Some Reflections about Electrical Status Epilepticus During Slow-Wave Sleep

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Taking a cue from some recent interesting reviews about electrical status epilepticus during slow-wave sleep (ESES) syndrome,[1] we underline the following points to foster the debate on this important topic in pediatric epileptology.

a) The first literature report of a condition with electroencephalogram (EEG) continuous paroxysmal abnormalities during sleep associated with cognitive impairment probably dates back to the 1942, when Kennedy and Hill described in a child the clinical picture of "dementia dysrhythmica infantum."[2] In the following decades Tassinari et al. defined this nosographic entity in a much more detailed and modern way, describing the EEG pattern characterized by diffuse spike-and-waves occupying at least 85% of the slow sleep duration, associated with neuropsychological deterioration, motor impairment (ataxia, dyspraxia, etc.), and focal or apparently generalized seizures.[3] This is the typical, and relatively rare, electro-clinical picture of ESES included in the International Classification of the Epilepsies among epileptic encephalopathies, where the massive EEG abnormalities impair development, leading to cognitive slowing or regression. This concept was, however, already implicit in Kennedy and Hill's paper.[2]

b) Over the years, numerous cases with frequent paroxysmal abnormalities occupying less than 85% of slow sleep and with heterogeneous clinical pictures, milder than the classic ESES, have been reported. Precisely these atypical ESES forms represent a challenge for pediatric epileptology, since they are much more frequent than the classic forms and harder to diagnose, and for them deciding whether to undertake a treatment aimed at improving the EEG (and if so, which one) is more complex.[1] In fact, even in atypical ESES the drug resistance is frequent and, unfortunately, the most effective pharmacotherapies (ie, corticosteroids) could have rather severe side effects.[1]

c) Landau-Kleffner syndrome (LKS) is often considered as an ESES variant,[3] but we believe they should be kept distinct. EEG in LKS shows bitemporal continuous paroxysmal abnormalities during slow-wave sleep, persisting during REM sleep, while in ESES spike-and-waves disappear during REM sleep. Clinically, LKS is characterized by acquired mixed aphasia, and not by global cognitive and behavioral deterioration as in ESES; seizures are less frequent and sometimes absent.[3] The long-term outcome of language in LKS is very heterogeneous and one of the main negative prognostic factors is a long duration of bitemporal continuous paroxysmal abnormalities during sleep. LKS is undoubtedly linked to ESES, also because it is possible to switch from one to the other, but they should remain distinct due to their respective electro-clinical differences.[4]

d) In particular in cases of ESES without an acquired structural etiology (eg, perinatal suffering), a genetic investigation is necessary (including array comparative genomic hybridization and next-generation sequencing multigene panel), due to the copy number variants (eg, Xp22.12 deletion) and the monogenic mutations (eg, *GRIN2A* mutations) that have been reported.[1] Regardless of the root cause, which is often unknown, there is much evidence

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from electrophysiological and brain imaging studies suggesting that thalamus may play a role in the ESES pathophysiology through a dysfunction of the thalamocortical circuit.[5]

e) Finally, we underline the importance of performing an EEG during sleep in all children showing a cognitive and/or behavioral decline, in order to exclude a possible ESES, even in the absence of seizures. According to our experience, an EEG picture of ESES is also possible in children who come to the attention of the neuropsychiatrist following the stagnation or regression of development.

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