ELECTRICAL STATUS EPILEPTICUS IN SLEEP SPECTRUM IN CHILDREN

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ABSTRACT
ELECTRICAL STATUS EPILEPTICUS IN SLEEP (ESES) REPRESENTS AN ELECTROENCEPHALOGRAPHIC PATTERN CONSISTING IN GENERALIZATION AND ACTIVATION OF EPILEPTIFORM DISCHARGES DURING NON-REM SLEEP. IT IS ASSOCIATED WITH A HETEROGENEOUS GROUP OF CLINICAL SYNDROMES AFFECTING CHILDREN IN VARYING DEGREES OF SEVERITY, WITH COMMON AND OVERLAPPING FEATURES. THE IMPORTANCE OF RECOGNIZING AND TREATING ESES LIES IN THE IMPROVEMENT OF THE CHILD'S PSYCHOEMOTIONAL AND COGNITIVE STATUS AND EVOLUTION. IN THIS ARTICLE, WE PRESENT DIFFERENT ELECTROCLINICAL CASES WITHIN THE ESES SPECTRUM, THAT HAVE BEEN DIAGNOSED AND MANAGED IN OUR CLINIC.

KEY WORDS*: ESES, CHILDREN, COGNITIVE, SLEEP*

DEFINITION OF ESES

Electrical Status Epilepticus in Sleep (ESES) represents an electroencephalographic pattern illustrating a marked potentiation of epileptiform activity during non-REM sleep, leading to near - continous bilateral or occasionally lateralized 1-3 Hz spike-wave (S-W) complexes, that occupy a significant proportion of the non-REM sleep. The exact proportion varies, according to the literature, between 25-85%, with no clear cutoff value being universally recognized[5]. We know that the epileptiform activity tends to be more marked in the first sleep cycles, especially in sleep-deprived patients. However, consensus on the definition of ESES, regarding the exact portion of sleep analyzed, still needs to be met.

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[5]Scheltens- de Boer M., Guidelines *for EEG in encephalopathy related to ESES/CSWS in children*, Epilepsia, 2009; 50(Suppl. 7):13–17

CLINICAL SYNDROMES

ESES manifests in clinical syndromes which tend to evolve according to common patterns, displaying an age-related evolution, summing the presence of seizures, the aforementioned sleep potentiation of epileptiform activity and neuropsychologic regression, as the interictal electroencephalographic discharges tend to persist even after seizure freedom[6].

Initially, ESES was associated with the syndrome of Continous Spikes and Waves during Sleep (CSWS), a childhood epileptic encephalopathy, the terms CSWS and ESES being used interchangeably. In time, however, ESES has been recognized in relation to more clinical entities, leading to a heterogeneous group, with common and overlapping features, all pertaining to a continuum along the ESES spectrum.

CSWS and Landau Kleffner Syndrome (LKS) are severe epileptic encephalopathies associated with regression in the child's development. While CSWS is associated with a global developmental regression manifesting in previously normal or abnormal children, Landau Kleffner Syndrome particularly consists in regression in language development.

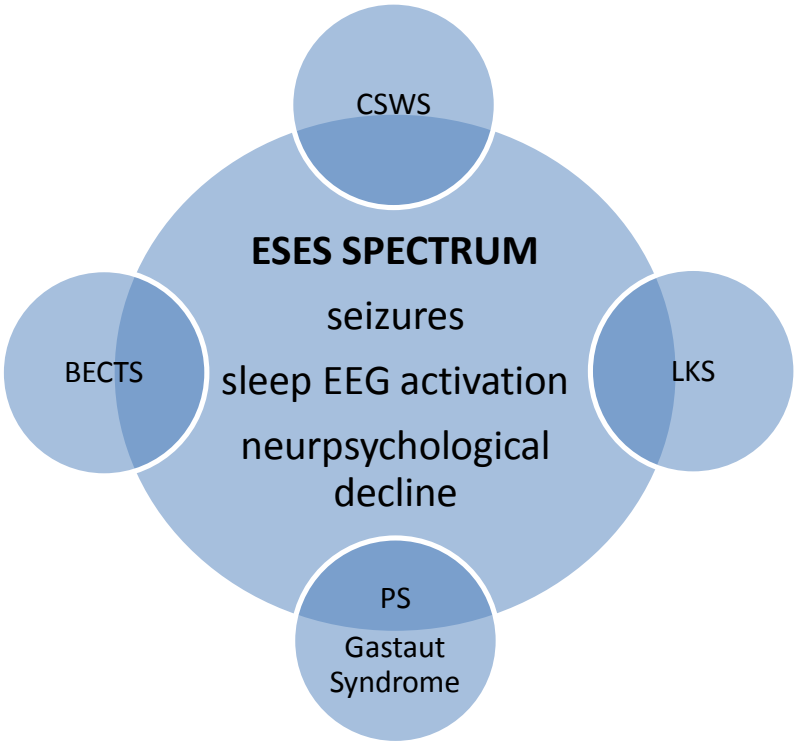
At the other end of the spectrum lie the pediatric focal epileptic syndromes, previously thought to constitute benign conditions, associated with a good outcome and no negative impact on the child's development .

Benign childhood epilepsy with centrotemporal spikes (BECTS) is the most frequent pediatric focal epileptic syndrome. Most seizures appear at the transition from wakefulness to sleep, are short and consist in unilateral sensory and motor manifestations in the oropharyngeal area. The EEG frequently shows a normal interictal tracing with superimposed usually unilateral clusters of high voltage spikes or sharp waves in the centrotemporal area, which tend to activate and generalize during non-sleep, up to a pattern consistent with ESES.

Panayiotopoulos Syndrome (PS) and Late Onset Childhood Occipital Epilepsy (Gastaut), separated by age of onset and clinical aspect of seizures, are both associated with unilateral epileptiform discharges in the occipital area, which also tend to activate during sleep.

All partial epileptic syndromes mentioned above are usually known to be associated with spontaneous improvement before puberty. However, careful psychological monitoring might reveal mild cognitive impairment, learning disabilities or behavioral problems, raising the need to monitor the child through sleep EEGs and psychological evaluations, periodically.

[6] Sanchez Fernandez I. et al., *Electrical Status Epilepticus in Sleep: Clinical Presentation and Pathophysiology*, Pediatric Neurology, 2012; 47(6):390-410



EEG ASPECTS

The following figures illustrate cases of patients with various clinical epileptic syndromes, united under the ESES spectrum, which have been diagnosed and managed in our clinic.

Case 1 - CSWS

Figure 1 shows an interictal EEG recorded during wakefulness (500µV sensitivity) in a 6 year old child with global developmental delay, polymorphic seizures and underlying hypoxic ischemic perinatal injury. The child was on valproic acid. Generalized aperiodic 3 Hz spike-wave complexes.

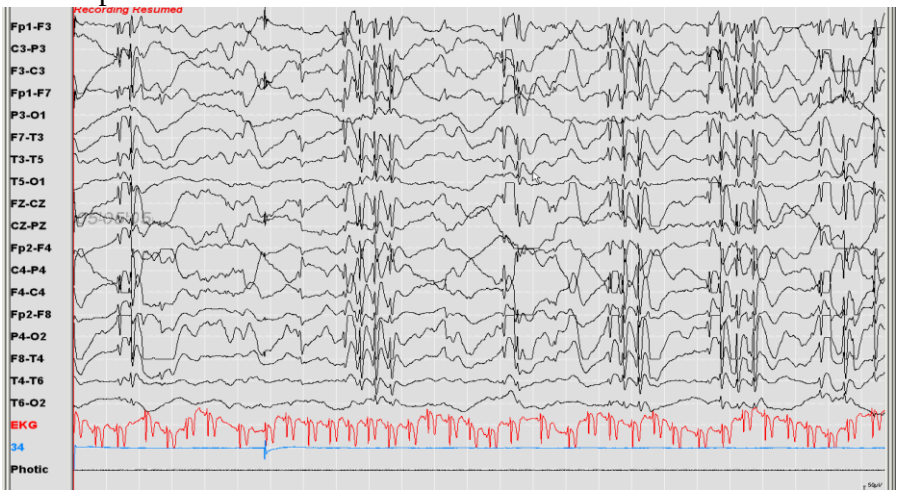


Figure 2: Sleep EEG (500µV sensitivity) - ESES/CSWS. Continuous generalized 2-3 Hz spike-and -waves.

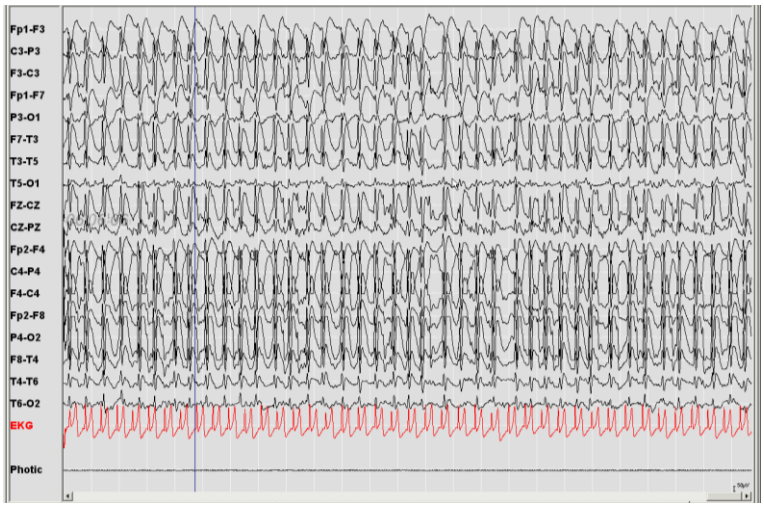
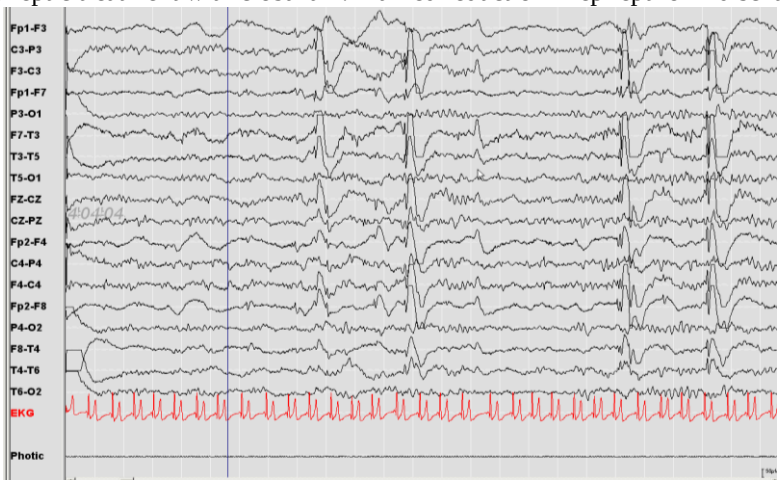
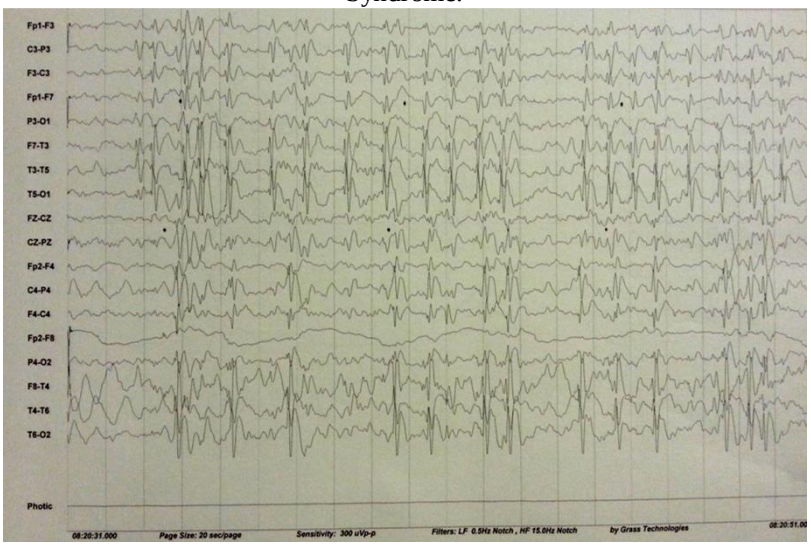


Figure 3: EEG (500µV sensitivity) aspect following 1 month of ACTH treatment and supplementation of antiepileptic treatment with clobazam. Marked reduction in epileptiform discharges.



Case 2 - LKS

Figure 4: Sleep EEG in a 11 year old child admitted initially to the ENT department for loss of expressive language and suspicion of hypoacus. Even though the child had no other clinical signs, nor epileptic seizures, the sleep EEG revealed near-continuous 1-2 Hz spike-wave complexes, prompting to Landau Kleffner Syndrome.



Case 3 - BECTS

Figure 5: Awake EEG (500µV sensitivity) in an otherwise healthy 6 year old boy with one focal sensory-motor seizure and one generalized seizure, with no identifiable lesions on the cerebral MRI. Spikes and spike-wave complexes over the left centro-temporal area.

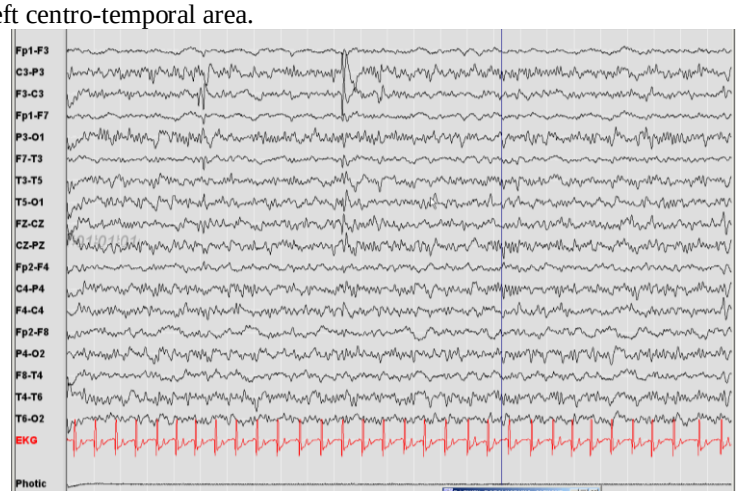
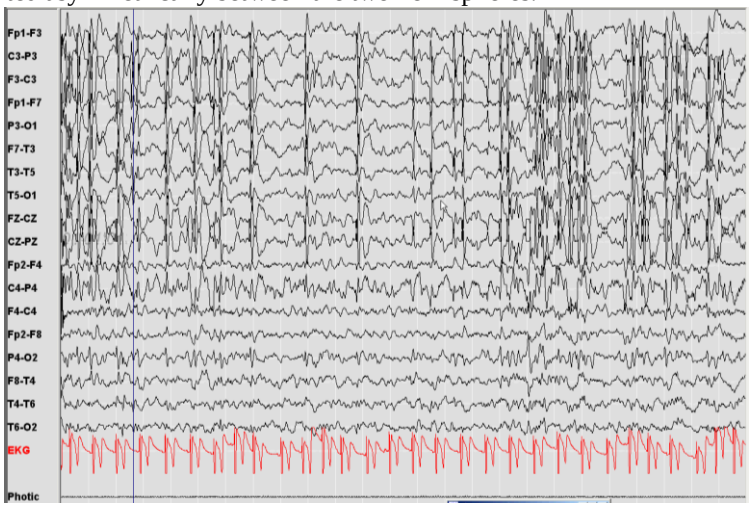


Figure 6: Sleep EEG (500µV sensitivity) - important activation of epileptiform discharges, which are near-continous but distributed asymmetrically between the two hemispheres.



PROGNOSIS AND MANAGEMENT

The syndromes associated with ESES have a common tendency towards remission of seizures into adolescence. However, the epileptiform EEG discharges may persist and there is a residual and often progressive cognitive and neuropsychological decline.

Psychological monitoring is therefore an important aspect of the management of children with ESES. Simple psychological assessment at various times might miss important information unless directed precisely at the evaluation of different abilities, while keeping in mind the overall development of the child and his medical history.

Because of the impact on the child's development and cognitive function, an electrical aspect consistent with ESES needs prompt treatment irrespective of clinical seizures, in order to improve neuropsychological functions and to prevent further decline.

The therapeutical options include corticosteroids, adrenocorticotrophic hormone and antiepileptic drugs such as benzodiazepines including clobazam, levetiracetam, valproic acid, ethosuximide[7]. The therapeutic schemes vary according to the underlying pathology and type of epileptic syndrome. Some options include: oral benzodiazepines combined with valproate

[7] Veggiotti P., Pera M. C. et al., *Therapy of Encephalopathy with Status Epilepticus During Sleep (ESES/CSWS syndrome), an update*, Epileptic Disord, 2012; 14 (1): 1-11

or steroids such as ACTH (80 UI daily with a taper of 3 months) or high dose prednisolone (2-5 mg/kg daily with a taper of 3 months)[8].

CONCLUSIONS

The article aims to bring into attention the electroclinical entity of ESES by illustrating it with cases from our pediatric neurology practice. The importance of performing sleep EEGs and psychological assessments comes from the understanding of the impact that aberrant brain electrical activity has on the child's development.

[8] Panayiotopoulos C.P., *A Clinical Guide to Epileptic Syndromes and their Treatment*, Springer; 2nd edition, 2007; 258- 259

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