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Abstract

Neurocognitive development in pediatric epilepsy

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Cognitive deficits are highly prevalent in children with epilepsy, ranging from 25 to 50% depending on age groups [1-3]. In addition to onset of epilepsy in infancy, other risk factors for cognitive deficits are resistance to anti-epileptic drugs (AED) and symptomatic etiology. The origin of the cognitive deficits is often multifactorial, and the challenge for the clinician is to identify the respective roles of the underlying etiology, the possible side effects of AED, and the epileptic activity (clinical seizures as well as interictal epileptiform discharges, IED). The role of epileptic activity is recognized in epileptic encephalopathies. Indeed, epileptic encephalopathy embodies the notion that the epileptic activity itself may contribute to severe cognitive and behavioral impairments above and beyond what might be expected from the underlying pathology alone, and that these can worsen over time [4]. A longitudinal study performed in a cohort of patients with epilepsy onset before age 3 has confirmed progressive adaptive behavior decline in epileptic encephalopathies starting in infancy, i.e. West syndrome, Dravet syndrome, and early-onset Lennox-Gastaut syndrome [5]. In some syndromes like Dravet syndrome, it is probably the repetition of seizures that contributes to cognitive decline, as IED are not abundant or even absent in the first stages of the disease. One study suggests that it is the occurrence of myoclonic seizures and absences but not convulsive seizures that are associated to cognitive decline [6]. In West syndrome, it is probably the interictal EEG abnormalities, i.e. the hypsarrhythmic pattern, which is detrimental for cognitive development. Thus, early control of spams is essential for cognitive outcome at least in West syndrome without underlying etiology identified [7]. In epileptic encephalopathies with continuous spike-waves during slow-wave sleep (CSWS), clinical and imaging studies evidence that interictal epileptic activity during sleep plays a major role in cognitive deterioration observed in these patients when awake [8]. Finally, first data of epilepsy surgery in children support the positive effect of surgery on cognition when performed early in the course of the disease [9].

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