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De Novo Typical Absence Status Epilpticus Of Late Onset

Abdellaoui Walid[1], Louanchi Malika[2], Ait-Oukaci Wassila[3], Toubal Nadia[2], Sadibelouiz Mustapha[4], Ait-Kaci-Ahmed Mahmoud[4]

[1]*neurology Department, Mostaganem Hospital, University Of Mostaganem*
[2]*neurology Department, Ibn Sina Hospital, University Of Annaba*
[3]*neurophysiology Department, Ait Idir Hospital*
[4]*neurology Department, Ait Idir Hospital, University Of Algiers*

Abstract:
Background*: Absence status epilepticus ASE is a type of non convulsive status epilepticus in which continuous or recurrent generalised epileptiform discharges are associated with a varying grade of consciousness. We describe the case of an elderly man who presented with typical ASE resolved after intravenous clonazepam.*
Clinical case report: *A 62-year old man had a history of idiopathic generalized epilepsy with tonic-clonic seizures. Treated since the age of 30 years with phenobarbital PB 100 mg/day. He was disoriented to time and place and a slowness in mental activity over 24 h after food poisoning. Laboratory tests and a CT scan of the brain were normal. Brain MRI did not show any structural lesions. EEG showed recurrent generalized paroxysms of 3 Hz spike-wave. Normal background activity. Normalization of mental status and EEG pattern after 1mg intravenous clonazepam (Benzodiazepine). Phenobarbital PB was substituted by valproic acid 1500 mg/day.*
Conclusion: *Typical diagnosis must be based on EEG pattern and clinical history. Efficacy of intravenous clonazepam in treatment of typical ASE.*
Key Words*: Absence status epilepticus ASE, Generalized paroxysms of 3 Hz spike-wave, Benzodiazepine.*

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I. Introduction

Absence status epilepticus involves mental confusion of varying intensity from a simple subjective slowdown of intellectual efficiency to catatonic stupor. This condition can persist for hours or days and is associated in one case out of two with bilateral periocular myoclonus which is not very ample [1,2]. Although the destructuring of consciousness actually proceeds from a continuum [3], a categorization of the intensity of disorders of consciousness into four stages was proposed by Lob et al. [4] then was regularly taken up thereafter [5]. Mild obtundation occurs in 20% of cases [4]. This stage is characterized by a simple slowing down of ideation and modes of expression. Frequently, discreet cognitive disturbances are only apprehended by the patient himself. A more or less extensive neuropsychological exploration [6,7] is then necessary to objectify the modification in the level of consciousness, responsible for a discreet decline in intellectual efficiency. Marked obnubilation is more usual (65% of cases). Severely disoriented patients are usually calm, immobile, speechless, perplexed and indifferent. They stand with their eyes half-closed and show no activity or intention of spontaneous activity. The fluctuating destructuring of consciousness is not organized in a cyclical way. At the clinical stage, an amnesic stroke, a psychiatric episode, toxic or drug-induced encephalopathy are often mentioned [8,9]. Sometimes, a generalized tonic-clonic seizure initiates or more frequently ends the episode [10]. A profound clouding of consciousness, or even a catatonic stupor, accounts for only 15% of cases.

The main difficulty is to evoke the diagnosis and to indicate an emergency EEG early. Delays in diagnosis are often significant, ranging from eight hours to several days [2]. EEG expression is polymorphic, realizing "virtually any pattern involving bilateral, rhythmic, non-reactive paroxysmal activity" [11]. In the series by Granner and Lee, in 59 patients seen consecutively, the average frequency of paroxysmal activity is between 1.0 and 3.5 Hz [12]. Morphological aspects strictly superimposable to those of typical absences are only present in 7% of cases. A focal predominance of paroxysmal activity is present in a quarter of patients. The correlation between the degree of clouding of consciousness and EEG aspects is not clear. The injection of a benzodiazepine during the examination (diazepam, 10 to 20 mg or clonazepam, 1 to 2 mg) constitutes a therapeutic test which is positive when it normalizes the EEG and relieves confusion. Ingestion of a single dose of clobazam at a dosage of 0.5 to 1 mg/kg may constitute an alternative in patients.

ambulatory elderly people [13]. In difficult cases (minor obscuration of consciousness, uncharacteristic EEG activity), it is imperative to perform neuropsychological tests before and after the injection of

benzodiazepines. The improvement in the neuropsychological score after injection confirms the diagnosis [9]. In subjects with recurrent absence states, valproate appears to be the drug of choice to prevent recurrence [14].

Nosographic place of absence states are part of a vast neurobiological continuum and can punctuate the evolution of any type of epileptic syndrome [15,16]. In certain patients, the absence state constitutes a particular evolutionary moment of worsening in the natural history of their epilepsy. In other patients, the absence state is the beginning of a non-specific neurological or systemic condition. The prognostic implications associated with absence states depend more on the epileptic syndrome in which they occur than on their specific electroclinical characteristics. A typical absence corresponds to a generalized seizure (discharge of spike waves at 3 c/s affecting the entire cortex) accompanied clinically in its usual form by a more or less profound alteration of contact. Typical absence states are characterized by the repetition of serial or continuous absences with, on a clinical level, a more or less intense confusional syndrome. Typical states of absence are very rare these days and are part of the context of idiopathic generalized epilepsy most often comprising absences [17]. The EEG expression is represented by synchronous and symmetrical bilateral discharges of wave spikes or wave polytips whose frequency is greater than or equal to 3 Hz. The immediate prognosis is excellent, the injection of a benzodiazepine sufficient to resolve the states. of badness. As opposed to typical absences, atypical absences are characterized by an often incomplete break in contact. The term atypical refers to any EEG pattern different from the classic generalized wave spike appearance at 3—3.5 c/s of the typical absence. Most often, an atypical absence is associated with diffuse slow wave spike discharges at 2—2.5 c/s, often longer than 20 seconds, irregular, predominantly anterior and sometimes asymmetric. Atypical absence status occur in the context of chronic epileptogenic encephalopathy (Lennox-Gastaut syndrome and related encephalopathies) and are characterized by a fluctuating confusional state comprising tonic and/or myoclonic elements and/or lateralized ictal manifestations. The term EEG involves continuous or intermittent discharges of irregular and sometimes asymmetrical slow wave spikes or polyspikes. The immediate prognosis is guarded due to recurrence and resistance to therapy [18]. States of absence "de novo" in adults are primarily characterized by toxic and/or metabolic provoking factors [8,19,20] . They occur in elderly patients without pre-existing epilepsy. The electroclinical expression and immediate prognosis are variable. These conditions usually have the meaning of acute symptomatic crises linked to a transient epileptogenic situation, such as, for example, withdrawal or chronic use of psychotropic medications. They do not recur if the etiopathogenic factors are controlled. Genton et al. [21] recently reported a new entity of uncommon but quite characteristic idiopathic generalized epilepsy. They isolated, among all their patients seen during an absence state, eleven patients fulfilling the usual criteria for idiopathic generalized epilepsy and characterized by the occurrence of recurrent, spontaneous absence states which represent in these patients the main type of seizure. Due to an unusual, unrecognized presentation and atypia on the EEG level, these patients with absence state are unrecognized and can make the costs of inappropriate treatment. The patients were often resistant to intravenous benzodiazepines but were all controlled by appropriate anti-absence treatments, sometimes requiring dual therapy when valproate proved insufficient. Absence states with focal elements are observed in the context of pre-existing partial epilepsy, most often extratemporal and result from secondary bilateral synchrony. The EEG includes bilateral and asymmetric ictal discharges. The immediate prognosis is variable. Some of these forms are difficult to differentiate from a frontal complex partial status epilepticus [22,23]. Etiological factors of absence states. The etiological factors of absence states are very diverse. The presence of recurrent and resistant "atypical" absence states occurring in a young subject with pre-existing epilepsy should lead to the search for ring chromosome 20 syndrome, especially if there is a clear context of intellectual deterioration [24]. Endocrine factors are favored in women during periods of genital activity: catamenial period [4,25], pregnancy, immediate postpartum [26], menopause. Medicinal factors are predominant in states of absence occurring "de novo" in the elderly [8,16,27]. The role of psychotropic medications has been highlighted by numerous authors [1,2,9,28,30]. The molecules involved are very diverse in order of frequency and attributability, we find benzodiazepines (during chronic intake and particularly during withdrawal), neuroleptics, tricyclic antidepressants, barbiturates, lithium, meprobamate, viloxazine, methaqualone, monoamine oxidase inhibitors. Many cases have also been attributed to the use of non-psychotropic medications: theophylline, cyclosporine, baclofen, metformin, cimetidine, ifosfamide, ceftazidime, diuretics, piperazine. Several observations occurred following radiological examinations using metrizamide. Metabolic disturbance, isolated or associated with medicinal factors, is common: hyponatremia, hypocalcemia, hypoglycemia, chronic renal failure. Syndromic classification errors with the prescription of narrow-spectrum drugs (carbamazepine, phenytoin, vigabatrin, gabapentin, tiagabine) in the context of idiopathic generalized epilepsy most often involving absences can lead to a paradoxical worsening of epilepsy in the form of recurrent states of absence whose characteristics are often atypical [31].

II. Clinical Case

A 62-year old man had a history of idiopathic generalized epilepsy with tonic-clonic seizures. Treated since the age of 30 years with phenobarbital PB 100 mg/day. He was admitted to the neurology emergency

room for an altered state of consciousness over 24 h after food poisoning .He was disoriented to time and place and a slowness in mental activity. Neurological examination on admission did not show any focal signs .Laboratory tests: routine blood tests, including blood glucose level, blood count, blood ionogram , kidney test, liver test, were normal.

CT scan of the brain were normal. Brain MRI did not show any structural lesions. EEG performed in the emergency room. EEG showed recurrent generalized paroxysms of 3 Hz spike-wave. Normal background activity. Normalization of mental status and EEG pattern after 1mg intravenous clonazepam (Benzodiazepine). Cognitive and behavioral state preserved as documented by neuropsychological tests. Phenobarbital PB was substituted by valproic acid 1500 mg/day.

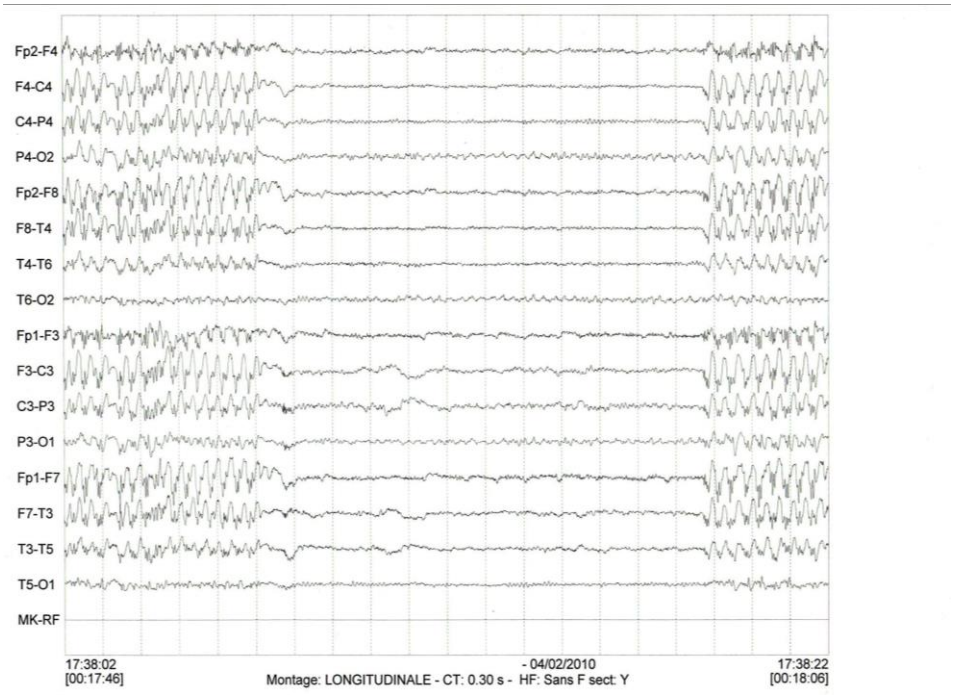

Figure 1. Generalized paroxysms of 3 Hz spike-wave

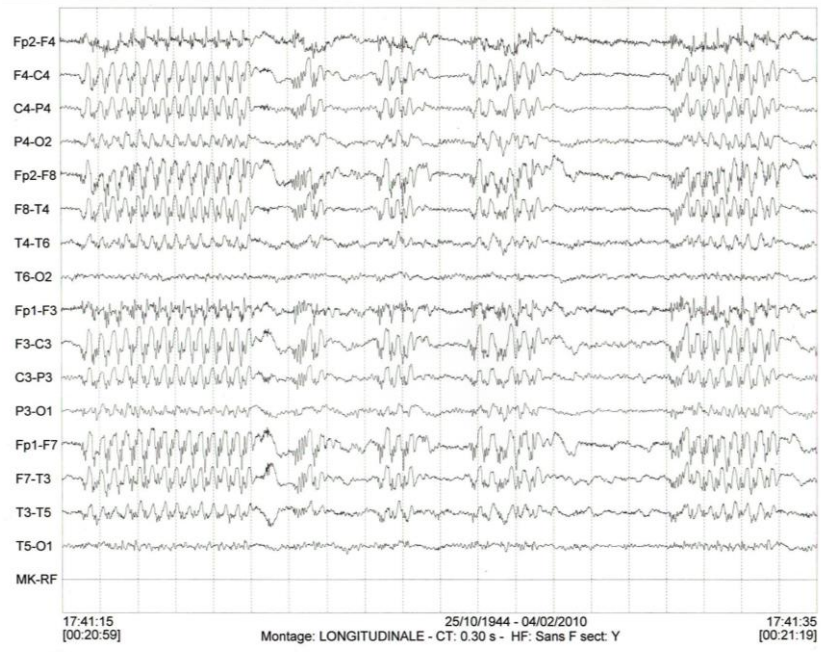

Figure 2. Generalized paroxysms of 3 Hz spike-wave (3 minutes later)

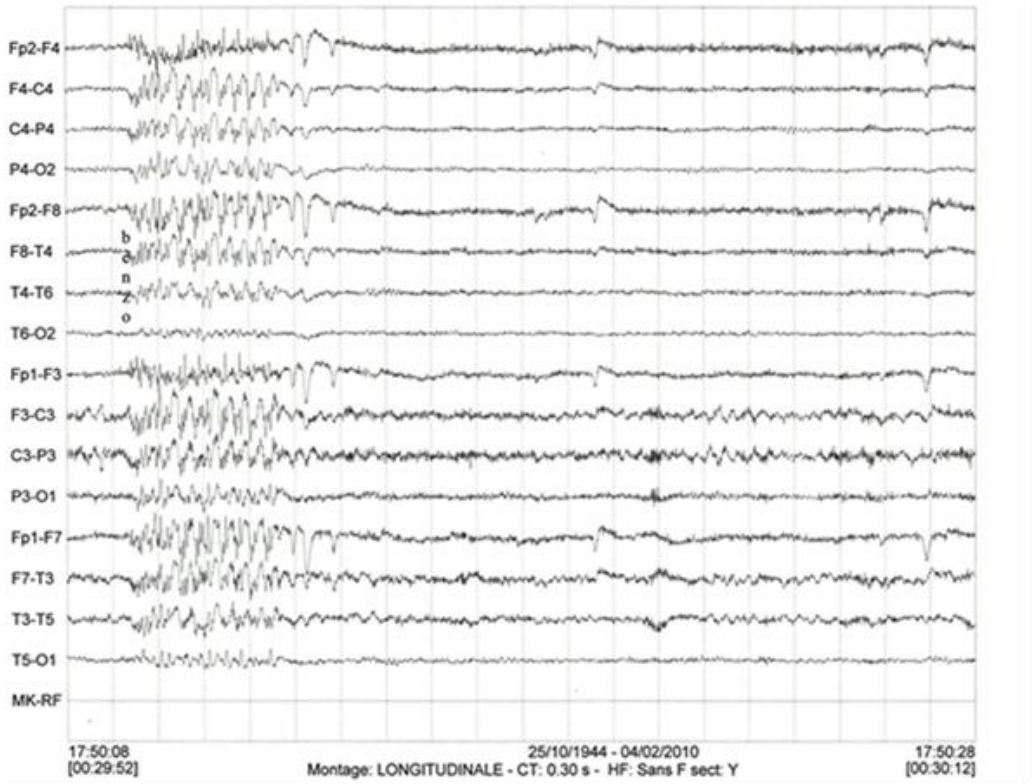

Figure 3. 1mg intravenous clonazepam (Benzodiazepine)

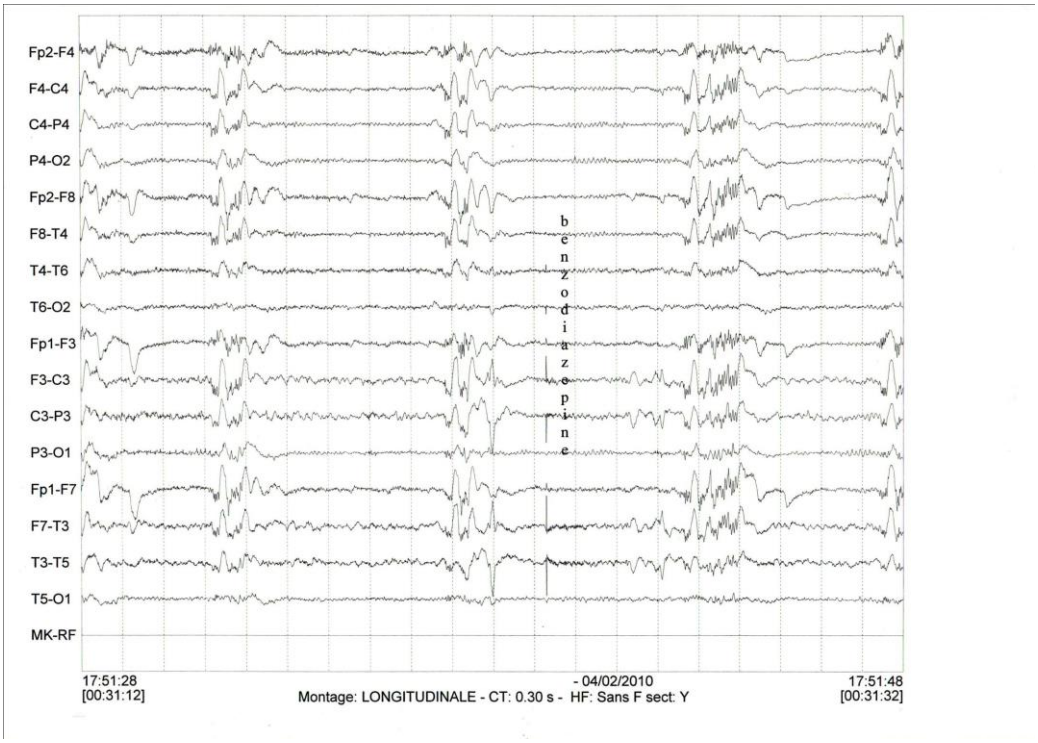

Figure 4. 1mg intravenous clonazepam (Benzodiazepine)

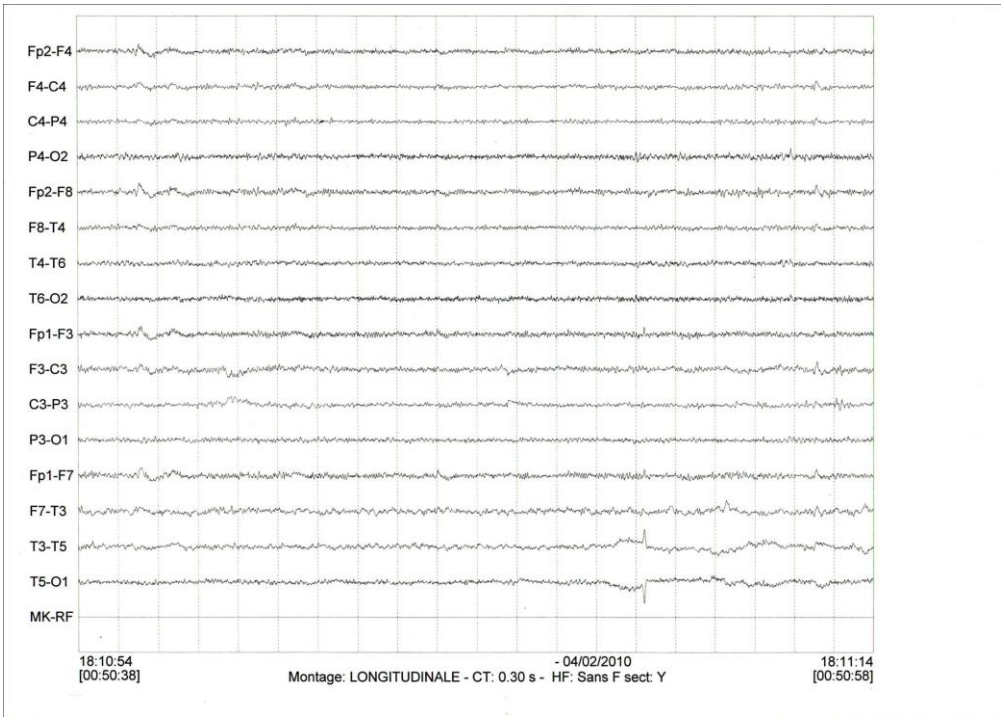

Figure 5. Normalization of EEG pattern

III. Discussion

Our patient presented a typical de novo absence state with late onset, with a history of idiopathic generalized epilepsy evolving since the age of 30 (made up of generalized tonic-clonic seizures). This de novo absence state occurs following to a probable metabolic disorder over 24 hours after food poisoning, typical by its neurophysiological aspect and by its clinical and electrical response to treatment with Benzodiazepine.

Absence status epilepticus can last from a few hours to several days. The cardinal clinical feature is clouding of consciousness (confusion) [32,33,34]. The extent of obscuration is extremely variable with, at one extreme, little more than diminished ideation and expression and deficits in activities requiring sustained attention, sequential organization, or spatial patterning. Amnesia may be mild or even absent. At the other extreme, we find stupor, immobility and muteness. In a typical case, the patient is responsive but moderately confused; simple voluntary actions are performed only after repeated requests. Higher cognitive functions are variably impaired. Motor disturbances occur in approximately 50% of cases, including myoclonus, atonia, rhythmic blinking of the eyelids, and tremors of the lips and face. Facial myoclonus, particularly of the eyelids, is common in typical absence states (and not present in complex partial status). Typically, the patient is in an expressionless state, with slow responses and a stumbling gait. There is often a poverty of speech, and present speech is often hesitant and monosyllabic, and there are long delays in verbal responses. The patient may appear perplexed and rather indifferent and is sometimes agitated. Motor automation is rare. Typical absence state episodes are usually triggered by factors such as menstruation, stopping medications, inappropriate medications (eg, vigabatrin, tiagabine), hypoglycemia, hyperventilation, flashing lights or sharp, lack of sleep, fatigue, stress. Absence state episodes often end in a tonic-clonic seizure but may end spontaneously. Much less often, a tonic-clonic seizure may precede the absence state period.

Typical absence state treatment can usually be quickly and completely removed by treatment with benzodiazepines administered as an intravenous bolus. The usual medications are diazepam at 0.2 to 0.3 mg/kg, clonazepam at 1 mg (0.25 to 0.5 mg in children) or lorazepam at 0.07 mg/kg (0.1 mg/kg in children). Bolus doses can be repeated if necessary. If this proves ineffective, intravenous valproate may be used. In rare cases, people may have repeated episodes of absence state, which may be interrupted at home with oral, rectal, or buccal benzodiazepines. Often, such an intervention will prevent the occurrence of a tonic-clonic seizure.

De novo absence status at the end of life, this curious syndrome manifests itself at the end of adult life. The main symptom is the sudden onset of confusion that can last for hours or even days. Many patients have symptoms of absence epilepsy early in life, who have experienced a long remission, although Panayiotopoulos has suggested that some cases have ongoing phantom absences that are overlooked [34]. Many cases are mistakenly observed as acute dementia or cerebrovascular disease, but the abrupt onset should suggest the possibility of an absence state, and the diagnosis is easily confirmed by EEG. In most cases, the active substance of psychotropic medications or withdrawal (especially benzodiazepines) appears to be the antecedent

cause of the episode, and it is perhaps best to consider this syndrome as a toxic or withdrawal syndrome and a form of acute symptomatic epilepsy and to include other cases of drug-induced absence states, drug discontinuation, or metabolic disorders such as hypoglycemia, hypocalcemia, or uremia in this category. The disease is quickly relieved by intravenous lorazepam (2 to 4 mg) and tends not to recur. Long-term antiepileptic drug treatment is usually not necessary.

IV. Conclusion

Typical ASE diagnosis must be based on EEG pattern and clinical history. Efficacy of intravenous clonazepam in treatment of typical ASE.