

# P-366 COMPARISON OF CORTICOSTEROIDS VERSUS CLOBAZAM TREATMENT OF EPILEPTIC ENCEPHALOPATHY WITH ELECTRICAL STATUS EPILEPTICUS FOR ELECTROCLINICAL REMISSION

İlknur Cankurt<sup>1</sup>, Recep Kamil Kılıç<sup>1</sup>, Tugba Hirfanoglu<sup>1</sup>, Esra Serdaroglu<sup>1</sup>, Ayşe Serdaroğlu<sup>1</sup>, Ercan Demir<sup>1</sup>, Ebru Arhan<sup>1</sup>

<sup>1</sup>Gazi University School of Medicine, Department of Pediatric Neurology, Ankara



### **INTRODUCTION**

ESES (electrical status epilepticus during slowwave sleep), is a self-limited epilepsy syndrome of which is seen in childhood, especially characterized by cognitive and behavioral impairments, with or without clinical seizures.(1,2) Numerous investigations have shown that continuous epileptic discharges during sleep not only increase the risk of clinical seizures but also disrupt sleep, memory consolidation, learning, and general cognition.(3,4 The severity of cognitive impairment and poor prognosis are correlated with the length of ESES; the longer ESES lasts, the more severe the neuropsychological damage and the worse the prognosis.(5) As a result, an early and effective treatment strategy is needed to improve the child's prognosis. The purpose of therapy is to improve cognitive function, as well as to eliminate the electrical state and control seizures. There is currently no consensus on how to treat ESES, which is a highly challenging epileptic syndrome in children. There is also no scientific consensus on how to measure EEG abnormalities and assess medication efficacy. (6-

# **OBJECTIVE**

The purpose of this study was to retrospectively compare electroclinical features, spike wave index (SWI) and subjective neuropsychological assessment as measures to evaluate treatment efficacy of clobasam versus corticosteroids after 6 months in children with Epileptic encephalopathy with electrical status epilepticus in sleep (ESES).

### MATERIAL-METHOD

We retrospectively included patients with ESES treated with either corticosteroids (intravenous methylprednisolone pulses or daily oral prednisolone) or oral clobazam for 6 months between January 2018- December 2021. Medical-files were evaluated for data concerning the caregiver responses regarding deterioration and improvement and with respect to clinical judgement. Clinical improvement was graded as full, more than 75%, more than 50% and less than 50%.

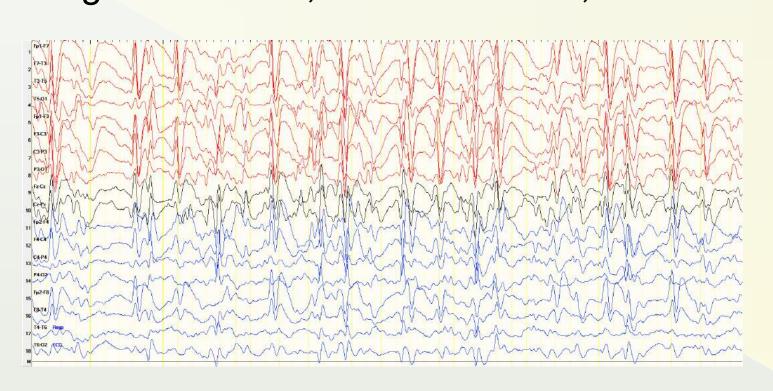




Figure I: 14 year old male before treatmet of pulse steroid

Figure II: Same patient 1 month after pulse steroid therapy

#### **RESULTS**

A total of 30 children received either corticosteroid or clobasam treatment for ESES and were included in the analysis. Eighteen received clobasam (1-1.2 mg/kg/day for 6 months) and 12 were treated with corticosteroids (monthly intravenous methylprednisolone or oral prednisolone). Nine out of twelve patients receiving corticosteroids had SWI reduction and six of them achieved a greater than 50% reduction in SWI. Sixteen patients receiving clobasam had SWI reduction and only six of them had an SWI reduction greater than 50% reduction (p = 0.001). We found that more patients receiving corticosteroid treatment, as compared to clobasam treatment, had a clinical imporovement more than 50% (9/12 vs 6/18, p: 0.001).

#### REFERENCES

- 1. van den Munckhof B, Arzimanoglou A, Perucca E, van Teeseling HC, Leijten FSS, Braun KPJ, Jansen FE; RESCUE ESES study group. Corticosteroids versus clobazam in epileptic encephalopathy with ESES: a European multicentre randomised controlled clinical trial (RESCUE ESES\*). Trials. 2020 Nov 23;21(1):957. doi: 10.1186/s13063-020-04874-2. PMID: 33228736; PMCID: PMC7686710.
- 2. Arican P, Gencpinar P, Olgac Dundar N, Tekgul H. Electrical Status Epilepticus During Slow-wave Sleep (ESES): Current Perspectives. J Pediatr Neurosci. 2021 Apr-Jun;16(2):91-96. doi: 10.4103/jpn.JPN\_137\_20. Epub 2021 Jul 2. PMID: 35018175; PMCID: PMC8706590.
- 3. Cao D, Chen Y, Liao J, Nariai H, Li L, Zhu Y, Zhao X, Hu Y, Wen F, Zhai Q. Scalp EEG high frequency oscillations as a biomarker of treatment response in epileptic encephalopathy with continuous spike-and-wave during sleep (CSWS). Seizure. 2019 Oct;71:151-157. doi: 10.1016/j.seizure.2019.05.023. Epub 2019 May 30. PMID: 31351306.
- 4. van den Munckhof B, van Dee V, Sagi L, Caraballo RH, Veggiotti P, Liukkonen E, Loddenkemper T, Sánchez Fernández I, Buzatu M, Bulteau C, Braun KP, Jansen FE. Treatment of electrical status epilepticus in sleep: A pooled analysis of 575 cases. Epilepsia. 2015 Nov;56(11):1738-46. doi: 10.1111/epi.13128. Epub 2015 Sep 4. PMID: 26337159.
- 5. Pera MC, Brazzo D, Altieri N, Balottin U, Veggiotti P. Long-term evolution of neuropsychological competences in encephalopathy with status epilepticus during sleep: a variable prognosis. Epilepsia. 2013 Oct;54 Suppl 7:77-85. doi: 10.1111/epi.12313. PMID: 24099059.
- 6. Saltik S, Uluduz D, Cokar O, Demirbilek V, Dervent A. A clinical and EEG study on idiopathic partial epilepsies with evolution into ESES spectrum disorders. Epilepsia. 2005 Apr;46(4):524-33. doi: 10.1111/j.0013-9580.2005.45004.x. PMID: 15816946.
- The Principles of Perinciples of Pediatric Patients with ESES. Generalized Enilopsy or Focal Enilopsy Enilopsy Res. 2020 Nov:167:106351. doi:

## CONCLUSION

Methylprednisolone is a medium-acting hormonal substance that, along with prednisone, has been used extensively to treat complex types of epilepsy with significant efficacy. It also has specific therapeutic benefits in ESES in terms of psychosomatic impairment and enhancing neurodevelopmental deficits in minimizing children.(9) In conclusion, we discovered that corticosteroid regimen was superior to clobasam controlling seizures, reducing abnormalities, and increasing cognition. Children with ESES and cognitive impairment may be candidates for hormone therapy at a young age, but more research with bigger sample sizes is required to determine the time of hormone beginning and recurrence rates.

Compared with clobasam, corticosteroids were found to be more effective in clinical and electrographical assessments. Children with ESES and cognitive impairment may benefit from corticosteroid therapy, but bigger sample sizes and more follow-up studies are required to determine the recurrence rates.

Correspondence: Dr. İlknur Cankurt
Gazi University School of Medicine, Department of Pediatric Neurology, Ankara

email: ilknurcankurt@gmail.com