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Editorial

Advances in childhood epileptic encephalopathies

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This special issue of the Journal of Pediatric Epilepsy is devoted to review the state of the art in the field of epileptic encephalopathies, with especial interest in the description of cognitive, imaging and electroclinical correlates. The term "epileptic encephalopathy" helps us to group seizure disorders in which the epileptiform abnormalities themselves are believed to be the main factor contributing to a progressive cerebral dysfunction characterized by cognitive deficits, behavioral disturbances, and/or psychomotor dysfunction. The Classification and Terminology Task Force of the International League Against Epilepsy includes the following syndromes under this diagnostic label: Ohtahara syndrome, early myoclonic encephalopathy, West syndrome, severe myoclonic epilepsy in infancy (Dravet's syndrome), Lennox-Gastaut syndrome, electrical status epilepticus during slow wave sleep, Landau-Kleffner syndrome.

In the first manuscript, Dr. Korostenskaja presents an extensive review of the current literature on neuroimaging and childhood epileptic encephalopathies (CEEs). With the advent of advanced neuroimaging techniques, used alone or in conjunction with electroencephalography, the ability to characterize brain dysfunction in CEEs improved. Contributions from structural (magnetic resonance imaging, diffusion tensor imaging) and functional neuroimaging techniques (positron emission tomography, single photon emission, computed tomography, functional magnetic resonance imaging, magnetoencephalography) are summarized for each of the main syndromes: early myoclonic epilepsy, Ohtahara syndrome, West syndrome, Dravet syndrome, Lennox-Gastaut syndrome, Landau-Kleffner syndrome, epilepsy with continuous spike and waves during slow wave sleep, and myoclonic status in non-progressive CEEs. This is a systematic review devoted to reflect on how these techniques can now contribute to improve the early differential diagnosis, the monitoring of treatment effects and the prediction of functional evolution on these patients.

The second review manuscript, presented by Dr. Salinas, provides an extensive overview of the neuropsychological functioning and the developmental outcomes in CEEs. Although the presence of seizures can play a dominant role in the cognitive development of the child with CEEs, it is important to recognize the role of the underlying pathology independent of seizures. The main focus of the paper is to reflect on how cognitive development in CEEs is associated not only to the clinical features defining each of the syndromes (i.e., electrographic findings, imaging and genetics) but as well can be related to the effects of the selected treatment. In children with CEEs pervasive and frequent seizures interferes with normal learning and development, precluding establishment of normal circuits upon which cognitive and behavior functions depend.

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Following these two comprehensive reviews on the neuroimaging and cognitive profiles associated to epileptic encephalophaties, two manuscripts present actual clinical experiences (electroclinical features and evolution of patients) in two of the less known syndromes: epileptic spasms in clusters without hypsarrhythmia, and myoclonic status in non-progressive encephalopathies. Dr. Caraballo presents first their experience in a group of 41 patients with myoclonic status in non-progressive encephalopaties describing the electroclinical features and outcome. Descriptions of an epileptic syndrome characterized by the recurrence of long-lasting myoclonic status in children with a non-progressive encephalopathy are scarce. In the last paper of this special issue, Dr. Caballo presents an update of the electroclinical features and evolution of patients with epileptic spasms in clusters without hypsarrhythmia, with or without focal or generalized paroxysmal discharges on the interictal electroencephalography. He concludes that the distinct electroclinical profile shown in this group of patients represent an epileptic encephalopathy that may be considered a distinct epileptic syndrome rather than a variant of West syndrome.

As more refined methods are incorporated in the clinical evaluation of pediatric epilepsies, including advanced neuroimaging and cognitive evaluations, our definition of "epileptic encephalopathy" is being reconsidered. One of the aims of these new methods is to characterize the relative effect of seizures on cognitive development in pediatric epilepsies.