# Electrical status epilepticus during sleep: A study of 67 patients

*Sibel Öz Yıldız[1], Ceren Günbey[1], Kader Karlı Oğuz[2], Gökçen Konuşkan[1], Göknur Haliloğlu[1], Dilek Yalnızoğlu[1]*

[1]Hacettepe University Faculty of Medicine, Department of Pediatric Neurology, Ankara, Turkey
[2]Hacettepe University Faculty of Medicine, Department of Neuroradiology, Ankara

ICNC 2022
17th INTERNATIONAL CHILD NEUROLOGY CONGRESS
ANTALYA, TURKEY I OCTOBER 3-7, 2022

## INTRODUCTION

Electrical status epilepticus during sleep (ESES) is also known as continuous spike and wave during sleep (CSWS). A typical electroencephalogram (EEG) pattern, progressive deterioration in cognitive, behavioral and motor skills, and an increase in spike-wave discharges at a frequency of 1.5 to 3 Hz during sleep are the characteristics of this epileptic encephalopathy specific for childhood (1).

Electrical status epilepticus during sleep commonly manifests between the ages of 2 and 12, with a peak between the ages of 3 and 5 years (2). Regarding the etiology for ESES; congenital or acquired structural brain abnormalities such as neuronal migration disorders, hydrocephalus, thalamic pathologies, chromosomal/genetic disorders, disruption of sleep homeostasis, and non-specific immune response have been reported to play a role (3-5).

## OBJECTIVES

We aimed to study clinical features, etiology, neuroimaging findings, treatment, and outcome in patients with ESES.

## MATERIAL AND METHODS

The study included 67 patients diagnosed with ESES pattern on EEG ( 40 males, 27 females) between 2015-2020 at our center. We retrospectly reviewed data on clinical, EEG and neuroimaging findings, neuropsychometric test results, treatments and response to treatment. The study comprised patients who had epileptic activation more than 50% in sleep EEGs, cognitive assesment was reported via neuropsychometric tests and/or clinical judgement.

## RESULTS

The median age at the initial admission was 60 months (0.03-124). Main symptom at admission was seizure 86.5% (n=58). The median age at the onset of ESES, and follow-up duration was 97 months (22-193), and 28 months (4-69), respectively. At the time of ESES diagnosis, 48 patients had ongoing seizures; 13 had focal onset, 35 had generalized seizures. Of 67 patients, 25 were on monotherapy, 40 were on polytherapy, 2 patients were not on anti- seizure medication (ASM). After ESES diagnosis, ASM treatments were adjusted. Most common ASMs were valproate and benzodiazepines. Median duration of recovery from ESES was 24.5 months (range 4–69 months in 42/67 (62%) of the patients. The leading etiology was structural causes (n= 18 acquired, n= 4 congenital) detected in 22 patients (32.8%). When we evaluated ESES with respect to neuroimaging findings; ESES resolved in 53.7% of patients with abnormal neuroimaging findings, and 79.2% of patients with normal findings ($p<0,05$). After resolution of ESES, 65.9% of patients with abnormal neuroimaging findings had borderline IQ or intellectual disability (ID), 37.5% of those with normal neuroimaging had borderline IQ or ID ($p<0,05$). At final visit; 5 patients showed improved cognition, 28 patients had cognitive decline, and 34 patients remained similar compared to baseline evaluation.

## CONCLUSIONS

Electrical status epilepticus during sleep has a complex etiological profile, and cognitive impairment is seen in majority of patients. The treatment choices are based on the centers' experience, mostly retrospective studies, and case studies. Further studies are required to develop standard treatment approaches and reduce the risk of cognitive impairment in patients with ESES.

## REFERENCES

1.Koutroumanidis, M., Arzimanoglou, A., Caraballo, R., Goyal, S., Kaminska, A., Laoprasert, P., Oguni, H., Rubboli, G., Tatum, W., Thomas, P., 2017. The role of EEG in the diagnosis and classification of the epilepsy syndromes: a tool for clinical practice by the ILAE Neurophysiology Task Force (Part 2). Epileptic Disord. 19, 385–437.

2. Samanta D, Al Khalili Y. Electrical Status Epilepticus In Sleep. 2021 Jul 7. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2021 Jan.

3. Van den Munckhof B, Zwart AF, Weeke LC, Claessens NHP, Plate JDJ, Leemans A, Kuijf HJ, van Teeseling HC, Leijten FSS, Benders MJN, Braun KPJ, de Vries LS, Jansen FE. Perinatal thalamic injury: MRI predictors of electrical status epilepticus in sleep and long-term neurodevelopment. Neuroimage Clin. 2020;26:102227.

4. Kessi M, Peng J, Yang L, Xiong J, Duan H, Pang N, Yin F. Genetic etiologies of the electrical status epilepticus during slow wave sleep: systematic review. BMC Genet. 2018 Jul 6;19(1):40.

5. Hirsch E, Caraballo R, Bernardina BD, Loddenkemper T, Zuberi SM. Encephalopathy related to Status Epilepticus during slow Sleep: from concepts to terminology. Epileptic Disord. 2019 Jun 1;21(S1):5-12.

## CONTACT

Sibel Öz Yıldız, MD
e-mail: dr_sibeloz@hotmail.com