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Types and Proposed Etiology of Nonconvulsive Status Epilepticus in Egyptian Children

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Abstract

Introduction: The recognition of convulsive status epilepticus is clinically apparent and easily diagnosed. However, the recognition and diagnosis of non-convulsive status epilepticus (NCSE), especially in children, is more troublesome and complicated.

Objective: Study aimed to define the NCSE syndrome, its classification, and frequency of presentation, proposed etiology and symptomatology.

Patients and Methods: A retrospective study of records of 112 children with the diagnosis of NCSE presented between June 2006 and December 2020 were reviewed. Children were classified according to clinical presentation, EEG pattern and neuroimaging results into a) Typical absence NCSE, b) Atypical absence NCSE, c) Simple Partial NCSE, and d) Complex Partial NCSE.

Results: Of the 112 Patients, 46 were males and 66 females. Age ranged from 6.2-14 years. Clinically, 57% of the children suffer from complex partial NCSE, 31% from atypical absence NCSE, 7% from typical absence NCSE, and 5% from Simple Partial NCSE. Cryptogenic etiology is the most frequent (47%), followed by vascular insults (18%).

Conclusion: Children have a higher risk of NCSE than adults. In this study, most children are known epileptics; cerebrovascular insults were the second frequent cause of NCSE, after cryptogenic. Clinical suspicion, EEG, or long trace EEG permits early diagnosis and intervention.

Keywords: Nonconvulsive status epilepticus; Complex partial status epilepticus; Absence status; Petit mal status; Typical absence status epilepticus; NCSE; EEG

Introduction

The recognition of convulsive status epilepticus is clinically obvious and easily diagnosed [1]. However, the recognition and diagnosis of non-convulsive status epilepticus (NCSE), especially in children, is more difficult [2-4]. The NCSE is often underdiagnosed or diagnosed late because it requires a high index of suspicion and an electroencephalogram. The fact that SE could consist of "delirium, stupor, or coma, cough, or hiccup and a variety of psychic states which have their basis in critical EEG discharges resulting in more or less complete physical or psychic exhaustion" [5,6]. Children with NCSE can present with variety of clinical manifestations including coma, confusion, drowsiness, altered mood, fugue states, aphasia, or vegetative abnormal autonomic symptoms, hallucinations, and paranoid behavior [7]. All these symptoms should occur in the absence of evident convulsive seizure activity (tonic, clonic or tonic-clonic). Despite the lack of convulsive activity, NCSE is thought to result in neuronal injury, and so its recognition and treatment is critical [8]. No consensus exists on the phenomenology, nosology, and classification of NCSE. Some authors use the term to describe all cases with altered sensorium without convulsive movements. The EEG remains the most important investigative and diagnostic tool for confirming NCSE.

Classified NCSE as following [9]:

Generalized Absence SE

- Typical absence SE.
- Atypical absence SE.

- De novo absence SE of late onset.
- Absence SE in other epileptic syndromes.
- Partial SE:
- Non-convulsive CPSE
- Non-convulsive simple partial SE

Boundary zones

- Electrographic SE
- Prolonged postictal confusional state
- Episodic behavioral disturbances and psychosis.
- Clinical manifestations

Typical Absence Status Epilepticus

- The status usually occurs in patients with idiopathic generalized epilepsy.
- It lasts 12 hours or less in over 50 % of patients but can persist for days, weeks or months.
- An episode of TASE is often terminated by an attack of a Tonic-clonic seizure.
- About 20 % show slowing of ideation and expression only.
- The verbal functioning may be well preserved as compared to CPSE. In 50 percent of patients there is marked clouding of sensorium.
- These patients are immobile, able to perform simple tasks after repeated asking, and are often mute or near mute with long delay in verbal response or with monosyllabic answers.
- The appearance is blank or puzzled as if in a trance. About 50 percent of patients show subtle motor features such as myoclonus, atonia, eyelid myoclonia, trembling of lips, facial grimacing and smiling. The presence of symptoms like eyelid myoclonia, myoclonus … etc. are useful clues in differentiating TASE from CPSE [9-11].

Atypical Absence Status Epilepticus

- Occurs in patients with secondary generalized epilepsies, particularly, of the Lennox Gastaut type, and in other cryptogenic or symptomatic generalized syndromes, especially those associated with mental handicap and cerebral damage (12).
- This contrasts with TASE, which occurs in individuals with normal mentality and neurological examination.
- The clinical features of atypical and typical absence SE may show considerable overlap at times.
- Atypical SE, however, shows some characteristics:
- Episodes in atypical SE are longer and more frequent,
- The onset and offset are more gradual; and are often preceded by changes in mood, motor activity for hours or days before the status.
- The status tends to fluctuate, and may evolve into minor motor, myoclonic or tonic seizures.
- Initiation or cessation of atypical absence SE with a tonic clonic seizure is unusual [12-15].

Non-Convulsive Simple Partial Status Epilepticus (SPSE)

- In SPSE, there is no alteration in awareness or responsiveness.
- The location of the ictal discharge determines the clinical manifestations, which may be **sensory**, psychic**,** or autonomic.
- It may manifest as
- prolonged unmotivated fear,
- Autonomic upset e.g., episodes of tachycardia, pupillary dilatation, and incontinence.
- prolonged aphasia (due to discharges arising from the left basal temporal language area), prolonged blindness (status epilepticus amauroticus),
- Visual hallucinations, ictal hemiplegia.
- Prominent psychic symptomatology with severe autistic or schizophrenic features.

Complex Partial Status Epilepticus

- Although earlier studies concluded that CPSE was rare; recent reports indicate that it is probably under reported. It is possible that atypical absence SE being reported may in fact be cases of CPSE.
- Until recently, CPSE had been equated with temporal lobe status epilepticus.
- Seizure origin was extratemporal in most of properly investigated patients.
- Thus, CPSE is not analogous with temporal lobe origin; on the contrary, extratemporal sites, notably frontal lobe may often be the responsible site.
- Aicardi observed that bilateral continuous paroxysmal activity could occur in partial status, especially with frontal lobe lesions making it resemble an absence SE.
- CPSE is associated with a wide variety of clinical patterns, which may be indistinguishable from those of atypical absence SE.

Criteria for diagnosis of CPSE [17,18]

- Recurrent complex partial seizures without full recovery of consciousness between seizures, or a continuous

"epileptic twilight state" with cycling between unresponsive and partially responsive phases (lasting more than 30 Minutes).

- Ictal EEG with recurrent epileptiform patterns like those seen in complex partial seizures.
- A prompt observable effect of intravenous antiepileptics on both ictal EEG and clinical manifestations of status.
- Interictal EEG with a consistent epileptiform focus.
- The alteration in sensorium can vary from bland confusion, either continuous or intermittent, to agitated unresponsiveness with bizarre, almost psychotic activity (15,16,19)
- Speech patterns are often abnormal with perseveration, aphasia, echolalia, confabulation, or just slow responses.
- It may start with Motor features in the form of adversive posturing, tonic posturing, and simple automatisms.
- Autonomic disturbances include belching, flatulence, change in color, fever, or pupillary changes.

Objective

The objective of this study is to describe the syndrome definition, classification, and its presentation frequency, proposed etiology and symptomatology of pediatric patients with NCSE who attended the Mataryia Teaching Hospital in the period between June 2006 and December 2020.

Patients and Methods

3583 children with or without behavioral changes, were admitted during the period from June 2006 to December 2020 to El-Matryia Teaching Hospital; those were admitted with disturbed levels of consciousness. All patients with disturbed level of consciousness had thorough clinical examination, routine laboratory investigations, and brain imaging, according to the hospital routine protocol. Of the firstly reviewed 518 children cohort, who had continuous EEG records for at least 12 hours (long trace EEG), 406 were excluded due to either normal EEG record, or having either metabolic disorder (e.g., diabetic ketoacidosis, hyperammonemia etc.), acute vascular brain insult (cerebrovenous thrombosis, ischemic insult, hemorrhagic insult etc.), or Space occupying lesions; the remaining 112 children with proven non-convulsive status epilepticus (NCSE) represent the subjects of the current study. EEG recordings were performed on 21 channel EEG instruments using the international 10–20 electrode system. Continuous EEG monitoring records lasting at least 12 h were available for every patient. Thus, a retrospective study of records of the 112 children with the diagnosis of NCSE presented between June 2006 and December 2020 were fully analyzed; those children were classified, according to clinical presentation, EEG pattern and neuroimaging results, into a) Typical absence NCSE, b) Atypical absence NCSE, c) Simple Partial NCSE, and d) Complex Partial NCSE.

Results

3% of the admitted patients with disturbed consciousness found to suffer from NCSE; this low percentage was due to the unavailability of EEG, as only 14.4% of the total admitted children (3583) had EEG done at admission. The actual percentage may reach 21.6%, as this is the percentage of children with positive EEG for NCSE in the cohort (112/518), who performed EEG at time of admission. Of the 112 Patients, 46 were males and 66 females; age ranged from 6.2-14 years. Clinically, 57% of the children suffer from complex partial NCSE, 31% from atypical absence NCSE, 7% from typical absence NCSE, and 5% from Simple Partial NCSE (Table 1, Figure 1).

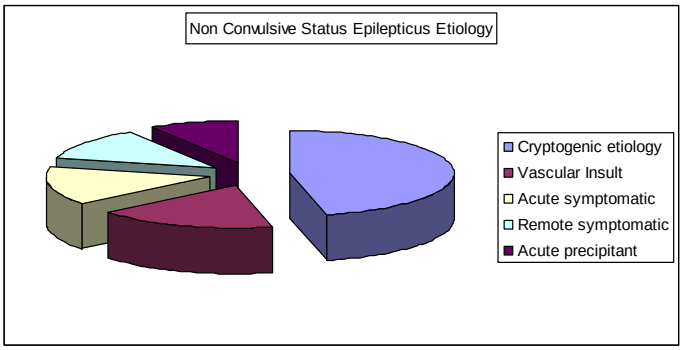

Figure 1: *Classification of study subjects according to dominant type of Non-convulsive Status Epilepticus.*

Cryptogenic etiology is the most frequent (47%), followed by presumed perinatal vascular insults (18%) (Table 2, Figures 2-6).

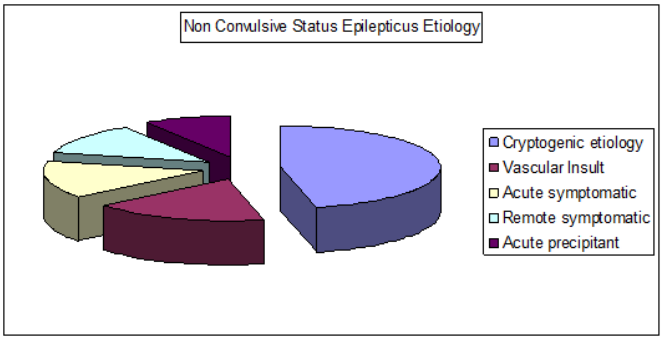

Figure 2: *Non-Convulsive Status Epilepticus Etiology.*

Discussion

The clinical presentation of NCSE is heterogeneous and the EEG is an essential diagnostic tool [23,24]. In patients with moderate or severe mental retardation, with psychiatric disorders and especially in critically ill patients, a high degree of clinical suspicion is often necessary to establish the diagnosis of NCSE

within the shortest possible time. Many reports use the term "NCSE" in an all-encompassing manner, applying this definition for patients with varied clinical manifestations and etiologies, in a way that impairs the distinction between focal and generalized forms, as well as the assessment of prognosis in different situations, thus, taking into consideration data such as etiology, age, clinical and electrographic presentation is of importance, given the fact that the course and prognosis of focal and generalized forms are quite different and possibly dependent on those aspects.

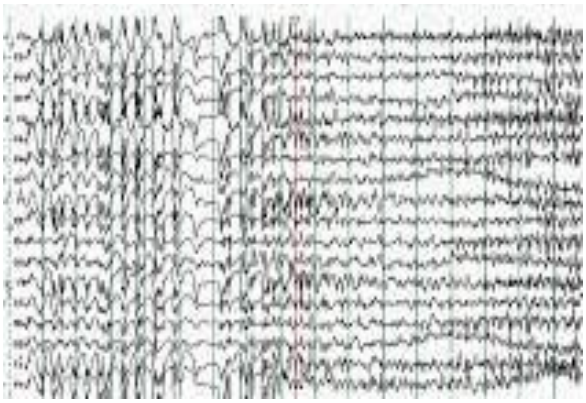

Figure 3: *EEG trace of Typical Absence NCSE*

Figure 4: *EEG trace of Atypical Absence NCSE.*

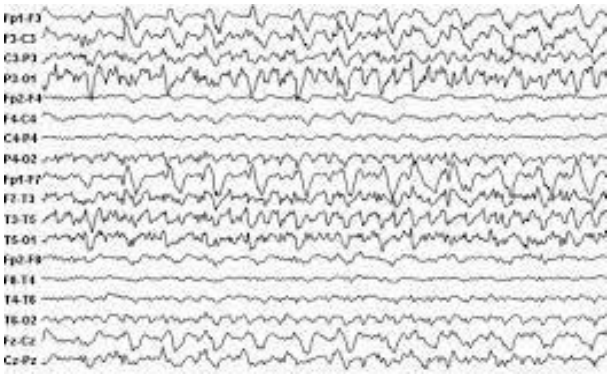

Figure 5*: EEG trace of complex partial NCSE.*

Figure 6: *Generalized periodic epileptiform discharge.*

In our study, NCSE occurs in children with or without a history of epilepsy, but most are known epileptics. Presumed Perinatal and pediatric strokes are frequent etiology of NCSE, so prolonged follow up in those groups of patients to early detect NCSE is mandatory. EEG can differentiate between subtypes when clinical presentation is non-decisive, that confirms the golden role of EEG in diagnosis and classification [20-24]. Clinical manifestations of NCSE in our study children include cognitive and/or behavioral impairments; symptoms characteristically fluctuate in intensity over time. Alteration of the conscious level characteristic of the syndrome can range from slowing of ideation to complete loss of awareness. Children are usually able to carry out simple tasks and even complex daily life activities. Speech may be slurred, incoherent, sometimes with echolalia and palilalia. Children may show behavioral, language and cognitive deterioration. Pediatric populations are at increased risk of NCSE development than adults, due to the addition of typical and atypical absence status epilepticus portion. Several neurological conditions, among them metabolic encephalopathy, drug intoxication, prolonged postictal status, and transitory global amnesia, require differential diagnosis with NCSE [7]. Behavioral changes that may occur in these cases can lead to an erroneous diagnosis of psychiatric conditions such as depression, psychosis, and hysteria; however, the most difficult differential diagnosis is by far between these two main types of NCSE, i.e., generalized with absence seizures and focal with complex partial seizures [26]. This distinction is especially difficult when dealing with focal NCSE of frontal origin, when the discharges tend to propagate in a relatively diffuse pattern [27]. A previous history of partial epilepsy and focal or lateralized EEG finding is of help for the diagnosis of complex partial SE, whereas the presence of generalized idiopathic epilepsy and generalized discharges supports the diagnosis of absence SE**.** The NCSE patients' EEGs in our work showed focal, multifocal, generalized, or periodic lateralized

epileptiform discharge. Periodic lateralized epileptiform discharges are mainly recorded from patients with CPSE, previous vascular insults or simple partial NCSE presented by disturbed behavior and other psychiatric manifestations [10].

Table 1: *Classification of study subjects according to dominant type of Non-convulsive Status Epilepticus.*

NCSE Types	Typical absence NCSE	Atypical absence NCSE	Simple Partial NCSE	Complex Partial NCSE
Number & %	8 (7%)	34 (31%)	6 (5%)	64 (57%)

Table 2: *Etiology of non-convulsive status epilepticus.*

NCSE	Cryptogenic etiology	Vascular Insult	Acute symptomatic	Remote symptomatic	Acute precipitant
No.=112	52 (47%)	20 (18%)	16 (14%)	14 (12%)	10 (9%)

Conclusion

Children have a higher risk of NCSE than adults to develop Non-Convulsive Status Epilepticus. In this study, most children are known epileptics; cerebrovascular insults were the second frequent cause of NCSE, after cryptogenic. Clinical suspicion, EEG, or long trace EEG permits early diagnosis and intervention.

Conflict of Interest

No conflict of interest

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