Pediatric Neurology Part I: Chapter 66. Epileptic encephalopathy with continuous spike-waves during slow-wave sleep including Landau-Kleffner syndrome (Handbook of Clinical Neurology)

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Epileptic encephalopathy with continuous spike-waves during slow-wave sleep (CSWS) is a spectrum of epileptic conditions best defined by the association of cognitive or behavioral impairment acquired during childhood and not related to another factor other than the presence of abundant interictal epileptiform discharges (IED) during sleep, which tend to diffuse over the whole scalp. It is part of the childhood focal epileptic syndromes, some cases being idiopathic and overlapping with benign rolandic epilepsy, and others being symptomatic of a structural brain lesion. Landau–Kleffner syndrome (LKS) is a particular presentation where acquired aphasia is the core symptom. Clinical, neurophysiological, and cerebral glucose metabolism data support the hypothesis that IED play a prominent role in the cognitive deficits by interfering with the neuronal networks at the site of the epileptic foci but also at distant connected areas. Therefore, the treatment should aim to suppress IED. This may be achieved using conventional antiepileptic drugs, but corticosteroids seem to have more pronounced and sustained efficacy. Outcome for epilepsy is usually good, CSWS being an age-dependent EEG pattern, whereas outcome for cognition, language, and behavior is variable. Rehabilitation represents an important part of the treatment and visual forms of language should be encouraged in children with LKS.

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