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Epilepsy with Continuous Spike-Waves during Slow Sleep: When and How Should We Treat?

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Epilepsy with continuous spike-waves during slow sleep (CSWS), which is one of the hallmarks of electrical status epilepticus during slow wave sleep (ESES), is a functional disorder with the following features: severe paroxysmal EEG disturbance; seizures that may be severe but self-limited; behavioral deterioration, with or without premorbid developmental disturbances; no demonstrable brain pathology sufficient to explain the behavioral deterioration; and stabilization or improvement of behavior once the epileptiform EEG abnormalities resolve [1]. Much serious in CSWS are cognitive and behavioral disturbances. In CSWS, paroxysmal activity permanently affects the frontal lobes and higher cognitive functions. At highest risk for permanent sequelae are those with the earliest and longest exposure to the active phase of CSWS. When and how should CSWS be treated?

CSWS can be characterized by frontal lobe dysfunction. The onset of paroxysmal electrical activity was closely related in time with the development of a neuropsychological and behavioral pattern characterized by inattention, impulsiveness, mood swings and perseveration, with deficits in time orientation, reasoning and learning strategies, which shared many features with adult frontal syndrome [2]. In CSWS, paroxysmal activity affects the frontal area and results in those impairments. The most common location for a lesion to produce secondary bilateral synchrony, which is representative EEG manifestation for CSWS, is in the frontocentral region. Frontal foci may easily elicit sustained bisynchronous discharges which often spread diffusely. Damage to the prefrontal cortex has been associated with an inability to organize and carry out goal-directed behaviors. In our previous 3-dimensional MRI volumetric studies, frontal and prefrontal lobe volumes revealed growth disturbance in all CSWS patients compared with those of normal subjects [3,4]. Moreover, in the patients with shorter seizure durations and CSWS periods, ratios were soon restored to a more normal growth ratio, whereas growth disturbances of the prefrontal lobes were persistent in the patients with longer seizure durations and CSWS periods. These findings suggest that seizure and the duration of paroxysmal anomalies may be associated with prefrontal lobe growth, which is associated with neuropsychological problems. Current research suggests that damage to the frontal regions during childhood may interrupt normal maturational processes and organization, resulting in impairments to neurobehavioral development. Integrative executive functions may thus rely on the health of frontal lobe tissue and connectivity with the rest of the cortex [5]. Children with CSWS may have cognitive and behavioral impairments related to frontal lobe dysfunctions. Moreover, the duration of CSWS period seems to be a significant prognostic factor.

In CSWS, there is strong evidence that cognitive functioning may dramatically improve if epileptic activity is reduced with antiepileptic drugs (AED) [6]. However, many CSWS children develop severe cognitive and language deterioration that is unresponsive to medical treatment as the disease progresses [7]. Further, cognitive and behavioral severity and persistence seem to be correlated with the duration and severity of EEG abnormalities. To prevent these disturbances and problems in CSWS, we should appropriately treat the patients to suppress CSWS as early as possible. Few AEDs may control CSWS, but none of them proved to be efficient in a randomized procedure. In recent reports, the effect of levetiracetam (LEV) has been observed in CSWS [8]. LEV has the advantage of preventing any worsening in CSWS. Based on these previous studies, the best treatment options may be required to remit seizures and EEG abnormalities as soon as possible to achieve the most favorable prognosis for CSWS patients. Aggressive treatment approaches to abolish the paroxysmal disturbance should be seriously considered in the high-risk group. LEV may represent an important addition to the treatments available for CSWS.

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