to identify retrorolandic lesions like the ones found in the cases described by Castaigne *et al.* (1970).

We might thus hypothesize that the continuous focal electrical activity during sleep found in our patient can be

regarded as a “functional lesion” whose clinical consequences can be correlated with the site of the abnormalities.

Study of other patients showing an EEG pattern of “unilateral continuous spikes and waves during slow sleep” and movement disorders, is necessary in order to improve descriptions of this, in our view, often unrecognized picture. □

### Legend for video material

The patient’s parents made this film in order to show her doctors the characteristics of her motor impairment and the modification of this picture of impairment during the course of the day. Note the different ways in which the same tasks (brushing teeth and taking biscuits) are performed in the morning, just after awakening, and in the evening. In the morning, motor activity involving the right arm is practically absent: the patient uses this arm only when specifically asked to do so. In the evening she uses her right arm spontaneously.

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