Continuous spikes and waves during sleep

Continuous spikes and waves during sleep (CSWS) is an epileptic encephalopathy characterized by generalised epileptiform activity and neurocognitive dysfunction with electroencephalogram (EEG) pattern of electrical status epilepticus during sleep.

Electrical status epilepticus during sleep (ESES) is now the term typically used to describe solely the electrographic findings, while the CSWS syndrome remains a clinical diagnosis of the constellation of neurocognitive deterioration from baseline, affecting motor, language, cognitive, or behavioral development, and a significant proportion of non-REM sleep disrupted by spike-wave discharges ¹⁾.

Causes and outcome are diverse and treatment is mainly empirical.

Etiology

A cohort in the largest monocentric observational study on treatment effects in children with CSWS, confirms CSWS as an age-dependent epileptic encephalopathy. Structural brain abnormalities are most frequent, but genetic causes are increasingly identified. More specific criteria for the diagnosis and treatment goals should be elaborated and implemented based on evidence. ²⁾.

Case series

In a retrospective descriptive analysis of clinical and EEG data of children with CSWS diagnosed between 1998 and 2018 at the University Hospital Heidelberg. Ninety-five children were included with a median age at diagnosis of 5.4 years. A structural/metabolic aetiology was found in 43.2%, genetic alterations in 17.9%, while it remained unknown in 38.9%. The proportion of patients with genetic aetiology increased from 10.3% (1998-2007) to 22.8% (2008-2018). On average, each patient received 5 different treatments. CSWS was refractory in >70% of cases, steroids and neurosurgery were most effective. No difference was observed between children with CSWS or Near-CSWS (Spike-Wave-Index 40-85%).

This cohort confirms CSWS as an age-dependent epileptic encephalopathy. Structural brain abnormalities were most frequent, but genetic causes are increasingly identified. More specific criteria for the diagnosis and treatment goals should be elaborated and implemented based on evidence.

Significance: This study is the largest monocentric observational study on treatment effects in children with CSWS, providing data for diagnostic and therapeutic decisions ³⁾.

References

1)

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2) , 3)

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